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Case report

Intradural extramedullary Ewing's sarcoma: A case report and review of the literature

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ARTICLE INFO

Article history:
Received 22 November 2015
Accepted 20 November 2016
Available online xxx

Keywords:
Intradural extramedullary Ewing's sarcoma
Lumbar spine
Surgery
Management

ABSTRACT

Introduction: Extra-skeletal Ewing's sarcomas are very rare lesions to the spine surgeon, with the intradural, extramedullary lesions being even rarer. Herein we present a patient with an intradural, extramedullary form of Ewing's sarcoma and review the relevant literature. The medical records, operative reports, radiographical studies and histological examinations of a single patient are retrospectively reviewed.

Case report: A 31-year old male presented with back-pain, right-leg progressive paraparesis, and inability to walk. Both motor and sensory disturbances were revealed on the right leg at the clinical examination. Lumbar MRI showed two lesions. The first one was an intradural, extramedullary lesion at the L2-L3 level, while the second was smaller, located at the bottom of the dural sac. The patient underwent gross total resection of the L2-L3 lesion after a bilateral laminectomy. Histological examination was compatible with Ewing's sarcoma, and was verified by molecular analysis. No other extra-skeletal or skeletal lesion was found. A chemotherapy scheme was tailored to the patients' histological diagnosis. The patient presented with local recurrence and bone metastasis 2 years after his initial diagnosis. A second operation was performed and the follow up of the patient showed no disease progression 18 months after revision surgery.

Conclusion: The spine surgeon should be aware of the existence of such rare entities, in order to timely fulfill the staging process and institute the proper therapy. The management of patients with extra-skeletal Ewing's sarcomas involves professionals as members of a multidisciplinary team, all of which should co-operate for the patient's optimal outcome.

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Abbreviations: IEES, intradural extramedullary Ewing's sarcoma; MRI, magnetic resonance imaging; RT-PCR, reverse transcriptase polymerase chain reaction; GFAP, glial fibrillary acidic protein; EMA, epithelial membrane antigen; EWS/FLI1, oncogenic transcription factor; RT, radiotherapy.

http://dx.doi.org/10.1016/j.pjnns.2016.11.006
0028-3843/

1. Introduction

Ewing's sarcomas are highly undifferentiated, primitive, malignant, small round-cell neoplasms, arising usually in children and young adults [1]. They are rare tumors that account for 10% of all primary malignant bone tumors in children and 3% of all childhood malignancies [2]. Moreover, they are very aggressive lesions; approximately 25% of patients have metastatic disease at the time of diagnosis [1]. Hereby we report an extremely rare case of an adult male with primary intradural extramedullary Ewing's sarcoma (IEES) of the spine and a skip lesion at the cul de sac of the dural sac, and review the pertinent literature.

2. Case report

A 31-year old male with unremarkable medical history, presented with back-pain, right leg progressive paresis (3/5 in all key muscle groups), causalgia and decreased sensation of the same limb, consistent with cauda equina syndrome. His lumbar spine magnetic resonance imaging (MRI) showed an intradural, extramedullary space occupying lesion behind the L2-L3 vertebral bodies, 5.2 mm × 1.3 mm in size, occluding the spinal canal and displacing the cauda equina to the left side and posteriorly. A second smaller lesion was found behind the L5 vertebral body with similar imaging characteristics. Both lesions enhanced inhomogeneously after contrast administration (Fig. 1a and b). The differential diagnosis included neoplasms such as the ependymoma, meningioma, neurinoma and spinal metastasis. Imaging of the neuraxis and thoraco-abdominal staging showed no evidence of other pathologic lesions.

L1-L3 laminectomy was undertaken and a fleshy mass was revealed after opening the dura that intermingled with the lumbar nerve roots. Gross total resection of the tumor was achieved and the specimen was sent for histopathological examination. No attempt was made to remove the second lesion, since there were no neurologic deficits and the surgical decision was to wait for the histology of the resected mass.

