Cervical Intramedullary Cavernoma in Children: case report and literature review

Cavernoma Intramedular Cervical em Criança: relato de caso e revisão da literatura

Case presentation: A 10-year-old male patient was admitted due to posterior cervicalgia, vomiting and progressive generalized weakness. Physical examination showed difficulty in ambulation and tetraparesis. Gadolinium-loaded magnetic resonance imaging (MRI) of the cervical medulla, showed a poorly contrasted mass with a hemorrhagic center. We undertook a posterior cervicotomity (C1-C3). Post-operative evolution was satisfactory with complete regression of the deficits 90 days after the surgery. Control MRI at intervals of 1 and 4 years confirmed absence of the tumor. Discussion: Cavernomas are vascular malformations, consisting of coarsely dilated vascular channels and coated by a single layer of endothelial cells, devoid of endothelium and myothelium. They represent only 5-12% of all vascular pathologies of the medulla, with only 10% affecting the pediatric population. There are only thirty cases of pediatric intramedullary spinal cavernomas in the literature, with predominance among males (2.1:1). Thoracic and cervical spinal cavernomas consist in 55% and 45% of the cases, respectively. In six cases (20%) cavernomatous lesions were associated with synchonic intracranial cavernoma. Among the reported cases, only one had poor evolution after surgery, whereas six patients persisted with prior symptoms. Conclusion: Spinal intramedullary cavernomas are rare entities, especially in the pediatric population, and are treated with surgery which improves prior neurologic deficits, besides preventing rebleeding.

Keywords: Cavernous angioma; Spinal intramedullary lesion; Child

RESUMO

A apresentação do caso: Paciente masculino, 10 anos, foi hospitalizado com quadro de dor em região cervical posterior, vômitos e fraqueza generalizada progressiva. O exame neurológico evidenciou dificuldade de deambulação e tetraparesia. A ressonância magnética (RM) da coluna cervical revelou lesão intramedular cervical em C1-C2 de intensidade mista, pouco realçada ao gadolíneo e com centro hemorrágico. Diante deste contexto, decidiu-se por abordagem cirúrgica por via cervical posterior à esquerda (C1-C3). O paciente evoluiu com regressão completa dos déficits 90 dias após a cirurgia. A RM de controle com 1 e 4 anos de intervalo não revelou resíduo ou recidiva tumoral. Discussão: Cavernomas são malformações vasculares, constituídas por canais vasculares grosseiramente dilatados e revestidos por uma única camada de células endoteliais, desprovidos de endotélio e miofibras. Representam apenas 5-12% de todas as patologias vasculares da medula, com somente 10% afetando a população pediátrica. Na literatura, foram publicados 30 casos de cavernomas intramedulares em crianças até 2018. Ao contrário da população adulta, o sexo masculino foi o mais acometido (2:1 casos). O acometimento da medula torácica e cervical foi de, respectivamente, 55% e 45% dos casos. Em 6 casos (20%), foram encontradas lesões cavernomatosas associadas na região intracraniana. Sobre o desfecho cirúrgico, houve piora clínica em apenas 1 caso, persistência dos sintomas pré-operatórios em 6 casos, e melhora clínica nos demais 23 casos. Conclusão: O...
Cavernomas are vascular malformations, characterized by grossly dilated, low-flow capillaries composed of only one layer of endothelial cells. They are mainly found intracranially and correspond to around 10% of all cerebrovascular malformations. Spinal ones are less common and constitute about 5% of the total. In children, spinal cord cavernomas represent less than 2% of non-traumatic spinal cord lesions, being relatively rare in the pediatric population¹.

On magnetic resonance imaging (MRI), cavernomatous lesions have a reticulated and mixed-intensity appearance. In the T2-weighted sequence there may be a hyperintense reticulated nucleus with a hypointense halo due to multiple bleeds and hemosiderin deposition without perilesional edema. This imaging pattern is pathognomonic and may appear as incidental findings in asymptomatic patients who underwent radiological examinations for other reasons. Computed tomography (CT) may show an isodense aspect with increased nonspecific density due to bleeding or microcalcifications²,³.

The clinical manifestations cover a wide spectrum and depend on the location, size and presence of associated bleeding. Genetic factors are important in the etiology of the disease and are associated with genes that have a role in angiogenesis in the neural tissue, justifying the formation of multiple cavernomas and a high rate of recurrence³. However, there are also new cases not associated with a positive family history⁴. Cavernomas have a higher tendency to bleed when compared to other vascular malformations of the central nervous system, causing acute symptoms and progressive neurological deterioration due to sudden bleeding. The risk increases immediately after a hemorrhagic episode and decreases over time. In addition, this risk is associated with other factors such as number of lesions and male gender²,³.

