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Case Report

Langerhans' cell histiocytosis involving posterior elements of the dorsal spine: An unusual cause of extradural spinal mass in an adult

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Abstract

Langerhans cell histiocytosis (LCH) is a clonal proliferation of Langerhans cells occurring as an isolated lesion or as part of a systemic proliferation. It is commoner in children younger than 10 years of age with sparing of the posterior elements in more than 95% of cases. We describe a case of LCH in an adult female presenting with paraplegia. MRI revealed a well-defined extradural contrast enhancing mass at D2-D4 vertebral level involving the posterior elements of spine. D2-5 laminectomy with excision of lesion was performed which lead to marked improvement of patients neurological status. Histopathology was suggestive of eosinophilic granuloma. We describe the case, discuss its uniqueness and review the literature on this rare tumor presentation.

Key words: Dorsal spine, langerhans cell histiocytosis, in adult patient, posterior elements of spine

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a clonal proliferation of Langerhans cells occurring as an isolated lesion or as part of a systemic (multifocal) proliferation.[1] The designation of LCH has replaced the previous nomenclature of the group of diseases termed histiocytosis X, which included eosinophilic granuloma (EG), Letterer-Siwe Syndrome and Hand-Schuller-Christian disease.[2] The incidence of spinal involvement in this disease varies from 6.5 to 25% of cases with LCH of the skeleton. Eighty percent of cases occur in children younger than 10 years of age.[3] In nearly 95% of cases of spinal LCH, there is destruction of the vertebral body, sparing the posterior elements.[4] Isolated involvement of posterior elements of spine in a child has been described by Garg *et al*.[4] Our's is the first to describe such a case in an adult who presented with progressive lower limb weakness. The etiology, pathophysiology, differential diagnosis and treatment of this disease are discussed with reference to our case.

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CASE REPORT

A 35-year-old housewife presented to us with complaints of progressive loss of power in both lower limbs since two months, tingling and numbness in both lower limbs, inability to pass urine and constipation of 2-week duration. She was symptom-free 2 months ago when she initially noticed mild weakness in left lower limb which spread to the right side in the next week. X-ray of dorsal spine done at local health center was reportedly normal. The weakness progressively worsened to the present state of no movement in the limbs 1 week preceding her presentation to us. On examination the power in both lower limbs was 0/5 with associated hypertonia (spastic paraplegia). Light touch and pinprick sensations were present but other sensations were grossly absent. There was no abnormality in both upper limbs. She was already catherized on presentation to us.

Magnetic resonance imaging (MRI) study of dorsal spine revealed a well-defined extradural contrast enhancing mass lesion at D2-D4 vertebral level on right side of spinal canal, extending into the D2-D3 and D3-D4 neural foramina causing compression on the cord with displacement of the spinal cord to the left. [Figures 1 and 2] T2W images showed subtle cord edema in the spinal cord at D2-D4 level. [Figure 3] A preoperative diagnosis of tuberculoma, organized hematoma, meningioma, schwannoma or metastasis was made based on radiological investigations. In view of neurological deficits and radiological evidence of spinal cord compression, a decision to operate the patient was taken.

In prone position, D2-D5 decompressive laminectomy was performed. The lesion was well circumscribed, moderately vascular and firm. It was easily separable from the dura and seemed to be arising from the lamina of the D3 vertebra. [Figure 4a] The tumor was removed in toto and the cord was lax at the end of procedure. [Figure 4b] Patient was extubated on table with no added neurological deficits.

Frozen section was suggestive of hemolymphoid malignancy. Histopathology showed fibrocollagenous tissue infiltrated by dense collection of lymphocytes, plasma cells and larger cells with vesicular nuclei resembling histiocytes, few eosinophils and occasional mitotic figures were also seen within the lesion suggestive of LCH. Patient was subjected for further investigations to find other foci in the body. Computed tomography chest, ultrasound breast and X-ray chest revealed nothing.

Patient was subjected to neurorehabilitation therapy with the help of physiotherapist and occupation therapist. Her power in the lower limbs improved progressively and by postoperative day 9, she was ambulatory with support. She had to strain at micturition and needed laxatives for almost a month after surgery. Her delayed MRI performed 6 months postoperatively showed total excision of lesion. [Figure 5] On follow-up of 1 year she was ambulatory with walker stick and her bowel bladder symptoms had improved. Examination revealed power of Grade 4+/5, reflexes were brisk but there was no clonus. Fifteen months after the spinal surgery she developed another lesion in the uterus for which hysterectomy was done. Patient was referred for systemic chemotherapy and is presently asymptomatic.

DISCUSSION

LCH is a disease caused by the monoclonal proliferation

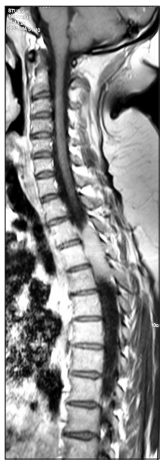

Figure 1: Postcontrast sagittal image of spine showing a contrast enhancing extradural lesion at D2-4 level with cord compression

