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Case Report

A rare case report of primary malignant melanocytoma of foramen magnum and literature review

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ABSTRACT

Background: Primary malignant melanoma is an extremely rare pathology that can occur anywhere in the brain and spinal cord. Patients often present with symptoms like that of brain tumors. This similarity and inability to make a definitive diagnosis from radiological imaging alone make it a challenging diagnosis. Gross total resection is the accepted mainstay of treatment, while histopathological biopsy can confirm the diagnosis.

Case Description: A young gentleman who was otherwise healthy presented with neck pain and right-sided weakness. Magnetic resonance imaging revealed an extra-axial mass at the level of the foramen magnum. The patient underwent lateral sub-occipital craniotomy with C1 laminectomy and maximum safe resection of the lesion. Intra-operatively, a firm, moderately vascular black-colored dura-based lesion was found. The frozen section revealed highly pigmented spindle cell neoplasm, and histopathology confirmed that it was malignant melanoma. Postoperative recovery was unremarkable.

Conclusion: Primary malignant melanoma is rare and very challenging to diagnose. It demands an early diagnosis and meticulous surgical management for a favorable prognosis.

Keywords: Black tumor, Foramen magnum melanoma, Melanocytoma of brain, Primary malignant melanoma of brain

INTRODUCTION

Melanoma is primarily a cutaneous malignancy, but melanocytes are present throughout the body, including the meninges.[8] Primary melanocytic tumors of the central nervous system (CNS) were first described by Virchow in 1859.[22] Previously referred to as pigmented or melanotic meningioma, Limas and Tio later named this tumor meningeal melanocytoma in 1972.[12]

Meningeal melanocytoma is a rare primary melanocytic tumor, representing only 0.06–0.1% of all brain tumors.[9] While these tumors tend to occur throughout the CNS, they have a predilection for the posterior cranial fossa, spinal cord, and Meckel's cave.[16] Primary CNS melanomas arise in patients ranging from 15 to 71 years of age, with a peak incidence in the fifth decade.[13]

Patients often present with symptoms such as headaches, neck stiffness, focal neurological deficits, hydrocephalus, and seizures, which can resemble those of other CNS tumors.[18] This

makes it challenging to diagnose melanocytomas based solely on radiological imaging.[1,6] A biopsy of the tumor specimen and histopathological examination is essential for a definitive diagnosis, helping to differentiate melanocytomas from meningiomas, schwannomas, medulloblastomas, paragangliomas, and gliomas.[5] Gross total resection is necessary to prevent recurrence, as few cases report local recurrence following incomplete removal of the tumor.[20]

CASE REPORT

A 36-year-old gentleman, right-handed, presented with complaints of neck pain for 3 months and right-sided weakness for 2 weeks. CNS and motor examinations were unremarkable. Magnetic resonance imaging (MRI) revealed a lobulated extra-axial mass lesion measuring 4.2 cm × 3.5 cm × 2 cm located in the posterior fossa inferior to the cerebellar vermis at the level of foramen magnum [Figure 1]. He underwent lateral sub-occipital craniotomy with C1 laminectomy and maximum safe resection of the lesion. Intra-operatively, a firm, moderately vascular black-colored dura-based lesion was found [Figure 2]. The frozen section revealed highly pigmented spindle cell neoplasm, and histopathology confirmed that it was malignant melanoma (spindle cell type). Postoperative recovery was unremarkable, with no neurological deficit. 2 weeks after surgery, he underwent external beam radiotherapy. The patient was also referred to a dermatology clinic due to suspicion of melanotic lesion on his legs, and a biopsy of the said lesions was taken, which was reported negative for melanoma, confirming this as a rare case of primary melanocytic tumor of CNS.

