Meningeal Melanocytoma: An Unusual Presentation of a Rare Tumor

Melanocitoma Meníngeo: Uma Apresentação Incomum de um Raro Tumor

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ABSTRACT

Meningeal melanocytoma is a rare type of benign pigmented tumor of the central nervous system that derives from leptomeningeal melanocytes, which originate from the neural crest. These tumors are commonly focal, but there are descriptions of multifocal forms in the literature, and reports of malignant transformation, with leptomeningeal dissemination. In this paper, a case of meningeal melanocytoma with leptomeningeal dissemination is reported and, based on a review of the literature, comments on the diagnostic and therapeutic difficulties relating to this disease are made.

Keywords. Meninges,Metastatic, Neural Crest, Melonocytic.

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RESUMO

O melanocitoma meníngeo é um tumor pigmentado, raro e benigno do SNC e deriva dos melanócitos leptomeníngeos, oriundos da crista neural. Este tumor comumente é focal, mas existem na literatura descrições de formas multifocais e relatos de transformação maligna, com disseminação leptomeníngea. Neste artigo, os autores relatam um caso de melanocitoma meníngeo com disseminação leptomeníngea e, baseados em revisão de literatura, comentam sobre as dificuldades diagnósticas e terapêuticas desta patologia.

Unitermos. Meninges, Metastático, Crista Neural, Melanocítico.

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INTRODUCTION

Meningeal melanocytoma (MM) is a rare type of tumor of the central nervous system (CNS) that comes from leptomeningeal dendritic melanocytes, which embryologically originate from the neural crest. These tumors, together with primary malignant melanomas, melanocytic schwannomas, meningeal melanoblastomas, pigmented neuroblastic tumors and melanomatosis, make up the group of primary melanocytic neoplasms of the CNS[1]. Melanocytes in the CNS are usually found at the base of the brain, in the brainstem and in the spinal cord. MM accounts for 0.06-0.1% of brain tumors and its estimated incidence is one in every 10 million inhabitants. It most commonly affects adult patients between the ages of 45 and 50 years, with slight predominance (1.5: 1) among females[2]. The term meningeal melanocytoma was coined after the demonstration of their melanocytic origin[3].

Most of these tumors are solitary, with slow growth and low levels of infiltrative action. However, there have been reports of transformation to malignant melanoma and of leptomeningeal or multifocal diffusion[4,5]. There is still a debate in the literature as to whether the multifocal form results from leptomeningeal diffusion[6-8]. We present a clinical case of a young patient with the leptomeningeal form of MM, and discuss the diagnostic and therapeutic aspects of this rare neoplasm.

CLINICAL CASE

The patient was a 25-year-old man who had been followed up at another service with a 10-month history of headache and intermittently blurred vision, associated with bilateral papilledema, with normal magnetic resonance imaging (MRI) of the cranium and a diagnosis of optic neuritis. The patient reported that the symptoms were partially alleviated with the use of corticosteroids.

The neurological examination showed only slight bilateral papilledema and Lhermitte sign, without other pathological neurological signs. Another MRI on the cranium showed slight widening of the sella turcica and presence of nodule-like lesions in the supra and infratentorial spaces, involving the brainstem, cerebellopontine angle region, cerebellum and suprasellar cistern (Fig. 1), along with an intraventricular nodular lesion in the frontal horn of the right lateral ventricle and a small lesion in the left sylvian fissure. The cerebrospinal fluid pressure was normal, but its protein content was 97.32 mg/dl (normal range: 15-45 mg/dl) and its white cell count was 6 cells/mm^3 (normal range: up to 5 cells/mm^3), with predominance of lymphomonocytes. It was negative for neoplastic cells.

The patient's condition progressed to hydrocephaly, which required emergency surgery to insert a ventricular-peritoneal derivation valve. In a second surgical procedure, he underwent a biopsy on one of these lesions, which was accessible in the region of the left sylvian fissure, with removal of rare blackened material of vascular nature. Cytologically, this material was suggestive of hemangioblastoma grade I. A biopsy on the meninges produced normal results. The patient progressed well after correction of the hydrocephaly, but with complaints of difficulty in passing urine.

During the clinical follow-up, an MRI evaluation was made on the spinal cord. In the cervical region, a nodular lesion was observed, in an anterior position at C5/C6 level (Fig. 1). In the dorsal/lumbar region, anomalous intradural and extra-medullary material was seen, especially in a posterior position, involving the entire length of the thoracic spinal cord. This material took on a nodular format at the level of the conus medullaris (Fig. 2). The patient then underwent L1/L5 laminectomy, during which an extensive blackened lesion involving the roots of the conus medullaris was observed, extending to the vertebral canal in an ascending manner. The material dissected easily from the roots, thus suggesting meningeal etiology, but total resection was not possible. The cytological and immunohistochemical analyses gave diagnoses of meningeal melanocytoma. The patient is following a course of oncological treatment.

