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Prognosis of Patients Victim of Spontaneous Subarachnoid Hemorrhage: A Comparison between Radiologic Scales

Prognóstico de pacientes vítimas de hemorragia subaracnóidea espontânea: comparação entre escalas radiológicas

Luiza Maria Dias Abboud Hanna[1] Sarah Eloisa Biguelini[1] Francisco Alves de Araujo Junior[1] Anderson Matsubara[2] Pedro Helo dos Santos Neto[3]

[1] Faculdades Pequeno Príncipe, Curitiba, Paraná, Brazil
[2] Hospital Universitário Evangélico Mackenzie, Curitiba, Paraná, Brazil
[3] Faculdade Evangélica Mackenzie do Paraná, Curitiba, Paraná, Brazil

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Address for correspondence Luiza Maria Dias Abboud Hanna, Rua Coronel Pedro Scherer Sobrinho, Cristo Rei, 152, Bloco 4A, Apartamento 211, Curitiba, Paraná, Brazil (e-mail: luizamariahanna@hotmail.com).

Abstract

Objective To analyze the population and the early mortality rate (up to thirty days) of patients victim of spontaneous subarachnoid hemorrhage (SAH) according to the Hunt-Hess clinical scale and the Fisher and modified Fisher radiological scales.

Materials and Methods We analyzed 46 medical records and skull computed tomography (CT) scans of patients with spontaneous SAH admitted between February 2014 and December 2017 at Hospital Universitário Evangélico Mackenzie, in the city of Curitiba, state of Paraná, Brazil. The method of the study was exploratory-descriptive, transversal and retrospective, with a quantitative approach. We analyzed epidemiological (gender, age), clinical (life habits, pathologies, Glasgow coma scale and Hunt-Hess scale) and radiological (Fisher and modified Fisher scales) variables, and the Hunt-Hess and the Fisher scales were correlated with risk of death. The data was submitted to statistical analysis considering values of $p < 0.05$.

Result There was a higher prevalence of spontaneous SAH among women (69.5%), as well as among patients aged between 51 and 60 years (34.7%). Regarding the grades on the scales, there was higher prevalence of Fisher 4, Modified Fisher 4 and Hunt-Hess 2. Evolution to death was higher among women (76.4%) and patients aged between 61 and 70 years (35,2%).

Conclusion Mortality was higher among patients classified as Fisher 3, Modified Fisher 4 and Hunt-Hess ≥ 3. The Fisher scale is better than the modified Fisher scale to assess the risk of mortality.

Keywords
- intracranial vasospasm
- subarachnoid hemorrhage
- aneurysm
- prognosis

Resumo

Objetivo Analisar o perfil epidemiológico, assim como a taxa de mortalidade precoce (até 30 dias do internamento) de pacientes vítimas de hemorragia subaracnóidea espontânea (HSAE) de acordo com a escala clínica de Hunt-Hess, e com as escalas radiológicas de Fisher e de Fisher modificada.

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Materiais e Métodos Realizamos análise de 46 prontuários e exames de tomografia computadorizada (TC) de crânio de pacientes vítimas de HSAE no período de fevereiro 2014 a dezembro 2017 admitidos no Hospital Universitário Evangélico em Curitiba-PR. O método de escolha foi o exploratório-descritivo, transversal e retrospectivo, com abordagem quantitativa. Foram analisadas variáveis epidemiológicas (idade, sexo), clínicas (hábitos de vida, patologias, escala de coma de Glasgow e escala de Hunt-Hess) e radiológicas (escala de Fisher e de Fisher modificada), e as escalas de Hunt-Hess e de Fisher foram correlacionadas com o risco de óbito. Os dados foram submetidos a análise estatística considerando valores de $p < 0,05$.

Resultados Houve maior prevalência de HSAE entre mulheres (69,5%), assim como entre pacientes com idade entre 51 e 60 anos (34,7%). Entre as escalas, houve maior prevalência nas graduações Fisher 4, Fisher modificada 4 e Hunt-Hess 2. Os pacientes que mais evoluíram a óbito foram os do gênero feminino (76,4%), e também aqueles com idade entre 61 e 70 anos (35,2%).

Conclusão A mortalidade foi maior em pacientes classificados com Fisher 3, Fisher Modificado 4 e Hunt-Hess ≥ 3. A escala de Fisher é melhor do que a de Fisher modificada para avaliar o risco de mortalidade.

Palavras-chave
- vasoespasmo intracraniano
- hemorragia subaracnóidea
- aneurisma
- prognóstico

Introduction

Spontaneous subarachnoid hemorrhage (SAH) caused by rupture of a cerebral aneurysm has a mortality rate of 48% in patients older than 60 years of age, whereas in younger individuals, the rate is of 19%.[1,2] Furthermore, 40% of the hospitalized patients die within a month of the event. Vasospasm, which occurs in 20% to 40% of the patients, is considered the most frequent complication in cases of spontaneous SAH.[3] The Fisher scale is widely used for the classification of spontaneous SAH, and it is based on the findings of skull computed tomography (CT); however, it fails to grade the risk of symptomatic vasospasm in those patients. The modified Fisher scale appears to have a higher predictive power for the occurrence of this complication.[3] Patients with SAH can also be classified according to their surgical risk, through the scale developed by Hunt & Hess, which is based on the clinical presentation of the patients.[4]

The objective of the present study is to evaluate the morbidity and mortality of individuals with spontaneous cerebral aneurysm rupture, based on the aforementioned scales, with the purpose of assisting the therapeutic management of these patients and comparing both scales according to their efficacy while predicting the occurrence of vasospasm.

Materials and Methods

The present study adopted an exploratory-descriptive and transversal method, with a quantitative approach. After signing the Data Use Consent Term and obtaining approval and consent from Faculdades Pequeno Príncipe (2,692,293) and the Ethics Committee at Hospital Universitário Evangélico Mackenzie (2,722,606), the data collection began.

The study sample was obtained through the analysis of the medical records of patients admitted to Hospital Universitário Evangélico Mackenzie (a reference center for neurosurgery), in the city of Curitiba, state of Paraná, Brazil, from February 2014 to December 2017, with a diagnosis of SAH caused by spontaneous cerebral aneurysm rupture. The initial sample consisted of 58 patients, and, after the first selection, 12 participants were excluded due to lack of information on the medical records or unavailable imaging exams.

First, the variables sex, age, vasospasm, death, clinical Hunt-Hess scale, Fisher and Modified Fisher radiological scales were subjected to a descriptive analysis. The frequency of each early death episode (up to 30 days of hospitalization) was assessed according to sex, age and vasospasm separately. Each scale was then analyzed separately regarding death and vasospasm.

Through the Logit model, the variable death was compared concomitantly with age, gender, hypertension, diabetes mellitus (DM), smoking, vasospasm, and the grades on the Hunt-Hess, Glasgow coma, and Fisher scales, followed by the comparison of death with the same variables, exchanging the Fisher scale with modified Fisher scale.

Finally, the odds ratio (OR) of each of the grades on the Fisher and modified Fisher scales was calculated to obtain the odds of the evolution into vasospasm.

The level of significance was set at 5% ($p = 0.05$) for all statistical tests, and the software used was the TIBCO Statistica (TIBCO Software Inc, Palo Alto, CA, US), version 13.4.0.14.

Results

The analysis of medical records showed a prevalence of female individuals, corresponding to 69.5% ($n = 32$)of the population studied, and individuals between 51 and 60 years of age, which is equivalent to 34.7% ($n = 16$) of the sample. Vasospasm affected 39.1% ($n = 18$) of the cases observed, 8 (44.4%) of which died. Regarding early mortality (death within 30 days of hospitalization), 17 individuals (36.9%) died, 76.4% ($n = 13$)

of which were female, as well as 35.2% ($n = 6$) of individuals in the age range of 51 to 60 years. A 100% frequency was observed in patients classified as Hunt-Hess 5 ($n = 4$).The highest occurrence of this outcome happened in individuals classified as Fisher 3 (equivalent to 54.5%; $n = 6$) and modified Fisher 4 (equivalent to 53%; $n = 9$).

When analyzing the OR of the frequencies obtained, there was a greater tendency of evolution to vasospasm in patients classified as Fisher 3 (2.3 times higher than those of other grades).

To analyze the relationship between mortality and the other characteristics described in the present study, the logit model was applied, in which the variables age, gender, high blood pressure, DM, smoking, vasospasm, Hunt-Hess scale, Glasgow coma scale and Fisher grades 1, 2, 3 and 4 are concurrently analyzed. When observing the results of the model used, we understood that there is a relationship between the aforementioned factors and early mortality among the patients, with 80.43% of reproducibility to confirm this hypothesis in different population groups ($p = 0.009$).

In a second evaluation, the Fisher scale grades were replaced by modified Fisher 1, 2, 3 and 4 in the same logit model, to verify if there was a difference between the Fisher and modified Fisher scales as predictors of mortality. The interpretation of the data, when using the modified Fisher scale, showed that there was a relationship between death and the conditions analyzed concomitantly, with a reproducibility of 77.78% to confirm this hypothesis in different population groups ($p = 0.01$).

Discussion

On the one hand, the epidemiological results of the present study were very similar to those of previous studies, especially regarding the gender and the age of the patients: women were more affected,[5–8] as well as individuals between 50 and 60 years of age.[5–8] Some studies correlate the higher risk among women with the decrease in the levels of female hormones after menopause.[5,9,10]

On the other hand, 58,6% ($n = 27$) of our patients suffered from high blood pressure, against only 38% of the sample in another research.[6] The same thing occurred with the smoking rate: 30.4% ($n = 14$) of our patients were smokers, against 64.3% of the patients i another study.[8] Both of these variables are considered risk factors for SAH.[11]

Regarding the grades of the patients at admission, there was no predominance of any grade on any of the scales.

As well as the epidemiological results, the mortality rate were very similar to that of another study,[12] in which it was higher among patients aged between 50 and 60 years and those who were female.[12] Althought some authors state that blood pressure does not increase the risk of death, 23,9% ($n = 11$) of the patients with high blood pressure in the present trial died. For individuals who suffer from diabetes, the risk was lower than 15%.

All patients classified as Hunt Hess 5 evolved to death, and the risk was directly proportional to the grade, such as in other studies.[12,15] In line with other studies, in the present study the patients classified as modified Fisher 4 had the higher risk of death,[3,12], as well as those classified as Fisher 3.

The OR proved that patients classified as Fisher 3 had the highest risk of evolution to vasospasm.[3]

There was no study in the literature comparing radiological scales with the risk of death. However, when considering all of those variables concomitantly, as the modified Fisher scale has 77.78% of reproducibility ($p = 0.01$), while the Fisher classification has 80% ($p = 0.009$), when using the OR, the evidence shows that Fisher 3 is better to predict this risk.

Conclusion

Our findings suggest that the risk of developing SAH is higher among women and people aged between 51 and 60 years. Among the pathologies, more than 50% ($n = 27$) of our patients suffered from high blood pressure. At admission, the most common grades were Fisher 4, modified Fisher 4 and Hunt-Hess 2.

The patients who are more likely to evolve to death are women, those aged between 61 and 70 years, and those classified as Fisher 3, modified Fisher 4 and Hunt-Hess 5.

Conflict of Interests

The authors have no conflict of interests to declare.

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Meningioma of the Fourth Ventricle: Literature Review*

Meningioma do quarto ventrículo: Revisão da literatura

Allan Dias Polverini[1] Rodrigo de Almeida Simon Sola[1] Guilherme Fonseca Bortoluzzi[1] Ismael Augusto Silva Lombardi[1] Giusepe Picone Junior[1] Carlos Roberto de Almeida Junior[1] Carlos Afonso Clara[1]

[1] Department of Neurosurgery, Hospital de Amor, Barretos, SP, Brazil

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Address for correspondence Rodrigo de Almeida Simon Sola, MSc, Rua Coronel Bento, 394, Vilas Boas, Campo Grande, MS, 79051-110, Brazil (e-mail: rodssola@gmail.com).

Abstract

Meningiomas are among the most common central nervous system tumors, with an incidence that ranges from 15% to 40% of intracranial tumors. Of these, only 0.5% to 3% are intraventricular, and the rarest of them occurs in the fourth ventricle.

Fourth-ventricle meningiomas originate generally from the choroid plexus and have no dural adhesions. Most often, they manifest in young patients, around 41 years of age, with a possible predominance in females, through intracranial hypertension and cerebellar syndromes. The treatment consists of surgical resection, which commonly presents good results due to the characteristics of the tumor. So, for better preoperative planning, the radiological differentiation of the most frequent tumors in this location is important.

The most common histologic subtypes are fibroblastic and meningothelial, both grade I according to the World Health Organization (WHO), although there are reports of tumors of grades II and III.

We report a case of meningioma of the fourth ventricle operated in our institution, and we have conducted a literature review, through which we found that 57 cases have been reported so far, with the first one reported in 1938.

Keywords

- meningioma
- fourth ventricle
- infratentorial neoplasms

Resumo

Os meningiomas estão entre as neoplasias mais comuns do sistema nervoso central, com incidência que pode variar entre 15% e 40% dos tumores intracranianos. Apenas 0,5 a 3% deles são intraventriculares, dos quais os mais raros são os do quarto ventrículo. Os meningiomas do ventrículo IV se originam, geralmente, do plexo coroide, e não têm aderências durais. Na maioria das vezes, manifestam-se em pacientes jovens, em torno dos 41 anos, com possível predominância no sexo feminino,

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por meio de síndromes de hipertensão intracraniana e cerebelar. O tratamento consiste em ressecção cirúrgica, que, comumente, apresenta bons resultados devido às características tumorais. Por isso, para um melhor planejamento pré-operatório, faz-se importante a diferenciação radiológica entre os tumores mais frequentes nessa região.

Os subtipos histológicos mais comuns são fibroblástico e meningotelial, ambos de grau I da Organização Mundial da Saúde (OMS), embora existam relatos de tumores de grau II e III.

Relatamos um caso de meningioma do ventrículo IV operado em nossa instituição, e realizamos revisão da literatura, mediante a qual verificamos que foram relatados 57 casos até o momento, sendo o primeiro de 1938.

Palavras-chave
- meningioma
- quarto ventrículo
- neoplasias infratentoriais

Introduction

Meningiomas are meningothelial neoplasms that originate in the arachnoid fibroblast, a squamous cell that covers the arachnoid villi and the Pacchioni granulations. They correspond to at least 15% of all intracranial neoplasms,[1] but may reach 40%,[2] and are classically adhered to meningeal structures.

Purely intraventricular lesions are rare and have a calculated incidence between 0.5% and 3% of intracranial meningiomas.[2] Out of these, 77.8% occur in the lateral ventricle trigone, with a higher frequency on the left side; 15.6% occur in the third ventricle; and 6.6%, in the fourth ventricle.[1,3,4]

It is believed that the meningiomas in the fourth ventricle originate from the choroid plexus, and do not present dural adhesions.[1,3,5] The first case was described by Cushing and operated by Sachs in 1938.[3,6,7]

We describe the case of a 31-year-old patient, who was operated at our service for resection of a tumor in the fourth ventricle whose anatomopathological diagnosis indicated meningioma; we also performed a review of the literature.

Case Report

A 31-year-old male patient, from the city of Caçu, state of Goiás, Midwestern Brazil, was referred to Hospital de Amor, in the city of Barretos, with a vertiginous condition that had been progressing for $\sim$ 8 months, associated with nausea and malaise. Upon neurological examination, he manifested dysfunction of cranial nerves IX, X, XI and XII and cerebellar syndrome, which presented in the form of dysbasia, ataxia and dysmetria, and was more pronounced on the left side. The subsequent investigation, with magnetic resonance imaging (MRI) (►**Figs. 1**, **2** and **3**), revealed a solid lesion of lobulated contour and intense enhancement after injection of a paramagnetic contrast medium in the Luschka foramen topography to the left and fourth ventricle. The lesion had thin cerebrospinal fluid fissure between the cerebellar vermis and the tumoral plane, with 4.7×4.0 cm in its major axes, compressing the bulb, without contact with adjacent meninx, with no dural tail identification. There was also a slight dilation of the supratentorial cerebral ventricles.

A bilateral suboccipital craniotomy was performed, with the installation of an external ventricular bypass and total macroscopic excision of the lesion (►**Figs. 4**, **5** and **6**), without intercurrences. During the intraoperative period, absence of tumor adhesions to the meningeal structures of the posterior fossa was confirmed. A microsurgical resection of the lesion was performed, and we opted for intralesional emptying with the aid of ultrasonic aspiration, followed by resection in fragments of the tumor remnant, without the use of electrophysiological monitoring, which was not available at the time, and avoiding traction maneuvers. Total macroscopic resection of the neoplasm was achieved.

In the postoperative period, the condition evolved with worsening of the dysfunctions of cranial nerves IX, X and XI and grade-III hemiparesis to the left, presenting a slow and progressive improvement. The patient underwent early tracheostomy and gastrostomy.

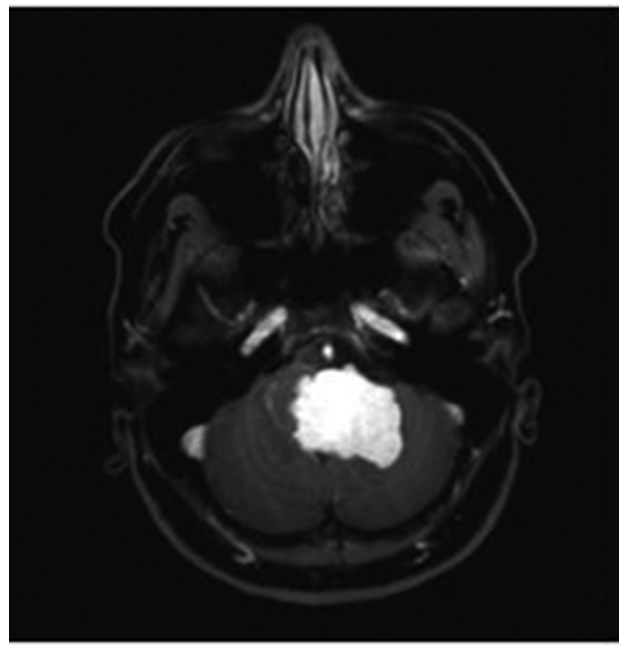

Fig. 1 Encephalon T1-weighted MRI scan with contrast showing tumor mass in the region of the fourth ventricle, with homogeneous enhancement and significant mass effect on the brainstem in the axial section.

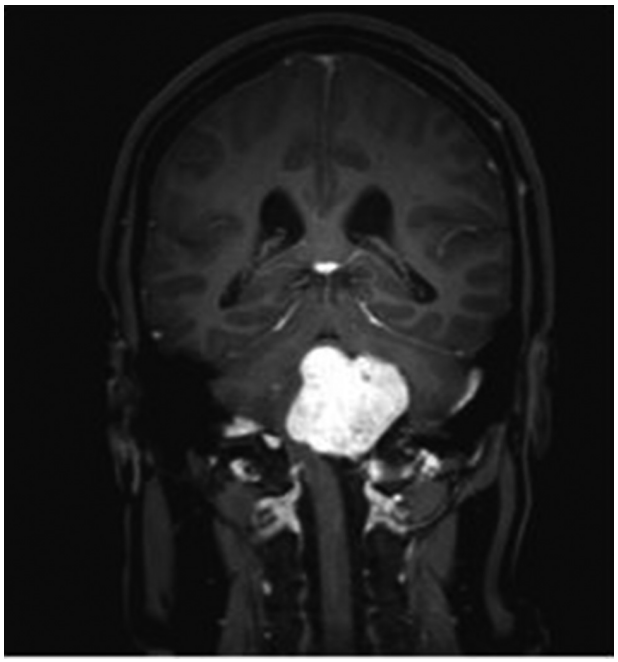

Fig. 2 Encephalon T1-weighted MRI scan with contrast showing tumor mass in the region of the fourth ventricle, with homogeneous enhancement and significant mass effect on the brainstem in the coronal section.

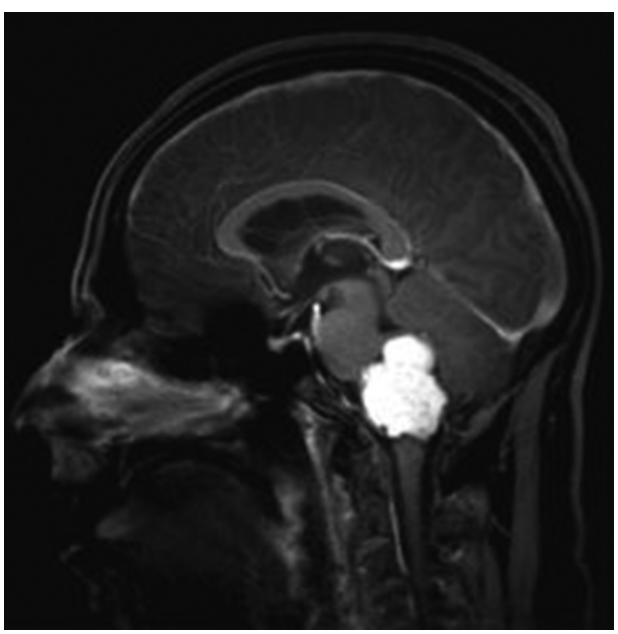

Fig. 3 Encephalon T1-weighted MRI scan with contrast demonstrating tumor mass in the region of the fourth ventricle, with homogeneous enhancement and an important mass effect on the brainstem in the sagittal section.

A picture of bronchopneumonia was diagnosed, with probable aspiration etiology, and urinary tract infection, which were treated satisfactorily, in addition to cerebrospinal fluid fistula associated with hydrocephalus, which was readily treated with a ventricular-peritoneal shunt (VPS) during the same hospitalization. The patient evolved with prolonged hospitalization and was enrolled into an intensive physical rehabilitation program.

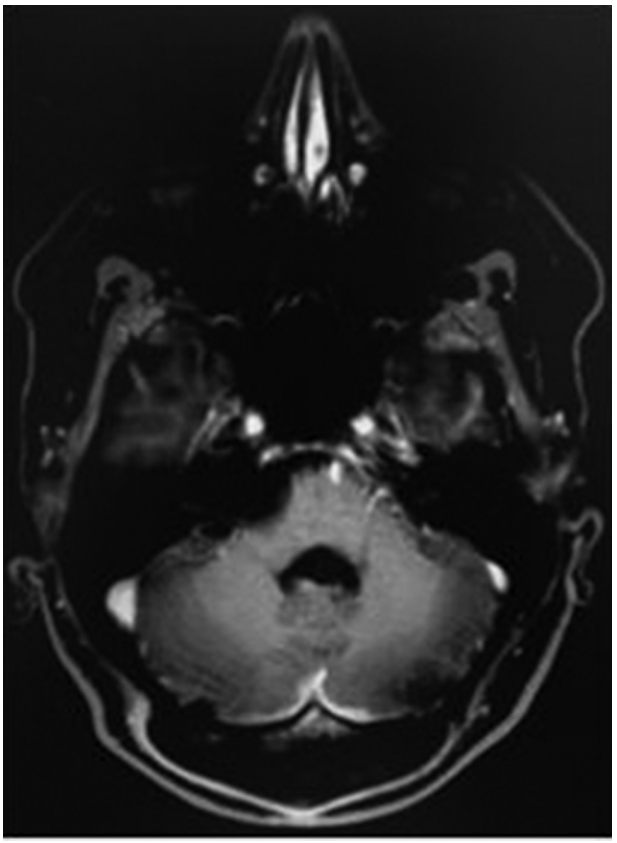

Fig. 4 Postoperative encephalon T1-weighted MRI scan with contrast in the axial section, showing complete resection of the tumor lesion, with centralization of the brainstem and free fourth ventricle.

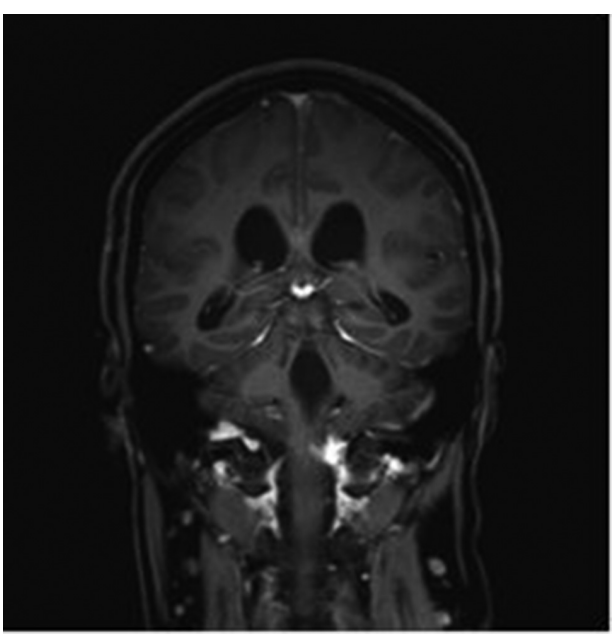

Fig. 5 Postoperative encephalon T1-weighted MRI scan with contrast in the coronal section, showing complete resection of the tumor lesion, with centralization of the brainstem and free fourth ventricle.

The patient followed with progressive improvement of the deficits after hospital discharge, and, around 9 months after the procedure, presented complete recovery of the

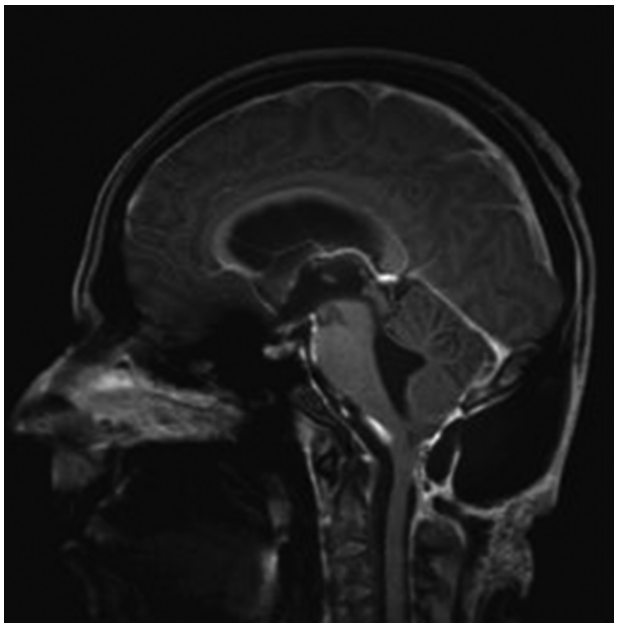

Fig. 6 Postoperative encephalon T1-weighted MRI scan with contrast, in the sagittal section, showing complete resection of the tumor lesion, with centralization of the brainstem and free fourth ventricle.

cranial nerve impairment, significant reversal of motor deficit, and significant improvement in the ataxic frame, maintaining a slight incoordination on the left side.

The histopathologic study revealed that it was a fibroblastic meningioma, grade I according to the World Health Organization (WHO). After 2 years of follow-up, he was referred to the hospital service in his hometown due to dysfunction of the DVS and history of viral meningitis treated in that city months before. The patient was then diagnosed again with meningitis and presented rapid worsening in the clinical condition due to sepsis, which caused his death despite the treatment.

