

Unusual Presentation of a Primary Ewing's Sarcoma of the Spine with Paraplegia: A Case Report

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ABSTRACT

Ewing's sarcoma is a primary malignancy of the bone affecting individuals in the second decade of life. Primary sarcomas of the spine are rare and the occurrence of Primary Ewing's sarcoma in the spine is very rare. Ewing's sarcoma occurring in the spine is divided into two types, Ewing's sarcoma of sacral spine which are very aggressive with poor prognosis and Ewing's sarcoma of the non sacral spine which is an extremely rare occurrence. Patient may present with neurological deficit when the tumour extends into the spinal canal causing spinal cord compression. Magnetic resonance imaging (MRI) is very sensitive in diagnosing the tumour and defining the extent of the tumour. Here we report an 18-year-old boy who presented with back pain and complete paraplegia of two months duration. The MRI gave a differential diagnosis of infective pathology due to the fluid collection in the paraspinal region, followed by primary malignancy as the second diagnosis. Patient underwent posterior spinal decompression and stabilization, and intraoperatively there was significant collection of pus whose culture showed no growth. The histopathology and immunohistochemistry studies confirmed the diagnosis of Ewing's sarcoma and patient was started on combination chemotherapy and radiotherapy.

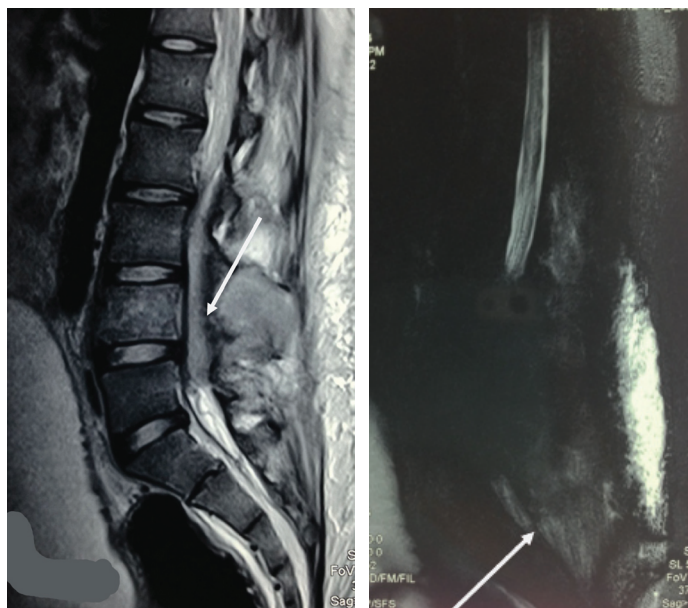
Keywords: Paraspinal, Primary malignancy of spine, Spinal cord compression

CASE REPORT

An 18-year-old male was admitted with severe back pain and inability to move both his lower limbs for the past two months. Back pain was insidious in onset, progressive and pain was present during rest. Two months back, patient noticed weakness of both lower limbs while walking which progressed to complete inability to move both lower limbs for the past one month. Patient gave history of intermittent low grade fever and weight loss in the past two months. He also gave history of bowel and bladder incontinence for the past one month. There was no contact history for tuberculosis. He had tenderness in the lower back and neurological examination revealed Grade 0 power in both lower limbs with hypotonia. Sensory loss started from mid thigh to involve the legs, ankles and feet of both lower limbs. The knee jerk and ankle jerk were absent and there was no response for plantar reflex. Patient had stage three pressure sore over his sacrum. His hemoglobin was 10.5 gm% with increase in the total WBC count and erythrocyte sedimentation rate. Other laboratory investigations were normal.

His radiographs showed no significant abnormality, MRI [Table/Fig-1-3] and post Gadolinium images [Table/Fig-4] showed hyperintense T2 signal from L4 body with soft tissue enhancement involving both the psoas muscles, fluid collection in the paraspinal muscles and an extension into the spinal canal from L2 to L5 vertebrae causing a complete block in the myelogram. The disc spaces were normal and there were no skip lesions on the MRI. The radiologist had given a differential diagnosis with infective spondylitis being the first diagnosis and malignant pathology being the subsequent diagnosis.

Patient was taken up for posterior spinal surgery, through a posterior midline incision L2 to L5 vertebra were exposed and there was frank pus in the paraspinal region which was collected for culture and sensitivity. Decompressive laminectomy was performed at L3 and L4 vertebra to enter the spinal canal. There was a layer of granulation tissue covering the dura as in tuberculosis pathology, which was carefully separated from the dura and sent for histopathological analysis. Transpedicular bone sample obtained from L4 vertebra and was sent for histopathological analysis. The levels were stabilized



[Table/Fig-1]: (1) MR sagittal image of lumbar spine showing involvement of L4 body, spinal canal and para spinal soft tissue component;

[Table/Fig-2]: Sagittal image showing myelogram block due to extension of tumour into the spinal canal

