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Primary Meningeal Melanocytoma Located in the Craniovertebral Junction: A Case Report and Literature Review

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Abstract

Primary meningeal melanocytoma is a rare benign tumor in the central nervous system (CNS), comprising less than 0.1% of all intracranial tumors. A 44-year-old man presented with occipital headache, nausea, and vomiting. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a well-defined intradural extramedullary mass lesion at the craniovertebral junction (CVJ). Gross total removal was achieved, and the patient improved symptomatically. The pathologic findings were consistent with meningeal melanocytoma. No tumor recurrence was seen on follow-up MRI two years after surgery. Cases of primary meningeal melanocytoma located at the CVJ are rare. The preoperative differential diagnosis of meningeal melanocytoma from meningioma is sometimes difficult because of their similar appearance on CT and MRI. Complete surgical removal is curative for most cases. We present a case of gross total removal of a meningeal melanocytoma located in the CVJ with references to the literature.

Keywords: meningeal melanocytoma, foramen magnum, craniovertebral junction

Introduction

Primary meningeal melanocytoma is a benign central nervous system (CNS) tumor derived from melanocytes. The most common location is the posterior fossa or upper spinal cord,[1–3] and a lesion located in the craniovertebral junction (CVJ) is rare.[4] As preoperative differential diagnosis of meningeal melanocytoma from meningioma can be difficult,[5–7] histopathologic and immunohistochemical examinations are necessary to make a definite diagnosis.[4,8] The optimal treatment of meningeal melanocytoma is complete surgical removal.[8,9] Here, we report a rare case of meningeal melanocytoma located in the CVJ and present a review of the literature.

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Case Report

A 44-year-old, previously healthy man presented with a month history of occipital headache, nausea, and vomiting. Magnetic resonance imaging (MRI) revealed a CVJ tumor, at which point he was referred to our hospital. For a few days immediately before admission, symptoms became more severe, but physical examination and laboratory tests were normal.

Imaging studies

Computed tomography (CT) showed a well-circumscribed intradural extramedullary mass lesion in the foramen magnum behind the bulbus medullae. There were no signs of calcification or hyperostosis of the adjacent bony structures. The mass was hyperintense on T1-weighted images but without clear contrast enhancement, and hypointense on T2-weighted images (Figs. 1A–1D). Cerebral angiography showed the lesion was avascular.

