Abstract: This is a case report describing an atypical presentation of Ewing's sarcoma involving the lumbar spine in an adolescent male, presenting to a tertiary spinal cord rehabilitation centre. A 17-year-old male presented with acute lower back pain and subsequent ascending lower limb paralysis associated with bowel and bladder dysfunction over a period of 2 weeks. Since initial plain radiographs showed no obvious abnormality, the patient was evaluated for causes of myeloneuropathy until an MRI spine showed the full extent of a lesion involving the L1 vertebra. Although it commonly affects the metaphyseal region of long growing bones, involvement of the non-sacral spine is rare in Ewing's sarcoma. Usually a late complication in the clinical course of events, paraplegia may be a presenting feature among these tumors. Hence, atypical progression of paraplegia in the adolescent age group with no prior history of trauma, and normal plain radiographs should also be evaluated early for malignant disease.

Keyword: Ewing's Sarcoma, Paraplegia, Adolescent, Myeloneuropathy

INTRODUCTION

Ewing's sarcoma which accounts for a quarter of all primary bone tumors in the pediatric population (1) has a peak incidence in the second decade of life and rarely occurs after 30 years of age. (2) Fewer than 5% of cases may present with compression of the spinal cord or the nerve roots emerging from it. The prognosis of these cases which initially present with pain and swelling of the affected bone is generally poor. Hence it is important to detect this condition early with appropriate imaging, confirmatory biopsy and accordingly initiate the necessary treatment regimen to prevent complications such as paraplegia. The occurrence has a slight male predominance, is more common in Caucasians, and seldom found among individuals of African and Asian lineage. While commonly affecting the femur, pelvis and long bones of the extremities, primary vertebral Ewing's sarcoma may be classified into sacral and nonsacral types with the latter constituting 9.9% of all cases. (3) This report highlights the importance of considering a neoplasm among adolescent patients presenting with ascending paraparesis, with no prior history of trauma and no obvious changes in plain radiographs of the spine. MR imaging done early in the work-up can help clinch the diagnosis and prevent unnecessary investigations looking for causes of myeloneuropathy. With early diagnosis, newer regimens of therapy which are known to increase 5 year survival rates to about 40%, can be initiated. (4)

CASE REPORT

A 17 year old boy presented with complaints of inability to move the lower limbs, associated with bowel and bladder incontinence with no history of trauma prior to onset of symptoms. About 3 months prior to presenting to our hospital, he developed sudden onset of pain in the lower back which progressively worsened over a period of one week. The pain was accompanied with mild weakness in both the lower limbs which lead to an unsteady gait. He also began to notice abdominal distension with difficulty in passing urine. A week later, he was admitted when he was completely bowel and bladder incontinent and required continuous bladder drainage. He had also become completely bedbound with loss of power and sensation in the lower limbs. Initial investigations at the local hospital did not reveal any significant information and he was managed conservatively. Around 2-3 days into his admission in the local hospital, his neurological level of weakness started ascending upwards and progressed from the waist towards the lower chest rendering him completely dependent on his family for most activities of daily living. He subsequently developed a sacral pressure ulcer and decided to visit our department for further management. There was no prior history of fever, loose stools or administration of parenteral injection prior to onset of symptoms. There was also no past history of tuberculosis, fever, significant loss of weight or other constitutional symptoms prior to initial onset of back pain. Patient denies smoking, consumption of steroids or any other drugs in the past. On admission he was found to be conscious and well-oriented to his surroundings, with stable vital parameters and normal cranial nerve function. Lower limb musculature was found to be wasted and associated with a flaccid tone compared to the normal bulk and tone of upper limb muscles. Motor muscle testing revealed Grade 0 power in all muscles of the lower limbs with loss of deep tendon reflexes at the knee and ankle joints. Beever’s sign was absent with bilaterally weak upper and lower abdominal muscles. Voluntary contraction of the external anal sphincter was also absent. Plantar reflexes were absent in both lower limbs as were the superficial abdominal, bulbocavernous and cremasteric reflexes. Perception of pin-prick and light touch sensation were absent below the umbilicus, as were joint position and vibration sensation in the lower limbs. Motor and sensory evaluation of the upper extremities was normal. While the review of systems did not reveal any other abnormalities, he had a stage 3 (National Pressure Ulcer Advisory Panel) sacral pressure ulcer. Patient...
was admitted with the primary goals of evaluating the cause of the neurological impairment and management of the sacral pressure ulcer. Simultaneously, he was trained with skills necessary to be functionally independent in activities of daily living including ambulation in a wheelchair and tricycle. Baseline investigations showed low hemoglobin (9.4gm/dL) along with an elevated total white blood cell count (15,450 cells/mm³). Aside from an elevated Erythrocyte Sedimentation Rate of 69mm/hr, other investigations including calcium, phosphorus and liver function tests were within normal range. A neurology consultation was sought to evaluate the possible cause of the subacute onset of myeloneuropathy. Investigations done included inflammatory and vasculitic markers, namely CRP, Anti-Nuclear Antibody (ANA), dsDNA, C-ANCA & P-ANCA, all of which were found to be insignificant in this patient. Along with this, an MRI of the dorsal spine with screening of the whole spine and T2 weighted flair axial imaging of the brain was also planned. A plain x-ray of the lumbosacral and thoracic spine did not show any obvious abnormality (Fig 1).