This case report was approved by the Ethics Committee of Hospital Santa Rosa (protocol number: 01/2011), and the patient gave her consent for publication.

DISCUSSION

Primary melanocytic neoplasms of the CNS are a rare group with the common characteristic of melanocytic nature. The first description compatible with MM

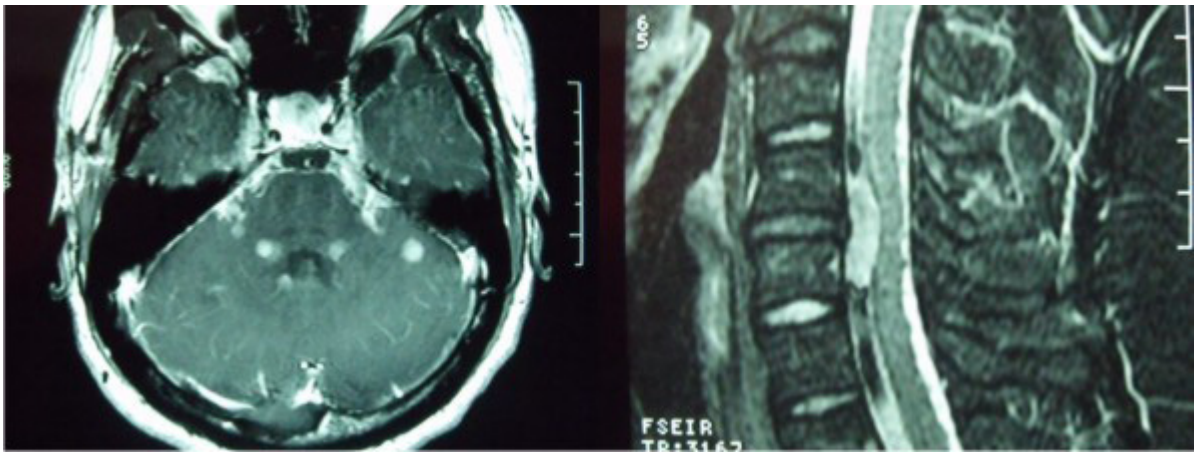

Fig.1. MRI images demonstrating multifocal lesions in supraselar cistern, cerebellopontine angle, involving the brainstem,and a extramedulary intradural lesion in cervical level C5/C6.

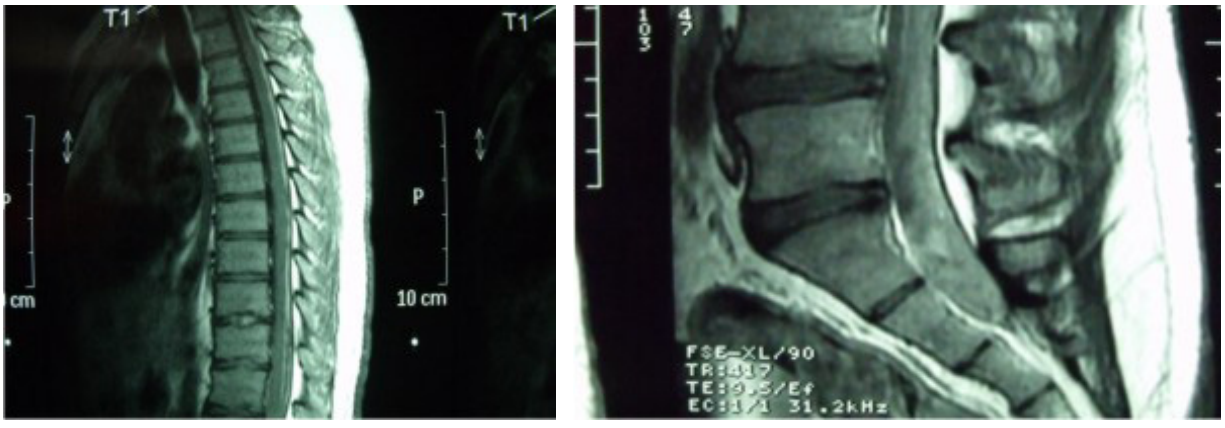

Fig.2. MRI images demonstrating the presence of anomalous intradural and extra-medularry material in thoracic spinal cord and cauda equina.

was made in 1912, in an autopsy[9]. According to a survey made in 2010[10], 112 cases have been described in the English-language literature. Depending on the distribution of melanocytes in the CNS, this neoplasm can occur in the brain and spinal cord regions. The brain forms have mostly been observed in supratentorial locations, and the spinal cord forms mostly in the thoracic region. Although MM has been described as a pathological condition of benign etiology, with a solitary nodular presentation and a tendency to recur at the excision site, some authors have questioned whether its behavior can be referred to as benign[4,5,7,11,12]. They cite reports demonstrating tumor recurrence, appearance of new lesions in regions other than the original location and cases of malignant degeneration, probably due to leptomeningeal dissemination. There are also reports of lesions appearing outside of the CNS, such as in the adrenal glands and liver[13,14].