Discussion

Meningiomas of the fourth ventricle are defined as those that have their origin in the local choroid plexus and occupy this ventricular cavity without meningeal implantation.[1,3,5] In 1963, Abraham and Chandy suggested a classification for posterior fossa meningiomas, without dural implant, consisting of three types: 1) choroid plexus meningiomas, which develop only in the fourth ventricle; 2) choroid screen meningiomas, which develop partially in the interior of the ventricle and partially in the cerebellar hemisphere and vermis; and 3) cisterna magna meningiomas, without dural implantation and with intraventricular extension, which are originate from the most lateral portion of the choroid plexus, outside the Luschka formen.[1,8,9] Those classified as types 1 or 2 are deemed true. The case herein reported is an example of ventricle meningiomas classified as Abraham type I, as there is evidence of cerebrospinal fluid fissure between the vertex and the medial portion of the right cerebellar hemisphere with the tumor interface, as well as a more lateralized disposition of the neoplasia on the left.

Some published series have reported a prevalence of female patients, with a 2:1 ratio.[1,4,10] In this review, we noted an equivalent distribution between genders, with a slight prevalence of females, with 28 cases (48.28%). There were 25 male cases (43.10%), and there was no identification of gender in 5 cases (8.62%).

The overall mean age of the reported cases is 41.64 years old. When analyzed separately by gender, the mean age is discretely different: the female gender is slightly younger (mean age: 41.25 years) than the male gender (mean age: 42.08 years). Some publications suggest a higher incidence in younger female patients.[1]

Patients with fourth-ventricle meningiomas usually manifest signs of insidious intracranial hypertension (IH), such as morning headache, nausea, vomiting and vertigo, as well as focal signs most commonly characterized by ataxic syndrome (cerebellar), long tracts,[2,3,10] and cranial nerve involvement, notably those whose nuclei are located in the point-bulbar segment of the brainstem, such as the presently reported case.

Among the differential diagnosis of fourth-ventricle tumors are metastasis, choroid plexus papilloma, hemangioblastoma, medulloblastoma and ependymoma.[3,5,11] The radiological differentiation of such lesions is important for the surgical planning, because it implies different levels of difficulty for total excision and, therefore, different initial forms of intraoperative management of the lesions. The characteristics of meningiomas, both in the MRI and in computeed tomograhy (CT), indicate well-circumscribed lesions with regular and mild edges, probably of slow growth, with homogeneous and very intense enhancement by the contrast agent.[3,6]

The treatment of choice is the microsurgical excision of the lesion, using intralesional emptying, with aid, if possible, given the characteristics of the neoplasia, ultrasonic aspiration or piacemeal resection when necessary, initially avoiding traction maneuvers, and paying special attention to the dissection along the floor of the fourth ventricle, usually with a clear cleavage plane, which, together with the fact that it does not present meningeal fixation, leads to the good results published in the literature, with complete excision of the lesion.[12] Considering the cases with resection volumes reported so far, only four did not indicate complete resection.

Data from the international literature suggest the prevalence of the fibroblastic and meningothelial subtypes, corresponding respectively to 40% and 24% of the cases.[1] In our review, the subtypes classified as WHO grade I correspond to 79.31% of the cases, with a prevalence of fibroblastic (29.31%) and meningothelial (15.52%) meningiomas.

Meningiomas classified as WHO grade II correspond to 17.24% of the cases, with a prevalence of clear cell (6.89%) and atypical (6.89%) meningiomas. Chordoid meningiomas corresponded to 3.44% (3) of the reported cases.[13,14] Only one anaplastic meningioma (WHO grade III) was reported, whose diagnosis was established in the recurrence of the disease after a long period of remission.[15] The histopathologic classification of the lesion was not available in 5 (8.62%) reported cases.

Through research in the database of indexed journals, as shown in►**Table 1**, we detected the existence of 57 cases of

Table 1 Cases reported in the international literature

Case	Publication	Author	Year	Age	Gender	Resection	Histology
1	1	Sachs et al[1,3,8,11,18–20]	1938	38	Female	Total	Fibroblastic
2	2	Voguel and Stevenson[7]	1950	65	Male	Necropsy	Meningothelial
3	3	Haas and Ritter[16]	1954	41	Male	Necropsy	Not available
4	4	Bustamente Zuleta and Londono[17]	1955	12	Male	Subtotal	Laminar trend
5	4	Bustamente Zuleta and Londono[17]		8	Male	Biopsy	Diffuse
6	5	Schaerer and Woosley[18]	1960	42	Female	Total	Not available
7	6	Chafee and Donaghy[19]	1963	38	Female	Total	Meningothelial
8	7	Hoffman et al[20]	1972	61	Male	Total	Transitional
9	7	Hoffman et al[20]		44	Female	Total	Transitional
10	8	Rodrigues- Carbajal and Palacios[21]	1974	49	Female	Partial	Meningothelial
11	8	Rodrigues-Carbajal and Palacios[21]		32	Female	Total	Meningothelial
12	9	Gökalp et al[22]	1981	30	Female	Total	Psammomatous
13	10	Tsuboi et al[23]	1983	30	Female	Total	Fibroblastic
14	11	Nagata et al[24]	1988	52	Female	Total	Fibroblastic
15	12	Matsumara et al[25]	1988	62	Male	Total	Fibroblastic
16	13	Nakano et al[26]	1989	58	Female	Total	Transitional
17	14	Jhonson et al[27]	1989	53	Male	Total	Osteoblastic
18	15	Diaz et al[28]	1990	5	Female	Total	Meningothelial
19	16	Ceylan et al[9]	1992	48	Male	Total	Angiomatous
20	17	Delfini and al[29]	1992	22	Male	Total	Fibroblastic
21	18	Lima de Freitas et al[30]	1994	32	Female	Total	Meningothelial
22	19	Iseda et al[31]	1997	67	Female	Total	Atypical
23	19	Iseda et al[31]		47	Female	Total	Transitional
24	20	Cummings et al[32]	1999	72	Male	Total	Fibroblastic
25	21	Chaskis et al[6]	2001	76	Male	Total	Fibroblastic
26	22	Akimoto et al[12]	2001	72	Female	Total	Transitional
27	23	Ooigawa et al[11]	2004	51	Female	Total	Transitional
28	24	Carlotti et al[33]	2003	23	Female	Total	Clear cells
29	24	Carlotti et al[33]		28	Female	Total	Clear cells
30	25	Bathoe et al[34]	2006	Not available	Not available	Not available	Fibroblastic
31	25	Bathoe et al[34]		Not available	Not available	Not available	Meningothelial
32	26	Liu et al[4]	2006	Not available	Not available	Not available	Mixed
33	27	Epari et al[14]	2006	20	Female	Total	Chordoid
34	28	Bertalanffy et al[35]	2006	Not available	Not available	Total	Not available
35	29	da Costa et al[36]	2007	45	Male	Total	Not available
36	30	Shintaku et al[15]	2007	61	Female	Total	Anaplastic
37	31	Wind and al[13]	2010	23	Male	Total	Chordoid
38	32	Burgan et al[37]	2010	14	Male	Total	Clear cells
39	33	Alver et al[1]	2011	61	Male	Total	Fibroblastic
40	34	Pichierri et al[8]	2011	30	Female	Not available	Meningothelial
41	34	Pichierri et al[8]		22	Male	Not available	Fibroblastic

(*Continued*)

Table 1 (*Continued*)

Case	Publication	Author	Year	Age	Gender	Resection	Histology
42	34	Pichierri et al[8]		22	Male	Not available	Fibroblastic
43	35	Zhang et al[38]	2012	23	Male	Total	Angiomatous
44	36	Qin et al[5]	2012	25	Male	Not available	Fibrous
45	37	Takeuchi et al[3]	2012	60	Male	Subtotal	Meningothelial
46	38	Zhang et al[10]	2012	40	Female	Not available	Psammomatous
47	38	Zhang et al[10]		43	Female	Not available	Clear cells
48	38	Zhang et al[10]		65	Female	Not available	Fibroblastic
49	38	Zhang et al[10]		60	Female	Not available	Fibroblastic
50	38	Zhang et al[10]		20	Female	Not available	Transitional
51	38	Zhang et al[10]		39	Male	Not available	Fibroblastic
52	38	Zhang et al[10]		50	Male	Not available	Atypical
53	38	Zhang et al[10]		9	Female	Not available	Fibroblastic
54	38	Zhang et al[10]		69	Female	Not available	Fibroblastic
55	38	Zhang et al[10]		57	Male	Not available	Atypical
56	39	Ødegaard et al[2]	2013	Not available	Not available	Not available	Not available
57	40	Liu and Kasper[39]	2014	60	Male	Total	Atypical

meningiomas of the fourth ventricle, and 40 publications or references to them are in articles in English or Spanish.

Conclusion

Fourth-ventricle meningiomas have little incidence, possibly with a slight prevalence in young women, commonly presenting with progressive symptoms of IH and cerebellar syndrome. The surgical treatment is usually effective and has good results, considering, for that, adequate preoperative planning obtained through the radiological characteristics of the tumors. The low-grade lesions are the majority, although there is a broad spectrum of diagnosed subtypes.

Conflict of Interests

The authors have no conflict of interests to declare.

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Chordoid Meningioma: Literature Review

Meningioma cordoide: Revisão de literatura

Carlos Roberto Massella Junior[1] Marcus Vinicius de Morais[1] Rafael Pontes Figueiredo[1] Andrezza Garcia Morales[2] Paulo Mácio Porto de Melo[1]

[1] Neurosurgery Service, Hospital Militar de Área de São Paulo, São Paulo, SP, Brazil
[2] Hospital da Faculdade de Medicina do ABC, São Paulo, SP, Brazil

Address for correspondence Carlos Roberto Massella Junior, HMASP, R. Ouvidor Portugal, 230 - Vila Monumento, São Paulo - SP, 01551-010, Brazil (e-mail: cr.massellajr@gmail.com).

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Abstract

Chordoid meningiomas (CMs) are a rare subgroup of tumors, accounting for ~ 0.5% of all meningiomas. Chordoid meningioma tumors correspond to World Health Organization (WHO) Grade II lesions and behave aggressively, with an increased likelihood of recurrence. There are few genetic studies about CMs, but we understand that there is deletion at many chromosomal loci. Histologically, CMs are characterized by strands and cords of meningothelial cells arranged in a mucinous stroma. Morphologically, it can mimic other chondroid and myxoid tumors within the brain and its vicinity, thus posing a diagnostic challenge. Chordoid meningiomas have an aggressive clinical course and a propensity to recur compared with classical meningiomas. The goal of the treatment is surgery, with total resection of the tumor; however, due to its high degree of recurrence, radiotherapy is often necessary as an adjuvant treatment.

Keywords
- meningioma
- chordoid
- brain neoplasms
- pathology

Resumo

Meningiomas cordoides (MCs) são um tipo raro de subgrupo de tumores, representando ~ 0,5% de todos os meningiomas. Os MCs correspondem ao grau II, pela classificação da Organização Mundial da Saúde (OMS), possuindo agressividade e alto grau de recorrência. Existem poucos estudos publicados sobre a genética desses tumores; porém, entendemos que há diversas deleções em alguns lócus cromossômicos. Histologicamente, os MCs são caracterizados por fios e cordões de células meningoteliais dispostos em um estroma mucinoso. Morfologicamente, podem mimetizar outros tumores condroides e mixoides, o que representa um desafio diagnóstico. Os MCs têm um curso mais agressivo que o meningioma clássico. O objetivo do tratamento é a cirurgia, com ressecção total do tumor, porém, devido à sua alta recidiva, se faz necessário o tratamento adjuvante com radioterapia.

Palavras-chave
- meningioma
- cordóide
- tumor cerebral
- patologia

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Introduction

Chordoid meningioma (CM) is a rare variant of meningioma that bears a striking histological resemblance to chordoma and has a greater likelihood of recurrence. The differential diagnosis includes chordomas, myxoid chondrosarcoma and chordoid glioma.[1] In the present study, we report a case of a female patient with a diagnosis of a recent onset of headache and visual cloudiness. Meningiomas represent almost 30% of primary intracranial neoplasms.

Chordoid meningioma is a rare subtype of meningioma, accounting to 0.5% of all meningiomas and associated with different behavior.[2]

Chordoid meningiomas are classified in Grade II by the World Health Organization (WHO).[3] Chordoid meningioma has a similar distribution between men and women. Chordoid meningioma can have a large distribution in the age range, and may range from 4 to 77 years old.[4] A little more than 100 cases of chordoid meningioma have been described in the English language literature, the majority of which are in the pathology and neurosurgery literature.

Material and Methods

Case Report

Female, 42-year-old, with complaints of sudden onset of headache and visual impairment. Upon neurological examination, the patient presented paresis of the left medial rectus muscle. The neurosurgery team requested a magnetic resonance imaging (MRI) exam, which showed an expansive lesion in the left frontotemporal topography. The lesion was causing an important compression of the medial rectus muscle (►**Fig. 1**). We opted for a left frontotemporal craniotomy, since it was possible to resect the lesion. The patient progressed well during surgery and, after the postoperative period, was discharged from the hospital feeling well and with no neurological deficits. (►**Fig. 2**)

We sent the material for pathological analysis, which revealed mesenchymal neoplasia with chordoid pattern in the left temporal frontographic topography (►**Figs. 1** and **2**). Therefore, the diagnosis was of chordoid meningioma. The immunohistochemistry analysis was positive for epithelial membrane antigen, therefore being typical of the chordoid meningioma (►**Figs. 3** and **4**). The literature review involved clinical case reports, randomized controlled trials and a series of cases describing clinical and epidemiological treatments for chordoid meningioma. The search was performed using the PubMed database and targeting all English language publications available involving chordoid meningioma. This review was organized applying the MeSH terms *chordoid meningioma, meningioma* and *chordoid*. The search about chordoid meningioma at PubMed/Medline retrieved 83 articles. For all these articles, we applied filters such as species (humans) and age (19 to 65 years old). Then, we retrieved 59 papers. Analyzing the titles of the articles, 59 were selected for summary review, considering relevance and relation to the present paper. Among those 59, 31 were selected and examined.

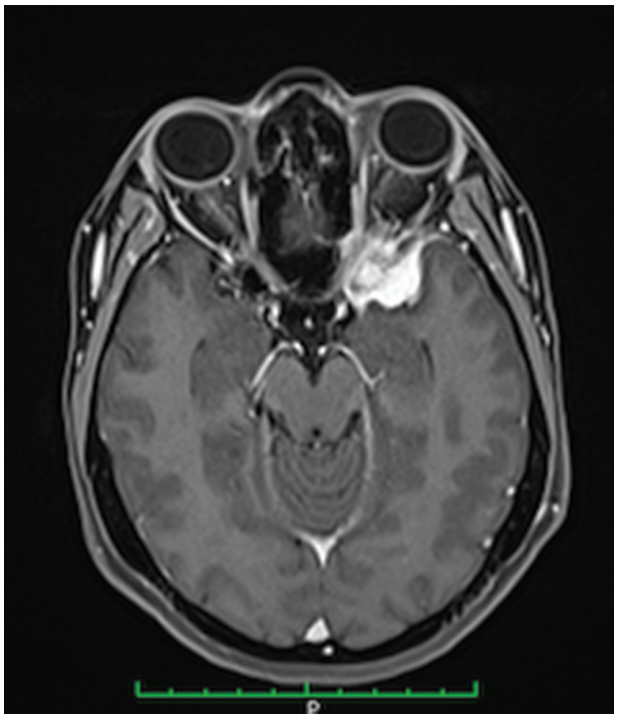

Fig. 1 Magnetic Resonance Imaging (MRI): Axial FLAIR showed an expansive lesion in the left frontotemporal topography. The lesion presented with important compression of the medial rectus muscle.

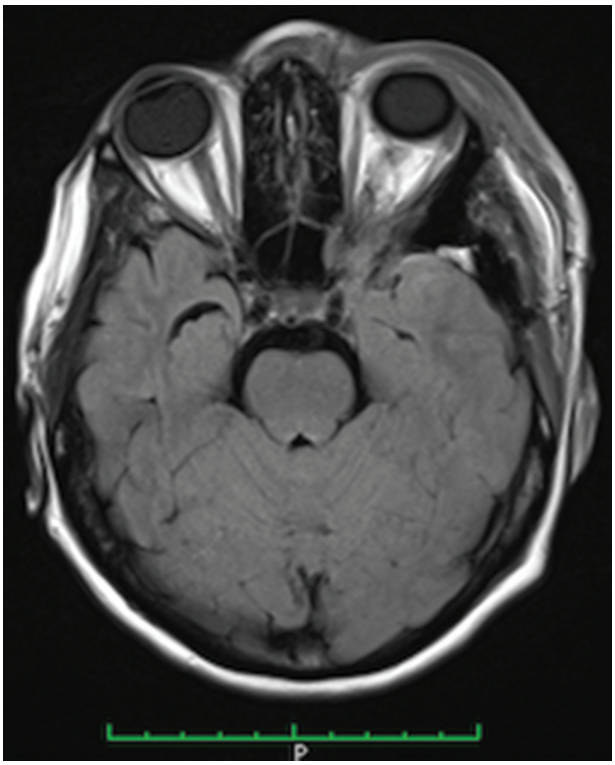

Fig. 2 Magnetic Resonance Imaging: Axial FLAIR - Evidence of satisfactory resection of the lesion.

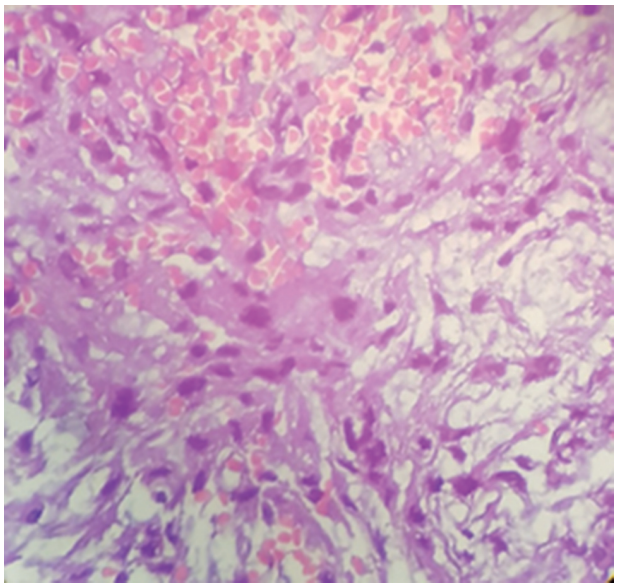

Fig. 3 Histology: Neoplastic cells in cordonal arrangement in the middle of an abundant mucoid matrix. Cells may be vacuolated, increasing the resemblance to the bullous appearance of chordoma cells.

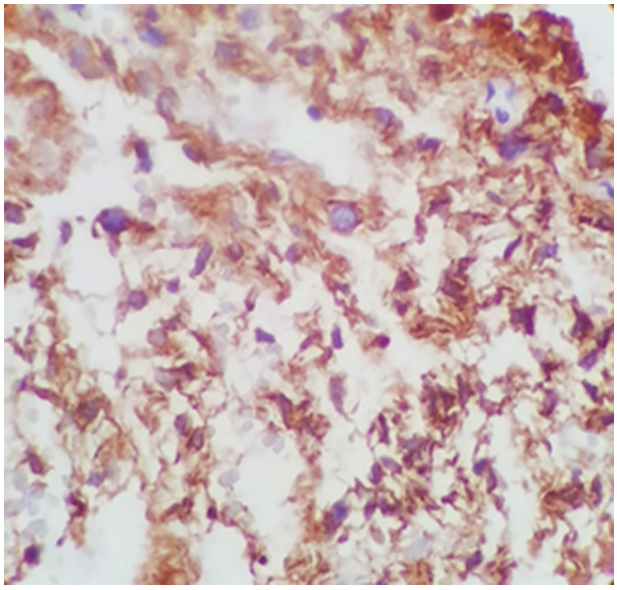

Fig. 4 Immunohistochemistry: Membrane epithelial antigen marks neoplastic cells. The interstitial material appears pale. Here it marks the cytoplasm of the neoplastic cells, with reinforcement at the cellular membrane level in areas.

Results

Meningiomas have preponderance for convexities. For instance, 17 to 25% occur in a frontobasal location. Within the frontobasal region, the olfactory grooves, the tuberculum sellae, the parasellar region and the petrous bone are preferred sites.[5] On the other hand, 5% occur along the cerebellar convexity, 2 to 4% at the tentorium cerebelli, and 2 to 4% within the cerebellopontine angle (CPA).[6]

Chordoid meningioma is an uncommon histopathological variant of meningioma frequently mistaken as chordoma, due to its histological similarity. Couce et al related 42 clinicopathologic studies about CM during the period of 1975 to 1997, and this study concluded that it was more common in the supratentorial region and in adults.[6,7]

There are only two studies about genetic alterations in CMs. The majority of meningiomas is known to have deletion at many chromosomal loci not yet studied in CMs, such as 22q, 18p, 14q and 1p. All of the literature cases showed to have either complete or partial deletion at 22q, 14q and 1p loci.[8,9] Additionally, all cases were variably positive for epithelial membrane antigen (EMA) and showed strong positive staining for vimentin, but were completely negative for glial fibrillary acidic protein (GFAP), in support of the diagnosis.[9] Because of the similarity between CMs and other lesions, especially tumors, careful assessment of each case is required. The main diagnoses are chondroid choroid, chordoid glioma, paraganglioma, metastatic mucinous carcinoma or metastatic renal cell carcinoma, among others.[10]

The goal of the therapy in CM is relieving the compression of critical anatomic structures as well as preventing tumor spread and recurrence.[11] The therapeutic preference for aggressive meningiomas are surgical resection, radiotherapy (RT) and chemotherapy, in cases of anaplastic meningioma.[10,11] Neurosurgery is the treatment of choice for meningiomas and, preferentially, with total resection.[10]

Discussion

Meningiomas are the second most common central nervous system neoplasm in adults and account for 15 to 20% of all primary brain tumors. Meningiomas are benign tumors; however 10% of the cases demonstrate a more aggressive clinical behavior, such as chordoid meningioma, which is a type of non-benign meningioma.[12] First described by Kepes in 1988, chordoid meningiomas are characterized by cords of eosinophilic, epithelial or spindle-shaped cells within a myxoid stroma, resembling chordoma. Variable numbers of lymphocytes and plasma cells characterize these tumors.

Chordoid meningiomas occur very rarely, corresponding to up to 1% of surgically removed meningiomas, even in large cohorts. Chordoid meningiomas are located supratentorially in 81.5% of the cases.[13] Unusual sites, such as the ventricular system, the foramen jugulare and the orbital area have been described in several case reports.[14,15] Histologically, CM consists of cords or trabeculae of eosinophilic and vacuolated cells embedded in an abundant mucoid matrix.[16]

However, sometimes the histological diagnosis is difficult because it is comparable morphology in other intracranial tumors, such as chordoid glioma, chordoma, extraskeletal chondrosarcoma, myxopapillary ependymoma and metastatic tumors.[8]

Immunohistopathological studies have shown that this tumor shows positivity for vimentin, EMA and D2–40, and occasional positivity for S-100 protein, on the other hand demonstrating negativity for GFAP and cytokeratin.[9–19]

The first choice of treatment for CM is surgical excision. The goal of surgery is a complete resection. The complete resection of a tumor is sometimes difficult because of its

location.[8–11] The surgical strategy is to perform total resection of the tumor, dura and bone, while protecting the surrounding structures. In contrast, when an incomplete removal is realized, due to the risk of injury to the surrounding vital neurovascular structures, postoperative adjuvant RT is an acceptable option.[16,19,20] Despite this recommendation, there is controversy surrounding the value of postoperative RT in atypical meningiomas. This treatment is commonly recommended for patients with atypical meningiomas following subtotal removal, but some investigators, acknowledging the greater recurrence risk of atypical meningiomas, recommend early irradiation, irrespective of the resection extent, to optimize local control and improve survival.[5] Once recurrent, grade II meningiomas carry a mortality risk and postoperative RT, after total removal, may prolong or prevent recurrence.[9] Perhaps, more comprehensive series are required to evaluate the role of brain invasion and postoperative RT in the prognosis of CM.[1–21] In our case, we reconvened the patient to continue the treatment and improve the prognosis.

Conclusion

Chordoid meningiomas are rare and different tumors of the meningiomas that present a high degree of recurrence, even when the satisfactory resection of the lesion is achieved. Recurrence is particularly worrisome after a subtotal surgical resection. Their histological similarity to chordoid neoplasms, especially chordoma, can make the diagnosis challenging. These two neoplasms have different biological behaviors, so it is important to be able to distinguish them. The goal of treatment is surgery, with total resection of the tumor. In cases of relapse or in cases of residual tumor, it is necessary to perform RT.

Conflict of Interests

The authors have no conflict of interests to declare.

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Spontaneous Convexity Subarachnoid Hemorrhage Caused by Internal Carotid Occlusion: Radiological Features

Hemorragia subaracnóidea espontânea na convexidade causada por oclusão da carótida interna: Características radiológicas

Benedito Jamilson Araújo Pereira[1,2] Radmila Agra Cariry Targino de Holanda[2] José Targino Neto[2] Luciano Ferreira de Holanda[2,3]

[1] Department of Neurosurgery, Centro Especializado em Neurologia e Neurocirurgia Associados (CENNA), Hospital Beneficência Portuguesa de São Paulo, São Paulo, SP, Brazil
[2] Department of Neurosurgery, Divisão de Neurocirurgia, Hospital Antônio Targino, Campina Grande, Paraíba, PB, Brazil
[3] Department of Medical Sciences, Faculdade de Medicina, Universidade Federal de Campina Grande (UFCG), Paraíba, PB, Brazil

Address for correspondence Benedito Jamilson Araújo Pereira, MD, Rua Martiniano de Carvalho 669, apto. 1.105, Bela Vista, São Paulo, SP, 01321-001, Brazil (e-mail: benedito.jamilson@hotmail.com).

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Abstract

A 79-year-old patient was admitted to the emergency room with transitory monoparesis in the left hand and dysphasia. The brain computed tomography (CT) and magnetic resonance imaging (MRI) showed a spontaneous right convexity subarachnoid hemorrhage (cSAH). Digital subtraction angiography (DSA) confirmed an asymptomatic occlusion of the right internal carotid artery (ICA) . Cases related to stenosis have already been described, but there is no similar report of a case related to occlusion, even though the pathophysiology of both entities is similar. Atraumatic SAH has been associated with intracranial and extracranial artery stenosis.

