Multifocal Spinal Meningeal Melanocytoma: An Illustrated Case Review

ABSTRACT
Primary melanocytic tumors of the central nervous system are rare. In this article the authors describe a case of C1C2 intradural extramedullary melanocytoma in a 43-year-old patient who presented with neck pain. C1-3 laminectomy was performed followed by excision of the lesion and an adjoining satellite nodule, along with the dural attachment. The histopathological features were consistent with a meningeal melanocytoma despite the presence of a satellite nodule. The patient has no evidence of recurrence during the six month follow up period. A brief review of literature pertaining to the radiological features, pathological findings, management and prognosis of this rare tumor is discussed.

KEYWORDS: Multifocal spinal meningeal melanocytoma, Intradural extramedullary, Cervical spine

INTRODUCTION
Spinal meningeal melanocytomas are rare, primary melanocytic tumors with a female preponderance and peak incidence in the fifth decade. Although benign, these tumors have a tendency for local recurrence. To the best of our knowledge, only two cases of multifocal melanocytomas have been reported so far in literature.

CASE SUMMARY
A 43-year-old male presented with history of dull aching neck pain in the suboccipital region for 6 months. The pain was non-radiating with no aggravating or relieving factors. Detailed neurological examination did not reveal any motor or sensory deficits. MRI cervical spine revealed a C1C2 intradural, extramedullary oval shaped mass located dorsal to the cord. The tumor was hyperintense on T1WI, hypointense on T2WI and showed homogeneous enhancement on contrast administration (Figure 1 A-C).

C1-3 laminectomy was performed through a posterior midline approach. Perioperatively, the duramater as well as the tumor were black in color (Figure 2 A-C). The tumor had a dural attachment and was deriving its vascularity from extradural vessels. Another 5 mm nodule with dural attachment was noted about a centimeter from the lower pole of the tumor. A complete excision of tumor and the nodule with their dural attachments was performed.

The perioperative impression was of a metastatic melanoma. A search for the occult primary with clinical examination, ultrasound abdomen and ocular examination did not reveal any obvious source. The histopathology of the mass showed spindle cells arranged in fascicles. The cells had elongated nuclei with vesicular chromatin and prominent nucleoli. However mitosis or necrosis was not seen (Figure 2 D-F).

Immunohistochemistry with HMB-45, S-100 and Vimentin were positive. The Ki67 labelling index was less than 1%. The possibility of meningioma with melanotic deposits was excluded by a negative Epithelial Membrane Antigen staining. The patient has no recurrence of the lesion at 6 months follow up (Figure 1D-F).

DISCUSSION
Melanocytes are pigmented cells normally found in the leptomeninges with highest concentration in the upper cervical spine and the ventrolateral medulla (3). This peculiar distribution of the melanocytes explains the preponderance of melanin containing tumors in these regions. Melanin containing tumors of the central nervous system include...
meningiomas, schwannomas, primitive neuroectodermal tumors apart from melanocytomas and melanomas.

The term ‘meningeal melanocytoma’ was first proposed by Limas and Tio in 1972 based on ultrastructural features to differentiate these benign pigmented tumors from those originating from meningothelial fibroblasts (3). Since then, less than 75 cases of meningeal melanocytomas have been reported in literature, half of which are spinal in location (1, 5).

Spinal meningeal melanocytomas occur commonly in the fifth decade with a female preponderance. They may be extra and/or intradural, presenting with radiculopathy, myelopathy or both. Rare instances of intramedullary melanocytomas have also been reported in literature (9). On MRI, they are usually solitary lesions that appear iso-hyperintense on T1WI, iso-hypointense on T2WI and show homogeneous enhancement on contrast administration. The degree of T1 and T2 shortening is related to the paramagnetic free radicals contained in melanin (indole, semiquinone and semiquinonimines) as well as the extent of fibrosis within the tumor. Melanomas, on the other hand, may display a heterogeneous signal as a result of hemorrhage within the tumor (4,10).

The World Health Organization has classified primary melanocytic tumors of the nervous system into diffuse melanocytosis, melanocytoma, malignant melanoma and meningeal melanomatosis (6). All melanin-containing tumors show presence of spindle or epithelioid cells arranged in sheets, bundles, nests or whorls surrounded by a fine network of reticulin fibres, with variable amounts of melanin pigment in the cytoplasm. The difference between melanocytoma and melanoma is the absence of nuclear atypia, mitotic figures, necrosis and microvascular invasion in the former and a high proliferation index (Ki 67 > 5%) in the latter. A positive immunoreactivity for S100 protein, Vimentin, HMB-45 and antimelanoma antibody with a negative immunostaining for Epithelial Membrane Antigen, Glial Fibrillary Acidic Protein and Neuron Specific Enolase favors the diagnosis of a melanocytic tumor (5). Malignant transformation of a melanocytoma to melanoma following complete excision has been reported in the literature (12).

Figure 1: Preoperative MRI of the spine showing an intradural extramedullary lesion that is hyperintense on T1WI (A), hypointense on T2WI (B) and enhancing homogenously on contrast administration (C). Post operative MRI performed at 6 months shows no residual tumor (D-F).
Gross total removal of tumor is associated with best prognosis. Local recurrences have been reported 1 to 12 years following incomplete excision (2,3,5). Radiotherapy as an adjuvant treatment modality for incompletely excised lesions has only 40% success rate in preventing local recurrence (7,9). Verma et al. reported a remission of 15 months in a thoracic melanocytoma following adjuvant chemoinmunotherapy (11).

Two cases of multifocal melanocytoma have been reported in literature till date. Shownkeen et al. reported a multiform

Figure 2: Perioperative photographs showing the black pigmentation of the duramater (arrow heads on figure (A) and the tumor (B) with a dural attachment. The excised tumor and the satellite nodule (arrow) are depicted in figure (C). Histopathological slides showing spindle cells containing melanin pigment arranged in fascicles (H&EX200) (D). Tumor cells have prominent nucleoli without mitosis (H&EX400) (E). Ki67 labelling index is less than 1% (IHC HRP PolymerX200) (F).
cervical melanocytoma at C3 level with additional 5 mm lesions at C4 and C5 levels. The long-term follow-up of the case did not find mention in the report (2). Ali et al. reported a case of multifocal melanocytoma involving thoracic spinal cord and bilateral cerebellopontine angles with diffuse leptomeningeal hyperpigmentation and hydrocephalus (8). A ventriculoperitoneal shunt and leptomeningeal biopsy was performed followed by an emergency spinal cord decompression through a posterior approach. Histopathological features of the thoracic lesion were consistent with melanocytoma while the cerebellopontine lesions were presumed to be melanocytomas based on the imaging similarities with the thoracic lesion. The patient expired a few weeks later due to brain stem compression which led the authors to conclude that multifocal meningeal melanocytoma has an aggressive clinical course resulting in poor prognosis.

The poor prognosis in the above case is largely influenced by the location of the lesion causing brainstem compression. In our case, the location of the lesions in the cervical spine and their complete excision along with the involved duramater has probably resulted in good prognosis, though longer follow up is needed to make any conclusive comments.

SUMMARY

Spinal meningeal melanocytomas are rare tumors with a benign biological behavior, having a tendency for local recurrence. Surgery is aimed at complete removal of tumor with its dural attachment. Radiotherapy is reserved for incompletely excised lesions and multiple lesions not amenable for surgical excision. More such cases need to be reviewed as the existing literature pertaining to the prognostic factors in multifocal melanocytomas is unclear.

REFERENCES