Acute Onset of Paraplegia Due To Melanocytic Cervico-dorsal Lesion: Case Report

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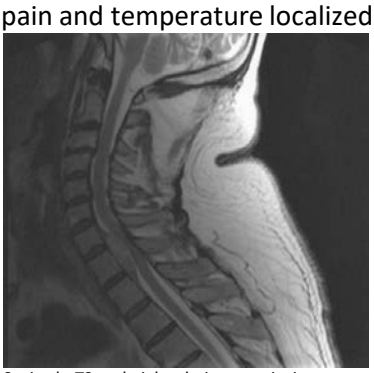
Introduction

Melanocytomas are rare pigmented tumors of the leptomeningeal melanocytes. They are benign but locally aggressive lesions and are very rarely associated with spinal localization; the pathogenesis remains unclear. Usually, spinal melanocytoma present with radiculopathy with or without progressive myelopathy. Acute paraplegia is a very rare clinical finding. We present a 62-years-old man who presented acute paraplegia due to C7-T1 intradural extramedullary anterior melanocytoma.

Case presentation

A 62 years-old previously healthy man was admitted to our hospital for acute onset of paraplegia. In the past 2 months he suffered of interscapular pain. The day of admission he presented lower limbs weakness which worsened rapidly to complete paraplegia. On neurological examination, the patient presented paraplegia with a sensory level to pain and temperature localized in T2, with preservation of proprioception.



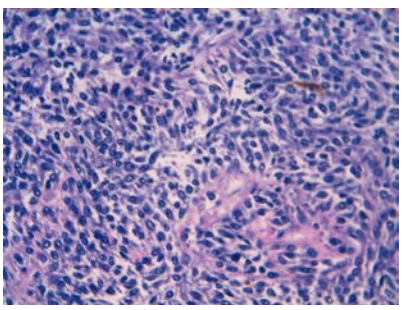
Sagittal T2 wheighted image: isointense extramedullary mass with dural attachment.

MRI showed an intradural extramedullary mass on C7-T1 level located anterior to the spinal cord with isointense signal and wide dural attachment as highlighted in T2 weighted image; mild heterogeneous enhancement in T1 post contrast images. The patient underwent emergency decompressive C7-T1 laminectomy with subtotal removal of the lesion. Intraoperatively, cervico-dorsal dura was discolored black and the lesion was firm and blackish in colour. Histopathologic examination revealed large cells, oval to polygonal in shape with prominent nucleoli, eosinophilic cytoplasm filled with melanin pigment. No significant increase in mitosis (1/10 HPF); low proliferation index (Ki 67: 2%). On immunoistochemestry: positivity with S100, Vimentin and HBM45 and negativity for EMA. Retrospectively, examination of the skin and the fundus of the eye did not reveal any melanotic lesions.

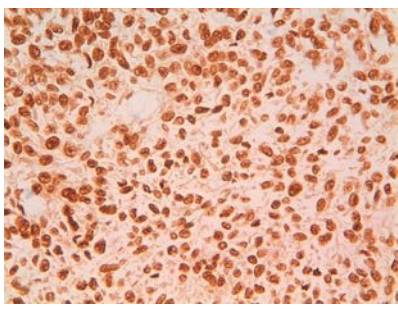
Post-operative MRI revealed expansion of cervicothoracic cord and the subtotal removal of the lesion. Neurological status improved in the postoperative period; after 1 week the patient was discharged to Rehabilitation Unit with a subtotal recovery of motor function.

Discussion

Melanocytes are cells of neural crest origin and are normally found in the leptomeningeal layers. Melanocytoma can commonly present in posterior fossa and in foramen magnum; in the spine, upper cervical region is the more involved. Compared to the metastatic lesions, primary melanocytomas has an incidence of 1 per million. Clinically, the tumor occurs commonly in the fifth decade and is more common in females.



Spindle-shaped cells with focal melanin pigmentation.



Immunohistochemical positivity for S100

The differential diagnosis include schwannoma, medulloblastoma and neurofibroma. Radiologically, MRI is the exam of choise. It could appear isointense or hyperhintense on T1 weighed images and iso or hipointense on T2 weighed images, with not homogeneus enanchment on postcontrast images; it is related to the degree of melanin content.
Usually melanocytoma presents with signs of progressive compression of adjacent nervous structures: myelopathy, radiculopathy, but also multiple cranial nerve palsies or chronic spinal arachanoiditis. Acute onset of symptoms is unusual, due to the benign nature of this tumor and its relatively slow growth. Even if acute paraplegia is reported as final symptom, it's a very rare and often unexpected onset.
Histologically melanocytomas feature variably pigmented spindle cells growing in tight nests. Melanocytomas are immunoreactive for S-100 protein, monoclonal melanoma antibody (HMB-45), and vimentin antibody; they are also negative for epithelian membrane antigen (EMA), which is an indicator of meningioma. Complete surgical excision is the treatment of choice; although these are benign tumors, risk of recurrence is noted; therefore, adjuvant radiation therapy is adivised in both comple and incomplete recection.

Conclusions

In this case we show an unusual presentation characterized by acute onset of paraplegia treated in urgent surgery with subtotal excision. Melanocytoma is rare, histologically benign tumor with good prognosis when compared to melanoma; however, local aggressive behavior has been recorded, especially in cases of subtotal gross resection. Even if the black dura and the severe paraparesis may forecast dark shadows over the clinical picture, the overall prognosis for spinal melanocytoma remains bright.

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