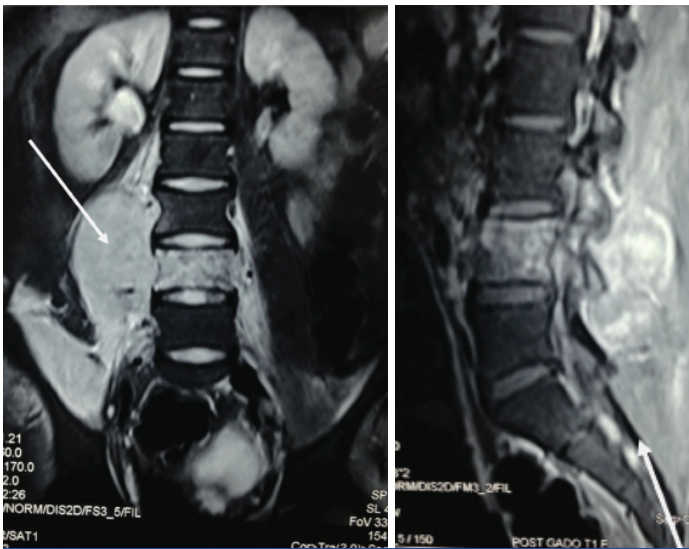
using pedicle screws and rods [Table/Fig-5,6]. The wound was then closed in layers.

Postoperatively the patient had good pain relief and was made to sit up with a brace on the second postop day. Neurology was reassessed and the motor power in both lower limbs were zero and sensory loss remained the same too. However, the patient had full recovery of bowel and bladder function.

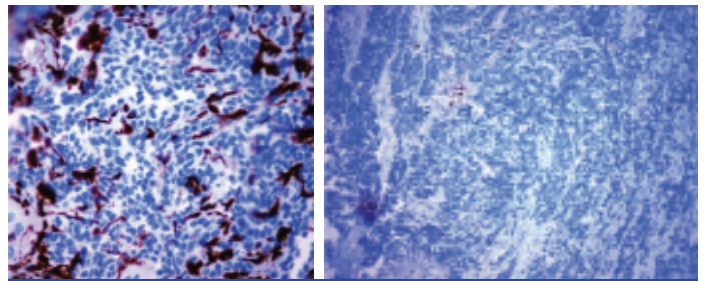
HISTOPATHOLOGY

Microscopic Description

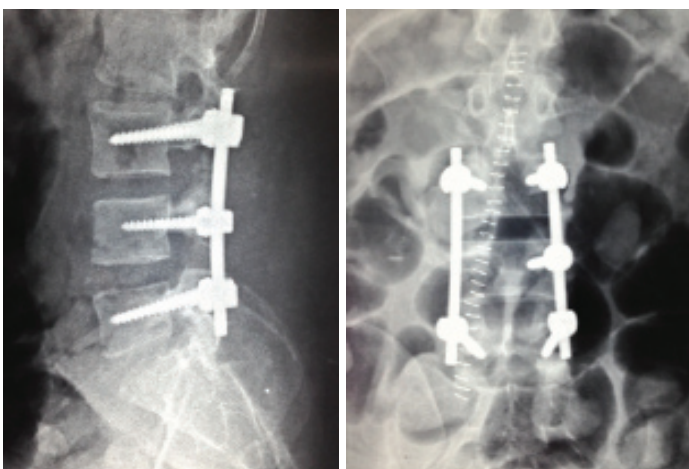
Sections show fibrocollagenous stroma and skeletal muscle fibres infiltrated by a malignant neoplasm arranged in diffuse sheaths, nests and pseudo acinar pattern. The cells have scanty cytoplasm, monomorphous population of round to oval nuclei with dispersed



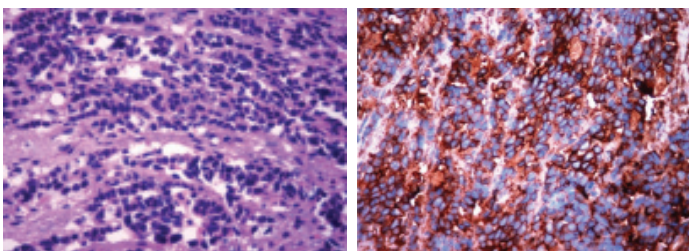
[Table/Fig-3]: STIR coronal images showing extension into the psoas muscle;
[Table/Fig-4]: Post gadolinium contrast image showing signal enhancement in L4 body and the paraspinal component



[Table/Fig-9]: Negative staining for LCA;
[Table/Fig-10]: Negative staining for Vimentin



[Table/Fig-5]: Postoperative lateral digital radiograph of lumbar spine with implants in situ; **[Table/Fig-6]:** Postoperative anteroposterior digital radiograph of lumbar spine with implants in situ



[Table/Fig-7]: Shows small blue round cells arranged in sheets 40X;
[Table/Fig-8]: Strong positivity for CD99

chromatin and inconspicuous nucleoli. Mitosis and apoptosis are seen. The tissue also shows extensive crush artefacts and areas of infarction [Table/Fig-7].