Keywords
- spinal hemorrhage
- carotid stenosis
- skull tomography

Resumo

Paciente de 79 anos foi admitida na sala de emergência, com monoparesia braquial transitória à esquerda e disfasia. O exame de tomografia cerebral (TC) e ressonância magnética mostraram uma hemorragia subaracnóidea na convexidade no hemisfério direito (cSAH). A angiografia por subtração digital confirmou uma oclusão da artéria carótida interna (ACI) direita assintomática. Casos como esses relacionados à estenose já foram descritos, porém, relacionados à oclusão, não há relato semelhante, embora a fisiopatologia de ambas as entidades seja semelhante. cSAH tem sido associada à estenose de artérias intracraniana e/ou extracraniana.[1]

Palavras-chave
- hemorragia subaracnódea
- estenose de carótida
- tomografia de crânio

Case Report

A 79-year-old patient was admitted to the emergency room with transitory monoparesis in the left hand and dysphasia. The brain computed tomography (CT) (►**Fig. 1A-C**) and magnetic resonance imaging (MRI) (►**Fig. 1D-F**) showed a spontaneous right convexity subarachnoid hemorrhage (cSAH). Digital subtraction angiography (DSA)

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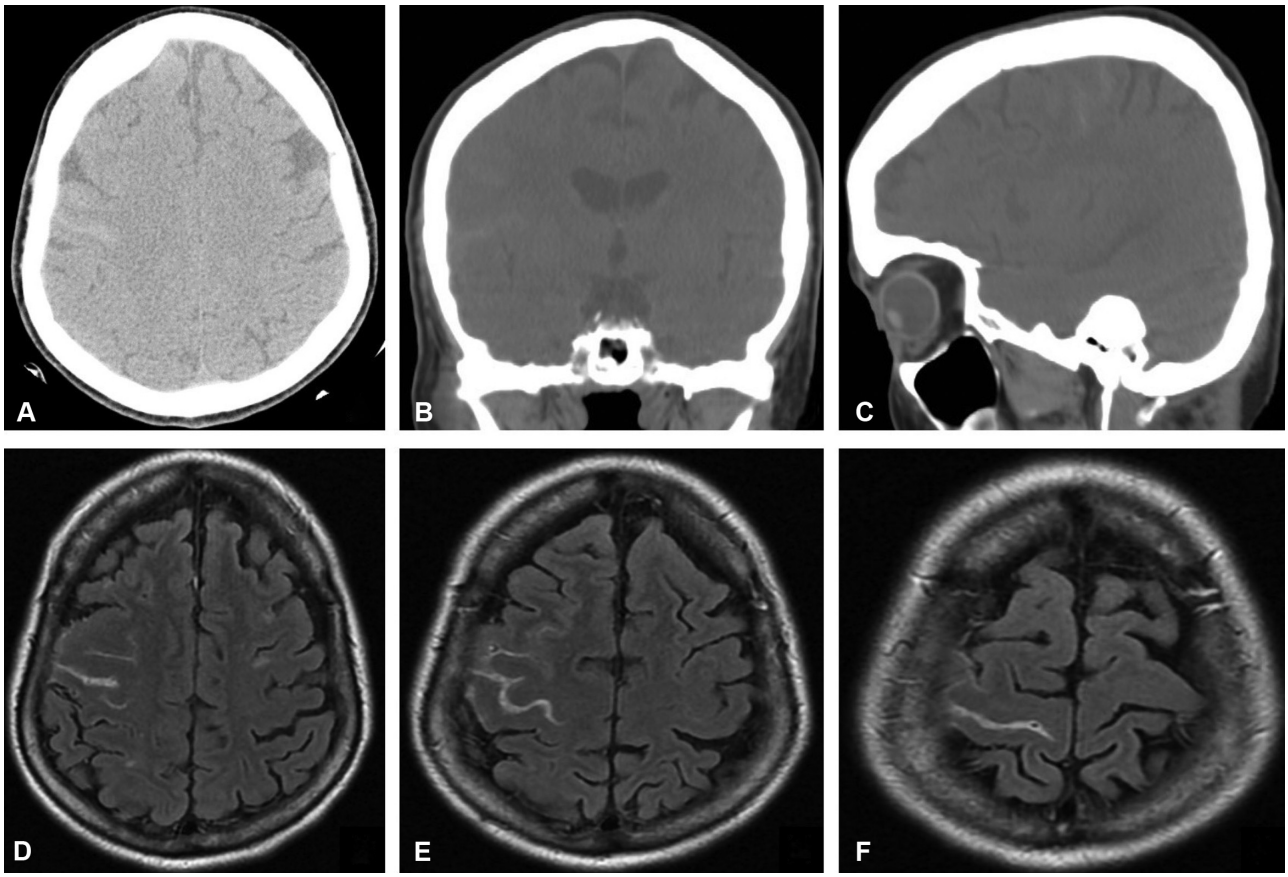

Fig. 1 (**A-C**) Axial, coronal and sagittal cranial computed tomography (CT) showing a hyperdense cortical lesion corresponding with a right convexity subarachnoid hemorrhage; (**D-F**) magnetic resonance imaging (MRI): axial T1-weighted gradient echo sequence image showing subacute right sulcal subarachnoid hemorrhage.

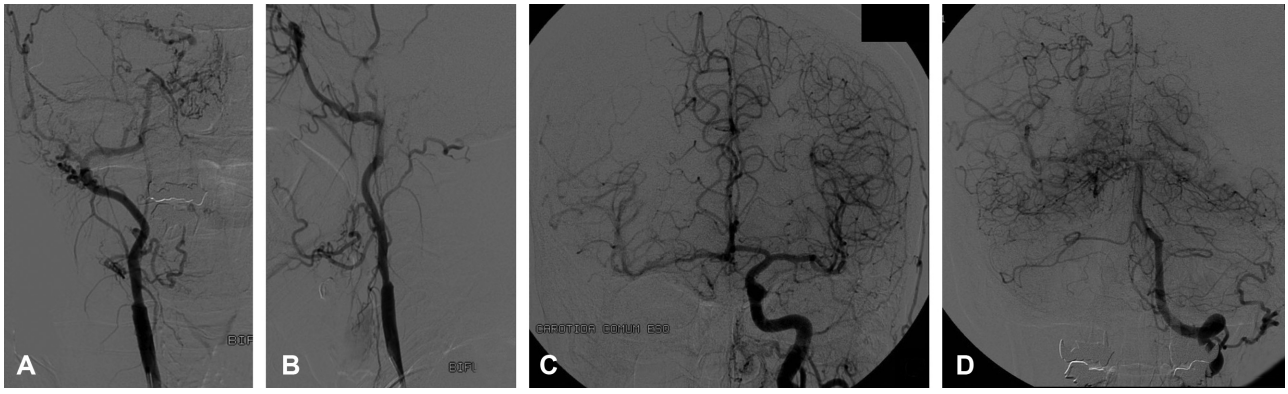

Fig. 2 Digital subtraction angiography (DSA): (**A-B**) antero-posterior and profile DSA of the right common carotid artery (CCA) showing occlusion of the right internal carotid artery (ICA); (**C-D**) vascularization of the right cerebral parenchyma by collateral circulation through the polygon of Willis, via the anterior and posterior communicating arteries respectively.

confirmed an asymptomatic occlusion of the right internal carotid artery (ICA) (►**Fig. 2**). Cases related to stenosis have already been described, but there is no similar report of a case related to occlusion, even though the pathophysiology of both entities is similar. Atraumatic SAH has been associated with intracranial and extracranial artery stenosis.[1]

Conflict of Interests

The authors have no conflict of interests to declare.

Reference

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Bruns Syndrome Caused by Intraventricular Neurocysticercosis: Literature Review

Síndrome de Bruns causada por neurocisticercose intraventricular: Revisão da literatura

Guilherme Gago da Silva[1] Nelson Pires Ferreira[1] João Pedro Einsfeld Britz[2]

[1] Department of Neurosurgery, Hospital São José, Santa Casa de Misericórdia de Porto Alegre, Porto Alegre, RS, Brazil
[2] Universidade de Caxias do Sul, Caxias do Sul, RS, Brazil

Address for correspondence Guilherme Gago da Silva, Departmento de Neurocirurgia, Hospital São José, Rua Sacadura Cabral, 242, apartamento 602, Petrópolis, Porto Alegre 90690-420, Rio Grande do Sul, Brazil (e-mail: Guilherme.gago@gmail.com).

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Abstract

Bruns syndrome is one of the clinical presentations of intraventricular neurocysticercosis, and it is characterized by episodes of headache, vertigo and vomiting. The intraventricular form of neurocysticercosis occurs in 7% to 20% of the cases, and it is more serious than the intraparenchymal form. The management is primarily surgical, associated with pharmacological therapy with anthelmintic drugs and corticosteroids. We report the case of a patient who presented Bruns syndrome due to neurocysticercosis.

Keywords
- neurocysticercosis
- cysticercus
- neurosurgery

Resumo

A síndrome de Bruns é uma das apresentações clínicas da neurocisticercose intraventricular, caracterizada por episódios de cefaleia, vertigem e vômitos. A forma intraventricular da neurocisticercose ocorre entre 7% e 20% dos casos, e é mais grave do que a forma intraparenquimatosa. O tratamento é principalmente cirúrgico, associado a terapia farmacológica com drogas anti-helmínticas e corticosteroides. Relatamos o caso de uma paciente que apresentou síndrome de Bruns por neurocisticercose.

Palavras-chave
- neurocisticercose
- cisticerco
- neurocirurgia

Introduction

Neurocysticercosis is the most common helminth infection of the central nervous system, caused by the larval form of *Taenia solium*. Bruns syndrome, described in 1906 by German neurologist Ludwing Bruns, is one of the presentations of intraventricular neurocysticercosis. It is characterized by a transient increase in intracranial pressure, caused by a floating mass in the ventricular system, producing hydrocephalus and leading to episodes of headache, vertigo and vomiting, which are triggered by sudden head movements.[1,2] The authors report the case of a patient who presented Bruns syndrome due to neurocysticercosis. A review of the published cases is discussed in the present article.

Case Presentation

The patient was a 46-year-old female, from the countryside of the state of Rio Grande do Sul, Brazil, with a history of untreated hydrocephalus, cognitive deficit and bacterial meningitis treated in the previous two years. Over the course of two weeks, she presented recurrent episodes of syncope, paresis in the four limbs, and loss of sphincter control, which improved between crises, and was associated with progressive visual loss, without improvement since the onset of the condition. Four days after the of onset of symptoms, she had an episode of seizure and vomiting. With the use of phenobarbital and phenytoin, she managed to control the seizures.

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The patient started an investigation with a neurologist at Santa Casa de Misericórdia de Porto Alegre, who requested a magnetic resonance imaging (MRI) scan. She sought the emergency department of the same institution after experiencing worsening of the paresis in the limbs and difficulty in walking. In the MRI scan, two cystic lobulated and septate lesions within the fourth ventricle were identified, causing ventricular expansion, with obstruction of the cerebral aqueduct, associated with an important dilation of the supratentorial ventricular system. Cystic lesions were also identified in the topography of the cisterna magna (►**Figs. 1** and **2**).

Upon physical examination, the patient was confused. Ocular opening was spontaneous, and the eyes obeyed bilateral commands.

A hypothesis of intraventricular racemic neurocysticercosis as the cause of Bruns syndrome was considered. After discussion with the neurosurgery team, the patient was admitted to Hospital São José and referred for surgical removal of the cysts. In the immediate postoperative period, the patient persisted with reduced level of consciousness and random eye movements. The postoperative tomography showed a diffuse reduction in the amplitude of the cortical sulci and gyrus, associated to loss of cortical-subcortical differentiation and reduction in the size of the basal cistern, an aspect related to diffuse cerebral edema. The ventricular system was effaced, and pneumoventricle in the frontal horns of the lateral ventricles was observed (►**Fig. 3**), which required the placement of and external ventricular shunt, and, subsequently, a ventricular-peritoneal shunt (VPS). After a positive Weinberg test, drug therapy with albendazole and dexamethasone was initiated. The patient developed into a persistent vegetative state and died 46 days later due to respiratory failure.

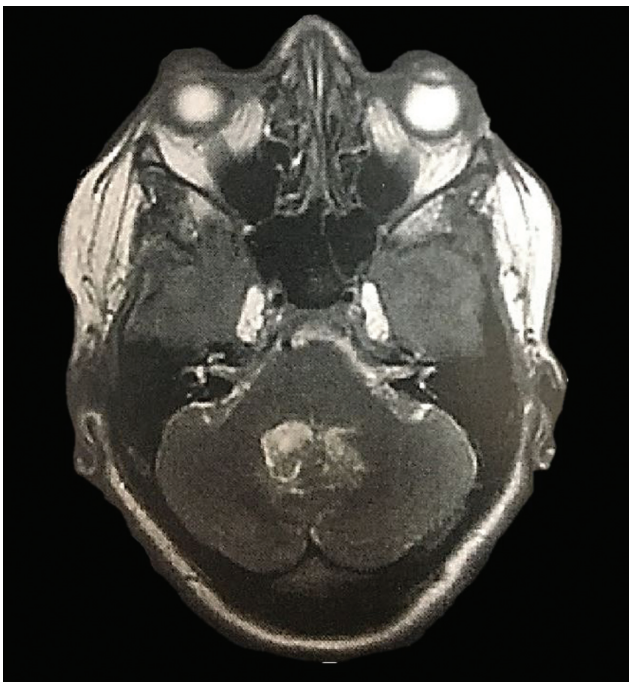

Fig. 1 Magnetic resonance imaging scan showing the presence of two cystic lesions within the fourth ventricle.

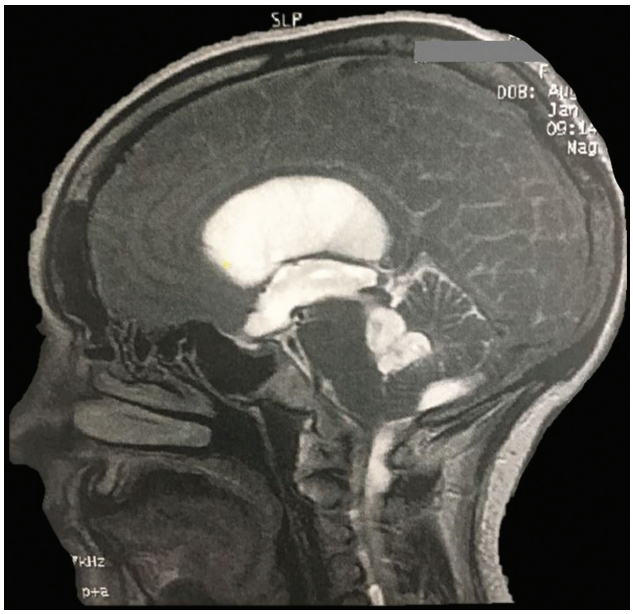

Fig. 2 Obstruction of the cerebral aqueduct and dilation of the supratentorial ventricular system.

Discussion

The clinical presentation of neurocysticercosis may vary depending on the host's immune status, the size of the cyst, and its location in the central nervous system. The

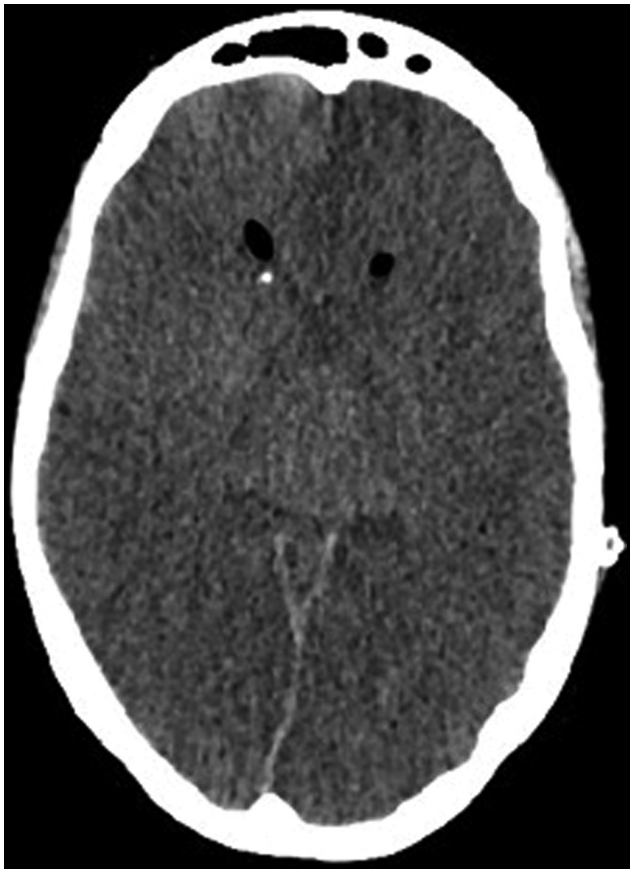

Fig. 3 Postoperative tomography.

most commom symptoms are epilepsy, headache, and focal neurological deficit, and the intraventricular form of neurocysticercosis only occurs in 7% to 20% of the cases, with the third and fourth ventricle being the most affected sites.[1,3]

The main causes of Bruns syndrome are mobile intraventricular lesions such as tumors, neurocysticercosis and colloid cyst of the third ventricle, with neurocysticercosis being the most common cause.[4] The syndrome consists of episodes of intense headache, vomiting and vertigo triggered by sudden movements of the head that produce transient hydrocephalus by displacement of the mobile ventricular mass. Some cases may present with rapid neurological worsening and death due to intracranial hypertension.[5] The diagnosis is difficult because the clinical manifestations are not very specific. The MRI is superior to tomography for the diagnosis of neurocysticercosis.[6]

When the ventricular form is accompanied by meningitis and hydrocephalus, mortality is greater than 50%, and most patients die within 2 years, even with the VPS. The prognosis of the intraventricular form is worse than that of the intraparenchymal form, and it follows a progressive course, due to obstructive hydrocephalus, intracranial hypertension and meningeal infection.[6] The clinical presentation of our case was less common because the patient only had visual loss on the neurological examination, despite signs of hydrocephalus and occlusion of the fourth ventricle in the MRI.

The management of intraventricular neurocysticercosis is different from that of the intraparenchymal form. Surgery is the primary therapeutic measure for the removal of the cysts. For cysts of the third and lateral ventricles, the endoscopic approach is the most recommended. In cases of involvement of the fourth ventricle, removal of the cysts may be more challenging, requiring open surgery. Ventricular shunts are commonly required for the management of obstructive hydrocephalus. Our patient underwent a similar management to that reported in the literature, with surgical removal of the cysts and associated drug therapy. The telovelar approach was chosen for the removal of the cysts

Table 1 Review of published cases of intraventricular neurocysticercosis

Author	Age/ Gender	Location	Clinical presentation	Management	Outcome
Aguilar-Amat et al[1]	29y/F	Intraventricular**	Headache, nausea, diplopia and gait instability	Albendazole, corticoids and surgery*	No deficits
Rodriquez et al[6]	43y/F	Fourth ventricle	Headache, vomiting and vertigo	Open surgery	–
Jensen and Post[7]	50y/F	Foramen of Monro	Headache, nausea, irritability and confusion	Endoscopic surgery	No deficits
Jensen and Post[7]	30y/F	Foramen of Monro	Headache, fever and confusion	Endoscopic surgery	–
Roongpiboonsopit et al[4]	69y/M	Fourth ventricle	Headache, vomiting and confusion	Surgery*	No deficits
Torres-Corzo et al[3]	22y/M	Third venticle	Headache and vomiting	Endoscopic surgery	No deficits
Torres-Corzo et al[3]	31y/F	Third venticle	Headache and papilledema	Endoscopic surgery	No deficits
Torres-Corzo et al[3]	17y/F	Third venticle	Headache and visual alterations	Endoscopic surgery	No deficits
Torres-Corzo et al[3]	28y/M	Third venticle	Headache and vomiting	Endoscopic surgery	No deficits
Torres-Corzo et al[3]	43y/M	Third venticle	Lethargy	Endoscopic surgery	No deficits
Torres-Corzo et al[3]	54y/M	Third venticle	Headache, vomiting and diplopia	Endoscopic surgery	No deficits
Torres-Corzo et al[3]	35y/F	Third venticle	Headache and Parinaud sign	Endoscopic surgery	No deficits
Shahani et al[2]	40y/F	Fourth ventricle	Headache, nausea and vomiting	Albendazole, dexamethasone, surgery* and VPS	No deficits
Shahani et al[2]	39y/F	Fourth ventricle	Headache, dizziness, nausea, vomiting and tinnitus	Albendazole, dexamethasone, surgery* and VPS	No deficits
Das et al[8]	24y/F	Fourth ventricle	Headache, vertigo and vomiting	Surgery*	No deficits
Dhiman et al[9]	11y/M	Fourth ventricle	Headache, vomiting and strabismus	Endoscopic surgery	Maintained strabismus

Abbreviations: F, female; M, male; VPS, ventriculoperitoneal shunt; Y, years old.
Notes: *Open or endoscopic surgery not specified. **No report of which ventricle was affected.

from the fourth ventricle. The flexible endoscopy approach is an option for patients with neurocysticercosis in the third ventricle.[3] The treatment with anthelmintic drugs may lead to worsening of the symptoms, and it should be administered concomitantly with glucocorticoids. The antihelminthic drug of choice is albendazole, which is superior to praziquantel because it does not interact pharmacologically with glucocorticoids and antiepileptic drugs.[2,7]

In a review of the PubMed database with the MeSH terms *neurocysticercosis* AND *Bruns syndrome*, eight publications of Bruns syndrome caused by neurocysticercosis were found, totalizing 16 cases. Headache was the main symptom reported (15 cases), followed by vomiting (9), confusion (3) and vertigo (3). Visual and ocular motility disorders occurred in six cases. Gait instability, tinnitus and fever were reported in one case each. The age of the patients ranged from 11 to 69 years, and surgical intervention was the choice in all cases. In the present case, syncope, paresis and visual loss were the initial symptoms. Signs of intracranial hypertension, such as vomiting, appeared a few days later. The review of the published cases is summarized in ►**Table 1**. The involvement of the third and fourth ventricles was similar, with 7 and 6 cases respectively, and in 2 cases the cysticercus was located in the lateral ventricles, in the foramen of Monro. Endoscopic surgery was the choice in most cases. The patient maintained the neurological deficit only in one case.

Conclusion

Bruns syndrome is a rare presentation of neurocysticercosis. It has a nonspecific symptomatology that can go unnoticed by the physician, becoming a serious condition and leading to death due to obstruction of the flow of the cerebrospinal fluid.

Conflict of Interests

The authors have no conflict of interests to declare.

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Percutaneous Treatment of Meckel Cave Arachnoid Cyst: Case Report, Surgical Strategy and Literature Review

Tratamento percutâneo de cisto aracnóide do cavum de Meckel: Relato de caso, estratégia cirúrgica e revisão da literatura

Jose Augusto Malheiros Filho[1] Antonio Gilson Prates Junior[1] Emmanuel Oliveira Vasconcelos e Sá[1] Luiza Cançado Guerra D'Assumpção[1] Lucas Rodrigues de Souza[1]

[1] Neurosurgery Service, Hospital das Clinicas, Universidade Federal de Minas Gerais, Belo Horizonte, MG, Brazil

Address for correspondence Antonio Gilson Prates Junior, MD, Hospital das Clinicas da Universidade Federal de Minas Gerais, Belo Horizonte, MG, 31270-901, Brazil (e-mail: antonio.ufop@gmail.com).

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Abstract

Arachnoid cysts are benign intracranial lesions. They are usually located in the middle fossa, but can be found in other locations. We present a case of symptomatic Meckel cave (MC) arachnoid cyst - a very rare location - and a treatment strategy not elsewhere described before for this condition. A 54-year-old female with trigeminal neuralgia with previous history of radiofrequency rhizotomy treatment 6 years before admission had been experiencing pain recurrence with progression, which required successive increases in carbamazepine dosage. Magnetic Resonance Imaging (MRI) showed dilatation of the right MC with extension to the petrous apex. The lesion was compatible with arachnoid cyst, and due to the worsening of the clinical condition, surgical treatment was chosen. Percutaneous puncture of the cyst through the foramen ovale with injection of intracystic fibrin sealant was performed. The patient woke up from anesthesia with pain improvement and was discharged asymptomatic the next day. After 12 months of follow-up, she remained pain-free. In the literature review, we found only eight cases reported as MC arachnoid cyst. These are likely to progress and become symptomatic owing to their communication with the subarachnoid space and a unidirectional valve mechanism. Pain improvement with this technique is probably secondary to the interruption of these mechanisms.

Keywords

- Meckel cave
- arachnoid cyst
- petrous apex cephaloceles
- trigeminal neuralgia
- percutaneous treatment

Resumo

Os cistos aracnoides são lesões intracranianas benignas. Geralmente estão localizados na fossa média, mas podem ser encontrados em outros locais. Apresentamos um caso de cisto aracnoide sintomático no cavum de Meckel (CM) – localização muito rara – e uma estratégia de tratamento nunca antes descrita para esta condição. Uma mulher de 54 anos com neuralgia do trigêmeo e histórico de tratamento com rizotomia por

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- cavum de Meckel
- cisto aracnoide
- cefalocele do ápice petroso
- neuralgia do trigêmeo
- tratamento percutâneo

radiofrequência 6 anos antes da admissão apresentava recorrência da dor com progressão, exigindo aumentos sucessivos na dosagem de carbamazepina. Ressonância magnética mostrou dilatação do CM à direita com extensão ao ápice petroso. A lesão era compatível com cisto aracnoide e, devido ao agravamento do quadro clínico, optou-se por tratamento cirúrgico. Foi realizada punção percutânea do cisto através do forame oval com injeção de selante de fibrina intracístico. A paciente acordou da anestesia com melhora da dor e recebeu alta assintomática no dia seguinte. Após 12 meses de acompanhamento, a paciente permaneceu sem dor. Na revisão da literatura, encontramos apenas oito casos relatados como cisto aracnoide do CM. É provável que estes progridam e se tornem sintomáticos devido à sua comunicação com o espaço subaracnóideo e a um mecanismo de válvula unidirecional. A melhora da dor com essa técnica é provavelmente secundária à interrupção desse mecanismo.

Introduction

Arachnoid cysts are benign intracranial lesions that account for 1% of nontraumatic intracranial lesions.[1,2] They are usually located in the middle fossa, but can be found in other locations - such as the suprasellar, the posterior fossa, the interhemispheric, the quadrigeminal cistern and the cerebral convexity. They are usually asymptomatic lesions, but may become symptomatic depending on location and size.[3] There are several treatment options, including endoscopic or microsurgical fenestration, microsurgical excision and cyst shunt. We present a case of symptomatic arachnoid cyst in a very rare location that was treated by a strategy not described elsewhere.

Case Report and Surgical Strategy

A 54-year-old female was admitted in our service with trigeminal neuralgia in the territories of the ophthalmic and maxillary trigeminal branches. She had a history of radiofrequency rhizotomy treatment 6 years before, and at that time, the patient presented improvement and good pain maintenance control with carbamazepine 400 mg a day. Ten months before seeking assistance, the patient recurred and experienced pain progression, which required successive increases in carbamazepine dosage up to 900 mg a day – dose to which she presented adverse reactions. Due to recurrence and worsening of the clinical condition, propaedeutic were requested. The magnetic resonance imaging (MRI) showed dilatation of the right Meckel cave (MC) with extension to the petrous apex. The lesion presented as hyperintense on T2-weighted imaging, hypointense on T1-weighted imaging (►**Figure 1**); suppression of the signal in the T2-weighted fluid-attenuated inversion recovery and absence of diffusion restriction on diffusion weighted imaging – description compatible with arachnoid cyst. There were no neurovascular conflicts or other lesions associated with the trigeminal nerve topography. Owing to daily episodes of excruciating pain and intolerance to drug treatment, surgical treatment was chosen.

