Case Report International Journal of Spine | 2022 January-June; 7(1): 19-23

Langerhans Cell Histiocytosis of Dorsal Spine in a Child Presenting with Deformity of the Back: A Rare Case Report and Review of Literature

Bharath HD[1], Krishnakumar R[1], Jeffy John[2]

Abstract

Background: Back pain in the pediatric population is a common complaint with wide differentials including mechanical cause, musculoskeletal involvement, infectious or inflammatory pathologies, and tumors. Transient back pain is common among children, and in majority of cases resolves without any treatment or with rest and mere hot/cold fomentations. However, pain disproportionate to physical findings and not reduced by pain medications, strongly suggest the presence of a serious underlying pathology.

Case presentation: A 13-year-old boy presented with a history of mild mid back pain lasting for a week associated with gradually developing deformity of the back, noticed by his parents. There is no history of any injury or fall prior to presentation. He had no history of any other constitutional symptoms. Upon radiological investigations he was found to have an isolated lytic lesion in D9 vertebra with differentials of tubercular/neoplastic/metastatic etiologies. Histopathological examination after biopsy was suggestive of Langerhans cell histiocytosis and was proven by immunohistochemistry. He was managed with conservative line of treatment with analgesics and other supportive care.

Discussion: LCH is considered a pediatric disease, and involvement of the pediatric spine in not uncommon. A child presenting with persistent and progressive back pain should be evaluated in detail even if the child is not presenting with any other constitutional symptoms to rule out rare causes of vertebral lytic lesions. There are no cases reported in the literature suggesting LCH of the spine in pediatric population presenting as the deformity of the back. Conservative treatment is a good choice for a patient with LCH without neurological deficits or spinal instability.

Keywords: Spine deformity, Children, LCH, Back pain

Introduction

Back pain in the pediatric population is a common complaint with wide differentials including mechanical cause, musculoskeletal involvement, infectious or inflammatory pathologies, and tumors. Vertebral and spinal neoplasms are rare in children and typically present with persistent and localized or generalized back pain that is worse at night time, constant or related to activities and symptoms lasting less than 3 months [1]. Transient back pain is common among children, and in majority of cases resolves without any treatment or with rest and mere hot/cold fomentations. However, pain disproportionate to physical findings and not reduced by pain medications, strongly suggest the presence of a serious underlying pathology.

Here we are presenting a case of a 13-year-old boy who presented with mild back pain and deformity of the back and found to have Langerhans cell histiocytosis (LCH) of the spine.

Case Report

A 13-year-old boy presented with a history of mild mid back pain lasting for a week associated with gradually developing deformity of the back, noticed by his parents. There is no history of any injury or fall before presentation. He had no history of any other constitutional symptoms. He has a past history of cardiac surgery (VSD closure) at the age of three years. The child was initially managed with oral analgesics and anti-inflammatory medications for 10 days after consulting a physician. After the medications, his pain completely vanished and the back deformity was started to decrease. He was later taken to an Orthopaedician who advised

[1]Department of Spine Surgery, Medical Trust Hospital, Cochin, Kerala, India.
[2]Department of Pathology, Medical Trust Hospital, Cochin, Kerala, India.

Address of Correspondence
Dr. Krishnakumar R,
Department of Spine Surgery, Medical Trust Hospital, Cochin, Kerala, India.
E-mail: krishnakumar.ram@gmaiall.com

Submitted: 01/03/2022; Reviewed: 26/03/2022; Accepted: 27/04/2022; Published: 10/06/2022

International Journal of Spine | Available on www.ijsonline.co.in | DOI:10.13107/ijs.2022.v07i01.034

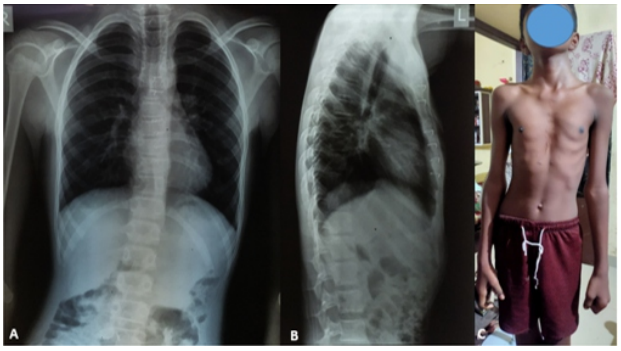

Figure 1. A, B, C. A: X-ray dorso-lumbar spine AP: showing deformity of the spine with list towards left. B: Lateral view showing anterior collapse of D9, C: Clinical picture showing deformity (listing towards left) (x-rays show sternal SS wires s/o cardiac surgery for VSD)

