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Case Report

Langerhans Cell Histiocytosis Masquerading as a Spinal Tumour in a Young Adult : A Case Report

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ABSTRACT:

Langerhans cell histiocytosis (LCH) is a rare clonal proliferative disorder of dendritic cells, predominantly affecting children. Adult spinal involvement is uncommon and may radiologically mimic malignant lesions.

A 33-year-old male presented with a three-week history of progressive mid-thoracic back pain without neurological deficits. MRI revealed an infiltrative lesion involving D3–D4 vertebrae with epidural and paraspinal extension, suggestive of a lymphoproliferative neoplasm. Surgical decompression was performed. Histopathological examination showed sheets of histiocytes with characteristic nuclear grooves in an eosinophil-rich background. Immunohistochemistry demonstrated positivity for CD1a and S100, confirming LCH.

Adult spinal LCH can mimic malignant tumours on imaging, posing a diagnostic challenge. Histopathology with immunohistochemistry remains the cornerstone for definitive diagnosis. Early recognition and appropriate multidisciplinary management are essential for favourable outcomes.

Keywords: Langerhans cell histiocytosis, Spinal mass, CD1a, S100

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare clonal disorder characterized by proliferation of dendritic cells with features similar to epidermal Langerhans cells.[1] It demonstrates a wide clinical spectrum ranging from isolated bone lesions to multisystem disease.[2]

Skeletal involvement is most common, but vertebral lesions are relatively rare, particularly in adults.[3] Unlike the classical pediatric presentation of vertebra plana, adult spinal LCH may present as infiltrative lesions with soft tissue extension, often mimicking lymphoma, metastasis, or infection.[4] This overlap frequently leads to diagnostic difficulty, making histopathological confirmation essential.[5]

CASE HISTORY

A 33-year-old male presented with gradually progressive mid-thoracic back pain for three weeks. The pain was dull aching, localized, and non-radiating, with no associated motor weakness, sensory disturbances, or bowel-bladder involvement. There was no history of trauma, fever, weight loss, or prior tuberculosis.

General physical and neurological examinations were unremarkable. Local examination revealed tenderness over the mid-thoracic spine. Chest X-ray (PA view) was normal.

Laboratory investigations including hemoglobin (14.2 g/dl), total leukocyte count (8600/mm³), bleeding time (2 minutes), clotting time (7 minutes), prothrombin time (14.2 seconds), INR (1.0), and serum creatinine (1.02 mg/dl) were within normal

limits. Serological tests for HIV, HBsAg, and HCV were non-reactive/negative.

Magnetic resonance imaging (MRI) of the cervico-dorsal spine revealed abnormal marrow signal involving D3 and D4 vertebrae, appearing iso-intense on T1-weighted images and hyperintense on T2/STIR sequences, with patchy diffusion restriction on DWI/ADC. There was marked post-contrast enhancement along with an enhancing epidural soft tissue component causing significant thecal sac compression and displacement of the spinal cord. A large paraspinal soft tissue component extending into adjacent spaces with encasement of the exiting nerve roots was also noted. These imaging findings were suggestive of an infiltrative neoplastic lesion involving the vertebra and adjacent soft tissue, with consideration of lymphoproliferative disorder.

In view of the risk of cord compression, the patient underwent D3–D5 laminectomy with decompression of the lesion. Intraoperatively, an extradural firm mass involving vertebral and paraspinal tissues was identified and excised.

Gross examination of the specimen: Received multiple fragmented grey-brown soft tissue pieces from extension of subcutaneous tissue, bone, intravertebral tumor (D3–D5 level), and tumor originating from soft tissue, measuring 2.5 × 2 × 0.8 cm, 3 × 2.5 × 0.8 cm, 3.5 × 2.2 × 1.1 cm, and 2 × 2 × 0.8 cm in aggregate, respectively.

