

Case report

Primary melanoma of the cauda equina: Case report and review of the literature

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ABSTRACT

The authors report the case of an 82 year-old woman with a primary malignant melanoma of the cauda equina resembling lumbar schwannoma in the MRI study. Melanocytic neoplasms are very rare but they should be included in the differential diagnosis of lesions involving the spinal nerves. The treatment of choice for these lesions is complete resection followed by radiotherapy. The outcomes reported in the literature are variable and are associated with the age of presentation, histopathological findings, extent of surgical resection and absence of metastatic lesions.

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Melanoma primario de la cauda equina: caso clínico y revisión de la bibliografía

RESUMEN

Se presenta el caso clínico de una paciente de 82 años con un melanoma maligno primario de la cauda equina, que en las imágenes de resonancia magnética parecía un schwannoma lumbar. Las neoplasias melanocíticas son muy raras, pero se deben incluir en el diagnóstico diferencial de las lesiones que afectan a los nervios espinales. Su tratamiento de elección es la resección completa, seguida de radioterapia. La evolución de estos pacientes es muy variable y va a depender de la edad del paciente, de los hallazgos histopatológicos, de la resección quirúrgica y de la ausencia de metástasis.

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Introduction

The primary melanoma of the cauda equine is an extremely rare neoplasm of the lumbar nerve roots. It is thought that this type of tumor is derived from the leptomeningeal melanocytes¹. It has been reported that these patients had a better prognosis than those with a supratentorial primary melanoma but they can behave aggressively too and carry a bad prognosis²⁻⁴. The treatment of choice is a complete surgical excision followed by radiotherapy with a variable response^{4,5}. To the best of our knowledge only nine cases of primary melanoma arising from a nerve root have been described in the world literature. In the present report we present the clinical, radiological and pathological features of this infrequent malignant lesion.

Case report

History

This 82 year-old woman presented with a one month history of intolerable low back pain radiating to the anterior surface of her left thigh. Her medical history was uneventful and no previous traumatic event was referred. Initially, she was treated with oral analgesia but she came to our outpatient clinic because of an exacerbation of her pain and progressive motor weakness.

Examination

Neurological examination revealed a 3/5 muscle power in the left thigh and 4/5 in the right thigh. Distal lower extremity muscles displayed 5/5 and all deep tendon reflexes were decreased. Sensation was also reduced over the left lower limb. The patient needed help with ambulation due to the pain and motor weakness. There was no bowel or bladder dysfunction.

Routine hematological and biochemical tests were normal. Magnetic resonance (MR) imaging revealed a homogeneous intradural mass encroaching on the cauda equina at L2 level. The tumor was hyperintense on T1-weighted images, hypointense on T2-weighted images and showed intense and homogeneous contrast enhancement (Fig. 1). MR imaging findings indicated a lesion depending on L2 left root, which suggested the differential diagnosis of schwannoma or meningioma.

Operation

An elective lumbar laminectomy L1-L3 was performed. A greyish mass was found in the intradural space in continuity with L2 left root and it was also adherent to other lumbar roots from both sides. The lesion was limited to the subdural space with no extradural space invasion. A subtotal resection of the tumor was completed due to its invasion into the upper conus medullaris and lower lumbar roots.



Fig. 1 – Sagittal MR images of the tumor at L2 level. The lesion is slight hyperintense on T1 and hypointense on T2 sequence with a mild and homogeneous contrast enhancement.

Pathological examination

Histopathological examination showed a high cellular neoplasm with epithelioid cells arranged in sheets (Fig. 2A). Pleomorphic nuclei were present and most part of the cells contained a brown cytoplasmic pigment identified as melanin. Immunohistochemical studies revealed a strong positivity for HMB45 (Fig. 2B), S100 (Fig. 2C) and MelanA (Fig. 2D). Based on these findings a Malignant Primary Melanoma was diagnosed.

Postoperative course

A search of primary lesions outside the central nervous system was performed with all the results negative and there were no evidence of metastases in the extension studies; including a normal postoperative whole body positron emission tomography. The patient suffered a worsening in her motor weakness affecting proximal muscles of both lower limbs, displaying 3/5 strength in thigh and knee flexion and extension bilaterally. Following surgery she started rehabilitation therapy and she was referred for palliative radiotherapy with good pain control but the patient died two months after surgery.

Discussion

Primary pigmented neoplasms of the central nervous system (CNS) are very rare. They include a spectrum of tumors ranging from the well differentiated meningeal melanocytoma to the malignant melanoma^{1,6-8}. Among these lesions primary malignant melanoma in the CNS accounts approximately for 1% of all melanoma lesions and the spinal location is very unusual. The first report of a melanoma limited to a spinal root was done in 1987 by Schneider et al⁴. Since then a total of nine cases have been reported in the literature; four cases at the cervical level^{1,9-11} and the other five at the lumbar level^{3,5,12-14} with no thoracic root compromise been reported