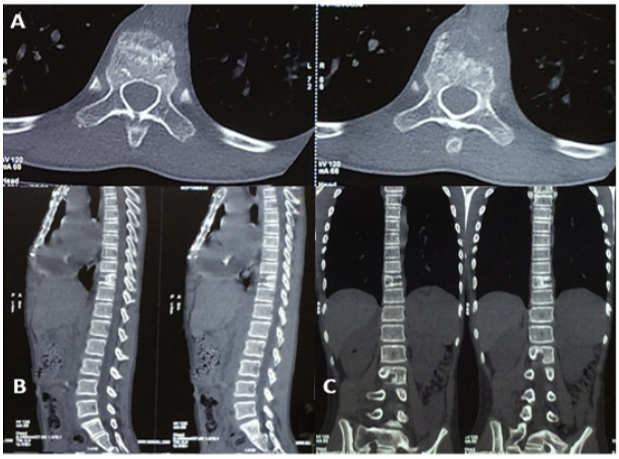

Figure 2. A, B, C: NCCT dorsal spine showing A(axial study) showing osteolytic area in anterior and left lateral quadrant of the D9 vertebral body, B(sagittal), C(coronal) showing osteolytic area in anterior aspect of D9 vertebral body and reduced left lateral height of the D9 vertebral body

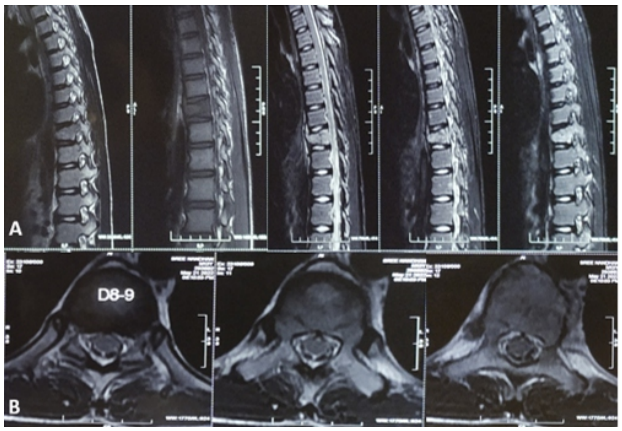

Figure 3. A, B: MRI showing Lumbosacral transitional vertebra L5, STIR mildly hyperintense and T1 isointense lesion in anterior aspect of D9 causing mild anterior and left lateral height reduction with paravertebral soft tissue thickening. Possibilities: LCH, TB, less likely metastasis

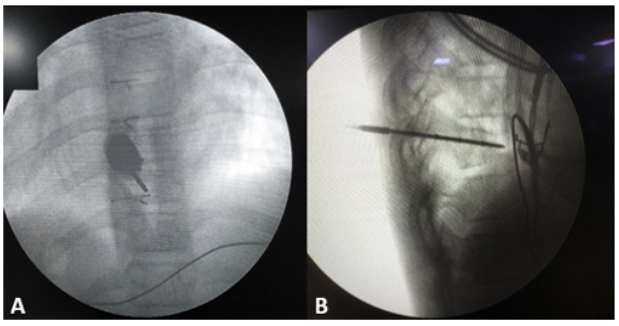

Figure 4. A, B (AP, Lateral view): Showing C-arm intra-op images of transpedicular biopsy needle in left pedicle of D9

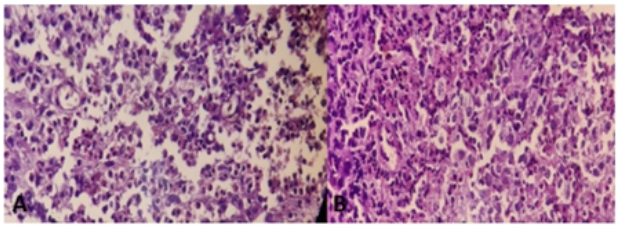

Figure 5. A, B: Histopathological examination showing diffuse inflammatory cell rich lesion composed of plenty of eosinophils and mononuclear cells with many histiocytic cells with reniform nuclei showing fine chromatin and longitudinal grooving. Mild to moderate nuclear pleomorphism, scant mitosis with admixed microvasculature and hemorrhage. No evidence of granuloma

