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RESEARCH ARTICLE

LANGERHANS CELL HISTIOCYTOSIS AS AN UNUSUAL CAUSE OF BACK PAIN IN PAEDIATRIC PRACTICE: A CASE REPORT

[1*]Rinsy, PV, [1]Jayakumar, C, [2]Raghavi, T. and [1]Sajesh Menon

Department of Pediatrics and Neurosurgery Amrita Institute of Medical Sciences, Kochi
Department of Neurosurgery and Neurosurgery Amrita Institute of Medical Sciences, Kochi

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ABSTRACT

Back pains in children are generally related to postural, traumatic or mechanical causes . Most children are fortunate to have a self limiting course. However paediatricians must have a high index of suspicion on unusual causes of back pain ,especially when the pain is disproportionate to history and clinical signs. We report a case of 5-year-old girl who presented with acute back pain following a trivial trauma. She was initially evaluated for tuberculosis spine, but soon diagnosed as Langerhans cell histiocytosis of spine and underwent surgery.

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INTRODUCTION

Low back pain in children has a wide list of differentials, including traumatic, mechanical, somatisation, infectious, inflammatory, and malignancy. Most of the presentations are self limiting and respond to analgesics. Usually any child with persistent pain, especially worse at night, disproportionate to physical findings and not relieved on analgesics are evaluated extensively for a more serious cause[1].But we present a case of a 5-year-old well girl who presented with acute back pain following a trivial trauma and found to have Langerhans cell histiocytosis (LCH) of the spine.

CASE REPORT

A 5 year old apparently well female child presented with acute back pain following a fall during play from a 3 feet slide. There was also a history of adopting unsupervised yoga postures at home. She was initially managed with local measures for pain at home. She had no other constitutional symptoms. She was further evaluated as excruciating pain persisted. On examination, she had no lymphadenopathy or hepatosplenomegaly. She could not bear weight and on local examination, there was tenderness and swelling over lumbar region. The rest of the musculoskeletal system and neurological examination was normal. Plain radiology showed compression at D9 vertebra (Fig 1).

***Corresponding author: Rinsy, PV,**
Department of Pediatrics and Neurosurgery Amrita Institute of Medical Sciences, Kochi.

Labs revealed normal blood counts, no anemia, normal ESR, negative inflammatory markers.Chest xray was normal,tuberculin test negative and gastric aspirate was negative for mycobacterium.USG abdomen and urine osmolality was normal.MRI spine showed pathological D9 vertebral collapse with prevertebral, paravertebral and epidural soft tissue (Fig 2). No cord compression.D7, D8-D10,D11 Pedicle screws and rod fixation with gross total excision of tumour involving D10 and dorsal stabilization was done under electrophysiological monitoring. Histopathology showed large cells having abundant foamy eosinophilic cytoplasm and pleomorphic convoluted/ cleaved nuclei with some showing prominent nucleoli, positive for Cd1a and S100 with a Ki 67 index of approximately 20%, favouring a diagnosis of Langerhans cell histiocytosis. Whole body PET CT imaging didnot show any metabolically active lesion both locally in the remaining vertebra and systemically. As she had single system LCH with low risk and age less than 5 years, she was planned for follow up and radiation only if it recurs.

DISCUSSION

Any back pain in children less than 4 years, persistent nocturnal pain, neurological symptoms, self-imposed activity limitations, and systemic symptoms are significant.[2]Usually infections such as osteomyelitis and discitis are associated with disturbing persistent nocturnal pain. Among bone tumors, benign neoplasms include osteoid osteoma, osteoblastoma, neurofibroma, and aneurysmal bone cyst.