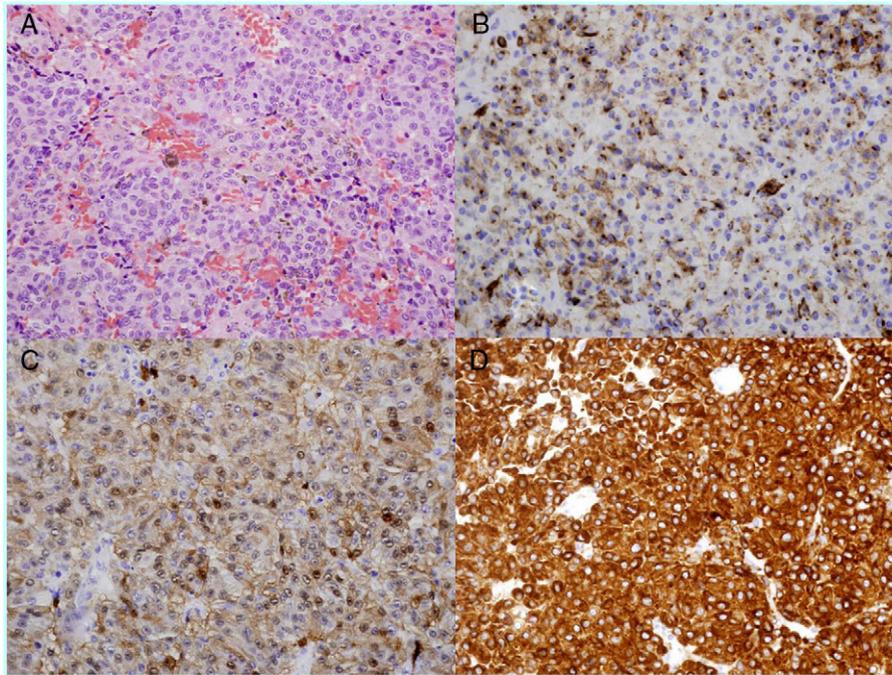


Fig. 2 – Histopathological images. A: Hematoxylin-eosin showing a pleomorphic high cellular neoplasm with brown cytoplasmic pigment identified as melanin. B: Immunohistochemical positivity for HMB45. C: S100 positivity. D: MelanA positivity.

yet. Although there are few cases in elderly patients^{1,10}, most primary meningeal melanomas present in the third and fourth decades of life with a variable prognosis^{4,8}.

Two theories have been proposed to explain the origin of the primary melanomas of the CNS. These lesions could originate from melanocytes derived from neural crest cells during embryogenesis or from melanoblasts accompanying the pial sheath of vascular bundles^{1,4,6}. The first step to diagnose a primary meningeal melanoma is excluding secondary or metastatic melanoma. An extension study is needed to prove the absence of melanoma in other sites of CNS and outside of the CNS. A whole body positron emission tomography/CT scan is very useful to detect any residual mass or the presence of melanoma at other sites in the body^{1,10}.

MR is the method of choice in the diagnosis of spinal tumors. Their location and their relation to adjacent structures are well defined with MR images. It can distinguish between extradural, intradural-extramedullary and intramedullary tumors in most cases, but the distinctions of the different types of tumors remains difficult. In our case, the tumor resembled a schwannoma arising from a lumbar root. Radiologically melanocytic lesions normally are hyperintense on T1-weighted sequences and iso or hypointense on T2-sequences. After the intravenous administration of a gadolinium-based contrast, these tumors show mild and homogeneous contrast enhancement^{1,6,11}. Hyperintensity on T1 sequences is due to the presence of melanin; but it can also be secondary to subacute hemorrhage or fat. It is well known that malignant melanoma can present foci of intratumoral bleeding resulting in variable signal intensity depending on the timing of bleeding. Moreover, MR imaging features of melanoma are determined by the degree of melanin and the

presence of acute or chronic hemorrhage^{2,6,12}. The non specific imaging characteristics and the rarity of these tumors, lead to their misdiagnosis and frequent exclusion from the initial differential diagnosis.

The pathological differential diagnosis includes various melanotic lesions that can affect the spinal roots: the cellular blue nevus, the melanotic schwannoma or MNST, the melanotic clear-cell sarcoma, the meningeal melanocytoma and the malignant melanoma^{1,11,13,14}. The main differential diagnosis is between melanocytoma, which is a benign lesion with a good prognosis; and the malignant melanoma which can behave in an aggressive pattern despite long survivals previously reported. The differences between them are the high cell density and proliferation index associated with malignant neoplasms as occurred in our case. Although there have been previous reports of malignant melanoma cases with long-term prognosis of eight years disease-free¹¹; fatal outcomes have been reported too with pulmonary metastasis 18 months after surgery³. In our opinion the outcome would be more influenced by the patient's age and the amount of surgical resection than the histopathological findings.

Conclusions

Primary melanoma arising from a nerve root is a very uncommon neoplasm that should be included during the differential diagnosis of lesions involving spinal nerves. Despite of its malignant appearance in histology some patients remain disease-free for years after the diagnosis. We believe that the good prognosis is related to the patient's age, the well-defined

borders of the tumor and the absence of dissemination or local extension at the time of diagnosis.

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