DISCUSSION AND LITERATURE REVIEW

Neurosurgeons rarely encounter primary intracranial melanocytic tumors. Virchow first described these tumors in 1859[22], but it was not until 1972 that Limas and Tio coined the term "meningeal melanocytoma" due to the lack of ultrastructural features of meningothelium and the presence of melanosome and premelanosomes that distinguished it from other meningeal tumors.[12] Since then, several theories have been formed to understand its etiology, including three histogenic hypotheses for primary cerebral melanomas – mesodermal theory, ectodermal theory, and neurogenic theory.[18]

In the CNS, melanocytes are preferentially localized at the base of the brain, around the ventral medulla, and along the upper cervical spinal cord.[21] This is consistent with their predilection to occur in the extramedullary intradural compartment of the cervical and thoracic spine and less frequently occur intracranially in the posterior fossa, Meckel's cave and cerebellopontine (CP) angle[24] and very rarely in the supratentorial compartment.[2]

According to the fifth edition of the WHO classification of tumors, melanocytic tumors are classified as diffused meningeal melanocytic neoplasms, which include meningeal melanocytosis, meningeal melanomatosis, and circumscribed meningeal melanocytic neoplasms which include meningeal melanocytoma and meningeal melanoma.[15]

As with any CNS tumor, the presentation of patients often depends on the location of the lesion. Those with melanomas in the CP angle cistern or posterior fossa typically show cranial nerve involvement and cerebellar signs such as axial or appendicular ataxia, as well as central vertigo. In contrast, patients with intracranial melanomas usually present with symptoms such as headaches, seizures, hemiparesis, or visual disturbances, which was the case with our patient, while spinal cord melanomas often lead to varying degrees of motor weakness, sometimes accompanied by bowel or bladder dysfunction.[10]

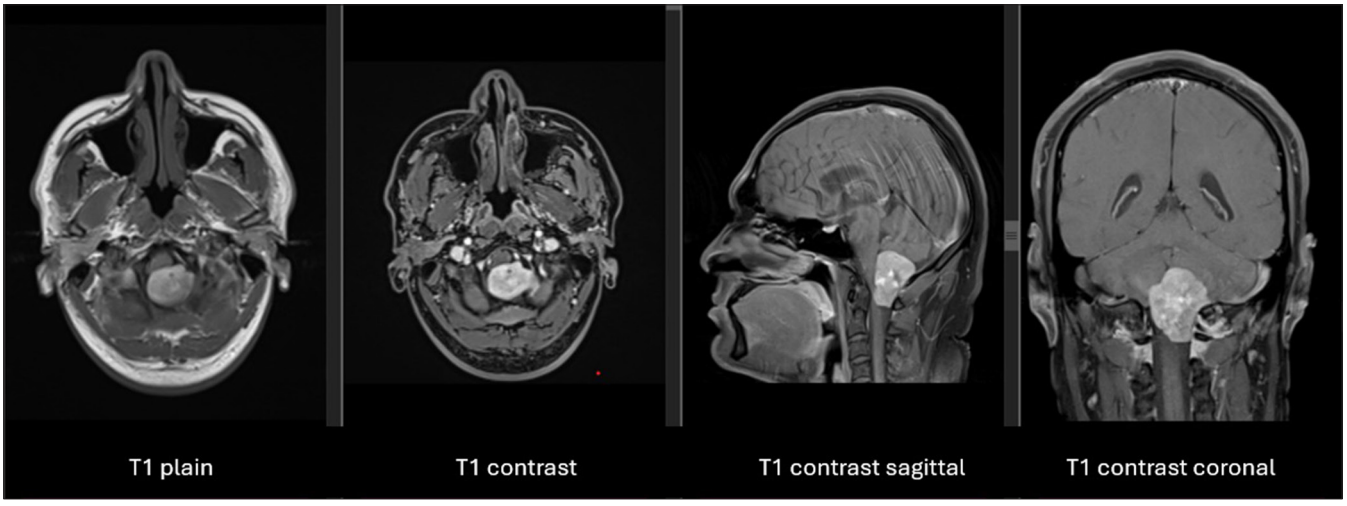

Figure 1: Magnetic resonance imaging showing extra-axial lesion at the level of foramen magnum.

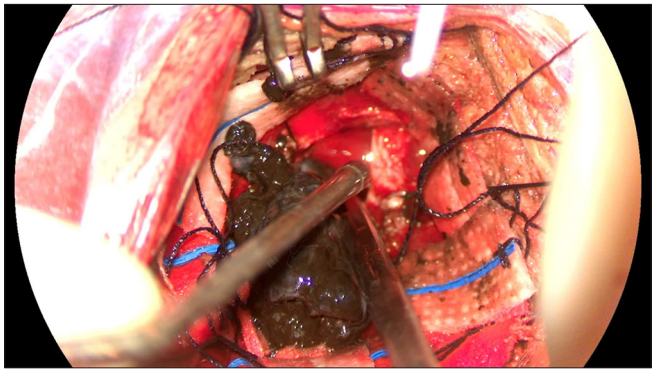

Figure 2: Intra-operative picture showing dark colored lesion, characteristic of melanoma.

