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Case Report

Primary Ewing's Sarcoma of the Spine with Conus Medullaris Syndrome
原發性尤文氏肉瘤造成的脊髓圓錐壓迫症

Laik Jayant Kumar [a, *], Kumar Ritesh [b]

[a] Department of Joint Replacement and Reconstructive Surgery, Tata Main Hospital, Jamshedpur, India
[b] Department of Orthopaedics, Tata Main Hospital, Jamshedpur, India

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ABSTRACT

Ewing's sarcoma (ES) is a primary neoplasm of the bone affecting age groups in the second decade of life. It commonly affects the metaphyseal ends of growing bones. Primary ES of the spine is rare and is commonly seen in the sacrum. The incidence of sacral ES in the spine is <5% whereas that of nonsacral ES is <0.9%. A high index of suspicion is needed to diagnose the condition and prompt surgical intervention combined with chemotherapy and radiotherapy treatment is needed to contain the progression of neurological deficit. We report a case of primary ES with sudden onset conus medullaris compression in a 17-year-old female.

中 文 摘 要

尤文氏肉瘤是骨骼的原發性腫瘤，主要影響年齡組是生命的第二個十年。它通常影響生長中骨骼的幹骺端。脊椎的原發性尤文氏肉瘤很罕見，常見於骶骨。骶骨的尤文氏肉瘤的發生率少於5%，而非骶骨的尤文氏肉瘤的發生率少於0.9%。診斷病情需要高度的懷疑指標，並且需求及時進行手術結合化療及放射治療以制神經功能缺損的進展。我們報告了一例原發性尤文氏肉瘤造成一名17歲女性突發性脊髓圓錐壓迫症。

Introduction

Primary Ewing's sarcoma (ES) affecting a vertebra is extremely uncommon. It is the second most common malignancy involving bones in the paediatric age group and it accounts for 4% of all paediatric malignancies.[1] The highest incidence of ES is seen in the second decade of life involving mostly the long bones of the extremities and the pelvis. Vertebral ES is divided into two groups: (1) sacral type, involving the sacrum and coccyx; and (2) nonsacral type, involving cervical, dorsal, and lumbar vertebrae.[2] Mostly, vertebrae are involved secondarily from metastasis of ES which originates elsewhere. It is extremely rare to come across primary vertebral ES if the sacrum is excluded.

Patients with ES present with neurological complications when the tumour tissue extends into the spinal canal compressing the cord or roots.

Case Report

A 17-year-old female patient was admitted with severe low back pain which increased on lying down and lessened with sitting along with a back rest. On the 3rd day of onset, pain radiated to the left leg and had no relief with nonsteroidal anti-inflammatory drugs. Examination revealed weakness of the extensor hallucis longus muscle with sensory deficit at the L5 dermatome. However, within the next 24 hours she developed gait instability with loss of bowel and bladder function. She stated that sensation in the perianal was lost 1 day prior to the clinical loss of bowel and bladder function. X-ray did not reveal any abnormality (Figure 1).

An urgent contrast enhanced magnetic resonance imaging (MRI) was done which suggested a tumour mass consistent with radiological features of ES (Figures 2 and 3).

Embolization of the tumour mass was done to reduce the vascularity followed by tumour mass excision with pedicular screw fixation. The tumour tissue was sent for histopathology. Immunohistochemistry showed positive expression of CD99 and FLI-1, but

* Corresponding author. *E-mail:* jklaik@gmail.com.