The malignant equivalent of MM is focal or diffuse melanoma (meningeal melanocytosis). This narrow differential spectrum implies difficulties in making a histological diagnosis. Factors such as symptoms lasting for more than one year, radiological similarity to meningioma, uniform cytological characteristics with predominance of fusiform cells and a diminished mitotic pattern favor a diagnosis of MM[15]. Immunohistochemistry aids in this differentiation through demonstrating that MM cases are characterized by positive reactions to anti-melanoma antibodies (HMB-45), anti-vimentin antibodies and protein S-100[2]. Our patient was positive for protein S-100 and a melanoma cocktail (HMB45+M2+M2-9E3), and also presented a neoplastic cell proliferation value (Ki-67) < 5%. No atypia, areas of necrosis or inva-

sion were observed. Although the differential diagnosis is of paramount importance, the result does not necessarily imply that these cases have stable evolution, since the reports in the literature demonstrate behavior that is very heterogeneous.

In a retrospective study on 47 patients was observed that five years after complete resection, there was local recurrence in 22% of the cases, while five years after incomplete resection, there was recurrence in 80% of the cases[16]. They also found that there were risks of malignant degeneration, which led to significant changes in the prognosis. This heterogeneity over the course of the disease, along with the small number of cases studied over the last few decades, have led to doubts about what the best therapeutic option for these patients would be. In 2004, these same authors reviewed 89 MM cases and observed that the five-year survival rate was 83% in the cases in which total tumor resection had been possible, but that it fell to 40% for the cases in which the resection had been incomplete. However, they observed that associated radiotherapy administered to patients with incomplete resection raised the five-year survival significantly, particularly in cases with a radiotherapeutic regimen of 45-55 Gy[17]. Complementary radiosurgery has also been used in a few cases described in the literature[18-20]. There is no evidence of benefits from the use of chemotherapy.

In the case reported here, several lesions diffused along the neuroaxis (notably in the cauda equina, thoracic spinal cord, cervical spinal cord, left cerebellopontine angle and suprasellar cistern) were observed in a patient who had presented clinical symptoms for more than two years. The material removed had benign characteristics, without indications of malignant transformation. This presentation of a leptomenigeal form, still with benign characteristics, was recently observed[21], and this contrasts with the few other published cases involving this clinical presentation, given that these other cases presented concomitant malignant degeneration of MM. This observation cannot be used for prognostic purposes, in view of the paucity of data in the literature, and neither can the extent of leptomeningeal diffusion shown in the present case.

CONCLUSION

Although MM is considered a rare neoplasm, presenting as a solitary nodule and with benign characteristics, a small number of papers has shown cases of MM with leptomeningeal dissemination and/or malignant transformation. These forms of MM with atypical evolution represent a diagnostic and therapeutic challenge, since there are no treatment protocol. In this case report, we present one clinical case with malignant behavior, and hope that this case makes a contribution through new clinical observations on this rare form of presentation of primary melanocytic neoplasm of CNS.

REFERENCES

1.Châabane M, Ellouze S, Hamrouni A, Mlika N, Ben Hammouda M, Khoudja F. Une tumer méningée rare: Mélacytome méningé. J Radiol 2003;84:415-6.

2.Franken SPG, Setz-Pels W, Smink-Bol M, Gijtenbeek JMM, Nanda D, Van Der Maazen RWM, Van Der Vliet T, Bussink J. Unusual case of bifocal leptomeningeal melanocytoma in the posterior fossa with seeding in the spinal canal. Br J Radiol 2009;82: e182-e188.
http://dx.doi.org/10.1259/bjr/30756805

3.Limas C, Tio FO. Meningeal melanocytoma: its melanocytic origin revealed by electron microscopy. Cancer 1972;30:1286-94.
http://dx.doi.org/10.1002/1097-0142(197211)30:5<1286::AID-CNCR2820300522>3.0.CO;2-V

4.Ali Y, Rahme R, Moussa R, Abadjian G, Menassa-Moussa L, Samaha E. Multifocal meningeal melanocytoma: a new pathological entity or the result of leptomeningeal seeding? J Neurosurg 2009;111:488-91.
http://dx.doi.org/10.3171/2009.3.JNS081096

5.González-Tortosa J, Ferri-Ñíguez B, Ros de San Pedro J. Melanocitoma meníngeo del ángulo pontocerebeloso: Um tumor benigno? Neurocirugia (Astur) 2009;20:372-80.

