Primary Ewings Sarcoma Of The Lumbar Spine Presenting As Cauda Equina Syndrome
R Shankar, A Rao, C Ballal, R Naik

Citation

Abstract
A 16 year old girl presented with low back ache and insidious progressive weakness of both lower limbs of 2 months duration with bilateral foot drop. She had bowel and bladder incontinence. MRI of Lumbo sacral spine showed lytic destruction with irregular soft tissue mass involving the spinous process L3 in to spinal canal and erector spinae muscles causing significant narrowing and extradural compression on the cauda equine (L3 4) roots. A CT guided FNAC was suggestive of PNET. She underwent L3,4 decompressive laminectomy with biopsy of tumor tissue. Immunohistochemistry showed CD 99 positivity suggestive of Ewings sarcoma. Postoperatively lower limb power improved. The patient has received chemotherapy but incontinence persisted.

INTRODUCTION
Primary Ewing’s sarcoma of spine is very rare with poor prognosis. The ESFT (Ewing sarcoma family tumors) also comprises a skin tumor and PNET. The possibility of primary PNET should be considered in the pediatric age group with spinal tumors presenting as Cauda equina syndrome. Usual presentation is in second or third decade of Life. They present with local pain, gait disturbance, motor deficit, or sphincter dysfunction. Peripheral PNET (pNET) of spine refers to all those tumors arising from the surrounding soft tissues vertebra or spinal nerve roots. The mean period of postoperative survival ranges from 18 to 20 months from various reports and case series. Nearly one third of the spinal PNET present with Cauda equina Syndrome. So far 15 cases of spinal PNET have been reported presenting with Cauda equina syndrome. We report a case of Ewings Sarcoma emanating from L3 spinous process presenting as Cauda equina syndrome.

CASE
A 16 year old girl presented with low back ache and insidious progressive weakness of both lower limbs of 2 months duration with bilateral foot drop. She had bowel and bladder incontinence. There was no history of trauma, tuberculosis and constitutional symptoms. She was conscious oriented and her vitals were stable. No pallor, clubbing ,lymphadenopath or edema was present. Neurological examination showed bilateral lower limb hypotonia, motor weakness grade 3/5 power around the hip and knee, sensory blunting (below L3 dermatome) with absent ankle and knee jerks. A diffuse tender area of fullness felt in the lower lumbar region more towards the right. Respiratory and cardiovascular systems were within normal limits. Blood investigations: Hb 11gm %, TLC-6800, N-55% L35% M6% E4 %,ESR -59, Plts 4.9, normal peripheral smear, LFT, RFT, Ca 9.2 , P-10.6, ALP- 187.

Figure 1
Figure 1 : MRI of LS spine showing extent of tumor and compressing the nerve roots (a-e).CT images of guided biopsy (f) and lytic destruction of L3 spinous process(g)

MRI of Lumbo sacral spine (Figure 1a-e) shows lytic destruction with irregular soft tissue mass involving the
Primary Ewing’s sarcoma of the lumbar spine presenting as cauda equina syndrome

Spinal Ewing’s sarcoma of spine is a very rare tumor. Typically, the posterior elements of the spine are involved and are mostly extradural and extramedullary. The incidence in the sacrum is rare compared to other parts of the spine with equal distribution. Complete surgical resection is difficult most of the times. Distant metastasis is seen in 38% cases, commonest being the lungs. They are CD 99 positive and associated with t(11,22) translocation. These highly aggressive tumors warrant high index of suspicion and prompt tissue diagnosis with surgical intervention at the earliest and chemotherapy. Also, there are no definite guidelines for optimal management. The prognosis remains poor with local recurrences and systemic complications with survival in the range of months to couple of years. Multimodal treatment offers hope of longterm and good quality survival.

DISCUSSION

Primary Ewing’s sarcoma of spine has male preponderance with posterior elements of the spine are commonly involved and are mostly extradural and extramedullary. The incidence in the sacrum is rare compared to other parts of the spine with equal distribution. Complete surgical resection is difficult most of the times. Distant metastasis is seen in 38% cases; commonest being the lungs. They are CD 99 positive and associated with t(11,22) translocation. These highly aggressive tumors warrant high index of suspicion and prompt tissue diagnosis with surgical intervention at the earliest and chemotherapy. Also, there are no definite guidelines for optimal management. The prognosis remains poor with local recurrences and systemic complications with survival in the range of months to couple of years. Multimodal treatment offers hope of longterm and good quality survival.

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Author Information

Ram HS Shankar  
Junior Resident, Department of General Surgery, Kasturba Medical College . Manipal University

A Divaker Rao  
Professor, Dept of Neurosurgery, Kasturba Medical College . Manipal University

Chandra Kumar Ballal  
Professor, Dept of Neurosurgery, Kasturba Medical College . Manipal University

Ramdas Naik  
Professor, Department of Pathology, Kasturba Medical College . Manipal University