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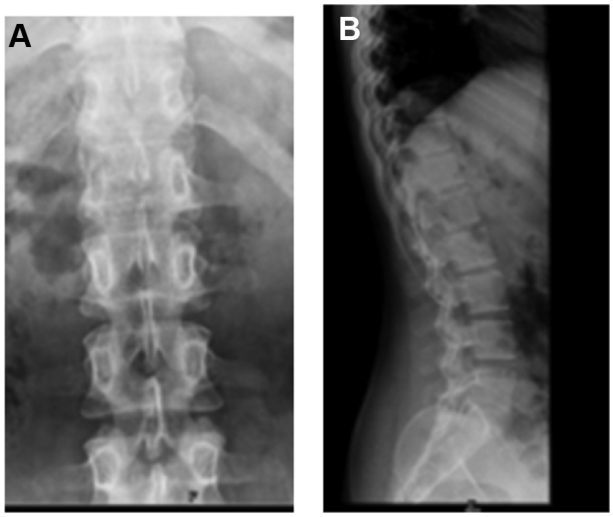

Figure 1. X ray of the lumbar spine spine. (A) Anteroposterior view; (B) lateral view.

the atypical cells were negative for desmin, LCA, or Tdt which confirmed the diagnosis of ES (Figure 4).

Neurological improvement and full recovery of bladder and bowel function was observed 1 week after surgery. The patient started walking with support at 2 weeks postoperation, and without any support 10 weeks after surgery.

Positron emission tomography–computed tomography was done before starting chemotherapy to exclude metastasis to the lungs.

The patient underwent chemotherapy as per the European Ewing's sarcoma protocol.[3] Although she is free from her neurological complications, her prognosis is still guarded.

Discussion

ES is a malignant, small, round cell tumour which commonly involves the pelvis, femur, humerus, ribs, and clavicle. ES is the second most common tumour in the paediatric age group involving bones.[1] Primary ES involving the spine is very rare (4%). Spinal ES is divided into sacral and nonsacral types.[2] The majority of ES of the spine involves the sacrum and coccyx. Nonsacral ES is a rare entity (0.9%),[4] and it is said that one will encounter this type of tumour once in one's whole career.

ESs are a group of tumours consisting of ES, Askin tumour, and peripheral primitive neuroectodermal tumours.[5–8] These tumours are grouped together because they share similar cellular physiology, as well as a shared chromosomal translocation between chromosomes 11 and 22, t (11; 22).

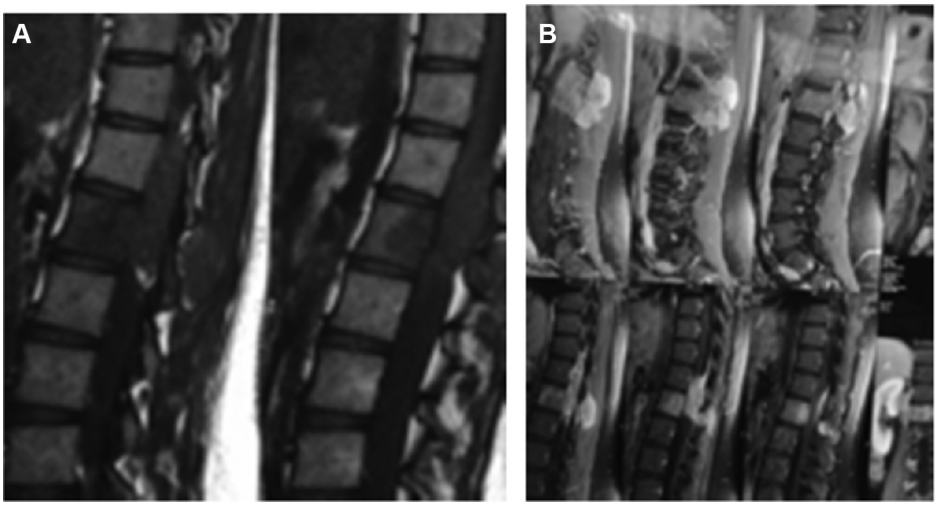

Figure 2. Contrast enhanced MRI showing an extensive tumour mass compressing the conus medullaris. (A) and (B) represent two different cut section of MRI in sagittal plane. MRI = magnetic resonance image.