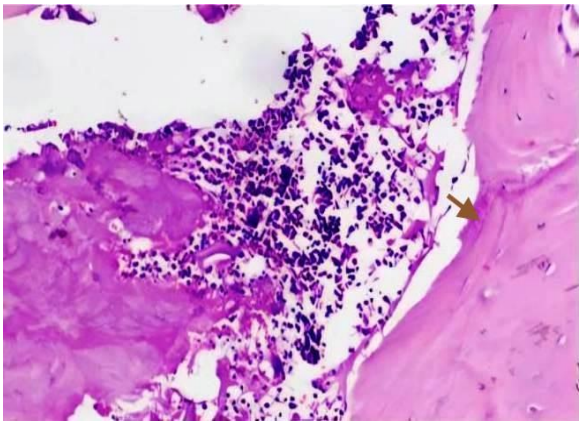

Figure – 1: Histopathological section showing sheets of Langerhans cells and eosinophil-rich fibrocollagenous background. Arrow demarcates bony trabeculae (H&E stain, × 40).

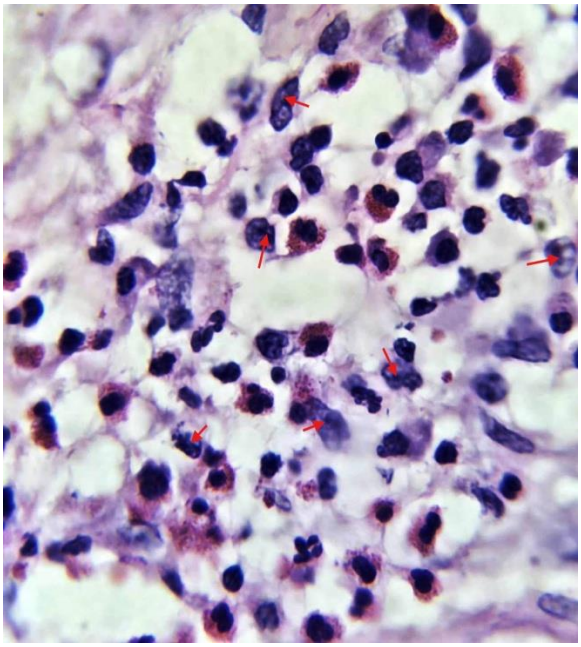

Figure – 2: Higher magnification showing Langerhans cells with irregular nuclear contours and longitudinal grooves (*red arrows*). Background also shows numerous scattered pink cytoplasmic eosinophils (H&E stain, × 1000).

Histopathological examination from all the tissue bits showed fibrocollagenous tissue with dense diffuse infiltration by histiocytes displaying abundant pale to eosinophilic cytoplasm and nuclei with characteristic longitudinal grooves and irregular ("coffee-bean") contours. The background showed a mixed inflammatory infiltrate composed of numerous eosinophils, few lymphocytes, few plasma cells, and sparse polymorphonuclear cells (Figures 1, 2).

Immunohistochemistry demonstrated strong membranous positivity for CD1a and nuclear-cytoplasmic positivity for S100 in the lesional cells, along with CD68 positivity in histiocytes (Figures 3, 4). The Ki-67 proliferation index was approximately 8–10%. These findings confirmed the diagnosis of Langerhans cell histiocytosis.

The postoperative period was uneventful, and the patient was referred for further systemic evaluation and management.

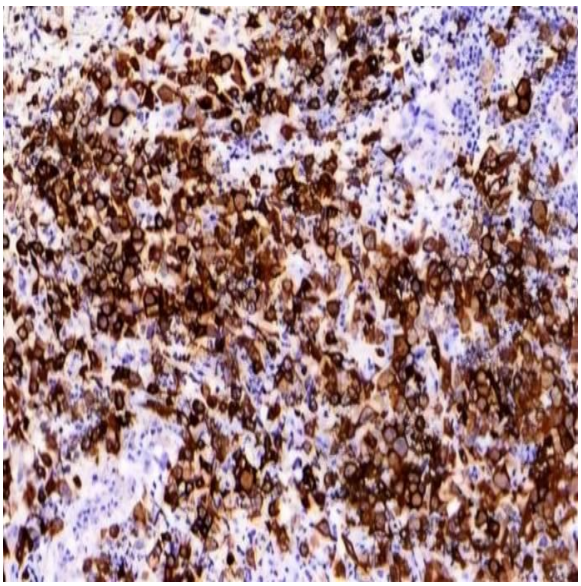

Figure – 3: Immunohistochemistry showing diffuse membranous CD1a positivity in tumor cells (×400).