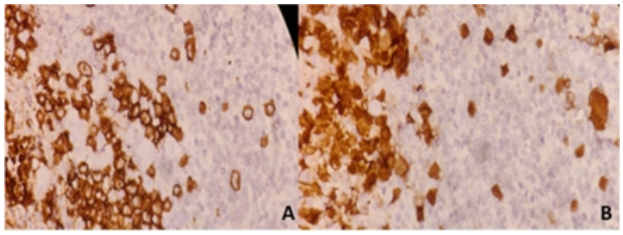

Figure 6. A, B: Immunohistochemistry study showing the lesional cells are immunoreactive for (A) CD1a, (B) S100

Figure 7. A, B: Immediate post-operative (A) and 2 week's post-operative follow-up (B) clinical pictures showing improvement in deformity of the spine

radiological investigations including plain X-ray, CT scan and MRI plain and contrast study of the spine. However, in view of persisting painless deformity, the child was evaluated for tuberculosis spine, neoplasms and other infective pathologies. On examination, child had no constitutional symptoms, lymphadenopathy or hepato-splenomegaly. On local examination, there was no tenderness over the dorso-lumbar spine. Deformity of the spine with listing towards left were present. The rest of the musculoskeletal system and neurological examination was normal.

Blood investigations revealed normal total and differential counts, erythrocyte sedimentation rate and C Reactive protein. Chest X ray showed normal lung fields. The spine X ray showed anterior collapse of D9 vertebral body with deformity of the dorso-lumbar spine showing mild list towards left (Fig.1. A, B, C). With a provisional diagnosis of lytic lesion in the D9 vertebra, NCCT dorso-lumbar spine (Fig.2. A, B, C) was done showing lytic lesion in the anterior aspect of the D10 with decreased height and minimal thickening of pre and paravertebral soft tissues, probability of infective spondylitis, tubercular. Magnetic resonance imaging (MRI) plain & contrast study (Fig. 3.A, B) of the spine was done which showed transitional lumbosacral vertebra L5, a STIR mildly hyperintense and T1 isointense lesion in anterior aspect of D9 causing mild anterior and left lateral height reduction with pre-vertebral soft tissue thickening, possibilities being LCH, TB, less likely any metastasis as no other lesions were found in rest of the screening.

Transpedicur biopsy of D9 vertebra was performed under general anesthesia (GA) under c-arm guidance (Fig.4. A, B) and the tissue was sent for microbiological examinations including gram staining, ZN staining, culture, Gene Xpert and TB-PCR, and histopathological examination. Microbiological staining studies did not reveal any organisms, culture were negative for any growth. Gene Xpert TB-PCR were negative for Mycobacterium tuberculosis. Frozen section study was suggestive of a lesion rich in inflammatory cells. Histopathological examination showed diffuse inflammatory cell rich lesion composed of plenty of eosinophils and mononuclear cells with many histiocytic cells with reniform nuclei showing fine chromatin and longitudinal grooving. Mild to moderate nuclear pleomorphism, scant mitosis with admixed microvasculature and hemorrhage. No evidence of granuloma (Fig. 5.A, B). In immunohistochemistry study the lesional cells are immunoreactive for CD1a, S100 and CD68 and non-immunoreactive for CD30 confirming the diagnosis of LCH (Fig.6. A, B). Medical oncologist opinion was obtained and PET CT scan was done to look for any other lesions. PET CT showed no abnormal increased FDG uptake in lytic lesion of D9 or in lymph nodes, spleen, liver, lungs, and adrenal glands and in rest of the skeleton up to the thighs. Impression given by PET CT was FDG non avid mixed lytic/sclerotic lesion in D9 vertebral body: metabolically quiescent solitary skeletal involvement, No FDG avid/CT demonstrable other skeletal/extra-skeletal involvement. The child was managed with IV analgesics and antibiotics during the time of admission, followed by oral antibiotics and anti-inflammatory medications for a week post discharge from hospital. His surgical wound healed well and he is completely symptom free now and has normal daily activities. His spine deformity is also gradually getting corrected (Fig. 7.A, B) and he is been kept under observation in regular OPD follow-ups.