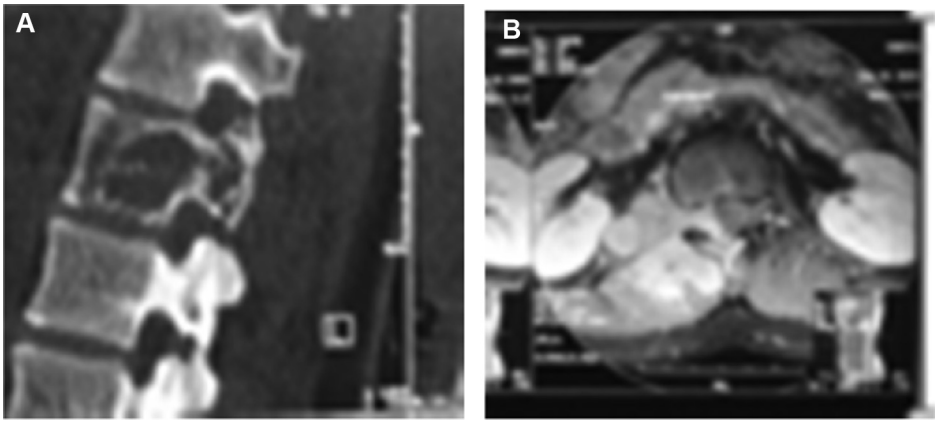

Figure 3. CT scan showing the destruction of the L1 vertebra with pedicular involvement. (A) represent pathology in sagittal section and (B) represent the same pathology in Axial view of CT scan. CT = computed tomography.

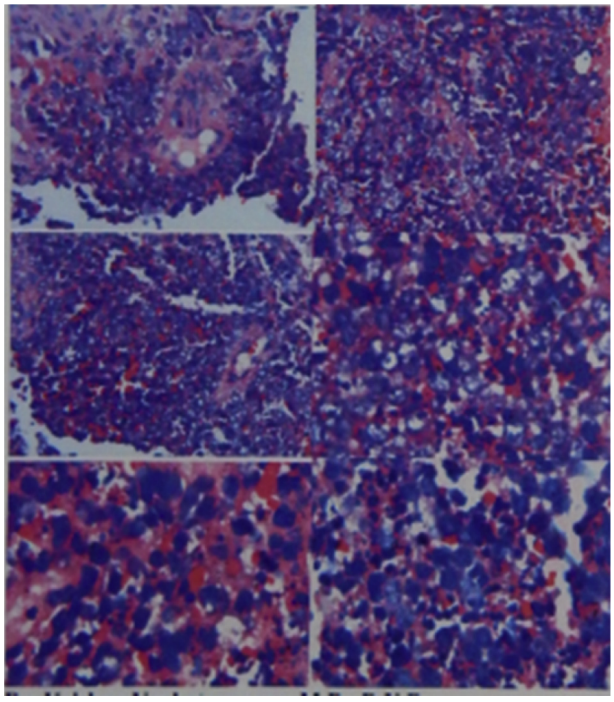

Figure 4. Section showing neoplasm composed of a monomorphic population of atypical cells with a high N:C ratio, opened up chromatin, and scant neoplasm. Plenty of necrotic debris is present throughout the lesion.

A very high index of suspicion is needed to diagnose ES of the spine involving the nonsacral area. In our case, the patient was admitted to another hospital with back pain but was discharged as a simple back pain. However, soon after she developed instability which led us to opt for an MRI which confirmed the provisional diagnosis of ES. Since both ES and lymphoma give similar pictures in the spine, histopathology was used to obtain a definite diagnosis.

Because the patient developed sudden conus medullaris features, it was decided to go for immediate surgery to decompress the spine by resecting the tumour mass.

As other round cell tumours such as lymphoblastic lymphoma have the same histological features, the tissue was sent for immunohistochemistry. Immunohistochemistry showed CD99 and FLI-1 positive, suggesting ES, and were negative for desmin, LCA, or Tdt which confirmed the diagnosis of ES.

The late presentation of the tumour can be explained by its unusual location; where the mass grew to a considerable size before reaching a critical size when it compressed the conus and produced pain and neurologic symptoms.

ES in the nonsacral area is a rare entity and, to the best of our knowledge, this is the first reported case of acute conus medullaris compression in a case of ES.

Conflicts of interest

The authors declare no conflicts of interest.

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