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CASE REPORT

Primitive Neuroectodermal Tumour and Extraskeletal Ewing's Sarcoma around the Lumbosacral Spinal Column

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ABSTRACT

We report on a woman who presented with a history of lower limb weakness and numbness for a few months. Based on cross-sectional imaging, at first she was provisionally diagnosed to have an aggressive tumour around the spinal column. Pathologically, the lesion was proved to be a primitive neuroectodermal tumour / Ewing's sarcoma. Herein, we discuss the clinical, radiological, and pathological features of this disease entity.

Key Words: *Magnetic resonance imaging; Neuroectodermal tumors, primitive; Sarcoma, Ewing's; Spinal neoplasms*

中文摘要

圍繞腰骶骼脊髓的原始神經外胚層腫瘤及骨外軟組織Ewing's肉瘤

黃鎮昇、朱曉青、譚國輝

一名女子感覺下肢無力及麻木數個月。根據病人的橫斷面顯像，起初診斷為有一個迅速蔓延的腫瘤圍繞著脊髓。此腫瘤病理學證實為原始神經外胚層腫瘤／Ewing's肉瘤。本文討論此病症的臨床、放射影像及病理學的特徵。

INTRODUCTION

Primitive neuroectodermal tumour (PNET) and related extraskeletal Ewing's sarcoma (EES) are small, malignant, round cell tumours belonging to the Ewing's sarcoma family. Most frequently they occur in the chest wall, lower extremities, and trunk. Rarely, they originate around the spinal column. Herein, we present an extremely rare example of PNET / Ewing's tumour arising around the lumbosacral spinal column. Reports on the radiological findings of this disorder are sparse and a local report is even more unusual.

CASE REPORT

A 41-year-old woman presented with progressive lower limb numbness and weakness for a few months. Neurological examination on admission revealed reduced lower limb power affecting L5 and S1 nerve roots and loss of sensation below the L1 dermatome.

Magnetic resonance imaging (MRI) of the spine demonstrated a large irregular extradural mass infiltrating the spinal canal at the L1 to S2 vertebral level, with obliteration and compression on the spinal

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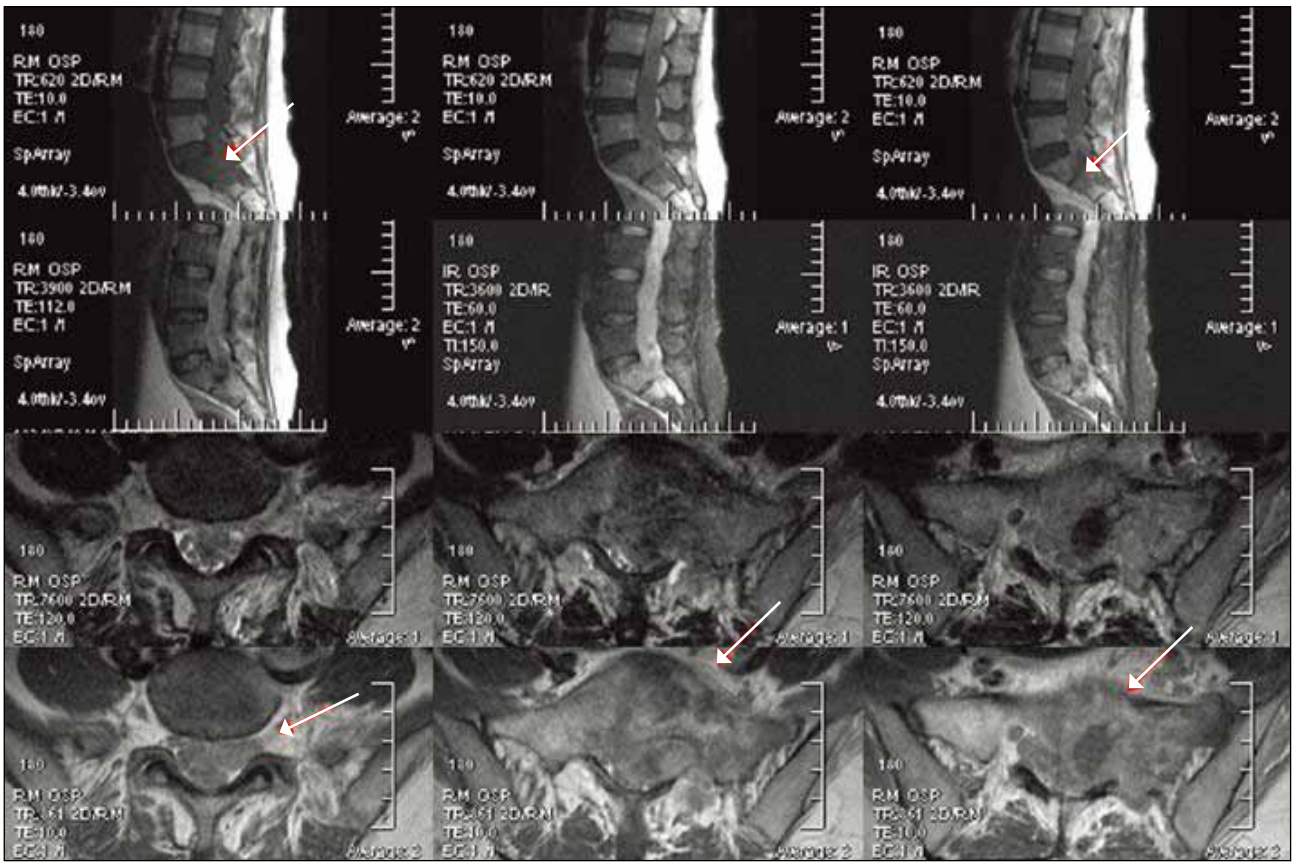

