

Primary meningeal melanocytoma

Khadilkar UN¹, Agarwal N², Deeshma², Bhat V³

¹Professor, ²Postgraduate, ³Assistant Professor, Department of Pathology, Kasturba Medical College, Mangalore, India

Abstract

Meningeal melanocytoma is an uncommon benign pigmented primary lesion that has to be distinguished as a pathological entity separate from other benign melanin containing tumours as well as its overtly malignant counterpart, melanoma. In the present study, a 58 year old lady presented with bilateral sensory and motor symptoms in the lower limbs. The MRI scan showed a dumb-bell shaped lesion in the spinal canal at 6th cervical vertebra to 1st thoracic vertebra (C6-T1) level mimicking a schwannoma and histologically proved to be melanocytoma.

Key words: melanocytoma, histology.

The term meningeal melanocytoma was first coined to describe a primary melanocytic tumour of the leptomeninges with prolonged clinical course and benign histology¹. Most of the reported cases of meningeal melanocytomas have occurred either in the spinal canal or the posterior fossa. Although more rarely located supratentorially and adjacent to cranial nerve nuclei, a number of tumours have been described in the Meckel cave¹. It comprises 0.06 % to 0.1 % of brain tumours. It behaves biologically like meningiomas and is prone to invade adjacent structures and recur locally².

Case report

A 58 year old lady presented with bilateral pin-prick sensation in the lower limbs of 2 months duration. She had inability to walk and gradually increasing weakness from 15 days. On examination, she was found to have decreased power in the right lower limb. MRI Scan showed a dumb-bell shaped lesion in the spinal canal at the level of C6-T1 extending into the thoracic cavity up to the apex of the lung and superiorly up to the periosteum of the first rib (Fig 1). The lesion enhanced homogeneously after the addition of Gadolinium.

Pathological findings

Grossly the tissue bits were brownish black, nodular and measured 4.5x3x2 cm. Microscopically the lesion was composed of variably pigmented cells, predominantly epitheloid and arranged in nests with a few whorls and fascicles composed of spindly cells. The cell cytoplasm was abundant, eosinophilic with numerous coarse melanin granules masking the nuclei. A melanin bleach revealed uniform, oval nuclei with granular chromatin and single distinct eosinophilic nucleoli. The stroma showed proliferating blood vessels. There was no

mitosis, necrosis or neural parenchymal invasion (Fig 2). Reticulin stain showed reticulin fibres surrounding nests of tumour cells (Fig 3) and S-100 protein and HMB-45 positivity was noted on immunohistochemistry.

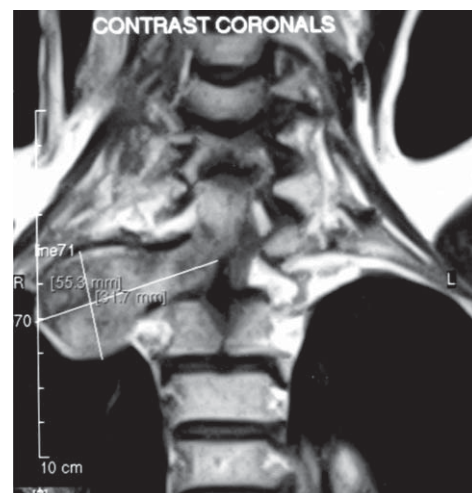


Fig 1: CT Scan showing dumb bell shaped tumour.

Correspondence

Dr. Urmila N. Khadilkar
Professor of Pathology
Kasturba Medical College
Mangalore-575001, India
E-mail: urmilank@yahoo.co.in

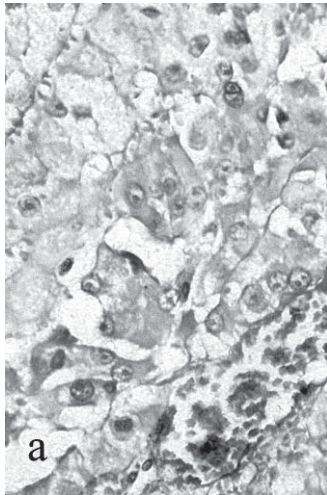


Fig 2a: Melanin bleach showing the distinct nucleoli in the cells.