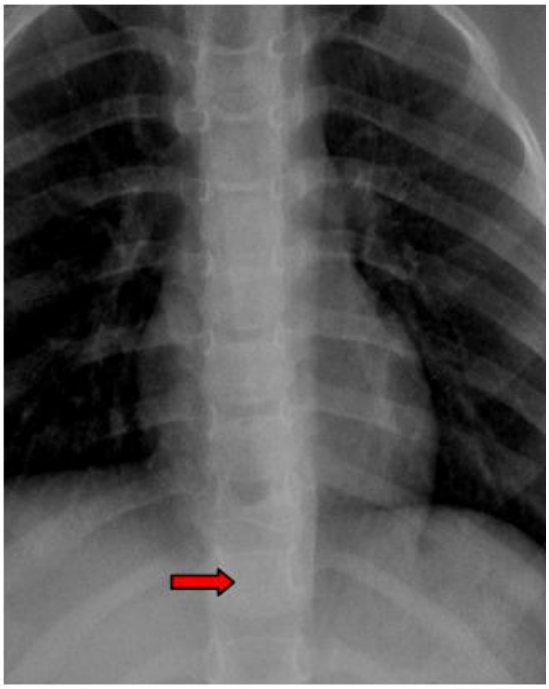

Fig 1. Plain radiograph showing collapse of D9 Vertebra

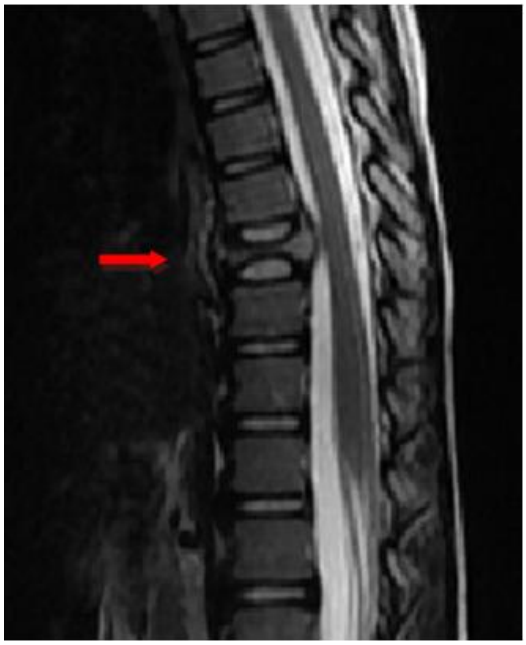

Fig 2. MRI Spine T2 weighted sequence sagittal view showing D9vertebral collapse with prevertebral ,postvertebral and epidural soft tissue. No cord compression

Malignancies include Ewing's sarcoma, lymphoma, neuroblastoma, astrocytoma, leukemia, LCH, and metastatic disease. Of which, LCH accounts for <1%. Langerhans cell histiocytosis (LCH) usually affects children of 1-5 years age with a male predilection, twice that of the females[3].It is an uncommon hematological disorder of the reticuloendothelial system associated with proliferation of Langerhans cells and mature eosinophils. Etiology is attributed to immune dysregulation or a neoplastic proliferation. Adults and older children usually present as bone involvement alone. Young infants usually present with skin rash, otitis media, fever, organomegaly, anemia, pain and pathological fracture of involved bone, and diabetes insipidus indicating multisystem involvement[4]. Though LCH can involve any bone, the usual sites are pelvis, ribs, skull, long bones, vertebra, and facial bones. The vertebral body is the most common affected part of spine in LCH. The incidence of spinal involvement varies from 6.5% to 25% in LCH of the bone[5]. Garg **et al**. have reported 45% lesions in the cervical spine; 32% in the thoracic spine; and 23% in the lumbar spine[6].Children with only skeletal lesions have been found to have a good prognosis when compared to children with systemic involvement[7]. In case of a predominant spinal presentation, along with spinal imaging,basic lab workup includes a complete blood count, liver function tests, erythrocyte sedimentation rate, tuberculin test, gastric aspirate for evidence of mycobacterium. Multisystem involvement is assessed by urine osmolality, chest radiograph, ultrasonography of the liver and spleen, bone marrow study and bone scan. Diagnosis of LCH is confirmed histopathologically. Light microscopy will show large histiocytes with grooved nuclei along with mononuclear histiocytes and multinucleated giant cells, lymphocytes, plasma cells, and neutrophils. Electron microscopy shows tennis racket" shaped cytoplasmic inclusions within histiocytes (Birbeck granules). Currently diagnostic value is high for demonstarting immunopositivity for CD1a marker and S100 protein.Biopsy specimen in our case was also positive for same, confirming the diagnosis.

The treatment options available are curettage, intralesional steroids, chemotherapy, and radiotherapy depending on size or number of lesions. Treatment aims at spinal stability, preservation of neurological function, and eradication of the lesion are the goals of treatment. Surgical options are considered over spinal orthoses if spinal segment is unstable and can cause cord compression. Chemotherapy itself rapidly reduces the intraspinal soft tissue mass and relieves the local and radicular pain even in patients with spinal cord and nerve root compression[5].

Conclusion

A child presenting with persistent and progressive back pain should be evaluated extensively even if it is not associated with any other constitutional symptoms to rule out rare causes of vertebral lytic lesion. As delay in the diagnosis of vertebral lytic lesion may have neurological sequelae in the growing children, any vertebral pain disproportionate to the signs should have low threshold levels for evaluation. Contributors: PVR reviewed the literature and drafted the initial manuscript. CJ was involved in the management of the patient, designing the article, revision of manuscript and will act as a guarantor for the paper.CJK and SJM were involved in the management of the patient .TR helped in image acquisition.

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