Discussion

Low back pain is common in pediatric and adolescent age group, which can be due to mechanical problems such as poor posture habits, physical activity, and neglected injuries. Fever, loss of appetite and other constitutional symptoms occur in the presence of an infection or tumorous conditions. The red flag signs warranting immediate evaluation are age less than 4 years, persistent nighttime pain, neurological symptoms, activity limitations, and systemic symptoms [1]. Back pain that occurs during night and awakens the child is usually associated with infections such as osteomyelitis and discitis. Benign neoplasms such as osteoid osteoma, osteoblastoma, neurofibroma, and aneurysmal bone cyst are few other causes of back pain in children. Malignancies like Ewing's sarcoma, lymphoma, neuroblastoma, astrocytoma, leukemia, LCH, and metastatic disease cannot be neglected unless proven otherwise [2].

LCH is a rare yet not uncommon disease associated with a proliferation of Langerhans cells involving the reticuloendothelial system. LCH comprises a spectrum of disorders. Localized disease is described as eosinophilic granuloma, while multisystem forms of LCH have names such as Hand-Schüller Christian disease, Letterer-Siwe disease, and systemic histiocytosis X. Our case was a solitary osseous lesion in D9 without systemic involvement. Although LCH most commonly occurs in the first two decades of life, it may affect patients of any age, from infants to elderly individuals. Annual 3-5 cases per million found among children. Among bone tumors, LCH accounts for <1%, and up to 80% of these children will present before the age of 10 years [3]. The most frequently affected site in children with LCH is the bone which is encountered in about 75%–80% of patients with LCH and may be the only affected site, especially in children older than 5 years of age [4]. The most frequent sites of skeletal lesions are the skull, femur, mandible, pelvis, and spine. LCH in the spine is reported to mainly involve the vertebral bodies, occurring in 6.5–25 % of cases, with the most frequent site being the thoracic vertebrae (54 %) in children, and the cervical vertebrae in adults [5, 6]. The incidence of spinal involvement varies from 6.5% to 25% in LCH of the bone [7]. In one study, they have reported 45% lesions in the cervical spine; 32% in the thoracic spine; and 23% in the lumbar spine [8]. Children with only skeletal lesions have been found to have a good prognosis when compared to children with systemic involvement [9]. In the vertebral body, LCH of spine appears as an osteolytic lesion causing complete collapse of the body, called as vertebra plana. The incidence of soft tissue extension in children with LCH of the spine is about 50%. In our case MRI showed mild pre and paravertebral soft tissue thickening with reduction of lateral height. Back or neck pain was the most common presenting complaint and was the only presenting symptom in one study. Torticollis and abnormal gait were the other symptoms of presentation [8].

Evaluation of these patients typically includes complete blood counts, liver function tests, ESR, CRP, radiological investigations including x-rays of whole spine, chest x-ray, CT and MRI (plain &

contrast) scans to look for osseous and extra-osseous involvement. Some studies recommend technetium bone scan or skeletal survey. Histopathological examinations classically show the picture of diffuse inflammatory cell rich lesions composed of plenty of eosinophils and mononuclear cells with many histiocytic cells. The diagnosis of LCH is confirmed by immunohistochemical staining for CD-1a and S-100 protein [10].

Spinal stability, preservation of neurological function, and eradication of the lesion are the goals of treatment. Spinal LCH is self-limiting and the prognosis is usually good. Various treatment modalities for spinal LCH have been established, including conservative management, intralesional steroid injection, radiation therapy, chemotherapy, and curettage with or without reconstructive surgery [8, 11, 12]. Mild, isolated lesions of the spine without neurological involvements or spinal instability can be managed with conservative treatment such as simple observation, non-steroidal anti-inflammatory drugs, immobilization or rest.

Surgery is generally reserved for selected cases with severe mechanical instability, deformity, or neurological deficit due to the disease per se or due to compression caused by the lesion [8]. As spinal LCH is mostly a self-limiting condition, especially in pediatric population, most authors recommend conservative line of treatment for disease without neurological or mechanical complications, as in our case.

