Rare Case of Young Patient with Intraventricular Angiomatous Meningioma

Caso raro de paciente jovem com meningioma angiomatoso intraventricular

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Abstract
Pediatric meningiomas are rare and account for only 2.2% of the central nervous system (CNS) tumors. In this age group, they are more frequently located in atypical sites, such as, mainly, the ventricular system, with a frequency of 8.8 to 13.6%. Adding this to the fact that the angiomatous subtype constitutes only 2.1% of all meningiomas, the rarity of the case reported here is corroborated. We report a 17-year-old female patient diagnosed with intraventricular angiomatous meningioma; she underwent surgical resection of the tumor in the body and frontal horn of the right lateral ventricle, and there were no neurological sequelae. With a follow-up of 26 months, there was no recurrence and the patient had clinical stability. Intraventricular tumors usually have slow growth and reach a considerable size until they cause symptoms and then are diagnosed. In addition, the deep location of the tumor and its proximity to eloquent areas make these tumors a neurosurgical challenge. The angiomatous subtype, due to the presence of hypervascularization (consisting of > 50% of vascular components), may, in some cases, hinder surgical resection as well as be erroneously diagnosed. However, surgical treatment aimed at total resection of the lesion remains the conduct of choice in the case reported here, especially in patients in the first two decades of life, in which the use of radiation is avoided. Specifically when it comes to the surgery, we chose a transcicalsal approach that allows a good transoperative visualization of the lesion when located in the body and frontal horn of the lateral ventricle.

Resumo
Meningiomas pediátricos são raros, representando apenas 2,2% dos tumores do sistema nervoso central (SNC). Em tal faixa etária, localizam-se com maior frequência em sítios atípicos, como, principalmente, o sistema ventricular – com frequência de 8,8
Introduction

Meningiomas have a progressively higher incidence with increasing age, with a mean age of presentation of 65 years old. Thus, they constitute the most frequently reported tumors of the central nervous system (CNS) in adulthood.¹ Children and adolescents cases are rare, representing 2.2 to 2.6% of CNS tumors.¹,² In this age group, they are more frequently located in unusual sites, such as in the ventricular system.³ Such intraventricular meningiomas have the particularities of being slow-growing and reaching a considerable size until they become symptomatic.⁵ In addition, the deep localization and the relationship with the underlying eloquent areas make tumor resection a neurosurgical challenge.⁵,⁶ In view of this, and that the angiomatous subtype – defined for presenting > 50% of the vascular components at the microscopic analysis – constitutes only 2.1% of all meningiomas, it is credited to the case reported herein.⁷ We are conscious of the fact that this is possibly the first report of a patient in the first 2 decades of life with angiomatous meningioma in the intraventricular site.

In this report, we aim to expose our neurosurgical experience in a case with rare variants and to conduct a review of the literature on the main aspects that we deem necessary to support our conduct.

Case Description

A 17-year-old female patient, previously healthy, presented orbital headache for 3 months, followed by blurred and double vision. On physical examination, convergent strabismus by right lateral rectus muscle paresis was found. In the other cranial pairs, sensitivity, strength and reflexes were preserved and there were no meningeal signs. It was then performed an investigation with cranial magnetic resonance imaging (MRI), which evidenced an expansive lesion in the frontal horn of the right ventricle, directly ahead of the foramen of Monro, with dimensions of 2.0 × 1.3 × 1.8 cm (AP x L x H) in the larger diameter sections. The lesion presented moderate hyperintensity with small hypointense foci in the T2 weighted images. On T1, it was isointense, and after the contrast showed intense and homogeneous impregnation, except for the same hypointense T2-weighted spots. There were no signs of dilatation of the supratentorial ventricular system (►Fig. 1 A-D). Such radiological features suggested the diagnosis of intraventricular meningioma.
meningioma, with calcification areas. It was considered an occasional finding, since the topography was not compatible with the presentation symptoms.

Surgical treatment was indicated, and this was performed at the same hospitalization, due to the risk of acute hydrocephaly. The approach was performed by the transcalous approach, with the following steps: dorsal decubitus and head in neutral position; bicoronal incision; right frontal paramedian craniotomy, with lateral extension of 5.5 cm from the midline and 5.5 cm from the coronary suture to the front; opening of the dura mater in “Ç” format based on the midline; under microscopy, dissection of the interhemispheric fissure and removal of the frontal lobe with positioning of the fixed spatula in Leyla support on the medial surface; identification of the rotating of the cingulate and pericalous arteries; 1.2 cm callosotomy starting from the transition between the knee and the corpus callosus body; identified the vegetating greyish lesion inside the right frontal horn, which presented softened consistency, aspirable after coagulation, and vascularized; resection of the lesion through coagulation and aspiration and by fragments, providing small residue adhered to the ependym of the thalamocauded droppings. Postoperative without intercurrences and absence of new deficits, remaining diplopia and strabismus. The histopathologic analysis and the immunohistochemical profile demonstrated extensive vascularization and low mitotic index (Ki67 < 2%). In addition, the results verified are: epithelial membrane antigen (EMA), positive; cytokeratin (CK), negative; progesterone receptor, negative; and glial fibrillary acid protein (GFAP), positive (Fig. 2 A-B). Thus, the diagnosis of angiomatous meningioma (World Health Organization [WHO] grade I) is confirmed. The postoperative MRI showed small residual focus along the striated thalamus (Fig. 3 A-D and 4 A-D). After 6 months, the patient underwent strabismus correction with an ophthalmologist at another institution. Currently, with ~26 months of follow-up, she is asymptomatic and without evidence of recurrence of the residual lesion.

