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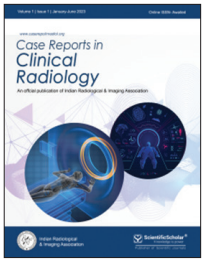

*Case Report*

# A rare cause of solitary rib lytic lesion in adult – A case report

K. Nadanasadharam[1], S. Shanmuga Jayanthan[1], G. Rupesh[1], S. Shanmuga Hariharan[2]

Departments of [1]Radiology and [2]Orthopedics, Meenakshi Hospital, Thanjavur, Tamil Nadu, India.

***Corresponding author:**
K. Nadanasadharam,
Department of Radiology,
Meenakshi Hospital, Thanjavur,
Tamil Nadu, India.

nadanasadharam912@gmail.com

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## ABSTRACT

Langerhans cell histiocytosis (LCH) is characterized by the proliferation of Langerhans cells, which can occur in single or multiple organ systems. The most common sites of involvement include skeletal system, central nervous system, lungs, skin, and lymph nodes. LCH involving the rib is one of the rarest site among adults. Herein, we report a case of a 40-year-old man with solitary LCH in posterior aspect of 4th rib.

**Keywords:** Langerhans cell histiocytosis, Solitary osteolytic rib lesion, Adult

## INTRODUCTION

Langerhans cell histiocytosis (LCH), previously known as histiocytosis X, is a rare neoplastic disorder. It is characterized by abnormal proliferation of Langerhans cells, which are antigen presenting cells. It can involve single or multiple organ systems. Although, it is relatively more common in the pediatric age group, it can also occur in adults. In adults, skeletal involvement is common whereas isolated rib involvement is rare. Only a very few case reports on adult LCH with rib involvement is described in the literature.[1] Herein, we report a rare case of LCH with solitary rib erosion in 40-year-old adult patient.

## CLINICAL HISTORY

A 40-year-old male presented with complaints of pain in the right paraspinal region at the level of 4th/5th intercostal space for the past 4 months. No history of trauma was present. No significant medical or surgical history was present. On local examination, tenderness was present at the level of 4th/5th intercostal space. There were no complaints of fever, chills, and shortness of breath or paresthesias. The patient had normal vitals. The laboratory blood tests were within normal range.

Chest X-ray PA view showed a solitary lytic lesion in the posterior aspect of the right 4th rib [Figure 1]. Non-contrast computed tomography (CT) and contrast-enhanced CT of the chest showed a solitary osteolytic lesion with minimal soft-tissue component in the posterior aspect of the right 4th rib [Figure 2]. There was no abnormal finding on the lung, mediastinum, and visualized portion of the abdomen. There was no other bone involvement. Basic laboratory tests were within normal limits.

The differential diagnosis, in this case, included fibrous dysplasia, aneurysmal bone cyst, solitary metastases, plasmacytoma, and LCH. Ground glass matrix, which is characteristic

