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Image Report

Posterior fossa melanocytic schwannoma extending to the cervicothoracic spinal cord: A clinical rarity

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ABSTRACT

Background: Melanocytic schwannomas (MSs) are rare, malignant peripheral nerve sheath tumors with only 200 cases reported to date. These pose imaging and pathological challenges for definitive diagnosis.

Case Description: A 25-year-old lady presented at our center with a prolonged history of gait disturbance, left ear tinnitus, headaches, and drowsiness. MRI findings showed a midline cystic lesion in the posterior cranial fossa extending caudally to the D1 vertebral body, with marked central hypointensity, and peripheral hyperintensity on T1-weighted images. A suboccipital craniotomy and debulking of the lesion were performed, showing a hyperpigmented, infiltrative tumor adherent to the surrounding structures. This was confirmed as a melanocytic schwannoma on histopathological analysis.

Conclusion: Posterior fossa MSs involving cervicomedullary region and extending distally to cervicothoracic spinal cord are rare and complex cases, particularly with regard to difficulty diagnosing preoperatively and surgical resection.

Keywords: Cervicothoracic, Melanocytic schwannoma, Neuro-oncology, Posterior fossa

INTRODUCTION

Melanocytic schwannomas (MSs) are a rare variant of benign Schwann cell neoplasms typically arising in the early middle age with a predilection for extracranial regions.[5] Although MS accounts for <1% of all neurolemmomas, the malignant potential of these tumors cannot be undermined.[1]

CASE DESCRIPTION

A 25-year-old right-handed lady presented to our neurosurgery clinic with progressively worsening complaints of gait disturbance, left ear tinnitus, headaches, and drowsiness for the past 6 months. On neurological examination, she exhibited hyperreflexia in both arms and left lower leg and was unable to walk in a straight line.

Preoperative MRI scans showed a solid-cum-cystic lesion with enhancement along the midline in the posterior fossa extending distally into the cervical spinal cord reaching to the level of upper

endplate of the D1 vertebral body, measuring approximately 13.3 × 4.6 × 3.4 cm [Figure 1]. T1-weighted images showed hypointensity centrally and hyperintensity in the peripheral region, with contrary findings on T2-weighted images. The lesion was causing effacement of the 4th ventricle resulting in hydrocephalus.

We performed a suboccipital craniotomy for resection of the lesion. For improved exposure, part of the C1 ring was removed and cisterna magna opened. A hyper-pigmented and black colored tumor with no clear margins was visualized [Figure 2]. The lesion was soft and fibrous in consistency with moderate vascularization. It was infiltrative and adhesive to the surrounding cranial structures including the brain stem. Therefore, it was decided to debulk as much of the tumor as safely possible without attempting the resection of lesion involving the cord distally. Intraoperative frozen section was inconclusive due to heavy pigmentation. On further analysis, histopathology confirmed a malignant neoplasm of melanocytic origin. Psammoma calcifications with a low Ki-67 index and equivocal HMB45 and Melan-A stains were seen. This made melanocytic schwannoma a more likely possibility than melanoma.

Postoperatively, the patient had an uneventful hospital course and discharged home on the 3rd day after surgery. On the most recent follow-up, her drowsiness and headaches had improved, with some difficulty in walking requiring support. She was referred to medical and radiation oncology services for further management.

DISCUSSION

MS may present in the craniovertebral junction (CVJ) with no pathognomonic signs as 29% of cases present asymptomatically.[2] Correlating clinical findings with radiology are also crucial to delineate MS from melanocytoma and melanoma; MS more likely involves spinal nerves or dorsal root ganglia.[4] While sporadic MS may show variable contrast enhancement, psammomatous MS shows calcific hyperdensities on CT.[7] However, MR imaging is the established gold standard through which the extent of melanization determines clinical suspicion. Sparsely melanotic MS can be mistaken for benign schwannomas, that is, hypointense on T1 and hyperintense on T2.

Spinal instability and progressive neuropathy are strong indications for operative intervention in craniocervical MS.[3] An optimal surgical approach should enable convenient tumor resection and reconstruction without collateral neurological damage. The posterolateral approach is considered the workhorse for craniovertebral MS through laminectomy or laminoplasty.[6] Ultimately, the goals of surgery should be