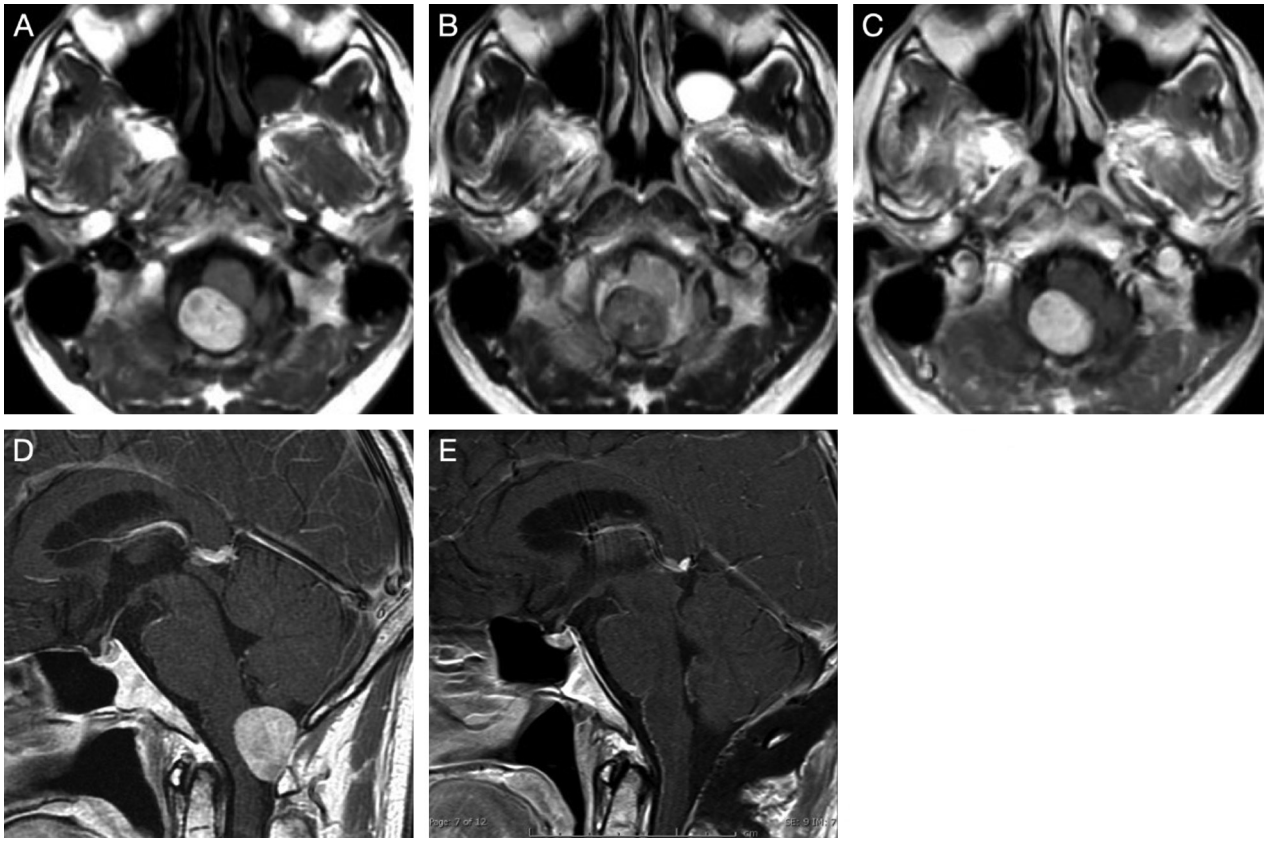

Fig. 1 MRI. (A) Axial T1-weighted MRI shows a well-circumscribed hyperintense lesion in the foramen magnum. (B) Axial T2-weighted MRI reveals hypointensity. (C, D) Axial and sagittal gadolinium-enhanced T1-weighted MRI shows an intradural extramedullary mass lesion in the foramen magnum is compressing the dorsal brainstem. The contrast enhancement is not evident. (E) Postoperative axial gadolinium-enhanced T1-weighted MRI show gross total removal. MRI: magnetic resonance imaging.

Operation and postoperative course

The patient underwent removal of the tumor via the midline suboccipital approach with C1 laminectomy. After a dural incision, a blackish-grey, well-circumscribed tumor, which was located of the foramen magnum behind the medulla oblongata was found (Fig. 2A). Most of the tumor was located in the subarachnoid region but partially attached to the dorsal dura mater. After dissecting from the surrounding tissue, gross total removal was achieved and dural attachment was coagulated (Fig. 2B). The postoperative course was uneventful, and the patient improved symptomatically. No radiation therapy or chemotherapy was given after surgery. A follow-up examination 2 years after surgery, MRI showed no evidence of tumor recurrence (Fig. 1E).

Histopathological findings

Histopathological examination revealed the tumor was multicellular and a relatively monomorphic population of spindle cells which formed a fascicled or nested growth pattern. There were variable amounts of pigment deposition consistent with melanin in the cytoplasm. There were few hemorrhages and necroses, but nuclear atypia, pleomorphism, or mitotic activity was not seen. Immunohistochemical study was positive for HMB-45 and S-100 protein. The MIB-1/Ki-67 labeling index was 4%. The histopathological findings were consistent with meningeal melanocytoma (Fig. 3).

Discussion

Primary meningeal melanocytoma is a rare benign tumor of the CNS. The prevalence rate is one per 10 million, and it accounts for less than 0.06–0.1% of all brain tumors.[2] The term melanocytoma was first introduced by Limas and Tio in 1972.[10] By ultrastructural studies, they determined this tumor was derived from melanocytes rather than meningothelial cells. The most common location is the posterior fossa or upper spinal cord because leptomeningeal melanocytes are most highly concentrated in these segments.[1,2]

