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.⁴,⁹ Here, we report a rare case of meningeal melanocytoma located in the CVJ and present a review of the literature.

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.
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. The term melanocytoma was first introduced by Limas and Tio in 1972. 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.
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)              | M   | 71  | Headache, Lt LE weakness            | 72        | Foramen magnum to C1             | Unknown  | Unknown  | Autopsy diagnosis | No        | 10              | No         | Alive   |
| 2   | Naul et al. (1991)                | F   | 68  | Headache                            | 1.5       | Foramen magnum                   | Iso      | Hypo     | CR         | No        | 18              | No         | Alive   |
| 3   | Uematsu et al. (1992)             | M   | 62  | Gait disturbance                    | 12        | Foramen magnum to C1             | Hyper    | Hypo     | CR         | No        | 18              | No         | Alive   |
| 4   | O’Brien et al. (1995)             | F   | 49  | Unknown                             | 24        | Foramen magnum to C3             | Unknown  | Unknown  | CR         | No        | 96              | No         | Alive   |
| 5   | Hirose et al. (1997)              | M   | 66  | Gait disturbance, Bt UE numbness    | 12        | Foramen magnum to C1             | Iso      | Hypo     | CR         | No        | 3               | No         | Alive   |
| 6   | Offiah et al. (2006)              | F   | 25  | Headache                            | 9         | Foramen magnum                   | Hyper    | Hypo     | CR         | No        | 6               | No         | Alive   |
| 7   | Fan et al. (2012)                 | M   | 48  | Headache, Lt weakness and numbness | 12        | Foramen magnum                   | Hyper    | Iso      | CR         | No        | 12              | No         | Alive   |
| 8   | Bhargava et al. (2013)            | M   | 55  | Four-limb weakness                 | 12        | Foramen magnum to C2             | Hyper    | Hypo     | CR         | No        | 10              | No         | Alive   |
| 9   | Yang et al. (2016)                | 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)                 | 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). 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. MRI typically showed hyperintense or isointense on T1-weighted images and hypointense on T2-weighted images. 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. 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. 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.

Histopathologic and immunohistochemical examination is necessary to make a definite diagnosis of meningeal melanocytoma. 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. MB-1/Ki-67 labeling index helps to differentiate melanocytoma from malignant melanoma, as they are low in melanocytoma.

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. Gross total removal is generally possible because the majority of the tumors are located in the intradural extramedullary compartment. Yang et al. 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.

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.

References

1) Brat DJ, Giannini C, Scheithauer BW, Burger PC: Primary melanocytic neoplasms of the central nervous systems. Am J Surg Pathol 23: 745–754, 1999
2) Yang C, Fang J, Li G, et al.: Spinal meningeal melanocytomas: clinical manifestations, radiological and pathological characteristics, and surgical outcomes. J Neurooncol 127: 279–286, 2016
3) Chen CJ, Hsu YI, Ho YS, Hsu YH, Wang LJ, Wong YC: Intracranial meningeal melanocytoma: CT and MRI. Neuroradiology 39: 811–814, 1997
4) Fan MC, Wang JF, Fu WW, Liu K, Li LD, Sun P: Primary meningeal melanocytoma located in foramen
magnum: a case report and review of the literatures. Chin Med Sci J 27: 115–120, 2012

5) Ahluwalia S, Ashkan K, Casey AT: Meningeal melanocytoma: clinical features and review of the literature. Br J Neurosurg 17: 347–351, 2003

6) Hamasaki O, Nakahara T, Sakamoto S, Kutsuna M, Sakoda K: Intracranial meningeal melanocytoma. Neurol Med Chir (Tokyo) 42: 504–509, 2002

7) Lee JK, Rho YJ, Jeong DM, Rhim SC, Kim SJ: Diagnostic clue of meningeal melanocytoma: case report and review of literature. Yonsei Med J 58: 467–470, 2017

8) Offiah CJ, Laitt RD: Case report: Intracranial meningeal melanocytoma: a cause of high signal on T1- and low signal on T2-weighted MRI. Clin Radiol 61: 294–298, 2006

9) Bhargava P, Grewal SS, Dewan Y, Jhawar SS, Jain V, Gupta B: Craniovertebral junction melanocytoma: a case report. Turk Neurosurg 23: 539–542, 2013

10) Limas C, Tio FO: Meningeal melanocytoma (“melanotic menigioma”). Its melanocytic origin as revealed by electron microscopy. Cancer 30: 1286–1294, 1972

11) Naul LG, Hise JH, Bauserman SC, Todd FD: CT and MR of meningeal melanocytoma. AJNR Am J Neuroradiol 12: 315–316, 1991

12) Uematsu Y, Yukawa S, Yokote H, Itakura T, Hayashi S, Komai N: Meningeal melanocytoma: magnetic resonance imaging characteristics and pathological features. Case report. J Neurosurg 76: 705–709, 1992

13) O’Brien TF, Moran M, Miller JH, Hensley SD: Melanocytoma Meningeal. An uncommon diagnostic pitfall in surgical neuropathology. Arch Pathol Lab Med 119: 542–546, 1995

14) Hirose T, Horiguchi H, Kaneko F, et al.: Melanocytoma of the foramen magnum. Pathol Int 47: 155–160, 1997

15) Clarke DB, Leblanc R, Bertrand G, Quarte GR, Snipes GJ: melanocytoma Meningeal. Report of a case and a historical comparison. J Neurosurg 88: 116–121, 1998

16) Liubinas SV, Maartens N, Drummond KJ: Primary melanocytic neoplasms of the central nervous system. J Clin Neurosci 17: 1227–1232, 2010

17) Jaiswal S, Vij M, Tungria A, Jaiswal AK, Srivastava AK, Behari S: Primary melanocytic tumors of the central nervous system: a neuroradiological and clinicopathological study of five cases and brief review of literature. Neurol India 59: 413–419, 2011

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