Figure. Magnetic resonance imaging (T1, T2 sagittal and axial) demonstrated a large irregular extradural mass (arrows) infiltrating the spinal canal at the L1 to S2 vertebral level with obliteration and compression onto the S1 and S2 region of the spinal canal. The mass extended through the left S1 sacral foramina to involve the left paraspinal muscle. The mass was heterogeneously enhanced (not shown).

canal at the S1 and S2 level. The mass extended through the left S1 sacral foramina to involve the left paraspinal muscle. The left L5 pedicle, S1 and S2 vertebral bodies were also involved, with loss of normal bone marrow signals. The lesion was isointense on T1-weighted imaging and hyperintense on T2-weighted imaging. With gadolinium contrast administration, the mass became heterogeneously enhanced (Figure). Myeloma and metastatic tumours were considered in the differential diagnosis of this lesion.

Laminectomy at L5 to S2 levels and excision of intraspinal tumour were performed and malignant Ewing's sarcoma / PNET arising around the sacral spine was diagnosed pathologically. Microscopy of the sections showed fragments of fibroadipose tissue infiltrated by confluent sheets of small tumour cells. The tumour cells were traversed by delicate fibrovascular septae and showed hyperchromatic irregular nuclei and scant, focally clear cytoplasm. Immunochemically, the tumour cells were non-reactive to LCA (lymphoid markers), actin (muscle marker) and AE1/AE3 & Cam 5.2 (epithelial markers). The cells were strongly reactive to CD99 and focally reactive to neural markers S100 NSE, CD56 and synaptophysin. Postoperatively, the reduced lower limb sensation and power persisted. The patient responded poorly to multi-agent chemotherapy, and finally succumbed two months after diagnosis.

DISCUSSION

Extracranial PNET / EES is a small, round cell, malignant tumour of bone, soft tissue and nerves, which mainly affects the chest wall, lower extremities, and trunk. PNET / EES around the spinal column is rare and sparsely reported. Descriptions of its radiological features are even more rare. It usually affects adolescents and younger adults, and more often they are males.[1,2]

Cross-sectional MRI is usually used in managing the Ewing's family of tumours, which appear as heterogeneously hypointense in T1-weighted images and heterogeneously hyperintense in T2-weighted

images.[2] Usually they have an irregular border and infiltrate adjacent structures. These features are sometimes non-specific, especially when the tumour is huge. They can also mimic a metastasis and myeloma; the conditions are therefore included in the differential diagnosis.

Apart from having a role in making the diagnosis, radiology assists management of PNET / EES lesions around the spinal column by: (1) discerning their size and any feature of malignant transformation, (2) being a guide to tissue biopsy, and (3) monitoring responses to therapy.

Extracranial PNET was first described in 1918 while EES was first described in 1969. Because of immunohistochemical, ultrastructural, and molecular biological similarities, PNET and Ewing's sarcoma have recently been classified into a group termed the Ewing family of tumours.[3] These are small, round cell tumours of bone, soft tissue, and nerves; morphologically they consist of germinal neuroepithelium. One of the specific pathological features of this tumour family is the membranous expression of the *MIC2* gene.[4]

Genetic studies involving fluorescent in-situ hybridisation or the reverse-transcriptase polymerase chain reaction can be very useful in diagnosing inconclusive / difficult cases. Identifying a Ewing's family tumour is crucial to management, because it carries a very poor prognosis. The diagnosis is usually clinched by clinical, radiological, and histological features. Radiologically, these tumours may be difficult to be differentiated from metastases, gliomas arising from ependymal cells, and myeloma. Often MRI can also be used to detect any cerebrospinal fluid seeding, which is another typical feature of such tumours.

The absence of a known primary and no Bence Jones protein in the urine rendered the alternative diagnoses less likely. In our patient, chordoma was another differential diagnosis, which was relatively unlikely in view of the patient's age. Certainly, the final definitive diagnosis must be supported by pathological features.

In terms of treatment, en-bloc resection still remains a curative option. In most cases however, patients have local extension at the time of presentation that inevitably precludes total resection. Ewing family tumours are chemo- and radio-sensitive. Thus, the management plan for tumours around the spinal column relies on a combination of these treatment options. Ewing family tumours around the spinal column have a poor prognosis; the 5-year survival rate being reported to be between 0% and 38%.[5]

CONCLUSION

Extracranial PNET / EES around the spinal column is a rare disorder and carries a poor prognosis. Radiology plays an important role in its management in terms of diagnosis and monitoring treatment. Identifying extracranial PNET / EES around the spinal column is important and is usually clinched by a combination of clinical, radiological, and histological features.

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