Literature reveals that the median age is 40 years for patients with intracranial tumors and 49 years for those with spinal tumors with slight female predominance (57.9%). Recurrence rates of 26.3% and a mortality rate of 10.5% have been observed in a 46-month follow-up period.[17]

Computed tomography (CT) scans reveal melanomas as hyperdense, extra-axial masses with irregular borders displaying homogeneous enhancement, which is often difficult to distinguish from meningioma.[4] On MRI, melanomas are classically hyperintense on T1-weighted images, secondary to the presence of melanin. Although T1 hyperintensity is considered to be a classic finding, this pattern is present in only 24–47% of lesions and, on the basis of histologic analysis, is associated with lesions with >10% of cells containing melanin.[25] Amelanotic melanomas appear isointense to hypointense on T1-weighted images and moderately hyperintense on T2-weighted images. Moderate perilesional edema is typically present.[23] A spoke-wheel pattern, which is a characteristic finding of meningioma on angiography, is also observed in cases of melanoma.[3] This radiologic resemblance of melanoma with other CNS pathology makes an early diagnosis difficult, which merits the intra-operative and histopathological findings for definitive diagnosis. Grossly, it appears black, dark brown, blood clot-like, locally invasive tumor. The adjacent meninges and skull are usually black stained.[14] Histologically, meningeal melanocytomas show spindle-shaped cells arranged in clusters with varying amounts of pigment.[19] Mitotic figures are usually absent but if increased mitotic activity is found, it is regarded as intermediate-grade melanocytoma of leptomeninges.[6] Electron microscopy can reveal key features such as melanosomes, the absence of desmosomes, interdigitations, and the basal lamina, helping to distinguish melanocytoma from other lesions such as meningiomas and schwannomas.[11] Gross total resection is the mainstay treatment for melanocytoma, ensuring a good prognosis and a 100% survival rate. Incomplete resection warrants a 5-year survival rate of 46%, but adjuvant radiotherapy can increase this to 100%[19], while metastatic cases have a mean survival of 5–6 months.[7] Postoperatively, full-body CT is warranted to rule out any metastatic lesions, followed by MRI brain at regular intervals for surveillance of recurrence. This case highlights the importance of early detection, meticulous surgical management, and vigilant follow-up in ensuring optimal outcomes for patients with this rare but challenging condition.

CONCLUSION

Primary malignant melanoma of the central nervous system (CNS) is an exceptionally rare and aggressive pathology that poses significant diagnostic and therapeutic challenges. Its clinical presentation often mimics that of more common brain tumors, which can complicate the process of reaching a definitive diagnosis. Radiological imaging alone is insufficient for a clear diagnosis, making histopathological examination following surgical biopsy a crucial step in confirming the condition. This case highlights the importance of considering primary malignant melanoma in the differential diagnosis of CNS lesions, particularly in patients presenting with unexplained neurological symptoms, such as neck pain and weakness. Early identification and prompt intervention are vital for improving patient outcomes. In this patient, the successful surgical resection of the tumor, coupled with the accurate histopathological confirmation, led to an unremarkable post-operative recovery, underscoring the importance of comprehensive surgical management. Given the rarity of this condition, a multidisciplinary approach involving neurosurgeons, oncologists, and pathologists is essential for optimal care.

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REFERENCES

1. Badhe U, Balasubramanian R, Varadhrajan S, Subramaniam R. A black mass at skull base-Foramen magnum melanocytic meningioma. Interdiscipl Neurosurg 2021;23:100899.
2. Beseoglu K, Knobbe CB, Reifenberger G, Steiger HJ, Stummer W. Supratentorial meningeal melanocytoma mimicking a convexity meningioma. Acta Neurochir (Wien) 2006;148:485-90.
3. Buetow MP, Buetow PC, Smirniotopoulos JG. Typical, atypical,