Percutaneous puncture of the cyst through the foramen ovale with injection of intracystic fibrin sealant was performed. The patient did not tolerate the sedation, so the procedure took place under general anesthesia. A 20-Gauge spinal needle was inserted through the ascending extraoral transoval route guided by lateral incidence fluoroscopy.[4] The introduction pathway started 3 cm lateral to the labial commissure and ascended until the intersection point between the coronal plane 3 cm anterior to the tragus and the sagittal plane of the pupil. After touching the skull base, the needle was directed under fluoroscopy to the foramen ovale, using as reference point the clivus intersection with the petrous pyramid. After entrance, the position of the needle was confirmed by fluoroscopy (►**Figure 2A**) and through

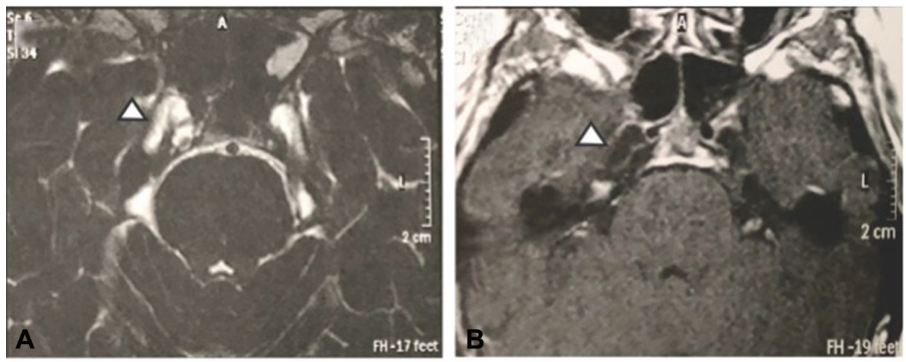

Fig. 1 Meckel cave (MC) arachnoid cyst on magnetic resonance imaging (MRI). (**A**) T2-weighted MRI image showing right MC dilatation with cerebrospinal sign (arrow). (**B**) T1- weighted MRI image showing right MC dilatation (arrow).

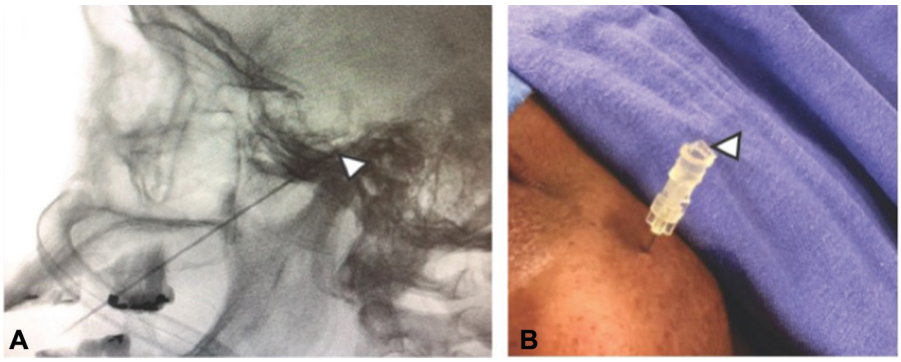

Fig. 2 Surgical technique. (**A**) Fluoroscopy-guided foramen ovale puncture. Note the tip of the needle in the intracranial compartment (arrow). (**B**) Puncture point 3 cm lateral to lip rhyme. Note the leakage of cerebrospinal fluid through the needle (arrow).

cerebrospinal fluid (CSF) leakage through its lumen (►**Figure 2B**). On account of active CSF drainage after the puncture, we chose not to inject contrast. Then 2 mL of Tissucol was injected and the needle was removed. The patient woke up from anesthesia with pain improvement and was discharged asymptomatic the next day. The MRI performed on the first postoperative day revealed reduction of the cyst dimensions and partial filling of the cyst by T2-weighted hypointense material extending through the cisternal segment of the trigeminal nerve (►**Figure 3**). After 12 months of follow-up, tapering of carbamazepine dose was achieved and the patient remained pain-free using only 400 mg a day.

Discussion

Arachnoid cysts are collections of intracranial CSF surrounded by arachnoid layers. They can be primary or secondary. The primaries are benign malformations originated from arachnoid division during development resulting in anomalous CSF collections. The secondary ones are less common and are associated with neoplasms, infections, bleeding, trauma or surgery.[2,3] Depending on their location and size, arachnoid cysts may become symptomatic.

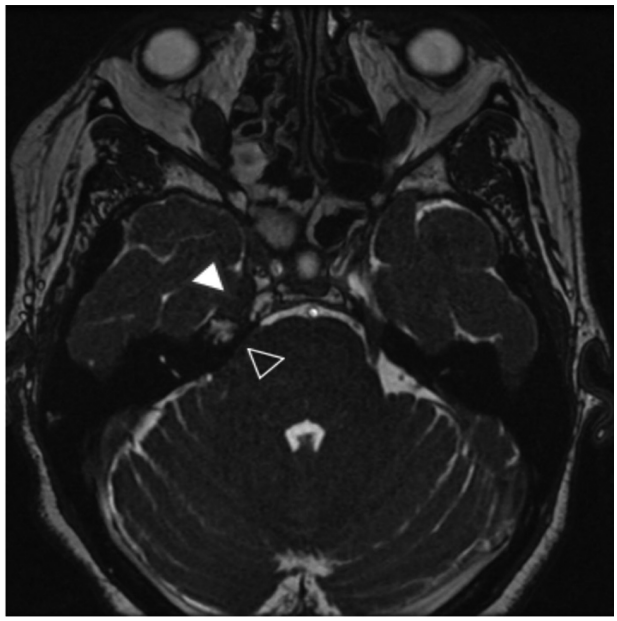

Fig. 3 Postoperative T2 -weighted magnetic resonance imaging showing partial filling of the cyst by with hypointense material (white arrow) extending through the cisternal segment of the trigeminal nerve (black arrow).

In a literature review, we found only eight cases reported as MC arachnoid cyst (►**Table 1**).[2,5–11] The average age of the patients was 41 years old – with a range from 1 to 58 years old – and there was a female predominance (5 out of 8). The most affected side was the right side (five out of eight). In all but one case, there was trigeminal neuralgia in 1 or more branches territories, associated or not with symptoms of other cranial pairs, such as diplopia, vertigo, and hearing loss. The patient in whom no neuralgia was reported was a 1.5-year-old child with exophthalmos and diplopia.[10] In only three cases the lesion was referred to as restricted to the MC, and in one of these, no MRI was performed and the lesion was not described in details in the article.[10] We can conclude that MC cysts tend to extend to adjacent regions, especially to the petrous apex - as occurred in the case reported in the present paper.

Of the reported cases, two were treated with drugs alone; five were treated surgically and one had spontaneous resolution. Of the five surgically treated patients, four underwent craniotomy treatment and only one underwent percutaneous treatment with cyst aspiration through the foramen ovale puncture.[2] In this patient, the control MRI presented cyst persistence and the author attributed the improvement to possible rhizotomy by injected contrast or needle injury.[2] Moreover, these data show the importance of preoperative differential diagnosis by MRI of lesions in the MC because in all patients undergoing craniotomy, the diagnosis of arachnoid cyst was made intraoperatively, excluding the possibility of less invasive treatment.[5,6,10,11]

There are reports of petrous apex cephaloceles. These are uncommon lesions, rarely described in the literature and sometimes radiologically indistinguishable from arachnoid cysts of the MC. They are considered by some authors to be the same entity[2,8]. Defined as cystic lesions with a cerebrospinal fluid-like sign at the petrous apex that protrude through the posterolateral wall of the MC, they can be

Table 1 Cases Reported in the Literature

Author	Age (years old), gender	Symptoms	Location, side	Treatment
Wörner et al[5]	44, M	Dysesthesia in V3, vertigo and diplopia	Meckel cave, CPA, R	Open resection
Batra et al[6]	55, F	Pain and paresthesia in V3	Petrous apex, Meckel's cave, R	Open fenestration
Bigder et al[2]	57, F	Pain in V2, temporal muscle atrophy and hemifacial paresthesia	Meckel cave, L	Percutaneous drainage
Fois et al[7]	42, F	Sensorineural hearing loss and neuralgia	Meckel cave, petrous apex, B	Medication
Jacob et al[8]	32, F	Diplopia and pain in V1	Meckel cave, R	Spontaneous resolution
Grasso et al[9]	40, M	Neuralgia in V2-V3	Meckel cave, middle fossa and greater wing of sphenoid, L	Medication
Beck et al[10]	1, M	Exophthalmia and diplopia	Meckel cave, R	Open resection
Jelsma et al[11]	58, F	Neuralgia and hemifacial hypoesthesia	Meckel cave, petrous apex, R	Open fenestration

Abbreviations: B, bilateral; CPA, cerebellopontine angle; F, female; L, left; M, male; R, right; V1, ophthalmic trigeminal branch; V2, maxillary trigeminal branch; V3, mandibular trigeminal branch.

composed of dura mater and arachnoid or just arachnoid – being described as meningoceles and arachnoid cysts.[12–15] However, dilatation may be restricted to the petrous apex without dilatation of the MC and they are usually not associated with trigeminal neuralgia, but with sixth cranial nerve neuropathy and otological symptoms.[8,12] Possibly, they are part of the same spectrum of disease of the arachnoid cyst in the MC differing only in presentation and progression.

Meckel cave arachnoid cysts are likely to progress and become symptomatic due to their communication with the subarachnoid space and a unidirectional valve mechanism that only allows CSF entry. This communication between the cyst and the CSF system may allow the transmission of pulsations to the nerve and cyst enlargement causing trigeminal neuralgia by a mechanism similar to neurovascular compression.[9]

We describe a noninvasive, effective and safe technique for the treatment of Meckel cave arachnoid cysts. We did not find in the literature a description of the same strategy. The improvement mechanism with this technique is probably the interruption of the communication between the cyst and the cerebrospinal fluid system, which prevents the transmission of pulsations to the nerve and enlargement of the cyst.

Conclusion

Meckel cave arachnoid cysts are rare lesions in which preoperative differential diagnosis is very important for proper surgical planning and for individualized treatment with less morbidity. Filling the cyst with fibrin sealant by percutaneous puncture was effective for symptomatic control of the patient with minimal associated morbidity. We consider that this technique should be considered for surgical treatment of this lesion.

Conflict of Interests

The authors have no conflict of interests to declare.

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History and Evolution of Epilepsy Surgery

História e evolução da cirurgia para epilepsia

Gustavo Passos[1]

[1] Department of Neurosurgery, Hospital de Clínicas da Universidade Federal do Paraná, Curitiba, PR, Brazil

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Address for correspondence Gustavo Passos, MD, Departmento de Neurocirurgia, Hospital de Clínicas da Universidade Federal do Paraná, Travessa Lange, 277 - Apartamento 1301, 80240-170, Curitiba, PR, Brazil (e-mail: dr.gustavopassos@gmail.com).

Abstract

The surgical treatment for epilepsy has a worldwide historical relevance for centuries. There are archaeological reports that date it back to ancient Egypt; however, the year 1886 is considered a landmark in the surgical treatment for epilepsy in the Modern Age, when the first surgery for the treatment of focal epileptic events was performed successfully. Since then, innumerable related articles have been published evoluonarily. Over the last centuries, new techniques and technologies provided better understanding, diagnosis and management for this disease. Thus, historical and evolutionary knowledge becomes important to let us better understand the current position of the surgery for epilepsy treatment and control.

Keywords
- history
- epilepsy
- epilepsy surgery
- temporal lobe
- depth electrodes

Resumo

A cirurgia para tratamento da epilepsia tem relevância histórica mundial há séculos. Há relatos arqueológicos que remontam ao Egito antigo; porém, é considerado marco no tratamento cirúrgico da epilepsia na Idade Moderna o ano de 1886, quando foi realizada com sucesso a primeira cirurgia para tratamento de crises epilépticas focais. Desde então, inúmeros relatos e artigos relacionados foram publicados. Evolutivamente, nos últimos séculos, novas técnicas e tecnologias nos propiciaram um melhor entendimento, diagnóstico e manejo desta enfermidade. Assim, torna-se importante o conhecimento histórico e evolutivo para que possamos compreender melhor a atual posição da cirurgia para tratamento e controle das epilepsias.

Palavras-chave
- história
- epilepsia
- cirurgia para epilepsia
- lobo temporal
- eletrodos profundos

Introduction

Epilepsy was not always being recognized as a pathology of neurological origin. In the past, it was believed to be related to demonic spiritual possession; Thus, it is understood that trepanation was not always performed for medical purposes, because the religious or spiritual character initially prevailed. However, the goal was always the same, the attempt to put an end to epileptic seizures.

Methodology

The PubMed database was used as the primary source of research with the keywords *epilepsy surgery history* and *epilepsy surgery evolution and advance*. In addition, some books were consulted, including: *Textbook of Epilepsy Surgery*, edited by H. Lüders,[13] and *Operative Techniques in Epilepsy*, edited by J. Girvin.

Brief History of Epilepsy and its Surgical Treatment

Trepanation (from the Greek *trupanon*, which means "perforating") is the removal of part of the cranial calvarium without causing lesions to the vessels, meninges, or the brain tissue itself under it. Cranial trepanation is described in manuscripts that date to before 1500 bC, and skulls from the Neolithic period (~ 10000 to 4000 bC) were found with

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signs of trepanation. The earliest report of a successful neurosurgical procedure was obtained through a carbon-dated skull from 5100 bC, which was found in a French archaeological site. Evidence of bone healing at the edges of the wound shows that the skull is from the first patient who survived the trepanation procedure.[1,2] Although it is not possible to know for certain. Why trepanation may have been performed to relieve intracranial pressure? Elevate fractures with cranial sinking? To remove bone fragments caused by penetrating trauma. It is known as a procedure that has been performed with different purposes since prehistoric times.[3,4] Historically, it was performed in the Middle and Far East, among Celtic tribes, among the Mayas, Incas, Aztecs and Brazilian Indians; it was very common in the Middle Ages and Equatorial Africa, where it is still performed today.[5–8]

The first description of epilepsy as a disease can be found in the *Corpus Hippocraticum*, a collection composed of 60 treaties dating to 400 bC, in Ancient Greece. Then, epilepsy was a controversial disease, often associated with possessions, genius and divinity, being even called the "sacred disease." It is in this book that Hippocrates, for the first time, talks about the hypothesis that epilepsy is located in the brain, and is not a disease of sacred origin. It would be caused by an excess of phlegm in the brain, which, when in contact with the blood, would cause epileptic seizures.[9] But the word *epilepsy* would be used for the first time by Avicenna (980–1037), a Persian polymath who wrote countless treaties on different subjects, 40 of them focused on medicine. The word derives from the Greek verb *epilambanein*, which means to be taken, attacked or dominated.

The age of epileptology, *stricto sensu*, began in 1861, when John Hughlings Jackson (1835–1911), the founder of "modern epileptology," correlated convulsive crises with a cerebral dysfunction. In the following years, new hypotheses were made, until in 1870 its concept changed: the crises would originate in the cerebral cortex.[10,11]

Studying unilateral motor crises, Jackson could conclude that the motor cortex, and not isolated muscles, as it was thought before, was responsible for limb movements. This idea was confirmed by research with cortical stimulation in animals performed by neurologist David Ferrier (1843–1928).

One of the first topographic maps of the somatotropic organization of the motor cortex was idealized and published by Fedor Krause in 1911. Wilder Penfield (1891–1976) perfected the technique, resulting in the famous homunculus of Penfield, a map with a cortical representation of motor and sensory functions. A more modern example of cortical topographic representations of specific functions is the work by Ojemann, which shows the cortical variant of human speech.

The transition from cranial surgery to brain surgery occurred between the second half of the 18th century and the first half of the 19th century, when the idea that epilepsy could be caused by a disorder originating in the cerebral cortex was established.[12] The transition from the pre-modern era (cranial surgery) to the modern era (brain surgery) occurred with the growing critical knowledge about trepanation, in association with the emerging theory of the somatotropic location of the brain areas, and the not less important evolution of asepsis and pain control during surgical procedures.

Epilepsy surgery was initially based on the visual identification of cortical lesions, usually of traumatic origin, and trepanations or trephine were used for the surgical access.

1886 marks the beginning of the contemporary age of epilepsy surgery, when the first surgeries were performed, aiming at the treatment of epileptic seizures in three patients. Sir Victor Horsley (1857–1916) was the neurosurgeon responsible for the surgeries performed in London. Victor Horsley and William MacEwen (1848–1924), from Glasgow, who at the same time also initiated the surgical treatment of patients with epilepsy, are considered the founders of British neurosurgery. In the same period, neurosurgeon Fedor Krause initiated the surgery for epilepsy in Germany. In the aforementioned procedures, there was collaboration between neurologists and neurosurgeons – in the United Kingdom, with Hughlings Jackson, and in Germany, with Hermann Oppenheim (1858–1919) –, which became indispensable for the success of the proposed surgeries.[13,14] Such events served as the basis for the formation of multidisciplinary groups for the treatment of epilepsy; however, in the second half of the 20th century, most of the centers specialized in epilepsy treatment introduced the joint model, with the collaboration of neurologists, neurosurgeons, neuroradiologists and neuropsychologists.

The scientific evolution and the increasingly concrete idea of achieving success in the control of epileptic seizures through surgery spread quickly throughout the world in the end of 19th century and early 20th century. With this, many countries all over the world began to devote resources to the development and research in this area. It is important to remember that, during this period, the surgeries performed for epilepsy control were mostly directed to epilepsy due to trauma or tumors. Until the 1920s, the surgeries were only performed for chronic and localized epilepsy, and the knowledge at the time allowed the physicians to program the region to be operated based only on the semiology and detailed clinical observation of the patients seizure. During surgery, the physicians needed to identify visually the cortical anatomical alterations to be resected, and they often used cerebral electrical stimulation to detect the motor area and the central sulcus, because electroencephalography (EEG) and electrocorticography (ECoG) were not yet available.

With the discovery of X-rays, in 1895, by German physicist Wilhelm Conrad Röntgen (1845–1923), pneumoencephalography, in 1919, by Walter Dandy (1886–1946), and cerebral arteriography, in 1927, by Portuguese neurologist and researcher Egas Moniz (1874–1955), there was a major breakthrough on epilepsy surgery, which enabled physicians to recognize the etiology and topography of the causes preoperatively using images. But it was only in 1929 that German neuropsychiatrist Berger (1873–1941) described the invention and application of the human electroencephalogram, which enabled the recognition of patterns associated with epilepsy and the location of areas with epileptogenic cerebral tissue. Thus, the use of electrophysiological knowledge in epilepsy surgery began, providing elements to aid in the

diagnosis and to determine the cerebral area to be resected prior to the surgical procedure.[15]

In 1934, the Montreal Neurological Institute, in Canada, was founded. Conceived by Wilder Penfield in conjunction with the McGill University, it was a hybrid and unique project for the time, with a hospital area for patients with neurological disorders, a research center totally focused on the nervous system, and a department of neurology and neurosurgery at the university. Penfield, who had great interest in epilepsy treatment, conducted his studies after getting excited about the data collected by pioneer Victor Horsley, and obtained neurosurgical training with Harvey Williams Cushing (1869–1939), who is recognized as the father of modern neurosurgery, and who had already performed surgeries under local anesthesia in patients with epilepsy. Cushing was the first to report the mapping of the sensory cortex with the aid of cortical electrostimulation. However, Cushing was mainly devoted to the study and treatment of brain tumors.[16,17]

In 1937, Canadian neurologist Herbert Jasper (1906–1999) joined the Montreal Neurological Institute. With his previous experience in EEG and with the electrographic foundations of the epilepsies already established, Jasper developed ECoG, which made it possible to map the location of the epileptic seizures during surgery, recording directly from the cerebral cortex and monitoring the responses of stimulation, thus delimiting the area to be resected with greater precision. In that same year, with the aid of intraoperative cortical stimulation and its motor findings, Penfield and Boldrey published for the first time the illustration of what would be perfected and then recognized, 11 years later, as the Homunculus (by Penfield and Rasmussen).

In followinf years, there was great development and many researches in the field of epilepsy, and the surgical treatment evolved along with that. The evolutionary progress and knowledge of the anatomopathology and the electroencephalographic parameters enabled the performance of surgeries totally based on the interictal EEG, according to the publication by Bailey and Gibbs,[19] in 1951, reporting the work with a series of patients with temporal lobe epilepsy undergoing surgical resection.[18,19] Concomitantly, the publications of Penfield, Jasper and other collaborators of the Montreal Institute presented their series of patients with temporal lobe epilepsy undergoing surgical ablation, their electroencephalographic alterations, and their results regarding crisis control.[20–22]

Thus, in the 1950s, there was a major worldwide spread of the surgery for epilepsy, mainly of procedures aimed at the resection of the temporal lobe. Surgeries unrelated to resections of lesions determined visually were initiated, but they were guided by clinical findings and preoperative electrophysiological exams.[23,24]

In 1953, Murray Falconer (1910–1977) published in London the technical standards for anterior resection of the temporal lobe, *en bloc*, which contributed to the research and development of the pathological bases involved in temporal lobe epilepsy. The results showed that a large number of patients with temporal lobe epilepsy had hippocampal sclerosis, which boosted the research and investigation of the pathological hippocampus and the relationship between hippocampal sclerosis and temporal lobe epilepsy.[25–27]

Bouchet and Cazauvieilh were the first to describe, in the early 19th century, the presence of hippocampal sclerosis in patients with epilepsy. However, whether this would be the cause or effect of epilepsy was still under debate until the 1960s, when it was possible to understand that this was an epileptogenic alteration.

In Brazil, the onset of the surgical treatment for epilepsy dates to the 1950s, and the most striking and pioneering contribution was by neurosurgeon Paulo Niemeyer Soares (1914–2004), who worked at Santa Casa da Misericórdia do Rio de Janeiro. Paulo Niemeyer was the first to propose and publish the amygdalohippocampectomy by transventricular access for the treatment of temporal lobe epilepsy in 1957-1958, with expressive results in crisis control.[28] This technique was accepted and is still used in many specialized centers. The first program aimed at epilepsy surgery in Brazil was established in the 1970s, at Universidade de São Paulo, by neurosurgeon Raul Marino Júnior.

Still in the 1950s, in Switzerland there was also a clear increase in the number of surgeries for epilepsy with Hugo Krayenbühl (1902–1985), who initiated and coordinated the department of neurosurgery at the University of Zurich.[29] At the same time, professor Krayenbühl was already performing a standardized technique for temporal lobectomy. One of his pupils – Mahmut Gazi Yasargil –, after researching and familiarizing himself with surgical microscopy, initiated, in 1969, the era of neurological microsurgery, designing and reinventing materials. Subsequently, Yasargil published a description of a microneurosurgical technique known as selective amygdalohippocampectomy, which consisted of the removal of the temporal medial basal structures using a transsylvian access, without the need for the removal of the anterior part of the temporal lobe.[30–32] This technique is used worldwide up to this days in many specialized centers for epilepsy surgery. After the introduction of the surgical microscope, several new techniques for epilepsy developed internationally.

The understanding and application of EEG and neurophysiology grew progressively, and, with this, the idea of establishing a preoperative diagnosis for an increasingly precise surgical planning caused variations and improvements to emerge in electroencephalographic monitoring.[33–35] New kinds of surface electrodes for EEG were developed, and the idea of obtaining information directly from the cerebral cortex with the subdural electrodes, already described in the 1940s, became widely used and disseminated in the following decades with the publications by Penfield and Jasper in Montreal.[36,37]

With the technological evolution and the advent of video in the EEG, prolonged electrophysiological monitoring became possible, facilitating the semiologic investigation of seizures.[38–40]

Following the idea of detecting the precise cerebral site involved with the onset of and to then proceed with surgical removal, and thus obtain the cure for epilepsy, French neurologist Jean Bancaud (1921–1993) postulated a way to monitor different areas and brain structures during a seizure, rather than using interictal encephalographic findings.

Bancaud and neurosurgeon Jean Talairach (1911–2007) were the first, between the 1950s and 1960s, in Paris, to describe in detail the implantation of deep cerebral electrodes with the aid of the device and coordinates of stereotaxy developed by Talairach, which is recognized as the founder of modern stereotaxy. In 1962, the technique was named stereoelectroencephalography (SEEG), and it would be the best way to correlate the semiology of the crisis with the anatomical temporal distribution obtained, and thus localize the ictal onset zone and the epileptiform dissemination network involved. In addition, there would be the possibility of cerebral stimulation, and thus of creating a three-dimensional model for investigation. The SEEG would be used in the preoperative evaluation of patients who are candidates for epilepsy surgery in France.[41–43]

Neuropsychology was extensively studied in the 1950s and 1960s, and it represents an evolution in epilepsy surgery. Brenda Milner (1918) and Juhn Wada (1924) were important researchers in this field, and they were able to collaborate with studies relating memory and language with mesial temporal resection in patients with temporal lobe epilepsy. Dr. Juhn Wada introduced and demonstrated the intracarotid use of sodium amobarbital to determine the cerebral dominance for the language function, the Wada test, as an important part of the preoperative study. Milner developed the basis of some neuropsychological tests that are still used today, and expanded the use of the Wada test to evaluate the functional memory reserve of patients undergoing surgical treatment for epilepsy. Penfield was the first to report and advocate the use of a multiprofessional group to perform the pre- and postoperative assessments, to better discriminate the pathological brain area with its psychological alterations and thus predict possible memory and language deficits with surgical treatment, besides being able to identify the presence of the cortical reorganization of cognitive functions. This was all to better guide the type and extent of resection to be performed in patients.[44–47]

From the 1970s onwards, with the establishment of the anatomopathological, electrophysiological and neuropsychological knowledge acquired, and the surgical results obtained in previous years, associated with the invention of computed tomography (CT) and other imaging methods, there was a grand global expansion in the surgical treatment of refractory epilepsies. The possibility of visualizing images of the human brain in three dimensions – and identifying structural lesions located in many patients with focal epilepsy, until then diagnosed as cryptogenic epilepsy – marked the onset of modern neuroimaging. The association of neuroimaging and EEG findings led us once again to surgeries based on directly visualized lesions, but now with preoperative visualization and electrophysiological correlation of the lesion with the crises. The CT quickly replaced the X-rays and pneumoencephalography, because it was much more sensitive in the detection of tumors, vascular lesions, posttraumatic alterations and infecto-inflammatory diseases.[48]

In the 1980s, magnetic resonance imaging (MRI) was introduced for the clinical practice, causing a revolution in several fields of medicine, and quickly replaced the CT in the diagnostic evaluation of epilepsies, as it demonstrated anatomical structures and cerebral pathologies in more detail.

