

Primary spinal meningioma in a 10-year-old boy

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The diagnosis of primary spinal meningioma is rare in children with an annual incidence of 8 cases per 1000000 people¹. Meningiomas are largely benign in nature and their recurrence is most associated to histologic type. The current treatment of choice is surgical resection.

The aim of this report is to present a case of a ten year-old boy diagnosed with a psammomatous meningioma in the topography of the thoracic column (T4), and to review some epidemiological, pathophysiological, clinical and prognostic features.

CASE

A ten year-old male patient with no relevant past medical history suddenly showed progressive spastic paraparesis of lower limbs with reduced sensitivity to pain, 40 days prior to hospital admission. Moreover, no infections had been observed prior to symptoms, nor relevant epidemiological antecedents or previous exposure to ionising radiation. Clinical examination revealed bilateral reduction in muscular strength of the legs and thighs (both power grade II) with normal bilateral deep tendon reflexes while no changes in sensitivity to heat, pain or proprioception were observed. The results of dermatologic examination were normal. Fundoscopy was normal. The child also had normal CSF tests. Brain CT scan showed

no signs of schwannomas or other tumors. A neuroaxis MR (Fig 1) showed a posterior intradural expansive mass at the level of T4 and T5 vertebrae, in an apparent extra-medullary, well delimited, homogenous, lobulated topography. The tumor measured around 1.8 × 1.2 × 0.9 cm and was characterized by T1 iso-signal, T2 hypersignal, and homogenous accentuated enhancement following contrast injection, and exerted a local expansive effect creating medullar dorsal compression with signs of local edema/trauma signs characterized by T2 hypersignal. The patient underwent laminectomy, and opening up of the dura exposed a whitish tumor compressing the adjacent medulla. The tumor presented a hard consistency, with little bleeding, a clear cleavage plane with the dura mater and weak adherences to the adjacent arachnoid. The tumor was fully resected. Anatomopathologic study revealed a psammomatous meningioma (Fig 2). Post-operative evolution was satisfactory, with recovery of muscular strength to power grade 4 in three days. He is currently undergoing motor rehabilitation with good clinical evolution.

DISCUSSION

Meningiomas are rare tumors in children, accounting for less than 5% of brain tumors in children and less than 2% of all

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MENINGIOMA ESPINAL PRIMÁRIO EM MENINO DE 10 ANOS DE IDADE

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Fig 1. MR showing [A] T1-weighted iso-signal tumor, [B] T2 hypersignal, and homogenous accentuated enhancement following contrast injection, and exerting a local expansive effect, [C] MR done one month after surgery with no signs of recurrence.

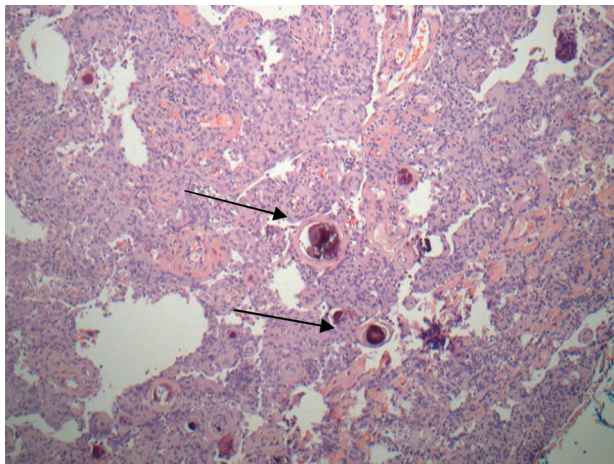


Fig 2. Histological aspect of the tumor - higher magnification showing proliferation of meningothelial cells with sparse psammoma bodies (arrows), hematoxylin-eosin, 40× magnification.

meningioma cases². Spinal meningiomas comprise about 25 to 46% of all spinal tumors³. Pediatric spinal cord tumors represent only 5% of tumors of the central nervous system, 25% of which occur in the intradural-extramedullary compartment⁴. Pediatric spinal tumors are very rare with an annual incidence of almost 1 per 1000000 children, presenting 5 to 10% of all central nervous system tumors in children⁵. One to 10% occur in the intradural-extramedullary space⁶. Reports in adult patients describe that the majority of cases tend to occur in women (around 2:1). However, this association is not replicated in children, where there are reports of an approximately 1.2:1 predominance of cases in males². A retrospective analysis conducted by Greene and colleagues of 20 patients showed a median age of presentation of the tumors of about 13 years old². Case reports in younger age groups included those in 14 and 20 month-old infants, while only a few reports of spinal meningiomas in chil-

dren of school age were found^{7,8}. Ferraz-Filho and colleagues reported a case of a primary extracranial meningioma of ethmoid sinus in a child and according to them approximately 20% of meningiomas presented extra-cranial extension in contiguity with presentation in structures such as facial sinuses⁹.