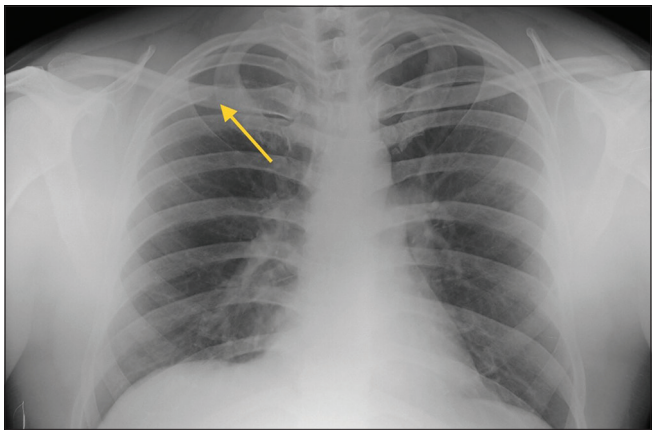

**Figure 1:** Chest X-ray PA view shows a solitary lytic lesion in the posterior aspect of the right 4th rib.

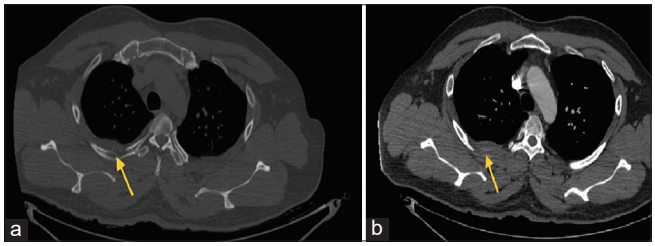


**Figure 2:** (a) Non-contract CT-chest shows a solitary osteolytic lesion in the posterior aspect of the right 4th rib. (b) Contrast-enhanced CT-chest shows minimal enhancing soft-tissue component adjacent to the osteolytic lesion.

of fibrous dysplasia, the most common lytic lesion of the rib, was absent in our case. Infection was ruled out, as there was no clinical history of leukocytosis or fever and with the absence of inflammatory periosteal reaction on CT imaging. The aggressive appearance of the lytic lesion with a narrow zone of transition warranted the exclusion of malignancy (such as LCH, solitary metastasis, and plasmacytoma).

As there was only minimal soft-tissue component, rib resection was preferred over imaging-guided biopsy for better tissue yield and diagnosis. Hence, for confirmation of the diagnosis and to relieve the persistent pain which was not responding to analgesics, partial resection of the 4th and 5th rib was performed. The histologic findings showed the proliferation of histiocytes and eosinophilic infiltration [Figure 3]. The immunohistochemical stain was positive for S100 and CD1a, consistent with the diagnosis of LCH [Figure 3]. PET-CT was performed following the diagnosis of LCH, to rule out multi-systemic involvement, and solitary involvement was confirmed in PET-CT. The patient was referred to a higher center for further management.

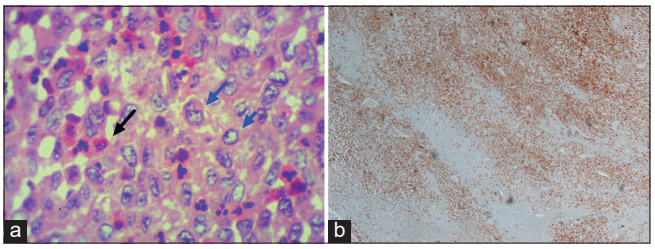


**Figure 3:** (a) H and E: Langerhans cell (Blue arrow) is characterized by ellipsoid shaped nuclei with a central groove ("coffee bean"), irregular grooved nuclear membrane, vesicular chromatin, and prominent nucleoli which are surrounded by eosinophils (Black arrow) (b) Langerhans cell histiocytosis, IHC: CD1a stain positive.

## DISCUSSION

LCH is a reactive and neoplastic proliferation of histiocytes – which are derived from dendritic cells – that exhibit antigen-presenting function (CD1a, S100, and CD207).[2,3] The electron microscopy may reveal Birbeck granules, which are characteristic.[2] The etiopathogenesis of this disease remains unknown, although both neoplastic and immune mediated hypothesis of monoclonal proliferation of cells is postulated. LCH is a very rare disease with a reported incidence of 0.2–2.0 cases per 100,000 children under 15 years old.[4]

Although LCH can involve any organ, osseous involvement is the most common site of involvement in adults. The monostotic (single site) form of involvement is more common than the polyostotic (multiple sites) type. LCH affects mostly the axial skeleton, with more than 50% of cases occurring in the skull.[5,6] The other sites in the descending order of involvement include: Long bones (17%), vertebrae (13%), pelvis (13%), and ribs (6%).[7]

Long bone involvement is more common in children, whereas, in adults flat-bones, such as rib or mandible, are more commonly affected.[6] In a study conducted by Islinger *et al.*,[8] among 211 adults and 330 pediatric cases of LCH, it was noticed that skull lesions were more common in children and ribs were more commonly affected in adults.