With the technological evolution, new sequences of images were being made available, and new techniques to define the morphology and volume of deep brain structures were developed. Currently, it is estimated that $\sim$ 70% of patients with focal epilepsy evaluated in specialized centers present alterations in MRI images.[49,50] With the evolutionary improvement of image quality in higher magnetic field devices, there was the possibility of identifying unrecognized lesions and to estimate anatomical alterations in a non-invasive way.

Still during the 1980s, new exams, not only imaging but functional exams, were developed and added to the preoperative diagnostic arsenal of epilepsies. Among them, there is positron emission tomography (PET), which uses a radiopharmaceutical, fluorodeoxyglucose, marked with fluoride 18 (18F-FDG),injected intravenously in the patient, to observe its distribution in the brain, detected by tomography. In the interictal period, there is a hypometabolism in the pathological area and a hypermetabolism in the ictal phase. In a correlated manner, single-photon emission computed tomography (SPECT) was developed, which uses technetium-99m as a radiopharmaceutical and presents an interictal hypometabolism and a hypermetabolism during the ictus.[51,52]

With these new forms of functional metabolic investigation, patients who did not have visible lesions in the MRI could be topographically diagnosed and, in conjunction with the video-electrographic findings, selected for surgical treatment. The aim of performing a high-quality and precise preoperative evaluation resulted in the development of new techniques that improved the safety and outcome of surgeries.[53,54]

In the 1990s, there was the introduction of functional nuclear magnetic resonance (fNMR). This technique explores the dependent effect on the blood oxygenation level, and ,in brain areas activated with specific functions, the percentage of deoxyhemoglobin is reduced compared with non-active areas, which generates an increase in the signal in the T2-weighted images, enabling a cerebral mapping.

The use of fNMR in memory and language areas is important in the programming of patients with refractory epilepsy selected for surgery.[55]

It is currently possible to obtain images of the interictal activity using EEG and fNMR together. The white matter and the connectivity of the tracts became visible with the introduction and evolution of tractography. This technology has contributed to the precise definition of the epileptiform zone, its connections and its relationship with eloquent areas.

Another recently available variant is subtraction ictal SPECT coregistered to MRI (SISCOM), which enables the fusion of the ictal SPECT image with the high-resolution images of the morphological MRI. Equally important for the non-invasive diagnosis of ictal focus is magnetoencephalography, the three-dimensional reconstruction of the interictal EEG tracing captured by countless surface electrodes fused with the three-dimensional images of the morphologic MRI. These more precise diagnostic techniques are specifically important in the so-called non-injured cases, that is, patients

refractory to medications, with no identifiable lesions in the MRI.

After the development of the MRI and its technological evolution, small congenital cerebral alterations could be identified and correlated with the primary ictal zone. The improvement and refinement of microsurgical techniques, and the knowledge and development in computer graphics with the use of neuronavigation associated with intraoperative MRI increased the success rates of the surgical procedures for epilepsy.[56]

The final decade of the 20th century was remarkable in the evolution of the surgery for epilepsy. In the 1980s, most of the surgery groups for epilepsy used basically one surgical technique for the approach, depending on the place where the surgeons were trained. In 1986, the first conference on epilepsy surgery was held in California, which managed to bring together almost all of the centers specialized in epilepsy in the world to present and compare their techniques and their results. In the years that followed, countless new publications and new specialized centers emerged, and most centers innovated and adapted different techniques for the treatment of different types of epilepsy. In the beginning of the 1990s, most of the world centers were already conducting research with deep cerebral electrodes and encephalographic recordings with subdural electrodes, and performing surgeries in many patients based only on the high-quality non-invasive investigation.

At the beginning of the 21st century, the first randomized controlled scientific studies that demonstrated the superiority of the surgical treatment in patients with temporal lobe epilepsy without drug control were published, confirming the already established consensus that surgery would be the best treatment for refractory epilepsies.[57]

Currently, the use of all available resources by a multidisciplinary group dedicated specifically to the surgical treatment of epilepsy is indispensable to make an accurate diagnosis, to precisely identify the primary ictal zone, and to perform the specific resection of this cerebral area, to obtain the highest rate of seizure control with the lowest risk of functional deficits.

Conflict of Interests

The authors have no conflict of interests to declare.

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Book Review - Do No Harm: Stories of Life, Death and Brain Surgery

Resenha do livro - Sem causar mal: Histórias de vida, morte e neurocirurgia

Gilvan Aguiar da Silva[1] Elton Gomes da Silva[2]

[1] Universidade Federal de Integração Latino-Americana do Brasil (UNILA), Foz do Iguaçu, PR, Brasil
[2] Sociedade Brasileira de Neurocirurgia do Brasil (SBN), Universidade Federal de Integração Latino-Americana do Brasil (UNILA), Foz do Iguaçu, PR, Brasil

Address for correspondence Gilvan Aguiar da Silva, Universidade Federal de Integração Latino-Americana do Brasil (UNILA), Foz do Iguaçu, PR, 85870901, Brasil (e-mail: gilvanaguiar@hotmail.com).

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Abstract

In the present article, we review the book *Do No Harm: Stories of Life, Death and Brain Surgery*, by British neurosurgeon Dr. Henry Marsh, a book that can offer a significant contribution to medical education regarding the ethics and vocation for neurosurgery.

Keywords
- neurosurgery
- medical education
- ethics
- medical errors

Resumo

Neste artigo, elaboramos uma resenha do livro *Sem causar mal: histórias de vida, morte e neurocirurgia*, escrito pelo neurocirurgião inglês Dr. Henry Marsh, uma obra que pode ser de contribuição significativa para a educação médica no que se refere à ética e à vocação para a neurocirurgia.

Palavras-chave
- neurocirurgia
- educação médica
- ética
- erros médicos

Introduction

Doctors are fallible, and although they try to stay protected under an armor of superiority and detachment, they can also feel anxiety and fear before surgery. In the book *Do No Harm: Stories of Life, Death and Brain Surgery*,[1] Dr. Henry Marsh, a renowned British neurosurgeon, with a particularly realistic approach and devoid of any passionate vision for the profession, bares the myths that patients have regarding physicians, revealing the human and also fallible face of the profession, in addition to the ethical dilemmas and the emotional exhaustion that these professionals suffer in the course of their careers.

This incredible saga of exciting stories of neurosurgery that Dr. Marsh faced during his career enables a deep immersion in what the life of a neurosurgeon is, and it is told with some excerpts of explanatory technical reports of how the surgeries or the issues that will be operated on arise, and there are also moving life histories of the patients on whom he operated throughout his career.

Unlike some biographies or non-fiction books that show a mythical view of the neurosurgeon as the typical student who has always been the best in school, and is intelligent and almost an enlightened being, right in the book's initial pages, Dr. Marsh tells the reader how the beginning of his career happened, and humbly emphasizes that he was no superstudent during high school. On the contrary, practically not having received scientific education, he was rejected by most medical schools in England, and ended up being accepted to a school of medicine with a class of students with poor results in high school (p. 82). He also tells his experiences in his previous work as a nurse in a

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home for the elderly, where he performed the most diverse tasks, which most people would consider unpleasant (p. 84), and how all of this was important for his career: "I am less afraid of failure; I was able to accept its existence and feel less threatened by it" (p. 92).

In this sense, facing moral and ethical dilemmas of ineffable complexity in his daily life, the author demonstrates his point of view that it is not difficult to know when to operate: the difficulty lies in deciding when not to operate, because, in some cases, surgeries have very high chances of failure, and the operated patient can have a very short, extremely dependent, painful, dramatic and hurtful survival, which makes the concept of success relative. Thus, he declares that: "The problem is when there is no certainty between operating or not. It is easy to be wise in retrospect" (p. 242–243), or, in a controversial way, he says: "In some cases, it is better to let someone die than to operate" (p. 131).

This form of reasoning can leave the false impression that the author is devoid of any ethics or feelings toward his patients, but the reader soon realizes that this is a terrible mistake, especially when he exemplifies his reasoning, describing how he eventually decided to operate on some patients who he knew to be terminally ill due to cowardice to confront the family and tell them the truth (p. 145), and how unpleasant these experiences were for all of those involved. He also describes a personal case that was quite moving, when his own mother was diagnosed with an incurable cancer and was sent home to die, a much less painful experience than it could have been in a hospital environment (p. 203).

Over the course of the stories, it is easy to see that although Dr. Marsh has become an extremely pragmatic and rational professional throughout his career, who does not believe in miracles, it is precisely this characteristic that enables him to develop a touching and profound involvement with his patients, for example, when he emphasizes his view that he does not feel rewarded by a grateful patient who returns several times to the office with gifts or compliments, but only feels really rewarded when his patients forget about him, for this means that they were healed (p. 42). It would be easy to write a book only with the cases of success and happiness, but it takes a lot of self-confidence and courage to take on the failures that are inevitable throughout anyone's life. Thus, by promoting this shock of reality and showing that physicians are also human and make mistakes, Dr. Marsh makes us reflect about the pressure to which these professionals are subjected, for they need to make quick decisions and are surrounded by the constant risk of making mistakes, because when neurosurgery errors occur, the results are usually catastrophic (p. 163).

One of these cases is that of a young athlete who ended up having the root of a nerve severed due to an error by one of his residents (but in which case he was also partially guilty) that made the patient unable to lift his foot, and he became limp and unable to run or ride a bike (p. 161, 180 and 183). There are still cases of patients who suffered strokes during operations and became incapable of speaking and understanding what is said, becoming almost disconnected from the real world (p. 192), or even the regret he feels for having let himself be convinced in the past to perform a psychosurgery (p. 125).

Other cases of errors or more dramatic complications are narrated, and in them the author exposes how the term "complications" is sometimes used as a euphemism for real chaotic scenes of uncontrolled hemorrhages or situations bordering on despair (p. 108 and 244).

In several passages, the author describes the burdens that come with the profession: he states that he behaved in an exultant manner before surgery at the beginning of his career, but also describes how difficult it is to maintain optimism and enthusiasm after many years into the profession, after having witnessed stories of despair, surgical complications, and weak results (p. 11). Therefore, it is expected that neurosurgeons feel anxiety and fear before operating (p. 47); the thing is that they cannot show this to the team or the patients: it is a weight that they need to carry alone.

In fact, some neurosurgeons, at the end of their careers, can suffer from psychological problems, such as anxiety or depression, due to the great stress to which they are subjected, with a routine of little time for the family and a lot of time surrounded by tragic stories. This becomes very clear, for example, on the day that Dr. Marsh had to interrupt a stroll in his rare time off to give a patient the terrible news that he would inevitably die, thus being shaken throughout the remainder of the day (p. 158), or even as all this routine of lack of time and stress led to the end of his first marriage (p. 21).

Many young medical students feel seduced by the specialty of neurosurgery, which is even perceived by some as the position of highest medical status, but few can imagine how dramatic and sufferable a lifetime devoted to neurosurgery can be when dealing with reality itself: "A typical career in neurosurgery involves death, discussions and sequelae" (p. 137), so it is not enough to have a calling for the profession, it is also necessary to have a realistic view of the environment in which you will have to live: "Neurosurgery is a horrible work, full of sorrows and disappointments, do not follow this path" (p. 194), advises the author.

Therefore, we conclude that this work, besides enabling the reader to have a notion of how some neurosurgeries are performed, provides incredible lessons and exciting stories, culminating in a profound analysis about medical ethics. Therefore, from the point of view of medical education and calling for neurosurgery, this work may be a reality check to subsidize the decision of students who still have doubts about the career they want to follow. In fact, the format adopted by the author, unlike the traditional academic writings of the field of science, makes the text extremely accessible, light and pleasant, a compelling read, and that also makes this work an instrument of scientific disclosure that can be used even as a complementary read in postgraduate or scientific initiation courses in the field of health sciences.

Conflict of Interests
The authors have no conflict of interests to declare.

Reference

1 Marsh H. Do No Harm: Stories of Life, Death and Brain Surgery. São Paulo, SP: Editora Nversos; 2017

Feasibility of Nano-based Neuroprosthetics in Neurosurgery

Viabilidade de neuropróteses nanotecnológicas em neurocirugia

Arthur Saniotis[1,2] Maciej Henneberg[2,3] Abdul-Rahman Sawalma[4]

[1] Department of Medical Laboratory Science, Knowledge University, Kurdistan Region, Iraq
[2] Biological Anthropology and Comparative Anatomy Research Unit (*BACARU*), Adelaide Medical School, University of Adelaide, Adelaide, SA, Australia
[3] Institute of Evolutionary Medicine, University of Zürich, Zurich, Switzerland
[4] Palestinian Neuroscience Initiative, Al-Quds University, Beit Hanina, Palestine

Address for correspondence Dr. Arthur Saniotis, PhD, Department of Medical Laboratory Science, Knowledge University, Kurdistan Region, Iraq (e-mail: arthur.saniotis@knu.edu.iq).

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Since their development, auditory, visual and pain-reducing neuroprosthetic devices have improved the lives of thousands of patients. Further advances in motor neuroprosthetics are attempting to restore motor functions in tetraplegic, limb loss and brain stem stroke paralysis subjects. This has been clinically feasible by simulating the conditions by which neuroplasticity is triggered.[1]

While the use of nanotechnology in neurosurgery is still at a developmental stage, over the next few decades medicine will witness an influx of nanotechnology techniques to neurosurgery in the areas of management, detection and treatment of gliomas, oncology, neuromodulation, neuroregeneration,[2] and minimally-invasive diagnostic and neurosurgical techniques.[3,4] For example, future neurosurgical procedures may use laser beams to seal tissue via the use of specific gold nanorods (GNRs), which would significantly minimize vascular injury and decrease postoperative healing time.[3]

The advent of nanotechnology in neurosurgery will challenge the current mechanistic approach that informs neuroprosthetics, through which it is not possible to apprehend the complexity of the cortical processes involved in cognitive, motor and sensory functions. One reason for this is our insufficient knowledge of the evolutionary processes that have shaped the human cerebrum.[5]

Second, current neuroprosthetic devices have a range of postoperative problems that include inflammation, glial-cell necrosis and scarring,[6] as well as degradation of electrode implants, among other problems. These are well-known to every neurosurgeon. Additionally, even the type of implantation method used by neurosurgeons,[7] as well as the time it takes to insert a neuroprosthetic device, can reduce their efficiency.[8]

Recently, we published an article[9] in *Frontiers in Neuroscience* in which we developed an idea for a novel nanotech neuroprosthetic device called "endomyccorhizae-like interface" (ELI). One reason for the development of ELI was to bypass the aforementioned neurosurgical problems.[9] Our design for ELI was inspired by *endomyccorhizae* (fungus/plant root symbiosis). During this process, fungi bundles project finger-like extensions (*mycelium*) that penetrate plant roots, forming a branching matrix (*arbuscules*). Similarly, in theory, ELI would send out multiple mesh fibers from a cation chamber. Briefly, the fiber tips would then attach to specific axonal membranes, thus achieving connectivity.[9] ELI's role would be in transmitting action potentials between neurons to which it is connected, as well as in improving the speed of receiving action potentials. In other words, ELI would endeavor to partially regain the normal connectivity speeds in interrupted neural pathways (that is, those evident in neurodegenerative disorders) without altering the regular tasks of neurons.[9] Uninhibited increase in neuronal activity may lead to functional difficulties. Therefore, ELI or other future nano-based neuroprosthetic devices will need to be

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able to react to inhibitory signals, thereby enhancing their sensitivity to signaling.

The ELI device would be delivered using a fine syringe needle through the orbital roof, which is lined with meninges, and inserted ~ 1 mm into the subarachnoid space with the cerebrospinal fluid (CSF).[9] The micro-diameter of the needle would ensure minimal invasiveness. Recently, researchers have used syringe needles with a diameter of 100 µm when injecting implanted electrical meshes into the brains of anaesthetized mice.[10]

Hence, this type of delivery would avoid the problems of injecting nanoparticles into the blood-brain barrier. The ELI would move using a propeller-like device. Upon entering the subarachnoid space, ELI would be in proximity with the cortical surface, and would spread over the neocortex by sensing the electrical activity of neurons. The energy to power ELI could be obtained from adenosine triphosphate (ATP)[11] or brain-cell (neurons and glial cells) electrolytes.[12]

We envisage that the quantification of the cognitive improvements achieved by patients could be made by measurements of mental aptitude with an appropriate test such as the IQ test or modified psychometric tests, based on recommendations by Snow et al[13] and Ferrara.[14] The tests would encompass a wide-ranging structure to assess not only the mental processes in patients (numerical/verbal/diagrammatic reasoning, tests for explicit/implicit memory), but also the interaction between patient and task in various situations and environments, as well as the response and reaction performance during kinesiology tasks to assess neural control in various clinical contexts.[14,15] Performance results would be recorded in computer-based formats, enabling scientists to deduce the novel context exploration (NCE)/neuron interaction in individuals.[14] Comprehensive testing would be ongoing to determine the long-term effects of NCEs.

While ELI is at a developmental stage, it exemplifies how nanotechnology may be able to significantly reduce the problems of current neuroprosthetic devices. A great deal of work is needed before ELI or other nano-based neural devices are engineered and become part of neurosurgery. The feasibility of such neuroprosthetics will require an understanding of the dynamics of diffuse neural networks and their integrative faculties.

The feasibility of using ELI seems obvious in neurodegenerative disorders that slow down, or interrupt, the communication among neurons. The device may also become useful in cases of recovery from strokes, brain injuries or therapeutic removal of a part of the cortex, when tumors are excised, for example. In those cases, it is necessary for uninjured cortical areas to take over functions normally performed in the damaged/removed cortex, and increasing the connectivity of the remaining cortex may improve the recovery. Were ELIs be able to be made sensitive to specific neurosecretions, they might provide replacements for dysfunctional neurotransmitter systems, such as dopaminergic systems.

Conflict of Interests

The authors have no conflict of interests to declare.

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Grisel Syndrome: Case Report

Síndrome de Grisel: Relato de caso

Bruno Roncaglio[1] Tadeu Gervazoni Debom[1] Marcus Alexandre Novo Brazolino[1] Thiago Cardoso Maia[1] José Lucas Batista Filho[1] Igor Machado Cardoso[1] Charbel Jacob Junior[1]

[1] Department of Orthopedics and Traumatology, Instituto de Coluna Vertebral, Hospital Santa Casa de Misericórdia, Vitória, ES, Brazil

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Address for correspondence Bruno Roncaglio, Departamento de Ortopedia e Traumatologia, Instituto de Coluna Vertebral, Hospital Santa Casa de Misericórdia, Vitória, ES, 29025-023, Brazil (e-mail: institutodecoluna@gmail.com; brroncaglio@gmail.com).

Abstract

We report a case of a rare disease, Grisel syndrome, which manifests as a kind of rotational fixation of the atlas on the axis, resulting from an infectious process of the upper airways. In the present report, we discuss etiology, clinical presentation, diagnosis, treatment and outcome after intervention.

Keywords
- atlantoaxial joint / injuries
- grisel syndrome / treatment

Resumo

Relatamos um caso de doença rara, a síndrome de Grisel, que se manifesta como um tipo de fixação rotatória do atlas sobre o áxis, decorrente de processo infeccioso das vias aéreas superiores. Neste relato, discutiremos etiologia, quadro clínico, diagnóstico, tratamento e evolução do caso após intervenção.

Palavras-chave
- articulação atlantoaxial / lesões
- síndrome de grisel / tratamento

Introduction

Grisel syndrome is a rare type of rotational fixation of the atlas on the axis (C1-C2), initially described in 1830, when Bell[1] reported a case of atlantoaxial subluxation secondary to a process of syphilitic ulcerative pharynx. Subsequently, it has been described as a rare complication of inflammatory processes secondary to otorhinolaryngological and gastroenterologic surgical interventions and to upper airway infections (UAIs). The etiopathogenesis has not been fully clarified, but it is described as an association with an infection or inflammatory process of the head and neck region with subsequent dissemination to the atlantoaxial joint, which would weaken at the level of its bone and ligamentous insertion. It has an incidence of 68% among the population younger than 12 years of age, and of 90% among those under 21 years of age.[2,3] The typical presentation is with cervical pain, deformity and limitation of movements (fixed torticollis), with the existence of a recent inflammatory or infectious process. The child adopts a cock robin position, due to the presence of the so-called paradoxical torticollis, which differs from the common muscular torticollis because, instead of shortening, there is stretching of the sternocleidomastoid muscle. Another finding from the clinical examination is the Sudek sign, in which the C2 spinous process is diverted to the same side of the head. In less than 15% of the cases, neurological complications occur, ranging from transient sensory alterations to tetraplegia, and even sudden death. The common differential diagnosis is meningitis, and retropharyngeal abscess, spasmodic muscle torticollis, trauma and adverse drug reactions should also be considered.[4] Imaging exams help in the diagnosis. Radiography and computed tomography (CT) can show an asymmetry between the lateral masses of C1 and an increase in the atlantodental interval.

The classification of Fielding et al[5] (►**Fig. 1**) enables the grouping of the different degrees of atlantoaxial subluxation

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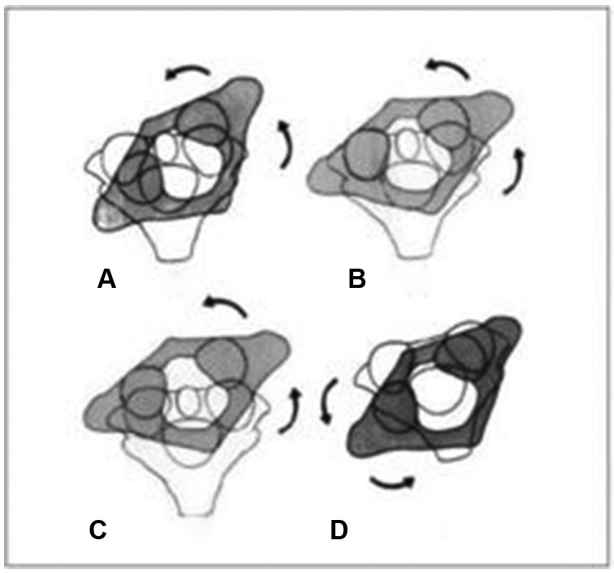

Fig. 1 (**A**) Type I; (**B**) type II; (**C**) type III; (**D**) type IV.

Fig. 2 Clinical image of a patient with head tilt to the right and rotated back to the contralateral side.

and assists in the prediction of the prognosis for potential complications, dividing the cases into: type I, fixed rotational subluxation; type II, anterior deviation = 3–5 mm; type III, anterior deviation > 5 mm; and type IV, posterior deviation. The treatment is conservative at first. Cases of failure, irreducible subluxation and recurrences have surgical indication.[5]

The objective of the present work is to report a rare case of Grisel syndrome, with inveterate atlantoaxial subluxation and its treatment with cranial halo, and to describe its physiopathology.

Clinical Case

A 9-year-old male patient was admitted to our service with cervical pain, deformity and limitation in range of motion for 50 days, with no history of trauma, which was treated as spasmodic torticollis. Regarding his previous history, the patient presented a picture of UAI in the three weeks preceding the onset of symptoms, with resolution after the use of symptomatic relief medications. Upon clinical examination, a limitation in the amplitude of the cervical movement was observed, with inclination of the head to the right and deviation of the contralateral chin, without neurological deficit (►**Fig. 2**). The patient was submitted to radiography and computed tomography (CT) scans, in which a C1-C2 rotational subluxation was visualized (►**Figs. 3**, **4** and **5**). The patient was submitted to cranial halo and traction in a surgical center, which was maintained for 15 days in a Stagnara chair with progressive traction, and subsequently used a halo vest for 12 weeks (►**Fig. 6**), obtaining C1-C2 reduction with a 5-mm index measured in the coronal and axial planes (►**Figs. 7** and **8**). The patient presented good evolution with the proposed treatment, and did not evolve with functional limitation in the range of motion of the cervical spine (►**Fig. 9**).

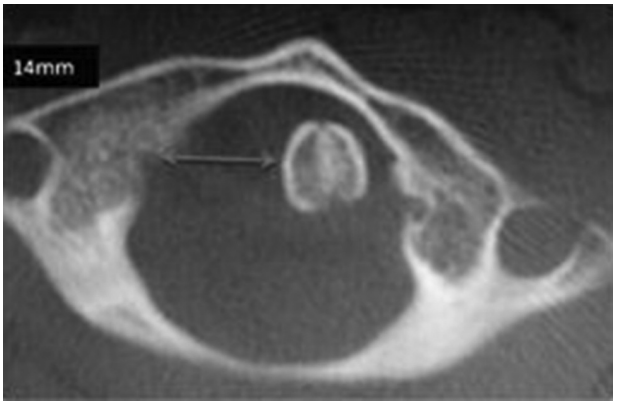

Fig. 3 Computed tomography; axial view showing rotational subluxation in C1-C2.

Discussion

Grisel syndrome is an eminently pediatric pathology, although there are cases reported in adults. In a series of cases, Watson-Jones[6] and Wilson et al[4] demonstrated that most cases are manifested in children under 10 years old. It results from a pathological relaxation of the ligaments around the C1-C2 joint, following an inflammatory, infectious or surgical procedure.[7,8]

The diagnosis of non-traumatic atlantoaxial subluxation requires radiological investigation. Simple cervical spine radiography may present an asymmetry between the joints in the anteroposterior projection and increase in the atlantodental interval in lateral projections. In children, the atlantodental interval normally measures less than 3 mm. An increase in this interval to more than 5 mm would suggest traumatic or non-traumatic subluxation.[8] The exam of choice in the evaluation of the bone cervical spine is the CT, while magnetic resonance

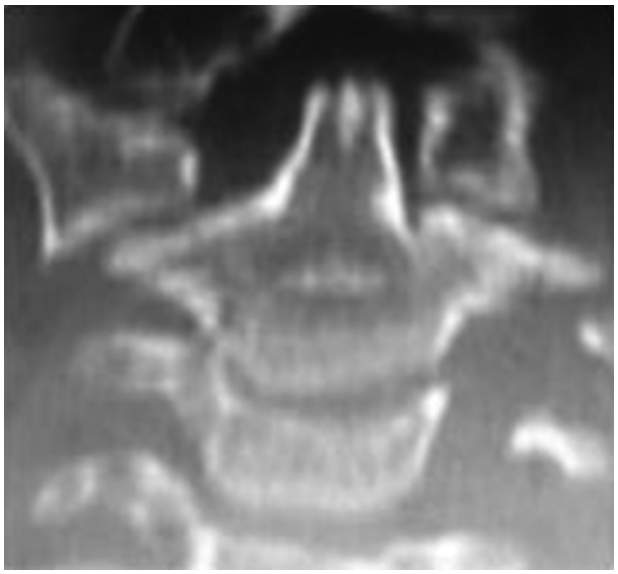

Fig. 4 Computed tomography; coronal view showing rotational subluxation in C1-C2.