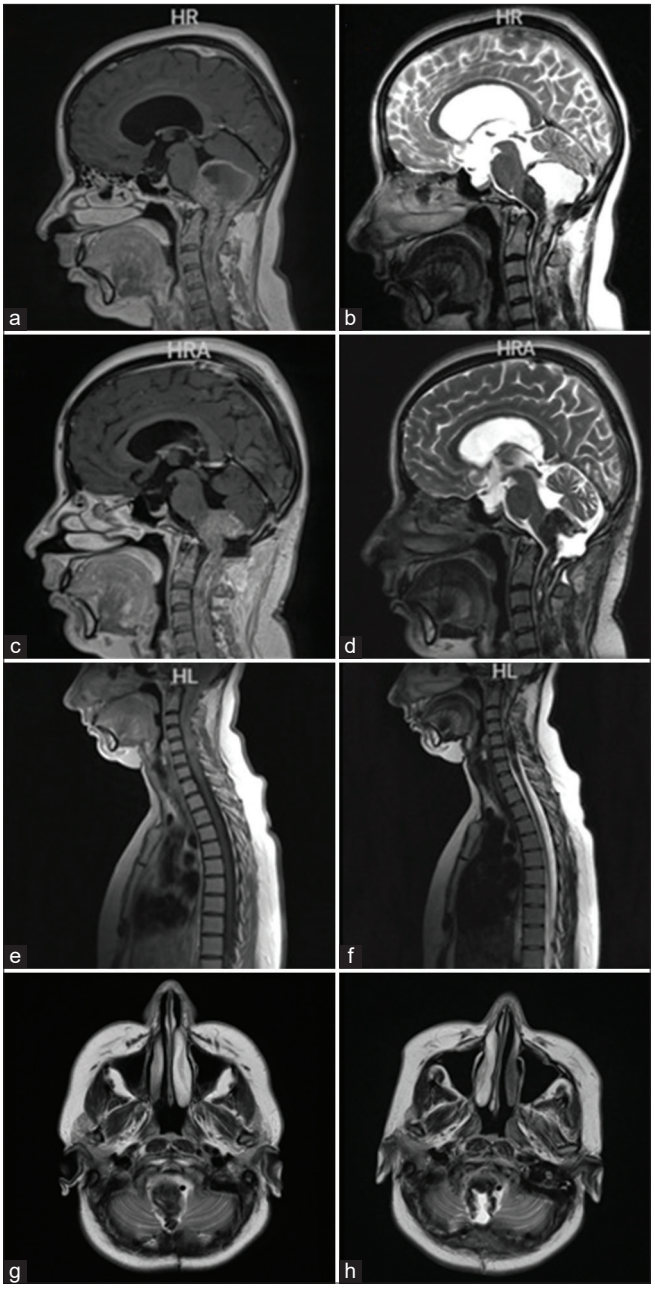

Figure 1: Preoperative MRI images: (a) Coronal T1-weighted image showing a posterior fossa lesion with a homogeneous center and peripheral high-signal intensity rim, seen to be extending down through to the cervicomedullary junction. Upward displacement of the brainstem can be seen as well as the basilar artery, with compression of the cerebellum. (b) Coronal T2-weighted image. Postoperative MRI images: (c) Coronal T1-weighted image with posterior resection cavity. (d) Coronal T2-weighted image. (e and f) Coronal T1-weighted and T2-weighted images showing extension of the lesion to the thoracic spinal cord. (g and h) Axial T2-weighted images depicting the lesion at the skull base and craniocervical junction, pre- and postoperatively.

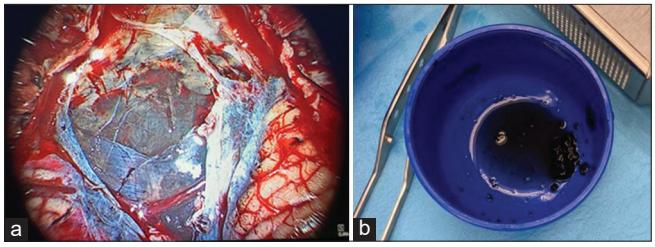

Figure 2: Intraoperative images showing the fibrous, adherent tumor bed (a) and highly pigmented tumor (b).

to achieve local tumor control, symptomatic relief, accurate histological diagnosis, and prolong survival time for such patients. Nonsurgical management such as radio-, chemo-, or targeted therapy is recommended for asymptomatic lesions with minimal growth or when complete tumor resection is not possible secondary to local infiltration.

Recent literature advocates for MS to be considered a malignant entity with incompletely excised tumors involving the neurovasculature having the worst prognosis. In the largest series, Torres-Mora *et al*. found rates of 42% distant metastases, 35% local recurrence, and 27% disease-specific mortality.[8] While metastasis can occur hematogenously or through CSF seeding, the recurrence-free period generally varies from 3 to 20 months for intracranial and intramedullary lesions.

CONCLUSION

Posterior fossa MS involving CVJ and spinal cord distally poses a significant neurosurgical challenge in terms of diagnosis and successful intervention. Careful preoperative evaluation and intraoperative prudence are crucial for optimal management.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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