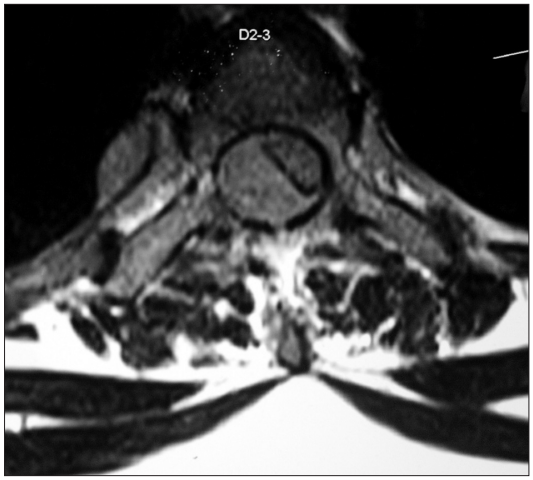

Figure 2: Postcontrast axial image of spine at D2-3 level showing the lesion occupying almost all of the spinal canal with displacement of the cord to left

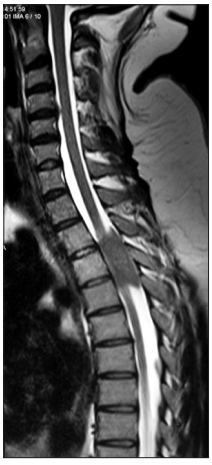

Figure 3: T2W sagittal image showing tumor compression of the cord with subtle changes in the cord signal intensity

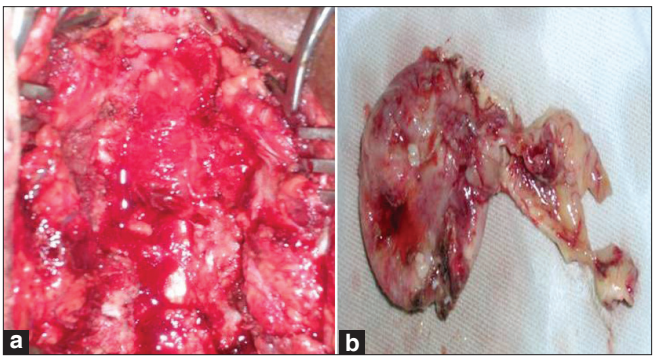

Figure 4: (a) Intraoperative picture after laminectomy showing the extradural tumor in situ and (b) specimen following excision

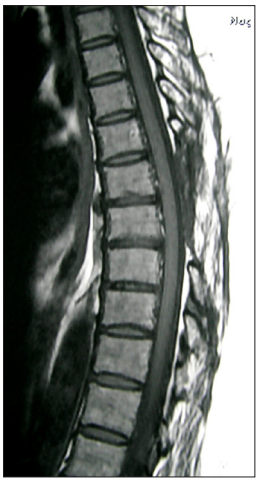

Figure 5: Postoperative postcontrast sagittal image of the cord showing complete excision of the lesion with postoperative changes

of Langerhans cells.[5] Lichtenstein (1953) grouped the Eosinophilic Granuloma, the Hand–Schüller–Christian disease, and the Letterer–Siwe disease, three different syndromes with the same histology, under the term "Histiocytosis X".[6] This term has been replaced nowadays by LCH.

LCH is a rare disease that is found mostly in children, with an estimated incidence between 0.2 and 2.0 cases per 100,000 children under 15 years of age and a peak incidence of age between 2–4 years. LCH has a higher incidence in patients with a persistent or transient systemic immunodeficiency, such as viral infections, leukemia, lymphoma, or genetic defect.[7] Even among the adults, this disease has preponderance in males, sometimes as high as 60–70%, and is more common in whites of northern European descent.[7] Our patient was a middle-aged adult female of South Asian origin and there was no history of previous medical illness.

There are three key subtypes of LCH: unifocal, multifocal, and systemic LCH. The most common presentation is unifocal (about 65%) and the bone is the most frequently affected tissue making up 90% of such cases.[8] The most common site involved is the skull and it accounts for more than 50% of cases.[7,8] In the spine it usually presents with local pain and stiffness only.[2] Neurological symptoms are extremely rare and are usually limited to mild paresthesias or radicular pain.[9] In our case the patient had severe neurological deficits in the form of paraplegia and bowel/bladder symptoms.

There are different ways to treat the unifocal LCH and there have been four approaches described in the literature. First option is not to offer any treatment at all, because in some eosinophilic granulomas self-limited growth has been reported, as was spontaneous regression. Second option is to attempt at least a partial resection or to perform a complete excisional biopsy of the lesion. Third option is a biopsy, followed by low-dose radiation to the lesion usually with 6–10 Gy with a local control rate of approximately 80%. Fourth option, is with intralesional corticosteroids, usually methylprednisolone 30–125 mg to limit immunoresponse, followed by sequential imaging. Finally, systemic chemotherapy may be indicated when LCH progresses systemically or is recurrent.[9]

In 95% of cases, LCH of the spine presents as a focal osteolytic vertebral lesion, with or without collapse of the vertebral body.[4,9] Posterior arch involvement is very uncommon.[2,4] The level of spinal involvement varies at different ages. While in children, the thoracic spine (54%) is commoner, in adults, 47% of reported cases involve the cervical spine, 33% the thoracic spine, and 20% the lumbar spine.[10] Our patient had exclusive involvement of the posterior elements of dorsal spine and there was no osteolytic component.

Our case was unique in several respects: 1. Adult female patient, 2. Posterior element involvement at the level of dorsal spine, 3. Absence of osteolytic lesion, 4. Severe neurological deficits and 5. Absence of comorbid factors. Eosinophilic granuloma should be considered as a differential diagnosis in all cases of spinal space occupying lesions even in the absence of typical features of LCH and the patient should be investigated for associated lesions in the body. Surgical excision should be offered when there is presence of neurological deficits. Patients should be watchfully observed after surgical excision. Radiotherapy/chemotherapy can be offered in recurrent disease or systemic involvement.

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