Hematoxylin – Eosin stains highlighted neoplastic cells with small, round or ovoid intensely stained nuclei and little cytoplasm with clear demarcation. Immunohistochemistry showed intense and diffuse positivity for CD-99, while did not stain for GFAP, EMA, cytokeratin 8 and 18, AE1/AE3, S-100 and neurofilaments. Reverse transcriptase polymerase chain reaction (RT-PCR) revealed the presence of the EWS/FLI1 fusion

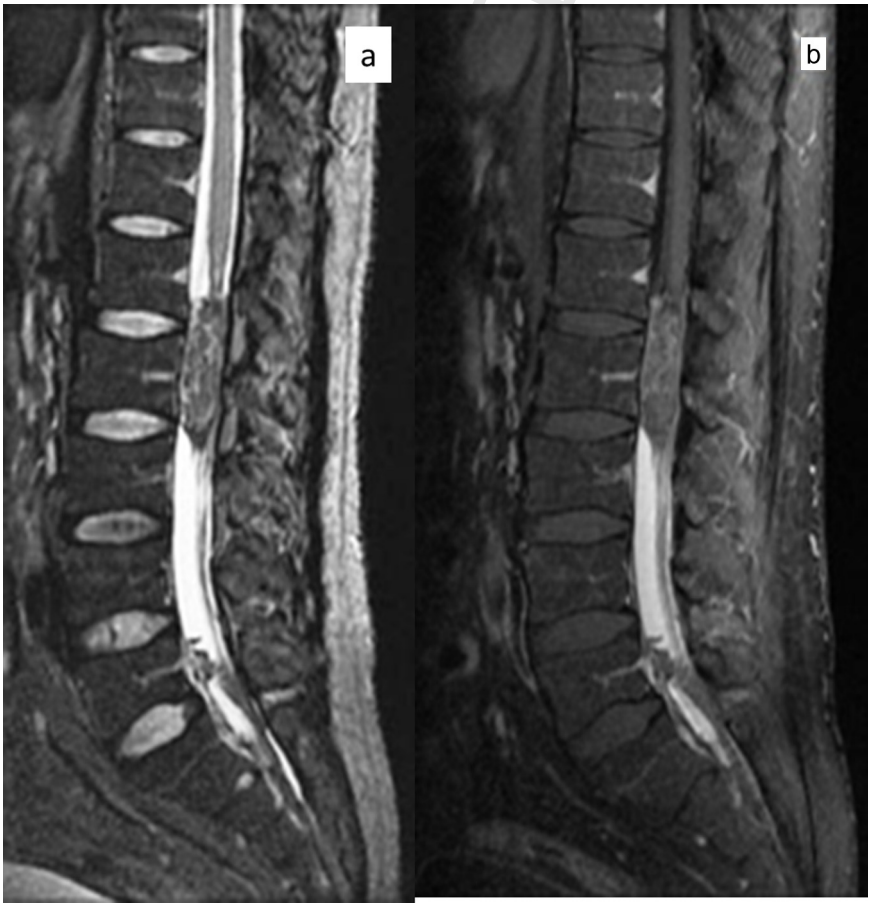

Fig. 1 – (a) Sagittal short tau inversion recovery MR image shows an intramedullary mass in the conus medullaris, extending from L1-L2 disc, along the L2 vertebral body up to L2-L3 disc. The mass has a heterogeneous low signal and there is a second, skip lesion at the level of L5 vertebral body. (b) Sagittal T1 MR image after intravenous administration of gadolinium shows patchy, moderate enhancement from the mass. Note that the CSF between the main and satellite lesion has abnormal high signal in T1 sequence that enhances, indicative of high protein content or blood.

Please cite this article in press as: Paterakis K, et al. Intradural extramedullary Ewing's sarcoma: A case report and review of the literature. Neurol Neurochir Pol (2016), http://dx.doi.org/10.1016/j.pjnns.2016.11.006