Case Report

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Vertebral Tumour Presenting with Progressive Flaccid Paraparesis and Areflexia in an Adolescent Girl

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Abstract

Ewing's Sarcoma (ES) is a rare malignancy but 2nd most common malignant tumor of long bones after osteosarcoma usually occurring in children and young adults. Rarely, it can affect vertebra or spine in adolescent masquarading as a progressive infectious lesion. The spinal ES usually occurs in lumbar vertebrae and presents with progressive neurological deficits in lower limbs. Ewing's Sarcoma has poor prognosis because of uncontrolled metastatic potential making early diagnosis and intervention critical for survival of the patient. We report here a case of ES in a 14-year-old girl with involvement of lumbar spine with local spread and invasion of cauda equina with radiological, histopathological and immunohistochemical features.

Keywords: Ewing's Sarcoma; Paraparesis; Tuberculous spineround cell tumor; Immunohistochemistry

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Introduction

Ewing's sarcoma is a rare malignant round cell tumor that was first described by Ewing. It can occur in any bone but commonly it is found in the diaphysis of long bones and pelvic girdle [1]. It accounts for 4-10% of all primary bone cancers affecting adolescents and young adults. ES can rarely arise from the spine itself, usually in a vertebra, and present with progressive neurological deficits [2,3]. Early diagnosis is imperative to initiate appropriate treatment protocol.

Case Report

A 14 year old female presented with low back pain and paresthesia of both lower limbs for the last 7 months. There was no bladder or bowel disturbance. Because of paresthesia she had a fall 6 month back and had an L2 vertebral compression fracture; imaging at that time reveals a paravertebral mass (**Figure 1**) with infiltration of spinal thecal sac and exiting nerve roots below conus medullaris and nerve roots of cauda equina. The initial MRI report suggested tuberculous spine for which ATD (anti-tuberculous drug) therapy was started as due to containment for coronavirus outbreak parents could not complete the biopsy and continued with ATD. There was no improvement after taking ATD for last 4 months. At present she developed severe low back pain and progressive weakness and numbness of both lower limbs when she was s brought to this hospital. On examination, there

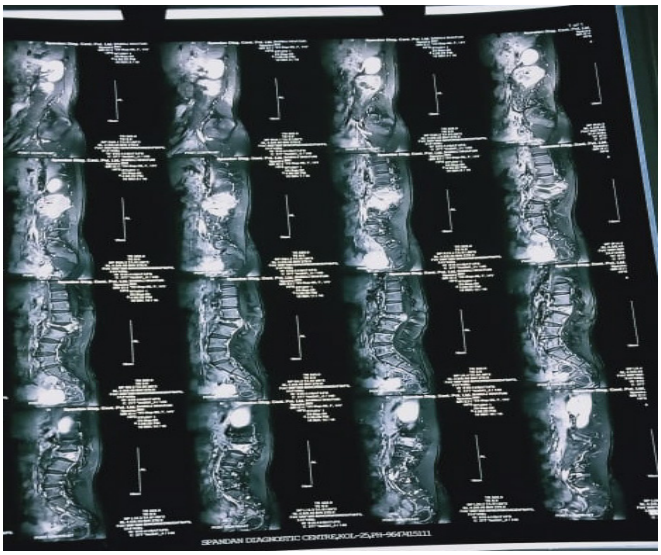

Figure 1 Initial MRI spine showing caries spine with destruction of L2 vertebra.