Fig. 2 (A) Hematoxylin and eosin staining, 200 x magnification: Image evidencing histopathological features of meningioma and significant vascular component. (B) Immunohistochemistry, Magnification - Analysis of 400 times: tumor cells presenting positive for epithelial membrane antigen.

Fig. 3 Postoperative magnetic resonance imaging: T1 with gadolinium, axial planes (A and B) and Coronals (C and D). Small residual focus near the foramen of Monro, adhered to the striate thalamus vein (identified in the transoperatory).

Fig. 4 Postoperative magnetic resonance imaging: T2 axial (A and B) and coronal (C and D). Small residual focus at the level of the foramen of Monro (arrows).
**Discussion**

Meningiomas are tumors that predominate in the 5th and 6th decades of life, having an average age of presentation of 65 years old.\(^1\) In general, they represent 36.4% of the primary CNS tumors and \(~\) between 24 and 30% in adults.\(^1,4\) On the other hand, in the pediatric population, the prevalence of CNS tumors varies between 0.4 and 4.6%.\(^2,3,9–13\) Gender equivalence also contrasts with what occurs in the adult population, which has a ratio between female and male gender of \(2:1.\)\(^1,14\) It is believed that this difference is due, especially in the prepubertal period, to the absence of the effect of hormones in the corticosteroid receptors of the cells of the meningioma.\(^10,15–17\)

In the first two decades of life, there is a higher incidence of grade II (atypical) and grade III (anaplastic) meningiomas, according to the WHO: 9.9 and 8.9%, respectively.\(^2,18\) They are characterized by being genetically and phenotypically more aggressive, with a high frequency of cerebral invasion.\(^9,14\) Among the most frequent grade I meningiomas, the angiomatous subtype occurs in 2.8% of the cases;\(^2\) and in 2.1% of all meningiomas at any age.\(^19\) This subtype is defined when the vascular component exceeds 50% of the total tumor area.\(^2,19\) However, differential diagnosis is necessary with hemangio-blastoma and hemangioectasyma, with essential immunohistochemical and morphology roles in the diagnostic confirmation: MIB-1/Ki67 low index and positivity for progesterone receptor, EMA, vimetin, cytokeratin and desmoplatelet.\(^7,19–23\)

Since these tumors are uncommon, the characteristics of angiomatous meningiomas are considered in few studies.\(^7,19,24,25\) They may present moderate to severe cerebral edema with a frequency of 74 to 88.9%,\(^7,19\) due to hyper-vascularization, increased capillary permeability and vascular endothelial growth factor (VEGF) secretion.\(^19\) In magnetic resonance imaging, they may present more signs of flow voids, rarely present necrosis, and they tend to have homogeneous enhancement to paramagnetic contrast.\(^19,23\)

Meningiomas in pediatric patients present in atypical sites more frequently than in adults: in the lateral ventricles, in the skull base, and in the posterior fossa.\(^2,3,10,15,26\) The intraventricular localization occurs in 11%, compared with between 0.3 and 3% in all ages and between 0.5 and 4.5% in adults.\(^2,10,27\)

Intraventricular meningiomas (IVMs) are in the lateral ventricles (more common on the left side) in 76% of the cases;\(^5\) 16% in the 3rd ventricle; and 7%, in the 4th ventricle.\(^5,27\) There are studies suggesting that lateral ventricles are the favorite site of pediatric IVMs.\(^26,28\) These originate from the choroid plexus, growing on the choroidal screen.\(^4\) The vascularization of the tumor depends on its location in the ventricle, and, in general, the main nutrient vessels depart from the choroidal arteries and are of small caliber.\(^27\)

Clinically, pediatric IVMs are usually asymptomatic, until they reach large dimensions in the lateral ventricles, where the risk of hydrocephaly is lower. On the other hand, when located in the 3rd or 4th ventricle, the obstruction of the cerebrospinal fluid (CSF) flow may result in manifestations in early stages of the tumoral growth.\(^5,27,29,30\) Therefore, symptoms – headache, nausea, vomiting, and visual disturbances\(^5,27,31\) – are more frequently related to tumor compression and to an insidious increase in intracranial pressure. Indolent cognitive deficits compromising memory and attention can also occur.\(^32,33\) Typical symptoms of acute intracranial pressure increase are uncommon.\(^29\) The clinic thus correlates with the location of the tumor within the ventricle, the size of the tumor and the direction of its growth.\(^27\) Finally, we emphasize that the clinical presentation of the patient reported – convergent strabismus by paresis of the right lateral rectus muscle – showed no correlation with the tumor, which still had a relatively small size and its location did not justify the signs and symptoms.