**DISCUSSION**

Development of the spine is a complex neuro musculoskeletal process which accounts for a range of malignancies which can affect it, each with unique clinical profiles. In this case, due to the age of the patient, the absence of trauma and normal x-ray imaging, the possibility of a benign process was detected much later than the presenting complaint of paraplegia with bowel and bladder dysfunction. The lack of an obvious swelling and review of previous x-rays of the spine showing no abnormalities, led to investigations for the cause of a myeloneuropathy such as acute disseminated encephalomyelitis, chronic inflammatory demyelinating polyneuropathy, multiple sclerosis or tropical spastic paraparesis. The presence of skin lesions, erythema nodosum like rash, serositis and positive ANA and dsDNA in the serum detected by laboratory were additional clues compatible with Ewing’s sarcoma of the spine in this case. The key to the diagnosis is to exclude conditions mimicking the same clinical symptoms such as acute disseminated encephalomyelitis, chronic inflammatory demyelinating polyneuropathy, multiple sclerosis or tropical spastic paraparesis. The radiological findings were consistent with the diagnosis of Ewing’s sarcoma involving the lumbar spine.

**Fig. 1** Plain Radiograph of Lumbar Spine

On reviewing the patient's clinical symptoms, a mild swelling was noted over the lumbar spine although he no longer had any sensation or pain in the region. The MRI Spine which could only be done a week after admission, showed an expansive lytic lesion involving the L1 Vertebral body with a large epidural soft tissue component almost obliterating the thecal sac from T11 to L2, widening the spinal canal with scalloping of the posterior margin of L2 extending through the neural foramina into the paravertebral regions and into the erector spinae muscles (Fig 2). The biopsy of our patient were negative for tuberculosis and other important differential diagnoses that must be considered in these cases. Cultures taken from the CT-guided biopsy of our patient were negative for tuberculosis and pyogenic infection. The clinical examination nor the plain radiograph of the lumbosacral spine showed any kyphotic deformity. There were no osteolytic or osteosclerotic changes visible in the plain radiograph of the spine. If the spine is an important investigation early in the course.

**Fig. 2 MRI Lumbar Spine**

Multiple samples of the above mentioned lesion were taken in a CT-guided biopsy for histopathological evaluation and culture. There were no immediate complications and the patient tolerated the procedure well. Biopsy of the lumbar vertebra showed fibrocollagenous tissue infiltrated by a tumour arranged in nests, sheets and trabeculae composed of small round cells with pleomorphic, hyperchromatic, mitotically active nuclei with nuclear moulding. Occasional perivascular tumor rose ttes were also observed. On Immunohistochemistry, the tumour cells were positive for CD99 and FLI1 and negative for Desmin and CD20. These findings confirmed the presence of a malignant round cell tumour suggestive of Ewing Sarcoma. There was no growth suggestive of pyogenic infection in the cultures taken from the biopsy site. A bone scan following intravenous administration of Technetium-99m methylene diphosphonate (MDP) showed increased tracer uptake in the L1 and L2 vertebra while normal tracer uptake was observed in the rest of the axial and appendicular skeleton, as well as both kidneys (Fig. 3).

**Fig. 3 Bone Scan Showing Tracer Uptake At Lumbar Spine (L1)**

An Oncology consultation was sought and based on the clinical, radiological and bone scan findings, the patient was given a tentative treatment protocol consisting of chemotherapy with 3 cycles of VAC (Vincristine, Adriamycin, Cyclophosphamide). After assessing response to this initial regimen, local radiotherapy and concurrent chemotherapy shall be planned. He will then require 17 cycles of VAC wherein Adriamycin shall be replaced with Actinomycin D from the 5th cycle onwards. In view of financial constraints, the family had decided to go home and return when they are ready to initiate the advised treatment protocol. At the time of being discharged, he was compliant with pronelying for the management of the sacral pressure ulcer. He was on continuous bladder drainage using a Foley’s catheter which will require to be changed once in ten days under aseptic precautions. He has also been trained to manage his bowel functions with digital stimulation and evacuation. The family has been trained to assist the patient with wheelchair transfers and ambulation. The patient was recommended on the further management and prognosis of the condition.
of acute back pain progressing fast to paraplegia – this could provide vital information that precludes the need for expensive tests in the evaluation of a myeloradiculopathy in this clinical setting. In the histopathological evaluation of biopsied specimens from the spine, differential diagnosis of a small round cell tumor includes Ewing’s sarcoma, neuroblastoma, primitive neuroectodermal tumors, rhabdomyosarcoma and malignant lymphoma. The translocation t(11; 22) (q24;q12) detected in over 90% of cases, helps differentiate Ewing’s Sarcoma from other small, round cell tumors.(8)

CONCLUSION
In conclusion we would like to reiterate the importance of early MRI imaging of the lumbar spine in a case of acute low back pain progressing to paralysis. Tumors such as Ewing’s sarcoma must be considered in the list of differentials in a case on non-traumatic paraplegia with normal plain radiographs, especially among patients in the second decade of their lives.(2)(9) CT-guided biopsy with histopathology and immunohistochemistry can confirm the diagnosis early; a bone-scan will be necessary to detect metastases and accordingly plan an appropriate treatment protocol involving surgery, chemotherapy and radiotherapy.

BIBLIOGRAPHY