Case Report

Primary Ewing's sarcoma of lumbar vertebra-A rare case report

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Abstract

Ewing's sarcoma is a rare bone tumor, first described by James Ewing in 1921. It is more common in bone than soft tissues. Involvement of vertebral bodies is extremely unusual. We report a case of a five year old girl with presenting symptoms of pain, difficulty in walking since one month, paraparesis and bladder and bowel incontinence since one week. Plain radiographs revealed sclerotic fourth lumbar vertebra. CT and MRI lumbosacral spine revealed involvement of fourth lumbar vertebra with bilateral paraspinal masses. Immunohistochemistry and Fluorescent in situ hybridization of biopsy from paraspinal mass revealed round cell tumor - Ewing's sarcoma.

Key Word: Ewing's sarcoma, ivory vertebra, paraspinal mass, primary spine tumor.

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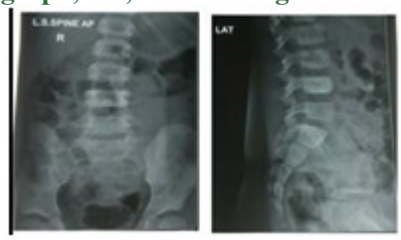
INTRODUCTION

Ewing's sarcoma is a rare, highly malignant tumour. Patient usually presents in second decade of life. It has predilection for long bones, femur being most commonly affected. However it can occur anywhere. Primary Ewing's sarcoma of spine is very rare, representing three to ten percent of all cases of Ewing's sarcoma. Microscopic examination demonstrates small, round blue cells with a uniform appearance. Electron microscopy, immunohistochemistry, and cytogenetic studies help differentiate ES from other small, round blue cell tumors. The translocation t(11;22)(q24;q12) is identified in most cases of Ewing's sarcoma. Here, we describe clinical presentation and radiological features of a case of primary Ewing's sarcoma of spine involving fourth lumbar vertebra.

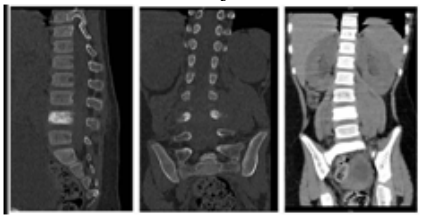
CASE HISTORY

A 5 year old girl presented with a three month history of pain and difficulty in walking. Paraparesis, bladder and bowel dysfunction were present at admission. No history of preceding viral fever or tuberculosis, no history of trauma. Physical examination of the patient disclosed paraspinal tenderness at lumbar region, inability to extend hips and knees completely, loss of bilateral knee and ankle jerks. No evidence of lymphadenopathy or pallor. No palpable mass in abdomen. Laboratory studies including complete blood picture, erythrocyte sedimentation rate, Mantoux, Hemoglobin were within normal limits.

Plain radiograph, CT, MRI findings are as follows.



Plain radiograph lumbar spine(AP and lateral views) showing sclerotic L4 vertebra with intact adjacent intervertebral disc spaces.



CT images showing body of L4 vertebra is hyperdense with decrease in height, with thickened trabecular pattern and involving pedicles. Bilateral paravertebral soft tissue masses extending from L1 to L5.

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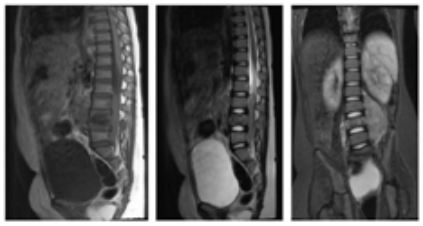

MRI images showing - Body of L4 vertebra is hypointense on T1W & T2W and hyperintense on STIR images. Diffuse paravertebral soft tissue mass present bilaterally from L1 to L5.Rest of the spine and spinal cord normal.

Patient was further evaluated by performing bone marrow aspiration, urine VMA/HVA levels, CT brain, the results of which were normal. Further skeletal scintigraphy was performed using Tc 99m MDP which localized pathology to L4 vertebra. Rest of survey unremarkable. A paraspinal mass biopsy was suggestive of malignant round cell tumor. Immunohistochemistry report was performed which said CD99 positive, CD117 positive, LCA and NSE negative - features consistent with Primitive Neuroectodermal tumor and final diagnosis came from Fluorescence insitu hybridization report where in there was positive EWSR1(22q12) gene in 66.6% cells examined - features confirmatory of Ewing's Sarcoma. The patient was subjected to 4 cycles of chemotherapy with vincristine, ifosfamide, etoposide, doxorubicin and there after the size of paraspinal masses decreased and patient's neurological status also improved.

Why an unusual presentation??

Feature	Classical ewing's sarcoma	Case in Discussion
M.C. Location	Lower limbs-45% , femur M.C.	Spine(3 – 10%)
Radiographic Findings	Permeative Laminated periosteal reaction	Sclerotic

Feature	Primary Ewing's Sarcoma of Spine	Case in discussion
M.C. Location	Sacrococcygeal	Lumbar(L4)
Age	12-24 Yrs	5 yrs
Pattern of Involvement	Multifocal	Unifocal(L4)
Radiographic Findings	Lytic Lesion(57%)	Sclerotic Lesion

DISCUSSION

Primary Ewing's sarcoma of spine is very rare[1]. It represents 3 to 10% of all cases of Ewing's Sarcoma. Most common location being sacrococcygeal region. In the study of primary vertebral Ewing's sarcoma (ES), the division of the spine into nonsacral and sacral is of utmost important because of the different behaviour of Ewing's sarcoma in these two regions in terms of response to therapy and survival rates. Patient usually presents at a mean age at presentation of 12-24 yrs[2] with the clinical trial of local pain, neurological deficits, and palpable mass. Other features include bladder and bowel dysfunction, sensory disturbance[3]. It is usually multifocal. Even when radiographs are normal the physician should be suspicious of underlying malignancy, especially when the patient is in the first two decades of life and symptoms, including severe local pain and tenderness, are not relieved by bed rest. The most common radiographic feature of Ewing's sarcoma of spine being lytic lesion[2] and complete flattening (vertebra plana) is next most common finding. Sclerotic lesions(ivory vertebra[5]) are rarely seen. CT better helps to know extent of involvement of vertebra. MRI is very sensitive in the early detection of ES in the spine[6]. MRI better delineates soft tissue extent and helps in preoperative planning to know surgical margins of tumor.

Differential diagnosis of ivory vertebra with para spinal soft tissue mass in paediatric age group -

1. Lymphoma
2. Metastatic Neuroblastoma
3. Metastatic Medulloblastoma
4. Osteosarcoma
5. Ewing's sarcoma

Because of biological heterogeneity, these tumors have variable sensitivity to radiation and chemotherapy[4]. Treatment consists of initial chemotherapy with vincristine, actinomycin, cyclophosphamide, doxorubicin to shrink the tumor before definitive treatment which consists of surgical resection or radiotherapy. Prognosis is better for non sacral type.

CONCLUSION

Ewing's sarcoma of spine is extremely rare (3-10% of Ewing's sarcoma), in which lumbar spine involvement is unusual. Though permeative type of bone destruction is described in Ewing's sarcoma, sclerosis is noted in the present case. Presentation in first decade and unifocal involvement are also rare in Ewing's sarcoma of spine.

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