Known risk factors for developing meningiomas include prior submission to radiation and the presence of type 2 neurofibromatosis, in which meningiomas are present in 25% to 40% of children with neoplasia¹⁰. The link to type 2 neurofibromatosis is becoming clearer, with the most common finding the loss of a tumor suppressor gene in the chromosome 22 (NF2). Other gene mutations may contribute to the progression of the meningiomas, leading to the anaplastic type. Ionising radiation is the most environmental risk factor predisposing to meningiomas¹¹.

Gezen and colleagues³ reviewed the main characteristics of 36 cases of spinal cord meningiomas. Regarding to the symptomatology, pain and sensory loss were the most common symptoms associated with some degree of weakness. Besides these symptoms, sphincter disturbances were also common in about 36% of the patients.

The slight predominance of this tumor in women over man in adults raises the hypothesis of influence of female hormones. Some tumors show receptors for estrogen and progesterone and some cases have a more fast growth on later stages of pregnancy. However, the exact link between hormones and development of meningiomas is still not clear¹².

There is correlation between symptomatology of patients and degree of peritumoral edema observed on MR studies. Edema is a common finding in meningioma cases with an incidence of between 40% and 78% depending on macrophage infiltration at lesion sites and on vascularization type - the more internal carotid artery branches nourishing the lesion, the higher the degree of edema.

Severe edema may cause significant and permanent neurological deficits. Mattei and colleagues conducted a retrospective study involving 55 cases of intracranial meningiomas and found a higher percentage of significant edema in tumors with cellular atypies and malignant histological traits, than in tumors with benign histological findings (9% versus 5%, level of significance “p”=0.0089)¹³. Similarly, Souto¹⁴ and colleagues analyzed 51 cases of intra-cranial meningiomas and observed a higher degree of perilesional edema in psammomatous, angiomatous and anaplastic variants. In view of this physiopathology, the use of steroids is indicated mainly to avoid neurologic impairment and to provide analgesia. Meningiomas are generally slow-growing tumors, spreading laterally throughout the subarachnoid space until becoming symptomatic.

A grading system proposed by the World Health Organization (WHO) is the most used in the classification of the meningiomas. There are 3 grades - benign (grade I), atypical (grade II) and anaplastic or malignant (grade III). The psammomatous subtype show uniform cells and an oval nuclei, probably due to glycogenation, and a low mitotic index¹².

Schaller and colleagues studied the correlation between histological type of neoplasms and degree of motor deficit after surgery with curative intent. Patients with psammomatous meningioma, who comprised 67% of the cases, were shown to present greater post-surgical motor deficits¹⁵.

A retrospective study of 55 patients with spinal meningiomas showed increased levels of total protein in CSF in 61% of the patients¹.

MR imaging is the gold standard for diagnosing meningiomas in children. The most common lesion pattern is the finding of T1 and T2 iso-intense masses associated with different degrees of peritumoral edema.

Surgical resection is the treatment of choice¹⁶, with the intention of complete resection of the tumor while preserving neurological functions and stability of the growing spinal canal. Due to the rareness of meningiomas in children, few surgical protocols are available, and recommended treatment is extrapolated from the literature on adult patients.

The five-year rate of recurrence of spontaneous meningiomas is around 40%, as tumors are slow growing. An-

nual neurologic and MR assessments are recommended for up to 10 years after diagnosis. Average five-year survival, considering all types and locations of meningiomas in children, is approximately 76%. However, little is known on this data owing to scant follow-up studies in patients with meningioma because of the rareness of this tumor in the pediatric population.

The scope of this report was to illustrate the diagnosis and treatment of a rare disease in children, the diagnosis of which is relatively straightforward using a good neurologic and diagnostic imaging exam. The prognostic is favorable in most cases following successful definitive surgical intervention. Follow up using clinical exams and serial imaging is crucial to enable early detection of and election of further surgical interventions when necessary.

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