This study aims to report a case of cervical intramedullary cavernoma in a 10-year-old male patient, as well as to analyze the cases described previously and to review the literature.

INTRODUCTION

A 10-year-old male patient was hospitalized with pain in the posterior cervical region, vomiting and progressive generalized weakness. Neurological examination revealed difficulty walking and tetraparesis. Cervical spine MRI revealed a cervical intramedullary lesion at the C1-C2 level of mixed intensity (Figure 1), not much enhanced by the gadolinium, with a hemorrhagic center. A posterior surgical approach with a left hemilaminectomy (C1-C3 level) was decided upon.

CASE PRESENTATION
Case Report

Figure 1. T1 (A) and T2-weighted (B) sagittal view of the cervical spine revealing intramedullary cervical lesion at C1-C2 level with mixed-intensity and hemorrhagic signal.

The spinal incision was performed on the posterolateral side of C2, where hemosideritic tissue was observed. Complete resection of the richly vascularized intra-axial lesion surrounded by numerous abnormal vessels was allowed. The histopathological examination was compatible with the diagnosis of cavernoma. The patient progressed with complete symptoms regression 90 days after surgery. Control MRI at 1 and 4-year intervals revealed no residual tumor or local recurrence (Figure 2).

Figure 2. Postoperative Sagittal T1-weighted MRI image of the cervical spine showing complete removal of the lesion.

LITERATURE REVIEW AND DISCUSSION

Thirty cases of pediatric intramedullary spinal cavernoma were found in literature, published until 2018. Included in this study were patients with 18 years of age or less who had a confirmed diagnosis of intramedullary spinal cavernoma and who had undergone surgical resection. Due to the small number of published cases, a standard epidemiological profile cannot be defined with precision, but it is already possible to identify important differences between the adult and pediatric profiles. In the adult population, the female sex is more affected (male / female ratio varies from 1:1.1 to 1:2 in the literature), while in the pediatric population it is more common in males, the ratio being 2:1. Of the 31 cases, 25 (80%) were in the range of 10 to 18 years, 5 (16%) between 1 and 10 years and only 1 (3%) younger than 1 year. Such an uneven distribution raises the hypothesis that hormonal influences during puberty lead to an increased risk of bleeding. In adults the incidence of intramedullary cavernomas is apparently higher in the thoracic spine, whereas in pediatric cases they are more evenly distributed, with 17 (55%) cervical and 14 (45%) thoracic cases in our review. As to the presence of another cavernoma in the neuraxis associated with the intramedullary cavernoma, this multiplicity was observed in 6 (24%) of the cases (all of the second cavernomas were found in intracranial locations); 19 (76%) did not present multiplicity; and in 6 cases there was no information available on the multiplicity of the cavernoma (Table 1). The surgical outcome was mostly satisfactory, as can be observed in Table 2.

The most common clinical presentation in children is acute neurological deficit, with incidence between 50 and 100% of cases, in contrast to adult cases in which the most common manifestation is slowly progressive neurological deficit. This presentation is quite varied depending on the size and location of the lesion and results from changes in blood flow caused by bleeding, micro bleeding or thrombotic processes that occur in the cavernomatous vascular tissue adjacent to the neural tissue. Rapidly progressive sensory and motor deficits in the limbs, paraparesis, hemiparesis, tetraparesis, lumbar pain,
progressive muscular atrophy, Brown-Séquard syndrome and autonomic symptoms (most commonly urinary retention) are examples of clinical manifestations reported in pediatric cases\textsuperscript{1,5,7,10,11}.

The cavernomas are angiographically occult malformations, however a small number of feeding arteries and drainage veins may sometimes be observed\textsuperscript{12}. On tomographic images, variable and non-specific findings are seen in the axial plane. It is possible to see an enlargement of the spinal parenchyma, in addition to acute hemorrhaging in some cases\textsuperscript{13}. The use of contrast medium may enhance the lesion\textsuperscript{13}. On MRI, the cavernomas present a characteristic image, in which a signal intensity around the lesion in T1 and T2 due to hemosiderin deposits coming from the previous hemorrhage\textsuperscript{12,13}. Gradient-echo sequences demonstrate low signal intensity within the hemorrhage\textsuperscript{14}