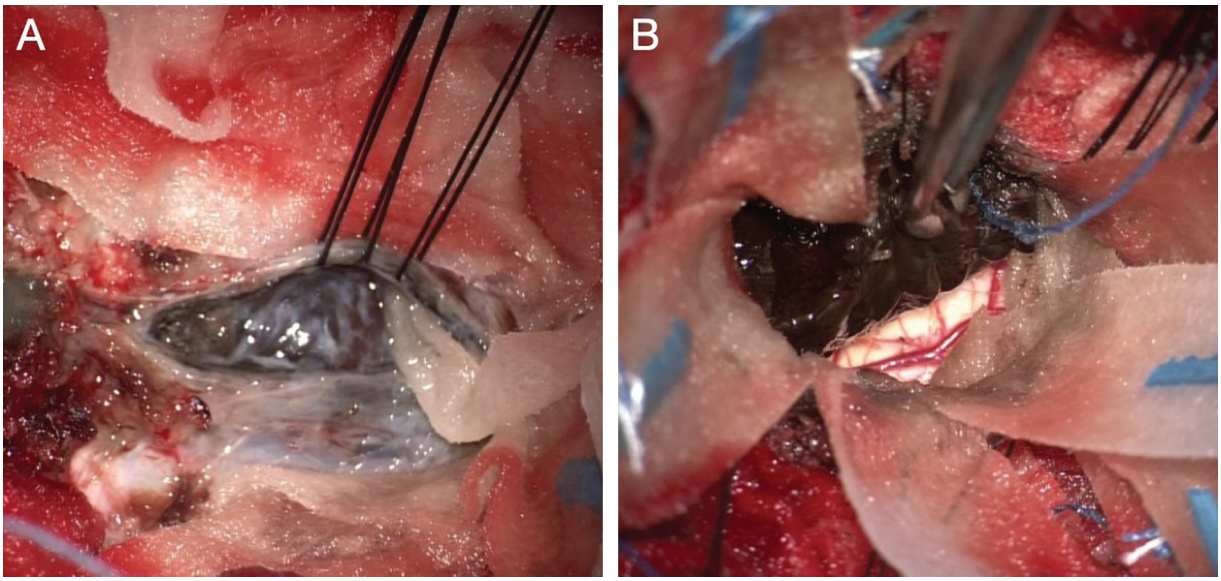

Fig. 2 Intraoperative microscopic view. A blackish-grey tumor can be seen (A). After peeling off the arachnoid mater around the tumor, gross total removal was achieved (B).