Intraventricular meningiomas usually present the classic radiological appearance of other meningiomas: well-defined globular form, but without dural tail. They are usually isointense to hypointense in T1-weighted images, hyperintense in T2-weighted image and undergo strong contrast enhancement.\(^16,27\) In particular, in the pediatric population, other more frequent intraventricular tumors may difficult the differential diagnosis: choroiid plexus tumors, ependymoma, primitive neuroectoelastic tumor, teratoma and astrocytoma.\(^30,34\) Choroiid plexus tumors usually affect children < 10 years old, and, at MRI, they have a multilobulated mass with intense contrast enhancement and fronds appearance. Ependymomas represent approximately one-third of CNS tumors in children < 3 years old and are characterized by necrosis, hemorrhage, cyst formation, and for presenting, in MRI, hypointense in T1 and hyperintense and heterogeneous in T2.\(^35\)

The surgical approach of a benign IVM is a neurosurgical challenge, in view of its deep location and its proximity to eloquent areas and vessels of the ventricles walls.\(^5,34\) The extension of the initial resection is an independent prognostic factor, with significant association with recurrence and malignization.\(^2\) The patient reported here did not present recurrence in the 26 months of follow-up, which is a result consistent with the literature. In a 2012 review, with 201 cases of several series, there were only 8 recurrences.\(^27\) However, in a meta-analysis with 677 cases of meningiomas in the first 2 decades, the numbers are more meaningful. There were 141 recurrences, with an average presentation of 3.6 years and with mortality for this event in 46 cases.\(^2\) Recurrence in this age group occurs basically in cases of atypical and anaplastic meningiomas, or after partial resection.\(^36\) Mortality and posturgical morbidity in postpubertal patients, as is the case of the patient reported here, are approaching the ones observed in meningioma cases in adults.\(^2\) The use of adjuvant radiotherapy should be avoided in young patients, and it may be possible to opt for serial evaluation and reoperation in case of recurrence.\(^2,3,8–11,14,18,37\)

In the literature, there are several surgical approaches for IVM resection: temporoparietal approach, transfrontal, medial posterior temporal gyrus, posterior inferior temporal gyrus, parieto-occipital, and transcalous.\(^5,34\) The choice is individualized and based on the location of the tumor within the ventricle, the tumor size and its vascular network, to always preserve the adjacent cerebral tissue, performing small
corticectomy resecting as little as possible.\textsuperscript{5,6,27,30,33,34}

The rationality behind the choice of the gem approach is determined by the option that allows the best access to the largest axis of the lesion, to minimize transcortical transgression, by the spectrum of neurosurgical deficits Pre-operative proximity with the aforementioned eloquent structures, besides the anatomical knowledge of the cortical and white substance.\textsuperscript{29} In the reported case, we chose the transcallosal approach to be the one that allows the best access to the frontal horn and lateral ventricle body. This approach avoids cortical injury; However, certain care is needed with the possible presence of tributary cortical veins of the superior sagittal sinus, which can be anticipated in the preoperative examinations, and with the corpus callosum, which should be distinguished from the gyrus rotation by the color change.\textsuperscript{5}

The posterior transcallosal approach has disconnection syndrome as one of its possible complications; however, the experience in our neurosurgery service demonstrates that the risk is minimal, since we have performed surgeries with resection of the two thirds posterior callosotomy of the corpus callosum with very low rate of this postoperative syndrome.\textsuperscript{5,30,33,38}

Despite the degree of difficulty, IVM surgery has shown low rates of morbidity and mortality in the last decades, and most postoperative complications – visual deficits and apraxias – are temporary.\textsuperscript{27,29,34} These low rates are consistent with the case reported here, which did not present postoperative complications or transoperative sequelae.

**Conclusion**

Intraventricular angiomatous meningioma is a rare entity, even more in patients in the first 2 decades of life. The clinic is unspecific in most cases, making it necessary to assess MRI for diagnosis and definition of the surgical approach, and the histopathological analysis is what defines the diagnosis of the angiomatous subgroup. Surgical resection is the treatment of choice. However, the objective of total resection should not be above the objective of preserving the functions and the quality of life of the patient.

**Conflicts of Interests**

The authors declare that there are no conflicts of interests.

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