Immunohistochemistry

The immuno stains revealed strong positivity for CD99 (strong membranous), (focal) NSE and although negative staining for Vimentin and FL-1 is noticed excluding other differentials with negativity for LCA, TDT, Desmin, Synaptophysin and CD 56, suggesting Ewing's sarcoma/ PNET [Table/Fig-8-10].

The serum lactate dehydrogenase level was 116 IU/L which indicated a favourable prognosis. The patient was started on chemotherapy with a regimen consisting of vincristine, cyclophosphamide and adriamycin followed by radiotherapy. The patient was discharged after completing the first cycle.

DISCUSSION

Ewing's sarcoma was first described by James Ewing in 1921. Ewing's sarcoma is derived from a primordial bone marrow derived mesenchymal stem cells. Internationally the annual incidence rate is approximately three cases per million children [1]. Data from the US National Cancer institute (NCI) Surveillance suggest that the incidence of Ewing's sarcoma is nine times greater in Caucasians than in African Americans [2].

Primary malignant sarcoma of the spine is very rare and account upto 14.9% of all primary bone sarcomas. Ewing's sarcoma is the second most common primary malignant tumour after oserosarcoma, and the reported incidence of Ewings's sarcoma of the spine was 4% [3]. Ewing's sarcoma originating from the spine is rare and classified into non sacral and sacral type. Ewing's sarcoma of the sacrum is more aggressive and responds poorly to treatment. Ewing's from the non sacral spine represent approximately 0.9% of all cases [4],thus making it an extremely rare presentation.

The Ewing's family of tumours comprise of Ewing's sarcoma, extraskeletal Ewing's sarcoma, primitive neuroectodermal tumour (PNET) of bone and soft tissue and chest wall tumour (A skin's tumour).The translocation t (11;22) (q24;q12) is identified in more than 90% of cases of Ewing's sarcoma [5,6]. Spinal involvement most commonly results from metastasis in advanced stages of the disease, while Ewing's sarcoma originating from the spine is rare and extremely rare if the sacrum is excluded [7].

The initial radiographs reveal no abnormality, thus favouring MRI for early detection. MRI also helps in assessing the extension of tumour into the spinal canal, extension into the soft tissue and also the relationship of the tumour to the nearby vascular structures. In our patient the radiologist had given a differential diagnosis on the MRI which was, infective spondylitis (probably tuberculous pathology), Ewing's sarcoma and neuroblastoma. This led to a diagnostic dilemma regarding the cause of the disease. Intraoperatively, significant collection of frank pus in the paraspinal region, and within the spinal canal a layer of tissue over the dura resembling the tuberculous granulation tissue was found. The pus was sent for microbiological culture and showed no growth of organisms. The tissue covering the dura was separated and transpedicular bone from L4 body were sent for histopathological examination which revealed the classical picture of Ewing's sarcoma as described above in the microscopic description and immunohistochemistry. Thus it is of utmost importance to consider Ewing's sarcoma a possible cause when a patient in the second decade of life presents with spontaneous back pain not relieved by rest, in the absence of constitutional symptoms and no significant changes in the radiograph. Radiographic changes are late to appear, usually after the neurological signs have become apparent. The most common finding on the radiograph would be lytic bone destruction involving the vertebra. However, the present patient who presented with complete paraplegia had no lytic changes that were visible on the radiograph.

When deciding on the treatment option for Ewing's sarcoma of the spine, the most important factor would be the presence or absence

of neurological deficit. If the patient is neurologically stable then a tissue diagnosis is required, a large bore needle biopsy may be considered. Once the diagnosis of Ewing's sarcoma is confirmed, neo adjuvant chemotherapy is started [8]. A neurological deficit warrants early surgical decompression of the cord which can provide maximum chance of recovery and also specimen for histopathology. Decompressive laminectomy alone removes the posterior tension band, which can render the spine unstable and risk of anterior column collapse and kyphosis. Posterior spinal stabilization reduces the risk of this complication.

After the surgery the patient regained his bowel and bladder control with good relief of back pain, however there was no change in the motor power of the muscles of the lower limbs. Following surgery post operative chemotherapy to control micrometastases and radiotherapy is very important. Neo adjuvant chemotherapy which is usually given as three or four drug regimen is more beneficial in shrinking the tumour, thereby increasing the chances of total excision, but also takes care of micrometastases and gives an idea about responsiveness of the tumour to adjuvant therapy [8].

In a study by Marco et al., [9] to evaluate the oncological outcome of patients treated with chemotherapy and radiotherapy for Ewing's sarcoma of spine, the disease free survival rate was 49% at five years and 36% at 10 y.

CONCLUSION

Primary Ewing's sarcoma of the non sacral spine is an extremely rare tumour, which requires high index of suspicion when the patient presents with only back pain and no other findings. Treatment with

a multi modal therapy of chemotherapy, radiotherapy and surgery at the earliest improves the survival rate, but the prognosis is poor when the disease arises from the sacrum, when the tumour has metastasized or when the serum lactate dehydrogenase level is elevated before starting the treatment.

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