Conclusion

There is paucity of literature on LCH of pediatric spine with deformity. LCH is considered a pediatric disease, and involvement of the pediatric spine in not uncommon. A child presenting with persistent and progressive back pain should be evaluated in detail even if the child is not presenting with any other constitutional symptoms to rule out rare causes of vertebral lytic lesions. Conservative treatment is a good choice for a patient with LCH without neurological deficits or spinal instability.

Declaration of patient consent : The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

Conflict of Interest: None; **Source of Support:** None

References

1. Bernstein RM, Cozen H. Evaluation of back pain in children and adolescents. Am Fam Physician 2007;76:1669 76.
2. Feldman DS, StraightJJ, Badra MI, MohaideenA, Madan SS. Evaluation of an algorithmic approach to pediatric back pain. J Pediatr Orthop 2006;26:353 7.
3. Brown CW, Jarvis JG, Letts M, Carpenter B. Treatment and outcome of vertebral Langerhans cell histiocytosis at the Children's Hospital of Eastern Ontario. Can J Surg 2005;48:230 6.
4. Khung S, Budzik JF, Amzallag Bellenger E, LambillioteA, Soto Ares G, Cotten A, et al. Skeletal involvement in Langerhans cell histiocytosis. Insights Imaging 2013;4:569 79.
5. Beltran J, Aparisi F, Bonmati LM, Rosenberg ZS, Present D, Steiner GC (1993) Eosinophilic granuloma: MRI manifestations. Skelet Radiol 22(3):157–161

1. Bernstein RM, Cozen H. Evaluation of back pain in children and adolescents. Am Fam Physician 2007;76:1669 76.
2. Feldman DS, StraightJJ, Badra MI, MohaideenA, Madan SS. Evaluation of an algorithmic approach to pediatric back pain. J Pediatr Orthop 2006;26:353 7.
3. Brown CW, Jarvis JG, Letts M, Carpenter B. Treatment and outcome of vertebral Langerhans cell histiocytosis at the Children's Hospital of Eastern Ontario. Can J Surg 2005;48:230 6.
4. Khung S, Budzik JF, Amzallag Bellenger E, LambillioteA, Soto Ares G, Cotten A, et al. Skeletal involvement in Langerhans cell histiocytosis. Insights Imaging 2013;4:569 79.

6. Kilborn TN, Teh J, Goodman TR (2003) Paediatric manifestations of Langerhans cell histiocytosis: a review of the clinical and radiological findings. Clin Radiol 58(4):269–278
7. Peng XS, Pan T, Chen LY, Huang G, Wang J. Langerhans' cell histiocytosis of the spine in children with soft tissue extension and chemotherapy. Int Orthop 2009;33:731 6.
8. Garg S, Mehta S, DormansJP. Langerhans cell histiocytosis of the spine in children. Long term follow up. J Bone Joint Surg Am 2004;86 A: 1740 50.
9. Ghanem I, Tolo VT, D'Ambra P, Malogalowkin MH. Langerhans cell histiocytosis of bone in children and adolescents. J Pediatr Orthop 2003;23:124 30.
10. Azouz EM, Saigal G, Rodriguez MM, Podda A (2005) Langerhans' cell histiocy tosis: pathology, imaging and treatment of skeletal involvement. Pediatr Radiol 35(2):103–115. doi:10.1007/s00247-004-1262-0
11. Puigdevall M, Bosio S, Hokama J, Maenza R (2008) Langerhans cell histiocytosis of the atlas in the pediatric spine: total reconstitution of the bone lesion after nonoperative treatment. A report of two cases. J Bone Joint Surg 90(9):1994–1997. doi:10.2106/jbjs.g.01352
12. Bertram C, Madert J, Eggers C (2002) Eosinophilic granuloma of the cervical spine. Spine 27(13):1408–1413

How to Cite this Article

HD Bharath, R Krishnakumar, John J | Langerhans Cell Histiocytosis of Dorsal Spine in a Child Presenting with Deformity of the Back: A Rare Case Report and Review of Literature | International Journal of Spine | January-June 2020; 7(1): 19-23.