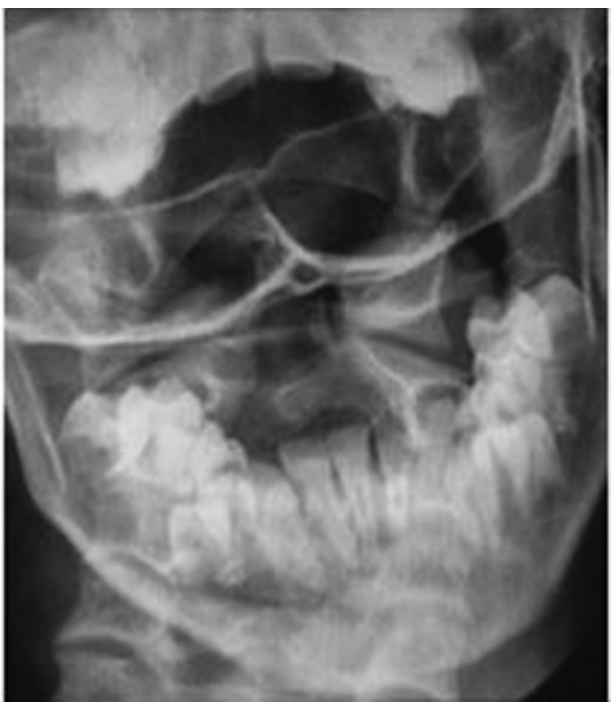

Fig. 5 Transoral radiography of the cervical spine showing subluxation in C1-C2.

imaging (MRI) provides information related to the structures of the surrounding soft tissues and spinal cord.

In the case in question, the patient presented an irreducible lesion, with an atlantoaxial interval of 14 mm, and type III in the Fielding et al[5] classification. Due to the delayed definitive diagnosis, there was no spontaneous reduction of the subluxation, and the patient was submitted to skeletal traction.

The prognosis with the conservative treatment is excellent, when the diagnosis is established in adequate time, that is, less than 3 weeks after the onset of the first symptoms. After this period, some form of surgical intervention may be required

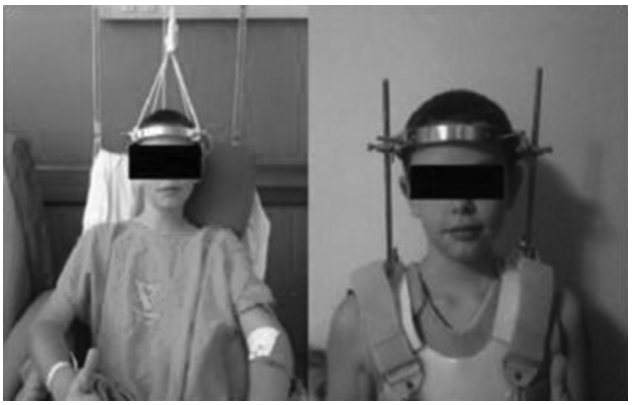

Fig. 6 Patient in traction and with halo vest for bloodless reduction.

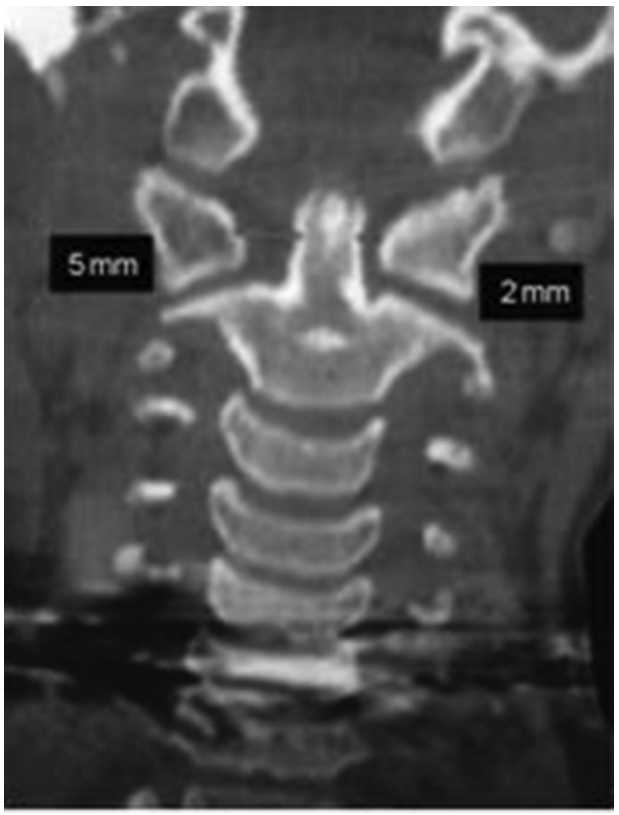

Fig. 7 Computed tomography; frontal view showing acceptable reduction of the subluxation in C1-C2.

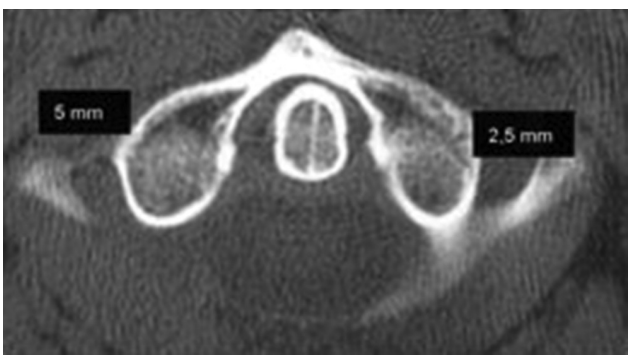

Fig. 8 Computed tomography; axial view showing acceptable reduction of the subluxation in C1-C2.

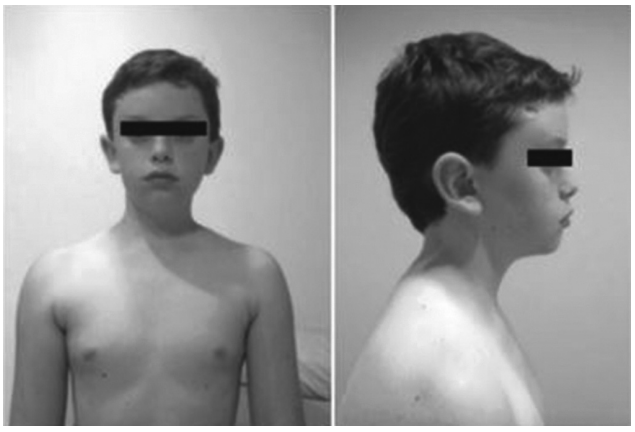

Fig. 9 Clinical image of the patient in anteroposterior and profile views after the reduction.

due to the irreducibility.[9] In most cases, the subluxation is reduced spontaneously.[10]

The treatment for Grisel syndrome is controversial. It usually starts with conservative and supportive measures, including resting, cervical collar, analgesics, anti-inflammatories, muscle relaxants and, when necessary, antibiotic therapy and abscess drainage.[3] The expected time for spontaneous reduction, in most cases, is after 7 days of treatment; after the reduction of the lesion and regression of the inflammatory process, the stability is recovered.[10] In cases in which spontaneous reduction does not occur, manual reduction under sedation, cranial traction and, lastly, surgical treatment with craniocervical arthrodesis may be attempted.[9,11,12] Wetzel and La Rocca[8] proposed a treatment protocol for non-traumatic atlantoaxial rotational subluxation based on the Fielding et al[5] classification. The conservative treatment for lesions of types I, II and III would be soft paste, rigid collar (Philadelphia) and halo-vest respectively. For type-IV lesions, they recommend surgical treatment, which is also indicated in cases of failure of the conservative treatment, recurrence of subluxation, and irreducible subluxations.[8] Gomes et al[13] evaluated retrospectively 35 cases of Grisel syndrome treated at a referral hospital, and they found 19 female children with a mean age of 6.9 years. Cervical pain was the main complaint of 85% of the patients, followed by deformity and loss of mobility. History of UAI was present in 80% of the cases, and trauma was also found in 14%. Only one patient suffered neurological deficit (deltoid and triceps paresis), with complete remission after the treatment. According to Fielding et al,[5] 14 patients were classified as type II, and there were no cases of type IV. The treatment performed with collar alone in twenty patients had good results. Surgical treatment was performed in six cases, with C1-C2 arthrodesis preceded by a reduction with halo and traction. Complications resulting from the treatment were scarce. Pilge et al[14] reported an 11-year-old patient with 2 months of evolution of atlantoaxial subluxation after surgery for cochlear implantation, in which a reduction was performed under general anesthesia and myorelaxants, followed by cervical collar. The reduction was satisfactory.

The patient in the present report was submitted to progressive load (halo) traction for 2 weeks, and used a halo vest for another 12 weeks. The case showed satisfactory evolution, obtaining a reduction of C1-C2, with an index of around 5 mm measured in the coronal plane.

Conclusion

Despite its rarity, it is fundamental that the medical community know this entity, in order to avoid subdiagnoses and consequent irreversible injuries.

Conflict of Interests

The authors have no conflict of interests to declare.

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Epidermoid Cyst of the IV Ventricle: Case Report

Cisto epidermoide do IV ventrículo: Relato de caso

Washington Clésio da Silva Ribeiro[1] Ricardo Macedo Camelo[1] Josias Inácio da Silva[1]
Moana Vergetti Malta[1] Rafael Costa Camelo[1] Arnon Castro Alves Filho[1]
Felipe Gomes de Barros Pontes[1] Adans Soares Porfírio[2] Daniel Fonseca Oliveira[2]
Bruno Clécio da Silva Ribeiro[3] Alessandro Henrique Rodrigues Ferreira[4]

[1] Department of Neurosurgery, Hospital Universitário Professor Alberto Antunes (HUPAA), Universidade Federal de Alagoas (UFAL), Maceió, AL, Brazil
[2] Neurosurgery Residency, Hospital Universitário Professor Alberto Antunes (HUPAA), Universidade Federal de Alagoas (UFAL), Maceió, AL, Brazil
[3] Faculdade de Medicina, Universidade Federal do Oeste da Bahia, Barreiras, BA, Brazil
[4] Faculdade de Medicina, Hospital Universitário Professor Alberto Antunes (HUPAA), Universidade Federal de Alagoas (UFAL), Maceió, AL, Brazil

Address for correspondence Washington Clésio da Silva Ribeiro, MD, Serviço de Neurocirurgia, Hospital Universitário Professor Alberto Antunes, Maceió, AL, Brazil (e-mail: washingtonrb@gmail.com).

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Abstract

Epidermoid cysts constitute congenital, benign and rare lesions, corresponding to ~ 0.2% to 1.8% of all intracranial tumors. Only 5% of the cases are located in the fourth ventricle. Despite their genesis in intrauterine life, they are usually diagnosed between the third and fifth decades of life due to their very slow growth pattern. The image weighted by the diffusion of the magnetic resonance is essential to establish the diagnosis. The ideal treatment consists of emptying the cystic content with complete capsule resection. In the present work, we report the case of a 31-year-old female with cerebellar syndrome that evolved with intracranial hypertension. The symptomatology was due to an obstructive hydrocephalus by an epidermoid cyst located inside the fourth ventricle, which was confirmed by the pathological anatomy.

Keywords
- epidermoid cyst
- fourth ventricle
- hydrocephalus

Resumo

Os cistos epidermoides constituem lesões congênitas, benignas e raras, que correspondem a cerca de 0,2% a 1,8% de todos os tumores intracranianos, sendo que em apenas 5% dos casos localizam-se no quarto ventrículo. Apesar de sua gênese na vida intrauterina, são geralmente diagnosticados entre a terceira e a quinta décadas de vida, em decorrência de seu padrão de crescimento muito lento. A imagem ponderada pela difusão da ressonância magnética é fundamental para a formulação do diagnóstico. O tratamento ideal consiste no esvaziamento do conteúdo cístico com ressecção completa da cápsula. Neste trabalho, é descrito um caso de uma paciente de 31 anos de idade com síndrome cerebelar, que evoluiu com síndrome de hipertensão intracraniana. A sintomatologia era decorrente de hidrocefalia obstrutiva por cisto epidermoide localizado no interior do quarto ventrículo, confirmado pela anatomia patológica.

Palavras-chave
- cisto epidermoide
- quarto ventrículo
- hidrocefalia

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Introduction

Intracranial epidermoid cysts (ECs) consist of slow-growing congenital lesions. Initially described by French pathologist Cruveilhier as the 'most beautiful of all tumors' based on their pearly nature,[1] ECs are originate from malformations associated with surface elements of the nervous system ectoderm during the neural sulcus closure, or from the formation of secondary cerebral vesicles.[2]

Because they grow slowly, they usually do not become symptomatic until the patient has reached the third or fifth decades of life. They develop through the progressive accumulation of keratin and cholesterol, produced by the peeling of the epithelium that surrounds the cyst.[3]

The occurrence of ECs is rare, making up to ~ 0.2% to 1.8% of central nervous system (CNS) tumors.[1] Classically, they are located more frequently in the following sites: the cerebellopontine angle (corresponding to half of the cases), the temporal fossa, and the suprasellar and quadrigeminal regions. Implantation in the cisterna magna region and the fourth ventricle is even rarer.[4] Approximately 5% of ECs affect this location.[5] According to Sengupta and Singh,[6] there are only about 100 reported cases of epidermoid tumors of the fourth ventricle.[6]

Structurally, ECs are formed by a white, pearly tissue, which is coated by a capsule adhered to adjacent structures. The cystic content is avascular, and presents a material resembling candle wax arranged in concentric lamels.[7] The lesion grows slowly, and has a flexible and deformable appearance, adapting to the space in which it develops.[8]

The tomographic aspect of this lesion is typical of a hypodense extra-axial mass, which is not impregnated by venous contrast. Upon magnetic resonance imaging, they may present a similar signal or a slight increase regarding cerebrospinal fluid (CSF) in the T1 and T2 ponderations. The main differential diagnosis is made in relation to the arachnoid cyst, and the distinction is usually made with sequences of fluid-attenuated inversion recovery (FLAIR) and diffusion. The arachnoid cyst follows the intensity of the CSF signal in all sequences, while epidermoid tumors are not hypointense in the FLAIR, showing areas of hypersignal in relation to the CSF. In diffusion sequences, epidermoid tumors typically have diffusion restriction, unlike arachnoid cysts.[9]

The ideal treatment is the removal of the cystic component with complete resection of the capsule, taking care not to compromise the neurological condition of the patient.[4] Although the contents of the cyst can be easily aspirated, the total removal of the tumor is not always possible due to the fact that the capsule is usually adhered to neighboring neurovascular structures.[10]

Aseptic meningitis, a complication that may occur postoperatively, can be avoided by preventing the extravasation of the cystic content into the surrounding subarachnoid space.[10] Irrigation of the surgical bed with corticoids has been reported as a form of aseptic meningitis prevention.[11]

The rate of recurrence is between 1% and 54%, and it can be avoided by devitalizing the remaining fragments of the capsule during surgery.[11] The surgical reapproach is usually performed for decompression. Malignant degeneration has been reported in cases of recurrent epidermoid tumors.[4]

The objective of the present article is to report a case of EC of the posterior fossa affecting the interior of the fourth ventricle with extension to the cisterna magna. Its presentation, radiological image and surgical management will be described.

Case Report

A 31-year-old female patient, who started with dysmetria, gait disorder, diplopia and dizziness one year previously, evolved with progressive headache over the last 3 months, associated with vomiting and loss of visual acuity. A computed tomography scan of the skull revealed a hypodense lesion located inside the fourth ventricle causing obstructive hydrocephalus. Initially, the patient was submitted to a ventriculoperitoneal shunt, with symptomatic improvement after the procedure, especially with regards to the headache and visual loss. With the stabilization of the neurological condition after intracranial hypertension treatment in emergency services, the patient was referred to our service for a complementary radiological study, aiming to enable the surgical approach of the lesion.

On the neurological admission examination, the presence of gait ataxia, positive Romberg sign, more pronounced dysmetria on the left, dysdiadochokinesia, nystagmus and paralysis of the sixth cranial pair on the left was verified.

On the magnetic resonance imaging, it was possible to observe an expansive extra-axial formation with epicenter in the cerebellar cistern measuring 4.1 cm × 4.9 cm × 4.2 cm, isointense to CSF in T1 and T2 (►**Figs. 1** and **2**). The aforementioned lesion did not present postcontrast enhancement (►**Fig. 1b**). The lesion compressed the bridge, the bulb, the IV ventricle, and the cerebellar hemispheres, and was more pronounced on the left. There was inferior insinuation of the lesion along the foramen magnum for ~ 9 mm. The heterogeneous signal in the FLAIR and especially the diffusion restriction made it possible to indicate EC as the main diagnostic hypothesis (►**Fig. 3**).

The patient underwent suboccipital craniectomy to access the posterior fossa. After opening the dura mater, a whitish and pearly lesion was found, emerging from the bulbocerebellar fissure (►**Fig. 4**). We performed the emptying of the lesion that occupied the interior of the IV ventricle (►**Fig. 5**). The surgical procedure went uneventful.

The postoperative period progressed satisfactorily, with improvement of the previous symptomatology. The results of the anatomopathological examination were consistent with EC. The patient was discharged on the 11th day after surgery. In a return consultation performed ~ 3 months after surgery, she was already walking without help, maintaining progressive improvement. The magnetic resonance imaging on this occasion revealed complete emptying of the lesion located in the IV ventricle (►**Fig. 6**).

Discussion

Epidermoid cysts constitute congenital, benign and rare lesions. They correspond to ~ 0.2% to 1.8% of all intracranial

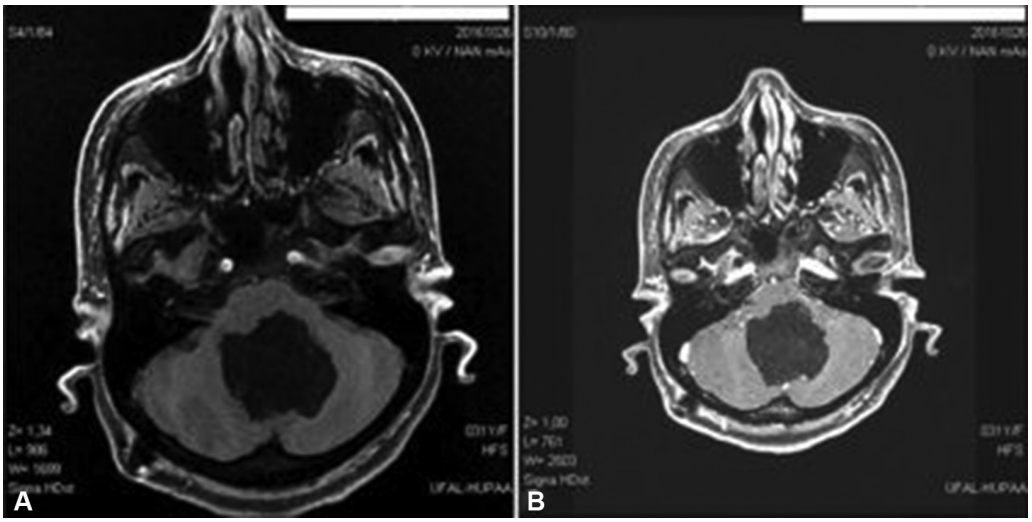

Fig. 1 Magnetic resonance imaging: axial cut showing cystic lesion inside the IV ventricle, measuring 4.1 cm × 4.9 cm × 4.2 cm. (**A**) T1 sequence without contrast evidencing a hypointense injury, slightly heterogeneous, and with a slightly larger signal from the cerebrospinal fluid. (**B**) Absence of enhancement after contrast administration.

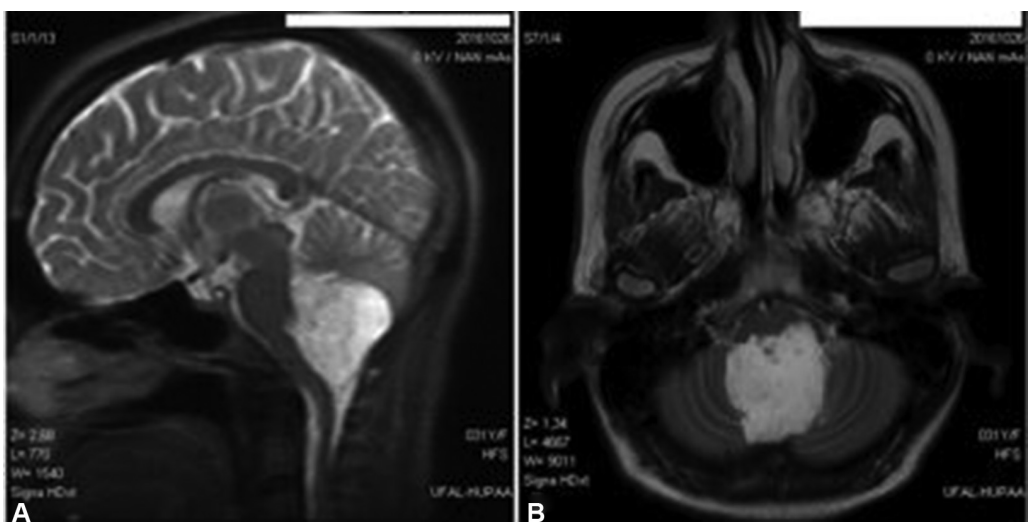

Fig. 2 Magnetic resonance with T2 weighting. The lesion is hyperintense, similar to the cerebrospinal fluid. (**A**) Sagittal cut; and (**B**) axial cut.

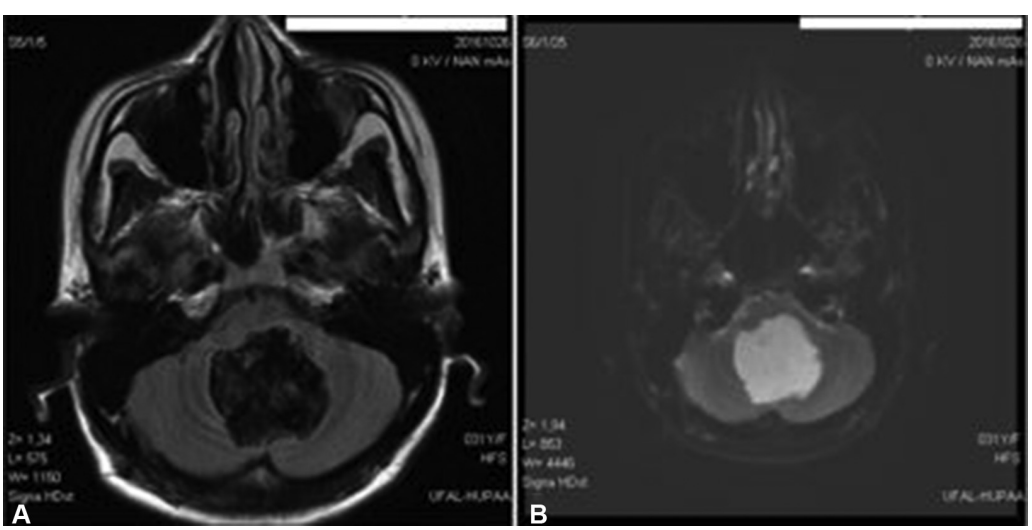

Fig. 3 Magnetic Resonance imaging: axial cut showing (**A**) heterogeneous signal in the FLAIR, and (**B**) a hyperintense signal in the restriction, making the epidermoid cyst the main diagnostic hypothesis.

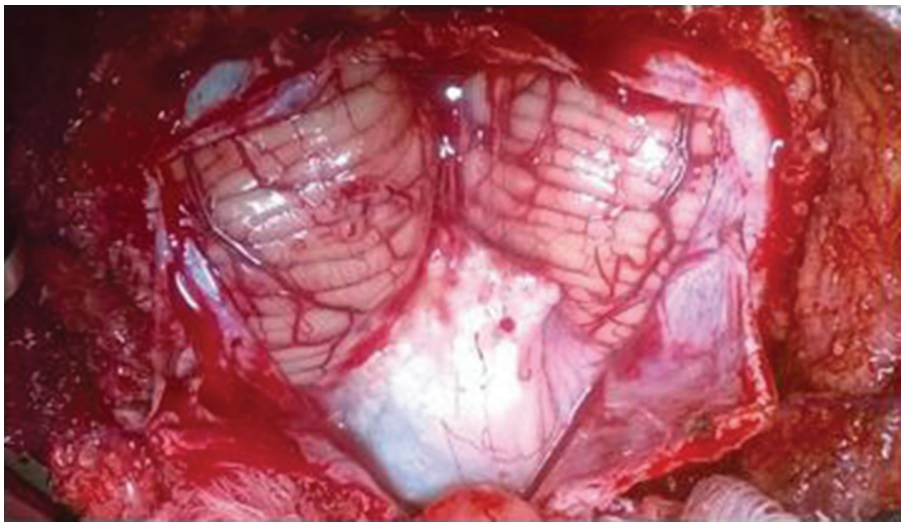

Fig. 4 Intraoperative finding. Whitish and pearly lesion emerging from the bulbo-cerebellar fissure.

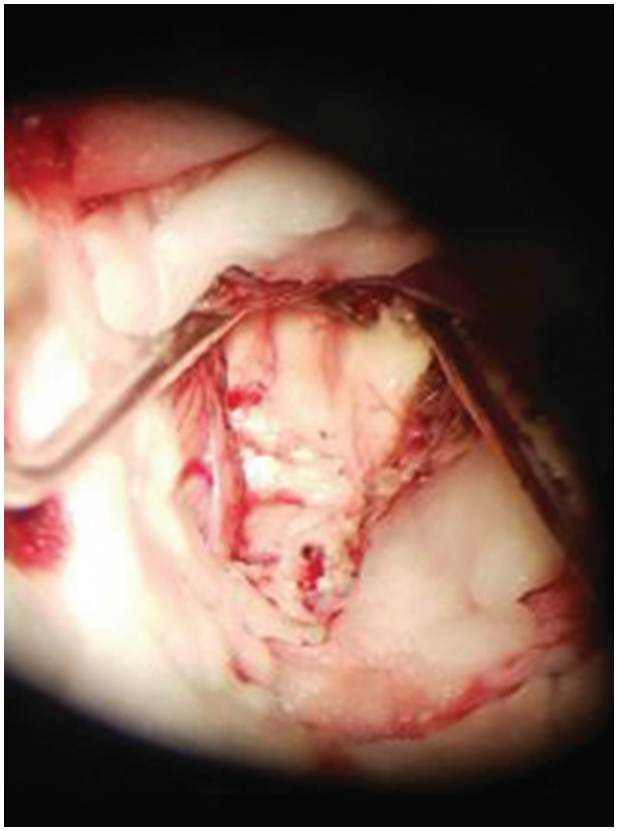

Fig. 5 Intraoperative findings. Emptying of the cyst, with a view of the floor of the IV ventricle (bridge and bulb) on the deep margin of the lesion.