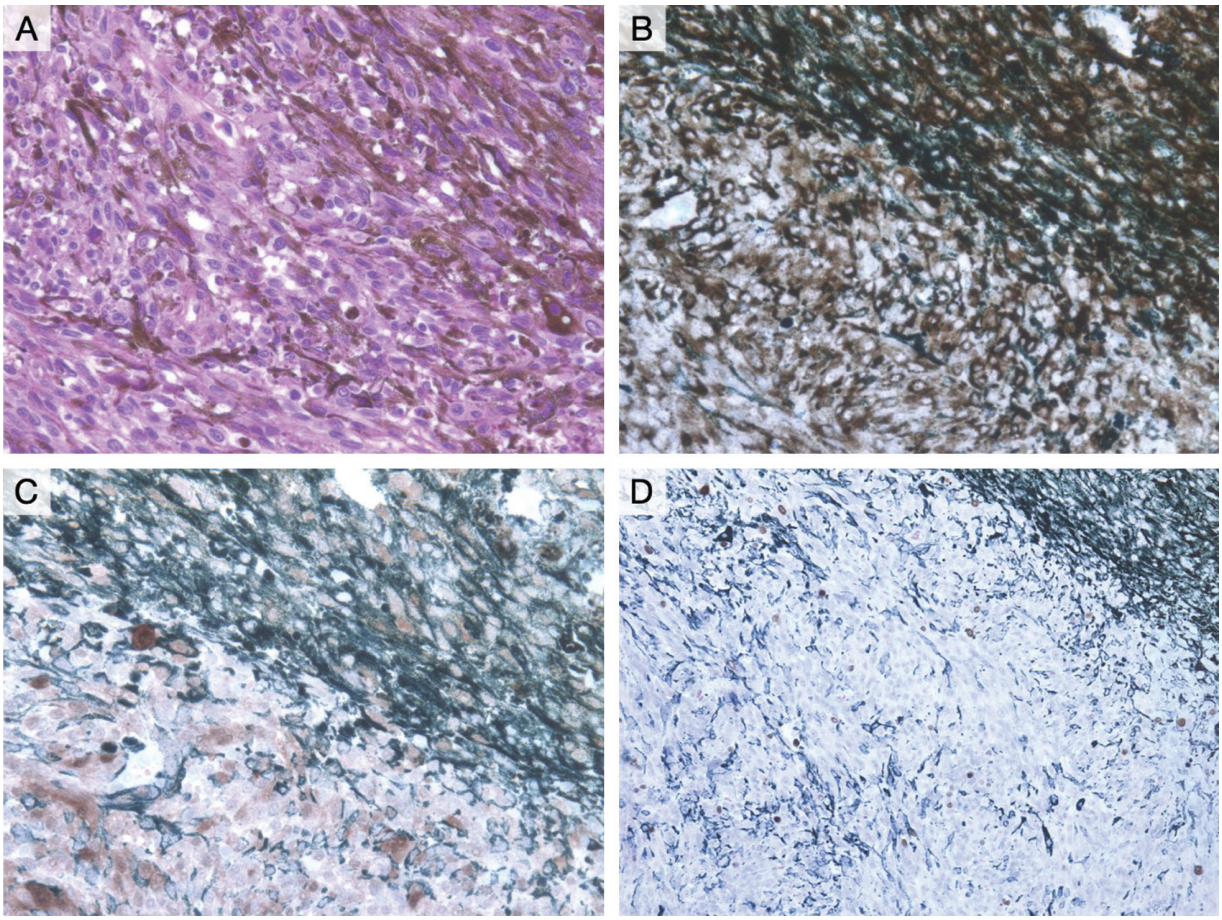

Fig. 3 Histopathological and immunohistochemical findings. (A) The tumor is composed of relatively monomorphic spindle cells that formed a fascicled or nested growth pattern. There are variable amounts of pigment deposition consistent with melanin in the cytoplasm (hematoxylin and eosin stain, 400×). (B) HMB-45 (+) staining of tumor cells. (C) S-100 (+) staining of tumor cells. (D) MIB-1/Ki-67 labeling index is low.

Table 1 Cases of primary meningeal melanocytoma located in craniovertebral junction reported in the literature

No.	Reference	Sex	Age	Symptom	DOS (mon)	Location	MRI (T1)	MRI (T2)	Operation	Radiation	Follow-up (mon)	Recurrence	Outcome
1	Limas and Tio (1972)[10]	M	71	Headache, Lt LE weakness	72	Foramen magnum to C1	Unknown	Unknown	Autopsy diagnosis				Death
2	Naul et al. (1991)[11]	F	68	Headache	1.5	Foramen magnum	Iso	Hypo	CR	No	10	No	Alive
3	Uematsu et al. (1992)[12]	M	62	Gait disturbance	12	Foramen magnum to C1	Hyper	Hypo	CR	No	18	No	Alive
4	O'Brien et al. (1995)[13]	F	49	Unknown	24	Foramen magnum to C3	Unknown	Unknown	CR	No	96	No	Alive
5	Hirose et al. (1997)[14]	M	66	Gait disturbance, Bt UE numbness	12	Foramen magnum to C1	Iso	Hypo	CR	No	3	No	Alive
6	Offiah et al. (2006)[8]	F	25	Headache	9	Foramen magnum	Hyper	Hypo	CR	No	6	No	Alive
7	Fan et al. (2012)[4]	M	48	Headache, Lt weakness and numbness	12	Foramen magnum	Hyper	Iso	CR	No	12	No	Alive
8	Bhargava et al. (2013)[9]	M	55	Four-limb weakness	12	Foramen magnum to C2	Hyper	Hypo	CR	No	10	No	Alive
9	Yang et al. (2016)[2]	M	51	Rt limb weakness, four-limb numbness	3	Foramen magnum to C2	Hyper	Hypo	CR	No	25	No	Alive
10		M	26	Vomiting, Bt UE numbness	2	Foramen magnum to C1	Hyper	Hypo	CR	No	31	No	Alive
11		M	39	Four-limb weakness	6	Foramen magnum to C1	Hyper	Hypo	CR	No	47	No	Alive
12	Lee et al. (2017)[7]	M	45	Headache	14	Foramen magnum to C1	Hyper	Hypo	CR	No	6	No	Alive
13	Our case	M	44	Headache, vomiting	1	Foramen magnum to C1	Hyper	Hypo	CR	No	24	No	Alive

Bt: bilateral, CR: complete removal, DOS: duration of symptoms, F: female, Hyper: hyperintense, Hypo: hypointense, Iso: isointensity, M: male, MRI: magnetic resonance imaging, LE: lower extremity, Lt: left, Rt: right, UE: upper extremity.