The natural history of intramedullary spinal cavernomas is not completely understood and in the pediatric population it is even more obscure. Current recommendations are based on case reports, small case series and reviews\textsuperscript{12}. The current consensus is that symptomatic cases should be treated surgically, while asymptomatic patients should be monitored clinically and possibly with MRI. Patients with mild symptoms should have their treatment indication based on surgical accessibility\textsuperscript{11}. The goal of the surgery should be the complete removal of the lesion. The postoperative result depends on some variables, such as duration of symptoms, type of

| Case | Author | Sex | Age | Site | Multiplicity | Outcome |
|------|--------|-----|-----|------|--------------|---------|
| 1    | Odem et al. (1957)\textsuperscript{15} | F   | 12  | C3-C4| No            | Improved|
| 2    | McCormick et al. (1988)\textsuperscript{16} | M   | 15  | T8-T9| No            | Unchanged|
| 3    | Scott et al. (1992)\textsuperscript{17}  | M   | 13  | C   | No            | Improved|
| 4    | Anson & Spezler (1993)\textsuperscript{9} | F   | 14  | C2  | No            | Improved|
| 5    | Anson & Spezler (1993)\textsuperscript{9} | F   | 17  | C2-C7| No            | Improved|
| 6    | Furuya et al. (1996)\textsuperscript{13} | F   | 14  | T10 | No            | Improved|
| 7    | Takahashi et al. (1996)\textsuperscript{18} | M   | 12  | T11 | -             | Improved|
| 8    | Tu et al. (1999)\textsuperscript{18}     | M   | 7   | C3-C4| -             | Improved|
| 9    | Tu et al. (1999)\textsuperscript{18}     | M   | 18  | C7-T1| -             | Improved|
| 10   | Deutsch et al. (2000, 2001)\textsuperscript{20} | M   | 8   | C2  | Yes           | Improved|
| 11   | Deutsch et al. (2000, 2001)\textsuperscript{20} | M   | 13  | T4  | Yes           | Improved|
| 12   | Nagib & O’Fallon (2002)\textsuperscript{21} | M   | 10  | T4  | No            | Improved|
| 13   | Nagib & O’Fallon (2002)\textsuperscript{21} | F   | 16  | T4  | No            | Improved|
| 14   | Bakir et al. (2006)\textsuperscript{22}  | M   | 14  | C6-C7| No           | Improved|
| 15   | Jallo et al. (2006)\textsuperscript{11/7} | M   | 8   | C2  | Yes           | Unchanged|
| 16   | Jallo et al. (2006)\textsuperscript{11/7} | M   | 13  | T4  | Yes           | Unchanged|
| 17   | Jallo et al. (2006)\textsuperscript{11/7} | M   | 18  | C7-T1| No           | Worse|
| 18   | Jallo et al. (2006)\textsuperscript{11/7} | M   | 18  | T1-T2| No           | Improved|
| 19   | Santoro et al. (2007)\textsuperscript{23} | M   | 11  | C1  | Yes           | Improved|
| 20   | Kharkar et al. (2007)\textsuperscript{22} | M   | 3   | T7-T8| -            | Unchanged|
| 21   | Noudel et al. (2008)\textsuperscript{24} | F   | 12  | T11 | -            | Improved|
| 22   | Miyoshi et al. (2010)\textsuperscript{25} | F   | 0.16| C4  | No            | Improved|
| 23   | Comips et al. (2010)\textsuperscript{26} | F   | 7   | C5  | No            | Improved|
| 24   | Comips et al. (2010)\textsuperscript{26} | F   | 10  | T9-T10| No        | Improved|
| 25   | Kodeeswaran et al. (2016)\textsuperscript{27} | M   | 15  | C5  | Yes           | Improved|
| 26   | Khabibiar et al. (2010)\textsuperscript{28} | M   | 16  | T12 | No            | Improved|
| 27   | Wei et al. (2014)\textsuperscript{29}    | M   | 15  | C3-C4| No            | Improved|
| 28   | Narayan et al. (2003)\textsuperscript{27} | M   | 17  | T7-T9| No            | Unchanged|
| 29   | Varoglu et al. (2008)\textsuperscript{30} | M   | 12  | C2-C7| No            | Improved|
| 30   | Sundlackoglu et al. (2003)\textsuperscript{27} | F   | 17  | T7-T8| -            | Improved|
| 31   | Authors’ case                        | M   | 10  | C1-C2| No            | Improved|
symptoms, location of the lesion and preoperative functional status. Longer duration of symptoms, sensory deficits, ventral location of the lesion and worse preoperative functional status are associated with worse surgical outcome.4.

The pediatric intramedullary spinal cavernoma is a rare entity, with few reported cases. After reviewing the literature, we concluded that there is a predilection for males and for the age group from 10 to 18 years. Its incidence in the thoracic and cervical spinal cord is similar and approximately one-quarter of the patients have multiple cavernomas. Clinically, the presentation is more often acute. Cavernomas can be effectively identified by MRI; and considered as the golden standard examination. When symptomatic, these lesions are treatable by surgical resection with a favorable outcome in most of the cases.

Table 2. Surgical Outcome

| Number of Cases | Surgical Outcome |
|-----------------|------------------|
| 1 (3%)          | Worse            |
| 6 (19%)         | Unchanged        |
| 24 (78%)        | Improved         |

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Case Report

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