6.Ferracini R, Gardini G, Brisigotti M, Lanzanova G, Manetto V, Lorenzini P. Metastasizing meningeal melanocytoma. Tumori 1980;66:405-8.

7.Koch HJ, Roeber S, Zimmermann UW, Schafer C, Villarubia V, Kuchelmeister K, et al. Spinal and cerebral leptomeningeal seeding from melanocytoma of the cerebello-pontine angle. Wien Med Wochenschr 2005;155:360-4.
http://dx.doi.org/10.1007/s10354-005-0199-8

8.Bydon A, Gutierrez JA, Mahmood A. Meningeal melanocytoma: an aggressive course for a benign tumor. J Neuroncol 2003;64:259-63.
http://dx.doi.org/10.1023/A:1025628802228

9.Barth A, Pizzolato GP, Berney J. Mélanocytome méningé intramédullaire. Neurochirurgie 1993;39:188-94.

10.Liubinas SV, Maartens N, Drummond KJ. Primary melanocytic neoplasms of the central nervous system. J Clin Neurosci 2010;17:1227-32.
http://dx.doi.org/10.1016/j.jocn.2010.01.017

11.Wang F, Xianghong L, Linghong C, Xiaolu P. Malignant transformation of spinal meningeal melanocytoma. J Neurosurg Spine 2007;6:451-54.
http://dx.doi.org/10.3171/spi.2007.6.5.451

12.Roser F, Nakamura M, Brandis A, Hans V, Vorkapic P, Samii M. Transition from meningeal melanocytoma to primary cerebral melanoma. J Neurosurg 2004;101:528-31.
http://dx.doi.org/10.3171/jns.2004.101.3.0528
13.Koenigsmann M, Jautzke G, Unger M, Théallier-Janko A, Wiegel T, Stoltenburg-Didinger G. 57-year-old male with leptomeningeal and liver tumors. Brain Pathol 2002;12:519-21.
http://dx.doi.org/10.1111/j.1750-3639.2002.tb00472.x
14.Oruckaptan HH, Soylemezoglu F, Kutluk T, Akalan N. Benign melanocytic tumor in infancy: discussion on a rare case and review of the literature. Pediatr Neurosurg 2000;32:240-47.
http://dx.doi.org/10.1159/000028945
15.Litofsky NS, Zee CS, Breeze RE, Chandrasoma PT. Meningeal Melanocytoma: Diagnostic Criteria for a rare lesion. Neurosurgery 1992;31:945-48.
http://dx.doi.org/10.1227/00006123-199211000-00019
16.Rades D, Heidenreich F, Tatagiba M, Brandis A, Karstens JH. Therapeutic options for meningeal melanocytomas. J Neurosurg 2001;95:225-31.
http://dx.doi.org/10.3171/spi.2001.95.2.0225
17.Rades D, Schild SE, Tatagiba M, Molina HA, Alberti W. Therapy of Meningeal Melanocytomas. Cancer 2004;100:2442-7.
http://dx.doi.org/10.1002/cncr.20296
18.Kurita H, Segawa H, Shin M, Ueki K, Ichi S, Sasaki T, Tago M, Kirino T. Radiosurgery of meningeal melanocytoma. J Neurooncol 2000;46:57-61.
http://dx.doi.org/10.1023/A:1006335616839
19.Hamasaki O, Nakahara T, Sakamoto S, Kutsuna M, Sakoda K. Intracranial meningeal melanocytoma. Neurol Med Chir (Tokyo) 2002;42:504-9.
http://dx.doi.org/10.2176/nmc.42.504
20.Beseoglu K, Knobbe CB, Reifenberger G, Steiger HJ, Stummer W. Supratentorial meningeal melanocytoma mimicking a convexity meningioma. Acta Neurochir (Wien) 2006;148:485-90.
http://dx.doi.org/10.1007/s00701-005-0705-1
21.Ayuga Loro F, Díez De Valle R, Casado López RM, Florensa Vila J. Melanocitoma meníngeo con comportamiento maligno. Neurología 2011;26:565-6.
http://dx.doi.org/10.1016/j.nrl.2011.01.023