Although primary meningeal melanocytoma may occur at the base of the brain, the CVJ is a rare location. Only 13 cases including the present case have been reported so far (Table 1).[2,4,7–14] Most of the patients have been male and aged from 25 to 71 years with a mean age of 49.9 years. The duration of symptoms ranged from several weeks to 6 years, and the predominant symptom was headache induced by the mass effect. Our patient also presented with headache and vomiting but did not develop obstructive hydrocephalus.

On CT scans, meningeal melanocytoma is characterized by well-circumscribed, isodense or slightly hyperdense extra-axial tumors that are homogeneously enhanced by the addition of contrast media.[15] MRI typically showed hyperintense or isointense on T1-weighted images and hypointense on T2-weighted images.[15,16] With gadolinium enhancement, most of these lesions appear uniformly enhanced, but there are some cases in which contrast enhancement is not clearly evident because of strong T1 hyperintensity on the unenhanced images.[2] Our case also showed hyperintensity on T1-weighted MRI, and gadolinium enhancement was not clear. These findings are strongly suggested for meningeal melanocytoma. However, preoperative differential diagnosis of meningeal melanocytoma from other meningeal tumors is often difficult because of the different content of the melanin pigment and the presence of hemorrhage and their similar appearance on images.[5,6] Tumor calcification and hyperostosis of adjacent bony structures have rarely been described in meningeal melanocytoma, but lack of these signs definitively does not rule out meningioma.[6]

Histopathologic and immunohistochemical examination is necessary to make a definite diagnosis of meningeal melanocytoma.[4,8] The tumor cells appear as a well-circumscribed, encapsulated, dark brown to black nodular lesion due to the abundant melanin production. They may be firmly attached to the underlying meninges like with meningioma. In general, the tumors are highly cellular and composed of monomorphic spindle or epithelioid cells arranged in whorls, sheets, bundles, or nests. Mitotic activity is usually low, and necrosis and hemorrhage are absent.

Immunohistochemical staining showed meningeal melanocytomas are positive for HMB-45 and S-100 protein, but negative for keratin, epithelial membrane antigen, glial fibrillary acidic protein, and neuron-specific enolase.[2,4,7,9,15] MIB-1/Ki-67 labeling index helps to differentiate melanocytoma from malignant melanoma, as they are low in melanocytoma.[1,2,9]

In our case, the tumor was composed of monomorphic spindle cells, which were arranged in fascicles and nests, and there was abundant melanin in the cytoplasm. Immunohistochemical studies were positive for HMB-45 and S-100 protein. The MIB-1/Ki-67 labeling index was 4%. Therefore, the histopathological and immunohistochemical diagnosis was meningeal melanocytoma.

Meningeal melanocytoma is a slow-growing benign tumor, for which complete surgical removal is recommended.[2,4–6,8,9] Gross total removal is generally possible because the majority of the tumors are located in the intradural extramedullary compartment.[16] Yang et al.[2] reported that meningeal melanocytoma has favorable prognosis, and after a complete excision, no tumor progression or focal recurrence was observed in their study. Of 13 cases summarized, all patients except for one case diagnosed by autopsy underwent gross total removal. The follow-up period ranged from 3 to 96 months (mean 24 months), and no recurrence was observed. All tumors located dorsal side of the CVJ seem to contribute to the ease of total removal. On the other hand, in patients with primary CNS melanocytoma arising from the other site, relapse and malignant transition have been reported, clinical follow-up could be necessary, and adjuvant radiation therapy is advised in cases of incomplete removal and recurrence.[4,7,15,17]

In conclusion, we report a rare case of meningeal melanocytoma located at the CVJ. Preoperative diagnosis is often difficult, and histopathologic and immunohistochemical examination is essential. The outcome should be good with complete removal, but regular follow-up is necessary because of local recurrence.

Conflicts of Interest Disclosure

The authors have no conflicts of interest to declare.

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