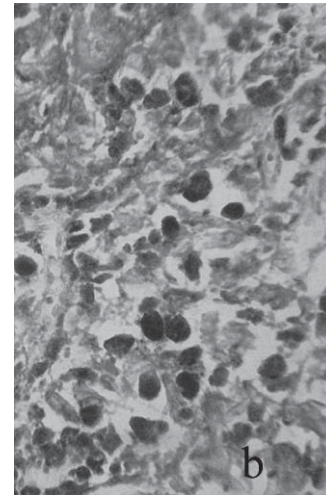


Fig 2b: Melanocytoma cells loaded with melanin pigment. (H&E x 400).

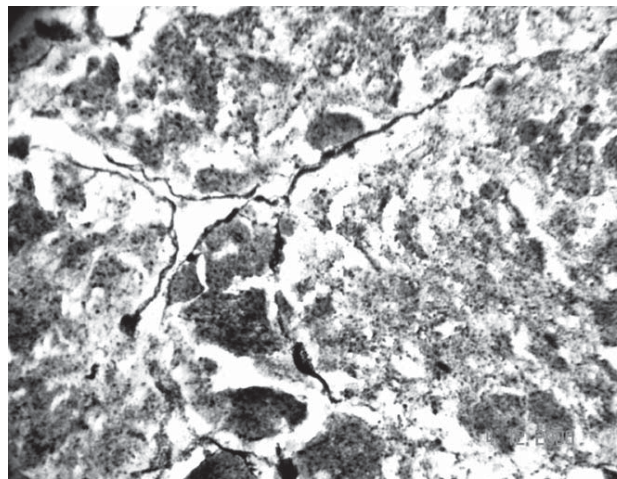


Fig 3: Nests of tumour cells surrounded by reticulin fibres. (Gordon & Sweet's x400).

Discussion

Meningeal melanocytoma is a benign tumour that originates from the melanocytes and presents as an extra-axial mass in the intradural, extramedullary space, attached to or arising from the leptomeninges³. On computer tomography (CT) scan these tumours are iso- to high density lesions that are enhanced by the addition of contrast media³. The signal features of iso- to high intensity T1- weighted images and low intensity T2 weighted images are considered unique and contrary to those generally seen among tumours of CNS. However the signal intensities vary depending on the degree of melanisation³.

On magnetic resonance (MR) meningiomas are difficult to differentiate from melanocytomas as the signal intensities are similar, whereas malignant melanomas differ in that they often display a heterogeneous signal as a result of the propensity to haemorrhage¹.

Both macroscopic and microscopic features of the tumour in the present study resembled those seen in meningioma and melanotic schwannoma. Although the tight nesting architecture of melanocytomas can be readily recognized, tumours with diffuse sheet like patterns or vasocentric fascicular patterns can be confused with other melanin-producing tumours, especially solitary pigmented lesions arising in the vicinity of spinal root exit zones⁴. Meningiomas feature tight cellular nests or whorls and occasionally melanocytes are trapped within a growing meningioma, but such cells are few and clearly do not represent a component of the tumour. Psammoma bodies have not been described in association with melanocytomas. Reactivity of meningioma cells for EMA helps in cases in which a ready distinction cannot be made on routine histology⁵.

Melanotic schwannomas often lack Antoni A and B patterns and more often show spindle and epithelial cells with nuclei having cytoplasmic pseudoinclusions. Because schwannoma cells are individually surrounded by basement membrane particularly at stromal interfaces, reticulin and immunohistochemical stains for laminin or collagen type IV can be useful⁵. An uncommon psammomatous variant of melanotic schwannoma, can be dumb-bell shaped involving the spinal foramina.

The clinically aggressive melanoma shows the same anatomical distribution and histology as melanocytoma except for the overt anaplasia, hypercellularity, increased mitotic activity and CNS invasion. Primary and metastatic melanomas show considerable histologic overlap although extensive necrosis, more obvious malignancy and higher mitotic indices are seen in metastatic melanomas.

It is important for pathologists to be aware of melanocytoma, which may mimic a primary or metastatic melanoma. It must also be distinguished from the histologically similar lesions of the CNS, including melanotic schwannoma and meningioma, and from tumours like medulloblastomas, pinealoblastomas and ependymomas which may also exhibit melanin.

Meningeal melanocytomas are biologically benign and can be cured by complete surgical resection although local recurrences have been reported in spite of gross – total resection and rare cases of cerebral and hepatic metastases have been reported^{3,6}.

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