Although plain radiography is usually the first step in the evaluation of the disease process, other imaging modalities such as CT, MRI, and bone scintigraphy better characterize bone lesions with higher sensitivity and specificity. Although MRI is more useful in the assessment of bone marrow and soft tissues, only a few studies have reported MRI findings of LCH in the skull, axial skeleton, and long bones in the literature.[9] In addition, MRI examination of the thoracic region is not exempted from physiological movements and susceptible artifacts, and hence, CT is considered to be the imaging modality of choice in the evaluation of a rib lesion.

The imaging features of LCH can vary depending on the site of involvement and phase of the disease. Bone lesions usually present as single or multiple round or oval-shaped lytic lesions, with well-or ill-defined margins. Rib lesions are usually expansile and lytic which appear aggressive and are often associated with enhancing soft-tissue components. There may also be a pathological fracture at the corresponding site. On MRI, LCH usually shows intramedullary increased T2 signal and decreased T1 signal with often associated edema in the surrounding soft tissue. Cortical destruction is often the most common radiological finding of the involved bone in adults (53% of cases).[9] Skull or vertebral body lesions may show punch-out defects or vertebral plana.

The differential diagnosis for aggressive lytic bone lesions includes metastasis or multiple myeloma and plasmacytoma in adults. The solitary lytic lesion in metastasis and multiple myeloma is rare with a high propensity for spine. Other differentials of lytic lesions in the rib include fibrous dysplasia and aneurysmal bone cyst.

Treatment is mainly focused on the most evidently affected organ without proper assessment of other systems, which can lead to under-diagnosis or incomplete staging. Recently, Girschikofsky *et al.*[10] and the histiocytosis association have developed a protocol for managing adult LCH with osseous involvement. For solitary lesions in bone, local treatment such as curettage or intralesional steroid injection would suffice whereas, for multifocal lesions in a single or multi-system involvement (i.e., two or more organ systems), systemic therapy should be considered. There is no standard first-line systemic therapy for LCH. Systemic therapy includes intralesional steroids, which may fasten the healing, radiotherapy, and chemotherapy. Intensive combination chemotherapy is indicated for an aggressive form of LCH. Drugs such as vinblastine, etoposide, cytarabine, and methotrexate are used as monotherapy or in combination for multi-systemic LCH. In cases of multifocal bone involvement, additionally, zoledronic acid is used.[10] The outcomes of LCH depend on its presentation. Those with the disseminated disease will have a mean lifetime of 1–2 years. Those with solitary involvement do have a better prognosis, but their quality of life is reduced.[7]

## CONCLUSION

Although LCH with isolated rib involvement is very rare in the adult population, it should be considered in the differential diagnosis, due to its good prognosis with the early detection and appropriate treatment.

## TEACHING POINTS

1. Isolated LCH of rib though rare in adults, it should be kept in differentials among other lytic lesions of ribs
2. Whole body PET-CT should be done as a routine screening to rule out multi-system involvement.

## DIFFERENTIAL DIAGNOSIS

| Differential diagnosis | Differentiating features |
|---|---|
| 1. Fibrous dysplasia | • Expansile lytic lesion<br>• Characteristic ground glass matrix |
| 2. Plasmacytoma | • Well-defined, "punched-out" lytic lesions with associated extraosseous soft-tissue masses.<br>• Occurs between 40 and 80 years of age. |
| 3. Lytic metastases | • Usually multiple lesions and spine is the most common location.<br>• Cortical destruction and periosteal reactions are characteristic. |
| 4. Aneurysmal bone cyst | • Expansile, eccentric lytic lesion<br>• On CT and MRI characteristic fluid-fluid levels are present. |
| 5. Osteomyelitis | • Endosteal scalloping, cortical erosion, or destruction occurs.<br>• In chronic or untreated cases, sequestrum, involucrum, and/or cloaca may be seen. |

**MCQs:**

1. All are markers of LCH except
   a. S-100
   b. CD 207
   c. HLA B-27
   d. CD 1a

Answer Key: c

2. DD for solitary lytic lesion includes all except
   a. LCH
   b. Metastasis
   c. Osteoid osteoma
   d. Plasmacytoma

Answer Key: c

3. Birbeck granules are characteristic of
   a. Ewing's sarcoma
   b. Metastasis
   c. Osteomyelitis
   d. LCH

Answer Key: d

### Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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