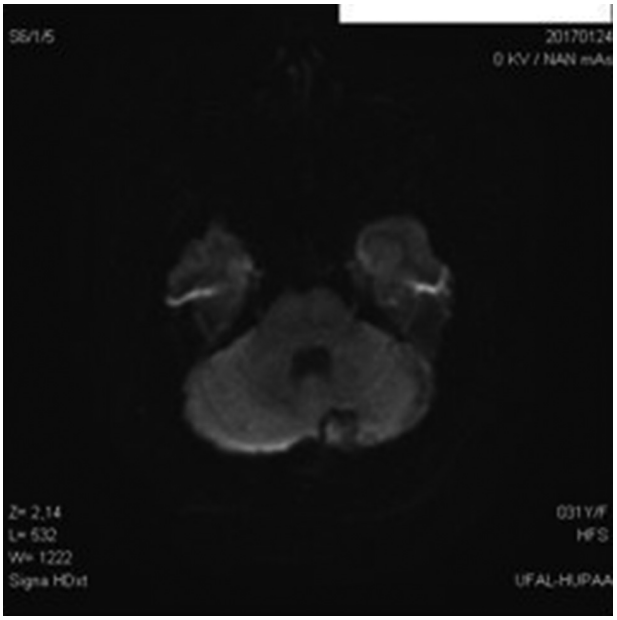

Fig. 6 Magnetic resonance imaging with diffusion weighting, evidencing the dissection of the cystic cavity with anatomical restoration of the IV ventricle.

tumors.[12] There is preponderance among females. The most frequent locations of these processes are: the cerebellopontine angle and the parasellar region; other rarer locations include the brainstem and the fourth ventricle.[13] They are located in the fourth ventricle only in 5% of the cases, denoting unusual topography. There are only ~ 100 cases of this location described in the literature.[5,6]

Epidermoid cysts are tumors of very slow growth, with a growth pattern like that of epidermal skin cells. They develop from the remaining elements of the epidermis during the closure of the neural sulcus and the disjunction of the surface ectoderm with the neural ectoderm, between the third and fifth weeks of embryonic life.[11] Despite its genesis in intrauterine life, the moment of diagnosis is usually established between the third and fifth decades of life, the age group of the patient in the case herein reported.

Clinically, cerebellar syndrome is the main responsible disease for the initial symptomatology, while intracranial hypertension syndrome is less common, since hydrocephalus, which occurred in the case herein reported, is of late onset, and is observed in less than 50% of the cases.[14] Although the extravasation of cystic content to the subarachnoid space has been shown to cause aseptic meningitis, no signs of meningeal irritation were observed.

Preoperative magnetic resonance imaging was fundamental for the establishment of the diagnostic hypothesis of EC. The lesion showed no contrast enhancement, and showed a signal similar to CSF in the T1 and T2 ponderations; however, it was restricted to diffusion, differentiating itself from other diagnostic possibilities, such as arachnoid and tumor cysts.

Two complications may alter the course of the disease: aseptic meningitis and malignant transformation. In order for aseptic meningitis to occur, a simple contact of the cystic content with the CSF can be enough. The treatment of this condition includes repeated lumbar punctures and corticoids.[15] This situation was not observed in the case of the patient in question.

On the other hand, malignant transformation is an extremely rare complication, and, when present, is associated with the development of squamous cell carcinoma. In the case of incomplete removal of the lesion, the growth of the remnant tumor is as slow as that of the primitive tumor, requiring annual follow-up through magnetic resonance imaging to evaluate its development potential.[15] Some studies have indicated radiotherapy as an alternative to the failure in surgical treatment or recurrence, and tumor control can be achieved through this method.[16]

Conclusion

Epidermoid cysts constitute congenital, benign and rare lesions, corresponding to $\sim$ 0.2% to 1.8% of all intracranial tumors, and they are located in the fourth ventricle only in 5% of the cases. Despite their genesis in intrauterine life, they are generally diagnosed between the third and fifth decades of life as a result of their very slow growth pattern. They develop from the malformations of the ectoderm during the closure of the neural sulcus, between the third and fifth weeks of embryonic life. The diffusion-weighted image of the magnetic resonance imaging was fundamental for the establishment of the diagnosis. The ideal treatment consists of emptying the cystic content with complete resection of the capsule, although it is not always possible when the capsule is firmly adhered to important structures. Radiological follow-up is necessary in the postoperative period to evaluate the recurrence, although malignant transformation is rare, and reoperation has, above all, a decompressive character for symptom relief.

Conflict of Interests
The authors have no conflict of interests to declare.

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Neurectomy of C2 for the Treatment of Occipital Neuralgia: Case Report

Neurectomia de C2 para tratamento da neuralgia occipital: Relato de caso

Marcelo José Silva de Magalhães[1,2] Bruno Lopes Nunes[2]

[1] Department of Neurosurgery, Hospital Vila da Serra, Belo Horizonte, Minas Gerais, MG, Brazil
[2] Hospital Aroldo Tourinho, Montes Claros, MG, Brazil

Address for correspondence Marcelo José Silva Magalhães, MD, Departmento de Neurocirurgia, Hospital Vila da Serra, Belo Horizonte, Minas Gerais, MG, 39402-315, Brazil (e-mail: marcelo7779@yahoo.com.br).

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Abstract

Occipital neuralgia (ON) is an uncommon cause of headache, and it is characterized by a stabbing paroxysmal pain that radiates to the occipital region. The present study includes a review of the literature and a case report. The etiology of this pathology can vary from traumas, infections, compressions of nerves or vertebrae, skull base surgeries, to degenerative changes and congenital anomalies. However, most of the time, the etiology is considered idiopathic. The diagnosis is essentially clinical. However, it is crucial that other types of primary headache are excluded. The treatment for ON may be based on nerve blocks, medications or surgeries. Neurectomy of the second spinal nerve is among the surgical techniques available.

Keywords
- neurectomy
- occipital neuralgia
- headache

Resumo

A neuralgia occipital (NO) é uma causa incomum de cefaleia, caracterizada por dor paroxística, em pontada, que se irradia para a região occipital. Este estudo inclui uma revisão da literatura e o relato de um caso. A etiologia desta patologia pode variar de traumas, infecções, compressões de nervos ou vértebras, cirurgias de base de crânio, até alterações degenerativas e anomalias congênitas. Contudo, na maioria das vezes, a etiologia é considerada idiopática. O diagnóstico é essencialmente clínico. No entanto, é fundamental que sejam excluídos outros tipos de cefaleias primárias. O tratamento da NO pode se basear em bloqueios nervosos, medicamentos ou cirurgias. Entre as técnicas cirúrgicas disponíveis, encontra-se a neurectomia do segundo nervo espinhal.

Palavras-chave
- neurectomia
- neuralgia occipital
- cefaleia

Introduction

According to the International Headache Society, occipital neuralgia (ON) is defined as a paroxysmal, stabbing pain that radiates to the occipital region.[1] This pathology was described in 1821 by Beruto et al, being considered an uncommon cause of headache.[2] Most cases of ON are of idiopathic origin. However, external causes such as infections, skull base surgeries, vertebral compressions, traumas and congenital anomalies may be considered as etiology.[2] Due to the variety of signs and symptoms that may manifest in patients with ON, this pathology may be mistaken for other types of primary headache.[3] Nevertheless, the diagnosis is considered essentially clinical.[3] As options for treatment, non-ablative surgical treatment techniques may be used, among which neurostimulation of the greater occipital nerve and medullary stimulation can be

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highlighted. Finally, in the ablative surgical treatment group, radiofrequency rhizotomy, Lissauer tract injury surgery, neurectomy of the greater occipital nerve, and neurectomy of spinal nerve C2 are found.[3] The objective of the present study is to report the clinical case of a patient with ON whose treatment of choice was neurectomy of the second spinal nerve.

Case Report

A 46-year-old patient, female, married, presented with a history of occipital headache of strong intensity associated with pain in the bilateral mastoid process and with burning sensation in the occipital region for ~ 8 years. She also reported the presence of paroxysms, in which the pain was exacerbated, with the presence of a trigger point bilaterally in the occipital region. The neurological examination showed a positive Tinel sign in the occipital nerve bilaterally, but without focal alterations. The patient was initially treated with: 150 mg of pregabalin every 12 hours; 25 mg of amitriptyline every 24 hours; 40 mg of fluoxetine every 24 hours; 500 mg of dipyrone + 5 mg of promethazine + 10 mg adiphenine every 8 hours; and 60 mg of codeine every 8 hours, without substantial improvement of pain. The patient was submitted to an MRI, in which secondary causes of headache were discarded. A laboratory review revealed an erythrocyte sedimentation rate of 36 mm after 1 hour. The patient was instructed about the treatment possibilities for occipital nerve neuralgia. After the suspicion of the diagnosis of ON, the patient underwent an anesthetic blockade in the region of the greater occipital nerve, with immediate relief of the symptoms, which corroborated the diagnosis. The patient underwent open-air rhizotomy of the greater occipital nerve with remission of symptoms for ~ 3 years, but with new symptoms present, which led her to undergo a new surgical intervention. Under general anesthesia, a skin incision was performed in the posterior midline, in the region between the external bulge and the spinous process of C7.[4] By means of careful dissection of the planes, the second spinal nerve was identified, ~ 3 cm from the midline (►**Fig. 1**). No tissue alterations were found in the proximal path of this spinal nerve, as well as no neuromas. A C2 neurectomy was performed bilaterally, proximal to the sensory ganglion (nerve extirpation, including the sensory ganglion). The patient was reevaluated four weeks after surgery, and showed improvement of the pain, without the aforementioned symptoms, only with discomfort in the cervical region and bilateral hypoesthesia in the occipital region. The patient did not present complications due to the surgical procedure during the first postoperative month. In the outpatient visits occurring in the third and sixth months, the patient presented improvement of the pain, returning to her work activities without any compromise.

Discussion

The greater occipital nerve originates from posterior branches of the segments of spinal nerve C2, after its emergence from the sensory ganglion.[4,5] It travels a recurrent path toward the lower edge of the lower oblique muscle of the head, circumventing it,

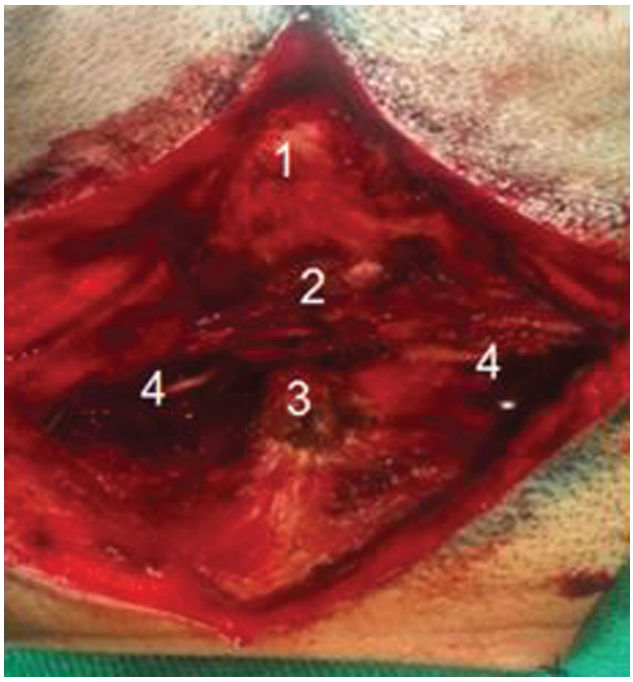

Fig. 1 Intraoperative photo of the occipital region. (**1**) External occipital bulge; (**2**) posterior tubercle of C1; (**3**) spinous process of C2; (**4**) second spinal nerve.

then travels in the upper direction, and, at this moment, is closely related to the semispinalis muscle of the head.[5] From this point on, it crosses the aforementioned muscle, maintaining its ascending path and establishing a new relationship when passing under the trapezius muscle. In its last portion, the nerve crosses the trapezius fibers, exteriorizing in the subcutaneous cell tissue.[5]

Occipital neuralgia is defined as pain in the occipital or cervical territory caused by stimulation and/or compression of the greater occipital nerve.[6] The pain has the characteristic of being unilateral and affecting the occipital region, which is innervated by the greater occipital nerve.[7] The patient can report it as pain in a burning or stabbing pattern, associated with the sensation of "shock," paresthesia, photophobia, nausea and vomiting.[8,9] Traumatic etiological, degenerative, oncologic or idiopathic factors are involved in most cases.[10]

The diagnosis of occipital neuralgia is essentially obtained by clinical examination, and no imaging exam presents good diagnosis efficiency.[11]

Different therapeutic approaches have been established for ON, which can be divided into conservative and surgical treatments. Among the options of conservative treatment are the use of cervical collar, analgesics, percutaneous nerve block, and use of botulinum toxin. As a non-ablative surgical treatment, neurostimulation of the greater occipital nerve is highlighted. Finally, in the ablative surgical treatment group are radiofrequency rhizotomy, Lissauer tract injury surgery, greater occipital nerve neurectomy, and neurectomy of spinal nerve C2.[2,12–14]

Conservative treatment of ON

The use of botulinum toxin has been shown to be efficient as therapy for ON, especially when associated with spinal cord

trauma.[8] It is noteworthy that it is already used in the treatment of other types of headache, such as migraine.[8] Infiltration of the greater occipital nerve can be used as diagnosis and treatment of this neuropathy. Infiltration can be performed with 0.5% bupivacaine, which may or may not be associated with methylprednisolone, in an anatomical point located 3 cm laterally and 2 cm inferiorly to the external occipital protuberance.[2,9] This nerve block showed positive results in the treatment of other types of headache. A hypothesis that justified good pain control is the proximity of the sensory neurons of the upper cervical region with neurons present in the trigeminal spinal nucleus.[11] Among the complications resulting from percutaneous nerve block due to its superficial location is intravascular injection.[1] In a study[2] involving 92 patients treated with percutaneous lidocaine and corticoid infiltration, good pain control was observed in 87% of the patients, with recurrence in 31.5% of them.[2]

Non-Ablative Surgical Treatment of ON

Neurostimulation in the greater occipital nerve has efficiency in the control of ON pain that ranges between 60% and 90%.[10] Among the complications of this surgical technique, we highlight the malposition of the electrodes and their migration, which can occur in 10% to 70% of the cases, with subsequent loss of stimulation and frequent indication of surgical revision.[10] A study[10] using an open technique for neurostimulation, despite the small sample, demonstrated that this technique is a safe, simple and effective modality to maintain direct contact between the electrode and the nerve trunk in a patient previously under anesthesia.[10] Medullary stimulation of C1-C4 has also been an option for the control of neuropathic pain in cases of ON.[15]

ON Ablative Surgical Treatment

In this treatment modality, open-pit rhizotomy of the greater occipital nerve is described for the treatment of ON.[5] Despite the small sample, one study[2] retrospectively evaluated 17 patients undergoing open-pit rhizotomy. No major complications were observed, but the potential risks are: infection, paralysis, cerebrospinal fluid fistula, and paresis of the trapezius muscle by the injury of the spinal accessory nerve.[2]

The surgery of the Lissauer tract injury to the C1-C3 dorsal roots has also been described as a treatment of ON. It is a modality of invasive surgical treatment, due to the need for cervical laminectomy. This surgery presents a higher complication rate when compared with other treatment modalities. Among the complications described are surgical wound infection, blood loss and spinal cord injury.[16]

Neurectomy of spinal nerve C2 is described as an option of ablative treatment of ON.[14] This technique has also been described in the arthrodesis of C1-C2, with the aim of minimizing pain or avoiding neuropathic pain postoperatively.[14] Among the reported complications are hypoesthesia and hyperesthesia of the occipital region.[14] It is important to emphasize that these described complications could be partly due to spinal cord trauma in this group of patients.[14]

Conclusion

Occipital neuralgia requires careful investigation to establish the diagnosis. Although there are several treatment methods, neurectomy of spinal nerve C2 may be an option for this pathology, having the advantage of being inexpensive. This technique requires anatomical knowledge for the proper recognition of the anatomical structures involved. Studies to better define the functional outcomes in the long-term are still necessary.

Conflict of Interests
The authors have no conflict of interests to declare.

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Compressive Neuropathy of the Ulnar Nerve in the Hypothenar Region by Lipoma: Case Report

Neuropatia compressiva do nervo ulnar na região hipotenar por lipoma: Relato de caso

Marcelo José Silva de Magalhães[1,3] Henrique Nunes Pereira Oliva[2] Bruno Lopes Nunes[3]

[1] Department of Neurosurgery, Vila da Serra Hospital, Belo Horizonte, Minas Gerais, MG, Brazil
[2] Faculdade Pitágoras de Montes Claros, Montes Claros, Minas Gerais, MG, Brazil
[3] Hospital Aroldo Tourinho, Montes Claros, Minas Gerais, MG, Brazil

Address for correspondence Marcelo José Silva de Magalhães, MD, MSc, Rua Francisco Versiane Ataide, 760, apartamento n° 1301., Montes Claros, MG, 39401-039, Brazil (e-mail: marcelo7779@yahoo.com.br).

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Abstract

Lipomas are well-defined tumors of the adipose tissue that often occur in the torso or the extremities of adult patients. These tumors usually develop painlessly and insidiously, but they may compress adjacent structures. The objective of the present study is to describe the case of a 68-year-old female patient with a giant lipoma located at the hypothenar region, with manifestation of compression of the common palmar digital nerves, the ulnar nerve, and the abductor muscle of the V finger. Regarding the symptoms, the patient felt moderate pain in the hypothenar region, with no Tinel sign, and no changes in the motor function or sensibility of the digits innervated by the ulnar nerve. Lipomas may present a varied range of histological characteristics, and malignant tumors may be a differential diagnosis. An imaging exam may aid in the diagnosis, which is confirmed by a histopathological study. For the present case, as recommended in the literature, a surgical procedure was performed for the resection of the tumor, which resulted in the control of the symptoms.

Keywords
- lipoma
- neuropathy
- ulnar nerve
- soft-tissue tumor

Resumo

Lipomas são tumores bem definidos, de tecido adiposo, presentes no tronco ou nas extremidades, e que ocorrem em pacientes adultos. Esses tumores geralmente se desenvolvem sem dor e de forma insidiosa, podendo, no entanto, comprimir estruturas adjacentes. O presente trabalho objetiva descrever o caso de uma paciente do sexo feminino, de 68 anos, com lipoma gigante localizado na face hipotenar, com manifestação de compressão dos nervos digitais palmares comuns, do nervo ulnar, e do músculo abdutor do V dedo. Como sintoma, ela apresentava quadro de dor moderada na face hipotenar, sem sinal de Tinel, e sem alterações da motricidade ou sensibilidade dos dígitos inervados pelo nervo ulnar. Os lipomas podem apresentar uma variada gama de características histológicas, e o diagnóstico diferencial pode incluir tumores malignos. O exame de imagem pode auxiliar no diagnóstico, o qual é confirmado por meio de estudo histopatológico. Para o presente caso, conforme preconizado na literatura, foi realizado um procedimento cirúrgico para a ressecção do tumor, que resultou no controle dos sintomas.

Palavras-chave
- lipoma
- neuropatia
- nervo ulnar
- tumor de partes moles

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Introduction

Lipomas usually present in adults as isolated tumors, with no presence of pain and with slow growth, more frequently in the thorax and extremities.[1] Lipomas are typically lobular and well-circumscribed, consisting of adipose tissue cells. They are separated from the surrounding adipose tissue by a thin fibered capsule.[1–3] Complementary exams are important because they assist in the determination of the type and location of the lipoma.[4] Lipomas can be classified as intramuscular, more frequent, and intermuscular, less frequent. In addition, intramuscular lipomas are divided into infiltrative and well-circumscribed.[2] Lipomas are commonly isolated and rarely multiple, presenting in varying shapes and sizes, being considered giant when its diameter exceeds 5 cm.[2,3]

The present work aims to describe the rare case of intermuscular lipoma in the left upper limb hypothenar region with ulnar nerve compression. We also provide a brief bibliographic review on the subjects involved in the studied case. For this, we used recent material available in virtual libraries, in addition to analyzing the medical records of the patient with unusual case presentation.

First, a review of the medical records of the patient was carried out to elaborate the case report. Then, a bibliographic research was conducted in the literature, seeking papers published in the last 47 years, in Portuguese, English and Spanish. The inclusion criteria of the researched studies were: the appropriate methodology applied, the update, and the similarity in some aspect with the present case. Exclusion criteria were: low relevance of some articles, nonapproach to the area of interest, and lack of important information.

The following descriptors were used: *lipoma*, *deepseated lipoma*, *lipomatous tumor*, *liposarcoma*, *lipoma of the extremities* and *intramuscular lipoma*. The digital libraries and open access electronic data sources accessed were the Scienific Electronic Library Online (SciELO) and the Virtual Health Library (VHL). The portal minhaUFMG was also used to access paid articles and a theme pertinent to this research.

Case Report

Patient M. A. O., 68 years old, female, housewife, sought outpatient medical care due to persistent headache. In her physical examination, it was detected a bulging area in the hypothenar region of the left upper limb extremity. No cyanosis or phlogistic signs were observed in the bulging region. The patient reported moderate-intensity pain in the left-hand thenar region, with irradiation to digits IV and V. No Tinel sign was found on the tumor lesion, and there were no sensitivity or motricity deficits identified.

A hand ultrasonography was requested, which evidenced an echogenic nodular well-defined image, without flow to the Doppler, encapsulated, measuring 24 mm, in the hypothenar region (►**Fig. 1**).

Upon presentation of the examination, we opted for hospitalization, which was preceded by the request for surgical risk exams. Local anesthesia was used with lidocaine without vasoconstrictor medication. As surgical access, we opted for linear incision in the V-digit axis, in the hypothenar region. Neurostimulation was performed during the procedure, in order to preserve the distal motor branches of the ulnar nerve. The lesion had a relatively well-delimited capsular structure, with a slightly oval shape and yellowish staining (►**Figs. 2a** and **2b**). After complete removal of the lesion, the common palmar digital nerves of the ulnar nerve and the abductor muscle of the V finger were identified. In the postoperative care, the patient presented pain improvement in the hypothenar region, and preservation of the motor function was confirmed.

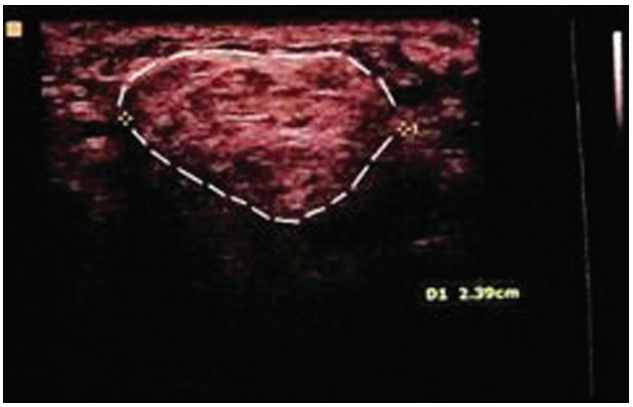

Fig. 1 Left hand ultrasonography evidencing a well-defined echogenic nodular image, without flow to the Doppler, encapsulated, measuring 2.39 mm, in the hypothenar region. Note the dashed lines skirting the lipoma.

The evaluation of the histopathologic sections of the tissue showed a benign neoplasia consisting of 12 g of unilocular mature adipose tissue (►**Fig. 3a**), with the presence of some muscular tissue (►**Fig. 3b**) and blood vessels (►**Fig. 3c**), interwoven by connective tissue with collagen fibers (►**Fig. 3d**) and fibroblasts, sometimes constituting a myxoid aspect. In addition, the absence of atypia and of signs of malignancy was observed.

Discussion

Lipoma is described as a benign adipose tumor, separated from the adjacent adipose tissue by a thin, well-circumscribed, well-defined film, that can occur in any region of

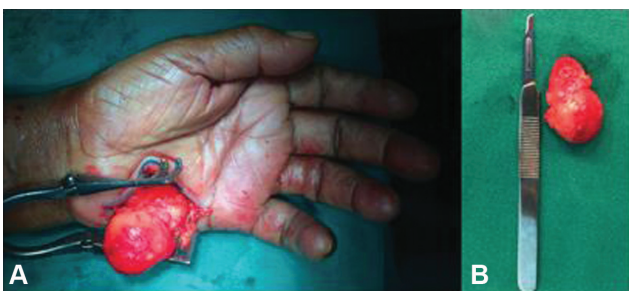

Fig. 2 (a) Perioperative photo showing voluminous lesion with a relatively well-delimited capsular structure, with slightly oval shape, and yellowish staining. (B) Photography of the lesion after its resection, confirming its oval and capsulated form.

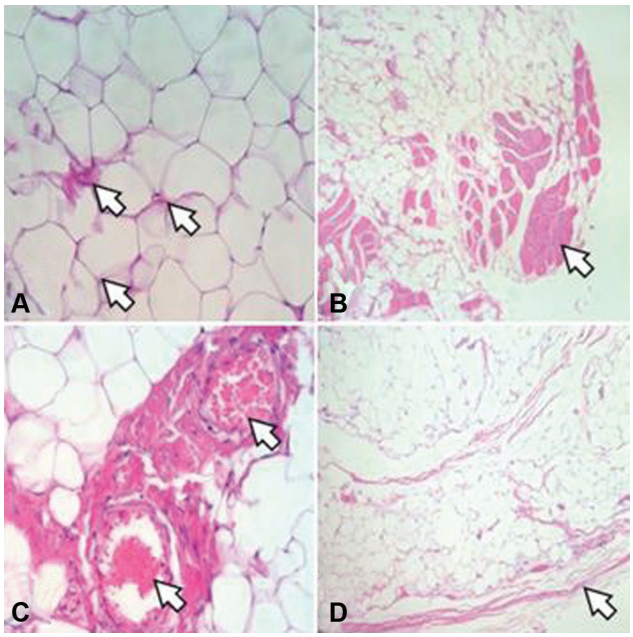

Fig. 3 Pictures of histological sections used for pathological study (Hematoxylin-eosin, enlargement x250). (A) Adipocytes with nuclear hyperchromatism (arrows) in medium to mature lipomatous background. (B) presence of muscle tissue (arrow). (C) blood vessels (arrows). (D) fibrated capsule (arrow).

the body. When poorly circumscribed and very infiltrated, it can be mistaken for well-differentiated liposarcoma. Well-differentiated liposarcomas, in turn, do not cause metastases, and they are considered of low degree of malignancy, but they present high recurrence rates.[5,6]

Among the main symptoms and signs of lipomas located in the hand are: local pain, paresis, hypoesthesia and local bulging by tumor growth in the underlying tissues (►**Table 1**).[5,6]

The best form of treatment for lipomatous tumors is complete surgical resection. Surgery should be performed after complementary exams that allow the surgeon to locate the tumor and plan their approach, since precipitated measures may contribute to the recurrence of the tumor if it is not properly removed during surgery.[5] Although surgery is commonly indicated, there is little consensus on which is the best surgical treatment for deep lipomatous tumors.[5,7] In the literature, in the case of suspicion of malignancy, the wide resection, with margins of ~ 1 cm, associated with annual follow-up with complementary imaging examination, is presented.[7]

Lipomas can be commonly visualized in ultrasound as distinct echogenic masses, having a varied reading; they may be: hyperechoic (20–52%), isoechoic (28–60%) or hypoechoic (20%).[8–10] When encapsulated, the capsule may be difficult to identify in this imaging examination mode.[11] In computed tomography (TC), lipomas are hypodense (~ 65,120 units

Table 1 Case reports of hand lipomas and summary of the main clinical findings[18–28]

Authors	Year	*n*	Tumor localization	Symptoms	Result of Surgical care
Rodriguez et al[18]	1970	15	12 in the hand and 3 on the wrist.	14 Asymptomatic tumors; 1 with pain; Limitation of movement.	Absence of recurrences and complications.
Ceballos et al[19]	2005	4	2 in the hand (1 in the palmar space, 1 in the thenar eminence) and 2 in the fingers.	Swelling and aesthetic discomfort.	Postoperatory period without intercurrences. No complications.
Kamath et al[20]	2006	1	Palmar region.	Swelling and discomfort for movement.	Recovery of the function and without recurrences.
Mohan et al[21]	2008	1	Between the thenar muscles.	Some difficulty in the seizure of objects (by important bulging).	Recovery of hand function.
Nadar et al[22]	2010	13	13 in the hand (5 on the back, 6 palmar, 1 on the wrist) and 1 on the forearm.	Bulging, pain, paresis and pruritus.	Remission of the feel-but. No recurrences were found.
Pagonis et al[23]	2011	1	Palmar region.	Compression of the median and ulnar nerves characteristic symptoms; Reduction of pulse movement amplitude; flexion limitation of the distal phalanx (I finger).	Remission of pain and recovery of motricity.
Chatterton et al[24]	2013	1	Thenar eminence.	Left thumb pain and swelling in the hand.	After excision of the lipoma, the patient went through a trapezectomy to treat arthritis of the carpometacarpal joints.
Ramirez et al[25]	2013	1	Third finger of the left hand.	Limitation of interphalangeal movement and digital paresthesia.	Complete motricity recovery and disappearance of paresthesia.
Radivojcevic et al[26]	2016	1	Ulnar region of the palm of the hand.	Pain and tingling in the fingers (IV and V).	Pain control.
Schmidt[27]	2017	1	Thenar region with extension to fingers.	Limitation of finger extension (I-III), paresthesia and pain.	Six months after surgery, function and sensitivity of the affected fingers were restored.
Ribeiro et al[28]	2017	1	Palmar region.	Paresthesia and pain in the fingers (I-III).	Reversal of the complaints of paresthesia and pain.