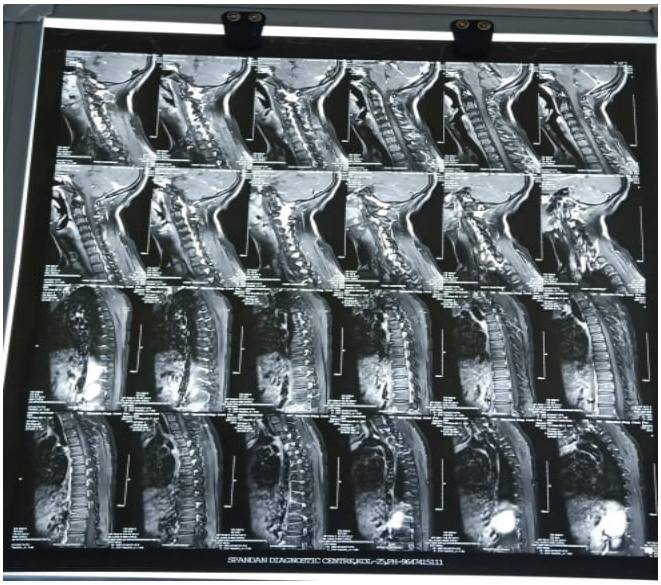

Figure 2 Recent CECT spine showing soft tissue mass seen arising from L2 vertebrae with right para vertebral extension.

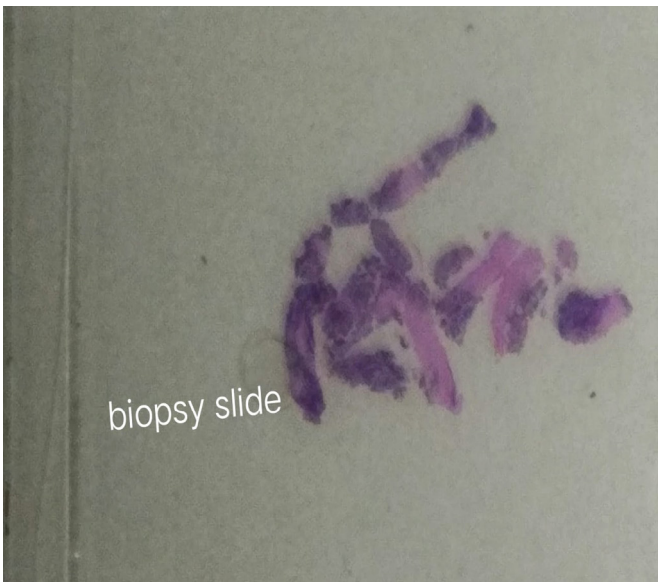

Figure 3 Biopsy slide with histopathology and immunohistochemistry (confirmed Ewing sarcoma).

was anemia, edema both legs, her vital parameters including pulse, blood pressure, temperature and respiration were within normal range. Neurologically, power of both lower limbs was grade 0/5, with severe hypotonia and absent deep tendon reflexes. Plantar responses were mute bilaterally. Touch, pain, temperature, vibration and position sensations were absent in both lower limbs.

Radiological features (**Figure 2**) after admission suggested a destructive lesion in L-2 vertebral body with a suspicion of malignancy. Routine hemogram and biochemical examination revealed no abnormality. USG guided biopsy was done from the L-2 vertebral lesion after obtaining consent. Microscopy (**Figure 3**) revealed nests of round to oval cells having hyperchromatic nuclei and scanty cytoplasm surrounded by broad bands of fibrous tissue with foci of necrosis. Immunohistochemistry showed positivity to Mic-2, NKX-2.2, synaptophysin and immunonegative to cytokeratin, desmin, LCA,TdT which suggestive of malignant round cell tumor- Ewing's Sarcoma.

Discussion

In our patient the initial interpretation of the MRI scan by the radiologist was that of a destructive lesion in the vertebral body of the L2 vertebra most likely to be due to an infective process like tuberculosis [4]. As biopsy could not be done due to containment for coronavirus outbreak, the final diagnosis was delayed. Subsequently when the patient attended our hospital, USG guided biopsy was done and the report confirmed the lesion to be Ewing sarcoma.

Conclusion

Any child presenting with progressive weakness of both lower limbs should be evaluated for dorso lumbar spinal pathology including malignant tumor of vertebra. Early diagnosis with biopsy and immunohistochemistry is essential in ES to improve survival by treatment with surgery, radiotherapy and chemotherapy in combination.

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