and misleading features in meningioma. Radiographics 1991;11:1087-106.
4. Chen CJ, Hsu YI, Ho YS, Hsu YH, Wang LJ, Wong YC. Intracranial meningeal melanocytoma. Neuroradiology 1997;39:811-4.
5. Dulai MS, Moes GS, Briley AL, Galperin IB, Smyth L, Cherry AM, *et al*. Gliosarcoma with melanocytic differentiation. Acta Neuropathol 2008;115:357-61.
6. Ferreira de Araújo MG, Almondes Santana Lemos LE, Negromonte Guerra PL, Marcia Dos Santos Lima Didjurgeit F, Batista Cezar A Jr., Faquini IV, *et al*. Supratentorial meningeal melanocytoma mimicking meningioma: Case report and literature review. Pathol Oncol Res 2023;29:1611482.
7. Greco Crasto S, Soffietti R, Bradac GB, Boldorini R. Primitive cerebral melanoma: Case report and review of the literature. Surg Neurol 2001;55:163-8; discussion 168.
8. Heistein JB, Acharya U, Mukkamalla SK. Malignant melanoma. In: StatPearls. Treasure Island, FL: StatPearls Publishing; 2025.
9. Jaiswal S, Jaiswal AK, Vij M, Behari S, Pandey R. Primary meningeal melanocytoma of cerebellopontine angle: A case report with 12 years follow up. Basic Appl Pathol 2011;4:99-102.
10. Kavin D, Purandare A, Wankhade R, Tapse M, Naren N, Diyora B. Primary melanocytic melanoma of foramen magnum. J Spinal Surg 2023;10:21-4.
11. Küsters-Vandevelde HV, Küsters B, van Engen-van Grunsven AC, Groenen PJ, Wesseling P, Blokx WA. Primary melanocytic tumors of the central nervous system: A review with focus on molecular aspects. Brain Pathol 2015;25:209-26.
12. Limas C, Tio FO. Meningeal melanocytoma ("melanotic meningioma"). Its melanocytic origin as revealed by electron microscopy. Cancer 1972;30:1286-94.
13. Liubinas S V, Maartens N, Drummond KJ. Primary melanocytic neoplasms of the central nervous system. J Clin Neurosci 2010;17:1227-32.
14. Long WL, Chen FY, Huang XL, Lu JX, Xu YN. Imaging and pathological diagnosis of primary intracranial malignant melanoma: A case report and literature review. Medicine 2023;102:e32767.
15. Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, *et al*. The 2021 WHO classification of tumors of the central nervous system: A summary. Neuro Oncol 2021;23:1231-51.
16. Puyana C, Denyer S, Burch T, Bhimani AD, McGuire LS, Patel AS, *et al*. Primary malignant melanoma of the brain: A population-based study. World Neurosurg 2019;130:e1091-7.
17. Rahimi-Movaghar V, Karimi M. Meningeal melanocytoma of the brain and oculodermal melanocytosis (nevus of ota): Case report and literature review. Surg Neurol 2003;59:200-10.
18. Rodriguez y Baena R, Gaetani P, Danova M, Bosi F, Zappoli F. Primary solitary intracranial melanoma: Case report and review of the literature. Surg Neurol 1992;38:26-37.
19. Singh Sodhi HB, Salunke P, Sahoo SK, Radotra BD, Kumar N. Primary ventral foramen magnum meningeal melanocytoma. Neurol India 2014;62:230-2.
20. Smith AB, Rushing EJ, Smirniotopoulos JG. Pigmented lesions of the central nervous system: Radiologic-pathologic correlation. Radiographics 2009;29:1503-24.
21. Tang K, Kong X, Mao G, Qiu M, Zhu H, Zhou L, *et al*. Primary cerebral malignant melanoma: A case report with literature review. Medicine 2017;96:e5805.
22. Virchow R. Pigment und diffuse Melanose der Arachnoides. Arch Pathol Anat Physiol Klin Med 1859;16:180-2.
23. Wadasadawala T, Trivedi S, Gupta T, Epari S, Jalali R. The diagnostic dilemma of primary central nervous system melanoma. J Clin Neurosci 2010;17:1014-7.
24. Wagner F, Berezowska S, Wiest R, Gralla J, Beck J, Verma RK, *et al*. Primary intramedullary melanocytoma in the cervical spinal cord: Case report and literature review. Radiol Case Rep 2015;10:1010.
25. Wong VK, Lubner MG, Menias CO, Mellnick VM, Kennedy TA, Bhalla S, *et al*. Clinical and imaging features of noncutaneous melanoma. Am J Roentgenol 2017;208:942-59.

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