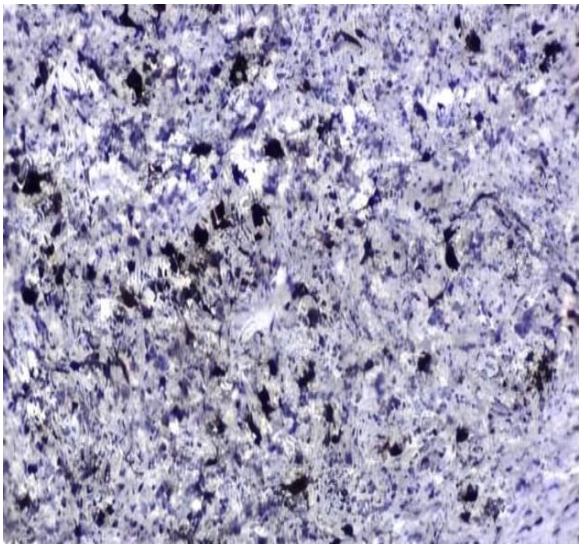

Figure - 4: Immunohistochemistry showing nuclear and cytoplasmic S100 positivity in lesional cells (×100).

DISCUSSION

LCH is currently regarded as a clonal neoplastic disorder of myeloid dendritic cell origin, supported by recurrent mutations involving the MAPK pathway, particularly BRAF V600E.[1,5] This has significantly altered the understanding of its pathogenesis and therapeutic approach.

Bone involvement is the most common manifestation of LCH; however, spinal lesions are relatively uncommon and often present atypically in adults.[3] Adult spinal LCH may manifest as infiltrative marrow lesions with epidural and paraspinal extension, closely mimicking malignant conditions such as lymphoma or metastasis, as observed in the present case.[4]

Radiological modalities such as MRI are crucial for assessing disease extent and neural involvement but lack specificity in differentiating LCH from other neoplastic or infective conditions.[4] Hence, histopathological examination remains the gold standard for diagnosis. The presence of Langerhans cells with characteristic nuclear grooves and an eosinophil-rich inflammatory background is diagnostic.[5]

Immunohistochemistry plays a pivotal role, with CD1a being highly specific and S100 serving as a supportive marker.[5] Additional markers such as langerin (CD207) may further confirm diagnosis where available.

The differential diagnosis includes lymphoma, metastatic disease, granulomatous infections, and other histiocytic disorders. Accurate distinction is essential due to differences in management and prognosis.

Management strategies are tailored based on disease extent. Isolated spinal lesions may be managed surgically, especially in the presence of cord compression, while multisystem disease requires systemic therapy.[6]

Recent advances have further transformed the management landscape of LCH. Molecular studies have confirmed that the majority of cases harbor activating mutations in the MAPK pathway, providing a rationale for targeted therapies.[7] Targeted agents such as BRAF inhibitors (e.g., vemurafenib, dabrafenib) and MEK inhibitors have demonstrated significant clinical responses , particularly in refractory or multisystem disease.[8,9] However, relapse following discontinuation remains a challenge, indicating the need for long-term therapeutic strategies.[7]

Recent outcome studies in adults have shown improved survival with modern treatment approaches, although progression-free survival remains variable depending on disease extent and risk organ involvement.[10] Additionally, ongoing research

continues to refine risk stratification and optimize individualized treatment protocols, including the role of targeted therapy and hematopoietic stem cell transplantation in advanced disease.[11,12]

This case highlights the evolving understanding of LCH as a neoplastic disorder and underscores the importance of integrating histopathology, immunohistochemistry, and emerging molecular insights for accurate diagnosis and management.

CONCLUSION

Langerhans cell histiocytosis (LCH) should be considered in the differential diagnosis of infiltrative spinal lesions in young adults, especially when radiology suggests malignancy. Histopathology with immunohistochemistry (IHC) remains the cornerstone of diagnosis. Early surgical intervention in cases of cord compression and timely multidisciplinary management are crucial for favorable outcomes.

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