Hounsfield).[10,11] Magnetic resonance imaging (MRI) is often the modality of choice in the case of lipomas, not only to confirm diagnosis, which is usually suggested by ultrasound and TC, as well as better evaluating the atypical features suggesting the diagnosis of liposarcoma. In MRI, lipomas show hyperintense in sequences T1 and T2. Furthermore, magnetic resonance imaging allows a better definition of the anatomy adjacent to the tumor.[11]

The histological aspect of lipomas is characteristic, with predominant amount of adipose tissue, with adipocytes and negative image of fat. There is also the frequent presence of a fibered capsule involving the superficial region of the tumor.[1] Less often, blood vessels are observed, often more abundant in the surrounding muscle tissue than in the tumor region, being muscular capillaries.[1,8] In addition, there is a beam of muscular fibers in the middle of the tumor tissue.[8]

Despite the range of possibilities for histological tissue types, which may vary from benign lipomas to liposarcomas, it is estimated that lipomatous neoplasms comprise half of all soft tissue tumors.[1,5] Due to eventual similarities between benign and malignant affections, histological analyses have been conducted, and studies report some characteristics that allow the physician to assist in the correct diagnosis and in the efficient referral to appropriate treatment. Advanced age, tumor of high dimensions, and localization at the extremity instead of the thorax, for example, are characteristics that suggest an atypical lipomatous tumor, instead of a giant lipoma.[5]

There are other types of lesions of a benign character that can be considered as a differential diagnosis in the present case. The importance of mentioning these tumors found in the hand is due to the complexity of the affected structures and, consequently, the implications, such as loss of sensitivity, pain or even motor impairment of the limb.[12] In the group of lesions that could be differential diagnosis are: synovial cyst, giant cell tumor of the tendon sheath, cysts of epidermal inclusion, fibromas, Schwannomas and neurofibromas.[12–17]

A table was elaborated relating rare and representative lipoma cases (►**Table 1**). Most of them present mild symptomatology and/or presence of local bulging without symptoms. Rare cases developed with neurological manifestations, and all revealed resolution of the pain and motor symptoms when present.[18–28]

Conclusion

The presence of lipoma in the hand, besides being uncommon, may not generate early clinical manifestations until the tumor reaches a larger volume and compresses the adjacent structures. The surgical procedure corroborates the effectiveness reported in the literature, being the recommended conduct. Due to the existence of lesions of different natures that may compromise the hand, surgery also allows obtaining material for histopathological confirmation.

Conflict of Interests
The authors have no conflict of interests to declare.

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Anterior Communicating Artery Aneurysm Uncommon Hemorrhagic Presentation: Case Report

Aneurisma de Artéria Comunicante Anterior Apresentação Hemorragia Incomum: Relato de caso

Dan Zimelewicz Oberman[1] Nicollas Nunes Rabelo[2] Rafael Rego Barros[1] Othavio Lopes[1] Jorge Amorim[1]

[1] Department of Neurosurgery, Hospital Força Aérea do Galeão, Rio de Janeiro, RJ, Brazil
[2] Department of Neurosurgery, Santa Casa da Misericórdia de Ribeirão Preto, Ribeirão Preto, SP, Brazil

Address for correspondence Dan Zimelewicz Oberman, MD, Department of Neurosurgery, Hospital Força Aérea do Galeão, Rio de Janeiro, Rio de Janeiro, RJ 21941-353, Brazil (e-mail: danzoberman@gmail.com).

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Abstract

Intracranial aneurysm rupture causes subarachnoid hemorrhage in 80% of the cases, and it may be associated with intracerebral hemorrhage and/or intraventricular hemorrhage (IVH) in 34% and 17% of the patients, respectively. However, on rare occasions, aneurysm rupture may be present causing isolate intracerebral hemorrhage or IVH without subarachnoid hemorrhage.

We describe an unusual case of an anterior communicating aneurysm rupture presented with IVH, without subarachnoid hemorrhage.

Although isolated IVH is rare, aneurysm rupture is a possible condition. Patients presenting with head computed tomography revealing IVH without subarachnoid hemorrhage should be promptly investigated with contrasted image exam to identify and treat possible causes, even in the absence of subarachnoid hemorrhage.

Keywords

- aneurysm rupture
- anterior communicating aneurysm
- intraventricular hemorrhage
- subarachnoid hemorrhage

Resumo

A ruptura do aneurisma intracraniano causa hemorragia subaracnóidea em 80% dos casos, e pode estar associada a hemorragia intracerebral e / ou hemorragia intraventricular em 34% e 17% dos pacientes, respectivamente. No entanto, em raras ocasiões, a ruptura do aneurisma pode estar presente, causando hemorragia intracerebral isolada ou hemorragia intraventricular, sem hemorragia subaracnóidea. Descrevemos um caso incomum de ruptura de aneurisma de comunicação anterior apresentado com HIV, sem hemorragia subaracnóidea. Embora a hemorragia intraventricular isolada seja rara, a ruptura do aneurisma é uma condição possível. Pacientes que apresentam tomografia computadorizada revelando hemorragia intraventricular, sem hemorragia subaracnóidea devem ser prontamente investigados com exame de imagem contrastada para identificar e tratar possíveis causas, mesmo na ausência de hemorragia subaracnóidea.

Palavras-chave

- aneurisma roto
- aneurisma de artéria comunicante anterior
- hemorragia intraventricular
- hemorragia subaracnóidea

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Background

Subarachnoid hemorrhage (SAH) is a life-threatening condition and accounts for 5 to 10% of all strokes in the United States.[1] Vascular abnormalities are well documented causes of non-traumatic SAH, with aneurysm rupture being the most common.[2,3]

Subarachnoid hemorrhage from intracranial aneurysm (IA) rupture is a leading cause of stroke disability and death in young patients, with a high mortality rate (50%) and up to 50% of morbidity in survivors.[4]

Intracranial aneurysm rupture causes SAH in 80% of the patients,[5] and it may be associated to intracerebral hemorrhage (ICH) and/or intraventricular hemorrhage (IVH) in 34% and 17% of the patients, respectively.[6,7]

However, on infrequent occasions, aneurysm rupture may be present, causing isolate ICH or IVH without SAH. Thai et al[7] reported a rate of only 1.6% of patients presenting with isolated ICH and/or IVH, leading a poor prognosis.

We describe an unusual case of an anterior communicating aneurysm rupture presenting with IVH, without t SAH.

Case Presentation

A 70-year-old male with a history of hypertension, type 2 diabetes, dyslipidemia, and previous episode of ischemic stroke presented to the emergency department (ED) of our institution with nausea, fatigue, and mild headache. His medications included Aspirin (Bayer AG, Leverkusen, Germany), atorvastatin, and antihypertensive. The patient denied the use of tobacco, drugs, or family history of a brain aneurysm. After an initial assessment, he presented nausea, vomiting, and drowsiness. On physical examination, his only significant finding was a Glasgow coma scale (GCS) score of 13, without any focal deficits. His blood pressure was 185×100 mm Hg and returned to normal ($< 140 \times 90$ mm Hg) after treatment with sodium nitroprusside in 7 hours. The remainder of the neurological examination was unremarkable.

A cranial computed tomography (CT) scan was initially performed and revealed a small bleeding in the right occipital horn of the lateral ventricles, early hydrocephalus, and a large ectatic basilar artery measuring 9 mm in diameter (►**Fig. 1**). No parenchymal or SAH was present. The patient, therefore, underwent to computed tomography angiography (CTA), showing a saccular aneurysm measuring $\sim 5.8 \times 6.7 \times 4.2$ mm in diameter arising from the anterior communicating artery (Acom) (►**Fig. 2**).

Digital subtraction angiography (DSA) findings confirmed the diagnosis of Acom aneurysm (►**Fig. 3**). Due to the size, morphology, and location of the aneurysm, we opted for treating it with surgical clipping.

Three days after the diagnosis of a brain aneurysm and before treatment, the patient presented deterioration of the level of consciousness and when the GCS got to 8, he was intubated. Twenty-four hours later, the patient underwent surgery. The aneurysm was clipped, and an extraventricular drainage was placed. Four days after the surgery, the patient developed severe pneumonia, and, despite treatment, the patient died 2 weeks later of septic shock.

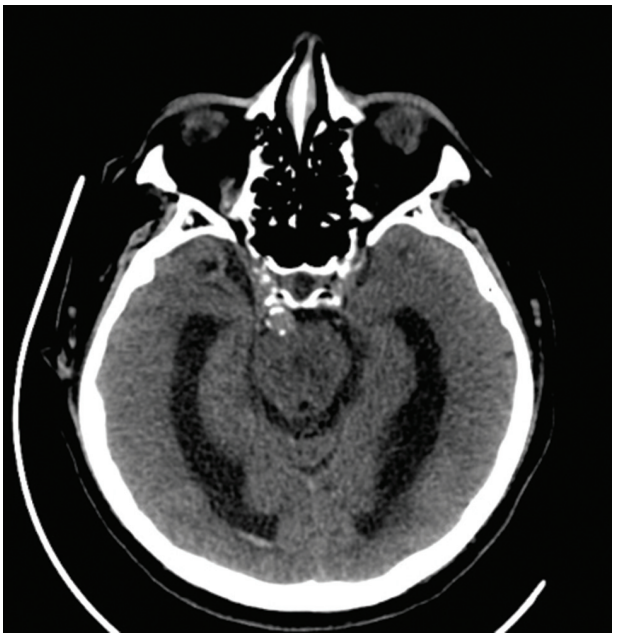

Fig. 1 Computed tomography scan without contrast. Axial plane (A), in which a slightly hyperdense lesion is seen in the occipital horn, without subarachnoid hemorrhage.

Discussion

Aneurysm rupture is more commonly seen as SAH in non-contrast CT scan, showing blood filling the subarachnoid cisterns. It can also be associated to subdural hematoma, ICH or IVH. Isolated IVH associated to aneurysm rupture is very

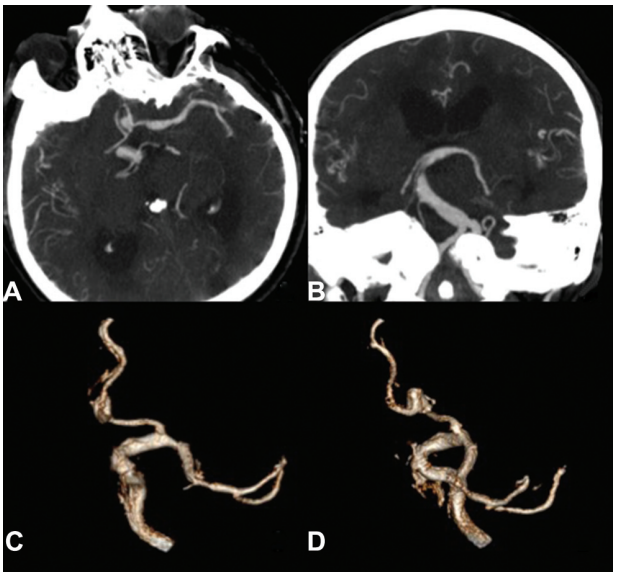

Fig. 2 Intracranial angiotomography. Axial plane (A) shows saccular dilation dependent on the anterior communicating artery with anterior-superior direction. Multiplane reconstruction of maximum intensity showing ectatic basilar trunk (B) and volumetric reconstructions (C and D) showing the anterior communicating aneurysm.

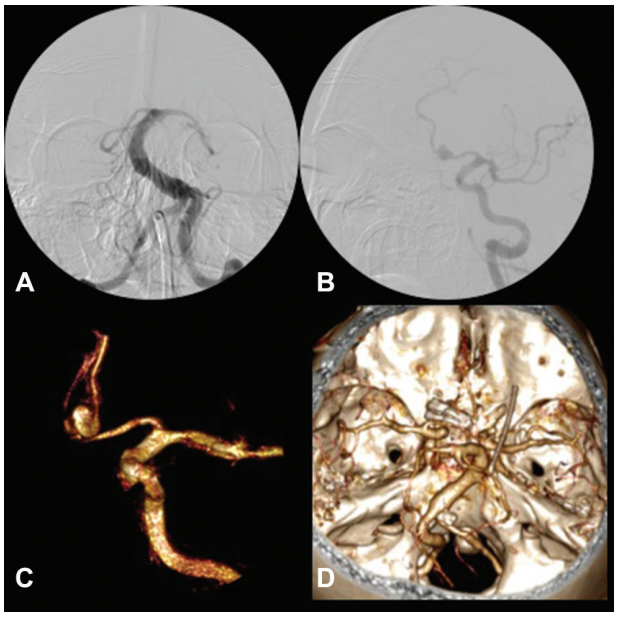

Fig. 3 Digital angiography oblique (A) and coronal (B) prior to surgical treatment. Volumetric reconstruction of digital angiography prior to surgery (C) and volume reconstruction of angiotomography (D) showing aneurysm clipped.

rare, and it is related to high morbidity and mortality, up to 40%.[8,9] Obstructive or communicating hydrocephalus occurs in 62% of patients, but only a third require extraventricular drainage.[9] The development of early hydrocephalus is an independent factor to poor prognosis.

Although many cases of isolated subdural hematoma or ICH associated to aneurysmal bleeding have been reported,[10,11] only few cases of pure IVH have been cited.[7,12,13] The two most common causes of isolated IVH are aneurysm rupture and arteriovenous malformation, while moyamoya disease and dural arteriovenous fistula are rarer.[8]

The features and location of the anuerysm can predispose to direct hemorrhage into the parenchyma or ventricular system. Considering the location of the Acom aneurysm in this patient and its anterior superior projection, occipital horn hemorrhage could be explained by direct rupture into the lamina terminalis and then into the ventricular system.[14]

The diagnosis of IVH without SAH is even more challenging depending on the timing when tests are performed. Computed tomography imaging is positive in over 90% of the cases of SAH on the day the hemorrhage occurs,[15] and the sensitivity of the test subsequently declines with the passing of time, reaching 50% by 5 to 7 days after the onset of symptoms.[16] Delay in performing a CT scan after bleeding might lead to false negative results and increase poor prognosis outcome. Thai et al[7] reported 6 patients had a sentinel event on average of 6.3 days before admission for head CT imaging. Due to the small amount of blood and an IVH without SAH in our patient, the diagnosis of aneurysm rupture and its treatment were delayed. This might have contributed to the decreased level of consciousness and poor prognosis of the patient.

Flint et al[8] found that catheter angiogram was performed in 52% of the cases of IVH, with the identification of the bleeding source in 56%. A causative lesion was identified in 44% of the patients, who were treated accordingly. The routine catheter angiography in IVH is warranted to identify potentially treatable causes of hemorrhage. This has direct implications in the management of these patients. Therefore, careful radiological examination is necessary to evaluate cases of isolated ventricle hemorrhage.

Conclusions

This is a case of IVH without SAH caused by rupture of an Acom aneurysm. After extensive neuroimaging investigation, an underlying aneurysm was discovered. Although isolated IVH is rare, aneurysm rupture is a possible condition. Patients presenting with head CT revealing IVH without SAH should be promptly investigated with contrasted image exam to identify and treat possible causes, even in the absence of SAH.

Conflict of Interests

The authors have no conflict of interests to declare.

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Does A Vertebra Fracture Cause A Tumor? A Dorsolumbar Paraspinal Desmoid Tumor: Case Report

Uma fratura de vértebra causa um tumor? Tumor desmoide paraespinhal dorsolombar: Relato de caso

Zeki Serdar Ataizi[1] Gulay Simsek[2] Fulya Colak Ataizi[3] Serdar Ercan[1]

[1] Department of Neurosurgery, Yunus Emre State Hospital, Eskisehir, Turkey
[2] Department of Pathology, Yunus Emre State Hospital, Eskisehir, Turkey
[3] Department of Radiation Oncology, Yunus Emre State Hospital, Eskisehir, Turkey

Address for correspondence Serdar Ercan, MD, Department of Neurosurgery, Yunus Emre State Hospital, Uluonder Mah. Salih Bozok Cd No:23, Tepebasi, Eskisehir, 26190, Turkey (e-mail: srdrercn@gmail.com).

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Abstract

Extraabdominal desmoid tumors are uncommon soft-tissue tumors. The etiology of the tumor is still unclear. Injury is one of the etiological factors of soft-tissue tumors. A 41-year-old female patient who had a traumatic vertebral body fracture on the thoracic spine was treated conservatively. Two and a half years later, she presented a painful, palpable swelling on the thoracolumbar region. In the present report, was discuss the patient, who underwent a surgery to remove the desmoid tumor (aggressive fibromatosis), within the context of the current literature. The literature on desmoid tumor caused by a trauma is rare. This is the first case that demonstrates an extraabdominal desmoid tumor following a spinal fracture. The swelling on the region of the trauma must be examined carefully and desmoid tumor must be kept in mind as a possible diagnosis.

Keywords
- trauma
- vertebral fracture
- extra-abdominal desmoid tumor

Resumo

Tumores desmoides extra-abdominais são tumores incomuns de tecidos moles. A etiologia do tumor ainda não está clara. A lesão é um dos fatores etiológicos dos tumores de tecidos moles. Uma paciente do sexo feminino, com 41 anos de idade, com fratura traumática do corpo vertebral na coluna torácica, foi tratada de forma conservadora. Dois anos e meio depois, ela apresentou um inchaço doloroso e palpável na região toracolombar. No presente relato, ocaso da paciente, que foi submetida à cirurgia para extração do tumor desmoide (fibromatose agressiva), foi discutido no contexto da literatura atual. A literatura sobre tumor desmoide causado por trauma é rara. Este é o primeiro caso que apresenta um tumor desmoide extraabdominal após fratura da coluna vertebral. O inchaço na região do trauma deve ser examinado cuidadosamente, e o tumor desmoide deve ser considerado como possível diagnóstico.

Palavras-chave
- trauma
- fratura vertebral
- tumor desmoide extra-abdominal

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Introduction

Desmoid tumors are slow-growing tumors of benign nature, which originate in the muscle, connective tissue, fascia, and aponeurosis and are rarely malignant but frequently invasive.[1] Desmoid tumors, which present a prevalence of 0.03% among all tumors, were first described by McFarlane in 1832.[2,3] The etiology of a large number of soft-tissue tumors is still unknown today. Several cases of desmoid tumors developing in the same region of previous isolated trauma are present in the literature.[4–6]

Case Report

A 41-year-old female patient presented with a growing painful mass on the left side, extending from the medial to the lower back region. The patient had fallen 2.5 years prior to developing the painful mass, and subsequently had a stable compression fracture of the 11th thoracic (T11) vertebral body (►**Fig. 1a**). The fracture was treated conservatively. No skin injuries, including laceration, abrasion, or ecchymosis, were observed on the thoracic and lumbar spine regions. However, 6 months later, an initially small swelling in the neighborhood of the T11 vertebral body occurred, which grew over time. Physical examination revealed a gross mass lesion in the left dorsolumbar region, which could be clearly seen through the skin and was not mobile due to the adherence of surrounding tissue. With the exception of the mass, physical examination of the thorax and abdomen went well, and the results of the laboratory evaluation were within normal range. Magnetic resonance imaging of the patient revealed a $10 \times 6 \times 7$ cm soft-tissue mass at the T11 and L2 levels, which was hypointense in T1 sequence, hyperintense in T2 sequence, and heterogeneous after administration of the contrast agent (►**Fig. 1b**). Destruction of the spinous process due to the tumor could not be detected. The patient underwent a surgical operation, and the mass was removed completely. Due to the extensive invasion and the insufficient cleavage, an extended surgical resection was performed. The histopathological evaluation of the mass revealed a desmoid tumor (►**Fig. 2**). The patient has been followed-up for 2 years with no recurrence.

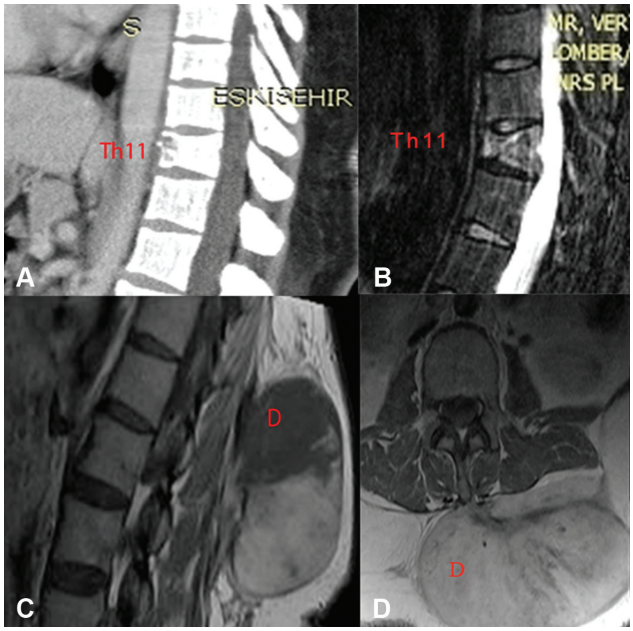

Fig. 1 (**A**) Computed tomography (CT) scan of spine after trauma. T11: compression fracture at the T11 level (**B**) Magnetic resonance imaging scan of T11 demonstrates the fracture-related edema. (**C**) Sagittal scan of vertebral body after 6 months. A desmoid tumor ($10 \times 6 \times 7$ cm) can be observed at T11 and L2 levels. (**D**) Axillary scan of vertebral body and desmoid tumor. Abbreviations: T: thoracic vertebra, D: desmoid tumor.

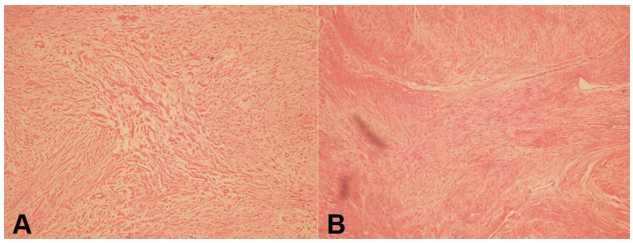

Fig. 2 (**A**) Layers of tumor cells with pale eosinophilic cytoplasm and spindle shaped. (**B**) The collagen in desmoid tumor may be keloidal.

Discussion

Desmoid tumors are benign and locally aggressive invasive tumors that originate in the muscle, connective tissue, fascia, and aponeurosis.[4] The cause of desmoid tumors is still unknown. However, these tumors are associated with genetic abnormalities, such as familial adenomatous polyposis and Gardner syndrome as well as with sex hormones in the pregnancy and postpregnancy periods, and also with isolated and surgical traumas and invasive interventions. Approximately 25% of the patients have had a previous trauma. In addition to desmoid tumors, lipomas and lymphomas are also mentioned among the tumors that are associated with trauma. The incidence of desmoid tumors is 0.03% among all tumors.[4,7–9] However, the percentages of patients with familial adenomatous polyposis and Gardner syndrome are 32% and 29%, respectively.[10] Desmoid tumors are more common among females.[11] Reitamo et al reported that 80% of desmoid tumors are seen in women, and 50% of them occur in the 3rd and 5th decades of life.[12] The majority of the cases are reported more frequently between adolescence and 40 years of age.[1]

Currently, there is no definitive and effective treatment for these tumors, which are usually treated by extensive surgical excision. The distant metastases are extremely rare,[13] although local recurrence can happen, and conservative therapeutic management must be ensured. Asymptomatic patients are treated conservatively. Radiotherapy, cytostatic agents, hormonal drugs, non-steroidal antiinflammatory drugs and antiviral drugs are alternative methods that can be used in the treatment of desmoid tumors.[2,14,15]

The association between the tumor and trauma is unclear. Ewing[16] has listed a criteria of tumor formation and suggested that trauma is theoretically a possible factor in tumor development. In light of this criteria, the case described in the present study exhibits a similar perspective of cause-effect

relationship, localization and neighborhood, histological type of the tumor, and latent period between the events. The uncontrolled cell proliferation in the region of the trauma explains the formation of desmoid tumors better.[4] In the case presently discussed, the radiological and physical examination was performed in the early period of the trauma and accepted normal, except for the T11 vertebral body fracture. The tumor supports the theory about tumor formation in the latent period of injury, occurring in the neighborhood of the injury zone. Previous studies have presented cases in which the trauma had occurred directly to the soft tissue. However, the trauma was indirect in our case. The valuable feature of our case is that it is the first reported case of a desmoid tumor formation developed after compression fracture of a vertebral body without any direct trauma to the skin.

In conclusion, in the presence of a rapid growth of the skin on the trauma region, desmoid tumors should be considered in the differential diagnosis of conservatively treated compression fracture. Despite the availability of a wide variety of treatment options, the best option for the treatment of desmoid tumors will be radical resection with surgery.

Ethical Publication Statement

The authors confirm that they have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

Authors' contributions

Zeki Serdar Ataizi and Serdar Ercan performed the clinical examination and surgical treatment, and were major contributors in writing the manuscript. Gulay Simsek and Fulya Ataizi performed the histopathological examination of the tumor, and analyzed and interpreted the data of the patient.

Conflict of Interests

The authors have no conflict of interests to declare.

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