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Langerhans Cell Histiocytosis of the Rib of an Adult Female Patient: a Case Report

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Langerhans cell histiocytosis (LCH) is generally considered a childhood disease that exhibits various nonspecific clinical and radiological manifestations that mimic infection or malignancy. Here, we present a case of LCH involving the rib in an adult patient. CT and MRI revealed an expansile lytic lesion with periosteal reaction on the left 8th rib, suggesting a malignant bone tumor. Surgical resection was performed and histopathological examination was consistent with LCH. Owing to its rare occurrence in adults and nonspecific aggressive features, LCH should be included in the differential diagnosis of an aggressive-appearing rib lesion in both adults and children.

Keywords: Adult; Computed tomography (CT); Langerhans cell histiocytosis (LCH); Magnetic resonance imaging (MRI); Rib

Case Report

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INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by the proliferation of Langerhans cells in one or multiple organ systems. LCH typically affects the pediatric and adolescent patient population but can also occur in adults. Skeletal involvement is most frequently encountered in adults with LCH, but rib involvement is not common. Various other conditions can involve the ribs; however, primary bone tumors of the rib are rare. Usually, common lesions located on the ribs are metastases from a primary malignancy elsewhere in the body, and there have been a few case reports of computed tomography (CT) findings of LCH in adults involving the rib (1). To the best of our knowledge, no studies to date have described the magnetic resonance imaging (MRI) features of these rib lesions, and here, we report a case of LCH of the rib in an adult patient.

CASE REPORT

A 40-year-old female patient presented with recent and progressive chest pain on the left side. She was referred to our hospital for analyzing an abnormal finding on a CT scan performed at the outside hospital. She had no previous trauma or known malignancy. Physical examination revealed no abnormalities except tenderness in the

left chest area. Laboratory data were within normal limits.

The non-contrast axial chest CT revealed a solitary lesion on the left 8th rib. It measured 1.6 × 0.6 × 1.7 cm and was a well-defined expansile osteolytic lesion with cortical thinning and disruption, suggestive of a pathological fracture (Fig. 1a). No chondroid or osteoid matrix was found in the lesion. Additionally, chest MRI using intravenous contrast enhancement was conducted for further evaluation. On axial T2-weighted MR image, a high signal intensity lesion on the left 8th rib with a bulging appearance and a partially ill-defined margin were noted, corresponding to the osteolytic lesion seen in prior CT images. The lesion showed low signal intensity on the T1-weighted image, and intense enhancement on the fat-saturated post-contrast T1-weighted image with pronounced perilesional bone marrow edema and extensive periosteal edema (Fig. 1b-d). The diffusion restriction was not clearly visible on the diffusion-weighted image or the apparent diffusion coefficient map (Fig. 1e, f). Because of the aggressive radiological appearance of the lesion, a malignant bone tumor, such as a bone metastasis, was initially considered. Accordingly, the patient underwent surgical resection of the rib mass.

Histopathological examination of the resected mass revealed a proliferation of Langerhans cells with prominent nuclear indentations and grooves, accompanied by eosinophil infiltration. Immunohistochemical staining was positive for CD1a and S-100 (Fig. 2). A diagnosis of Langerhans cell histiocytosis (LCH) was confirmed. The patient was transferred to another tertiary hospital, where

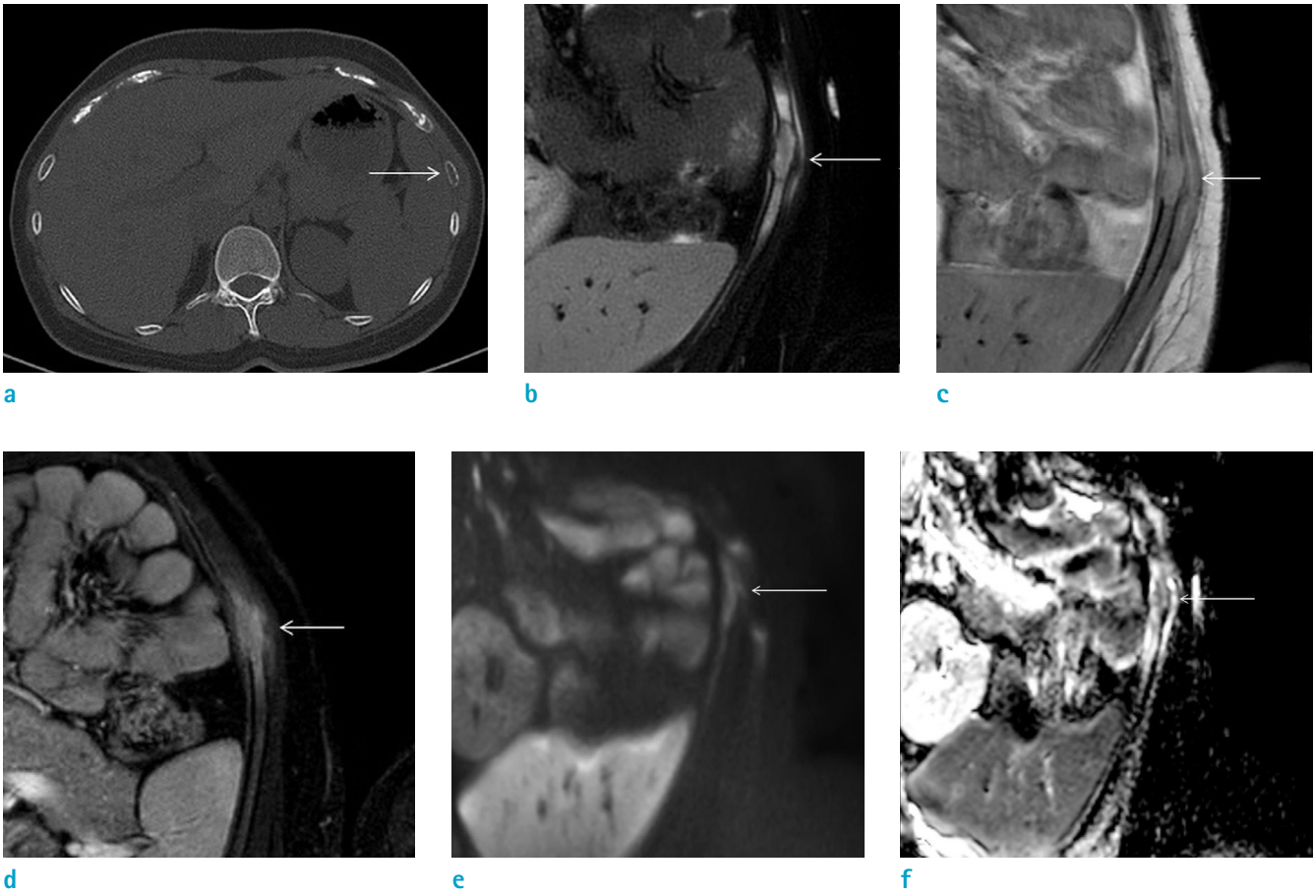
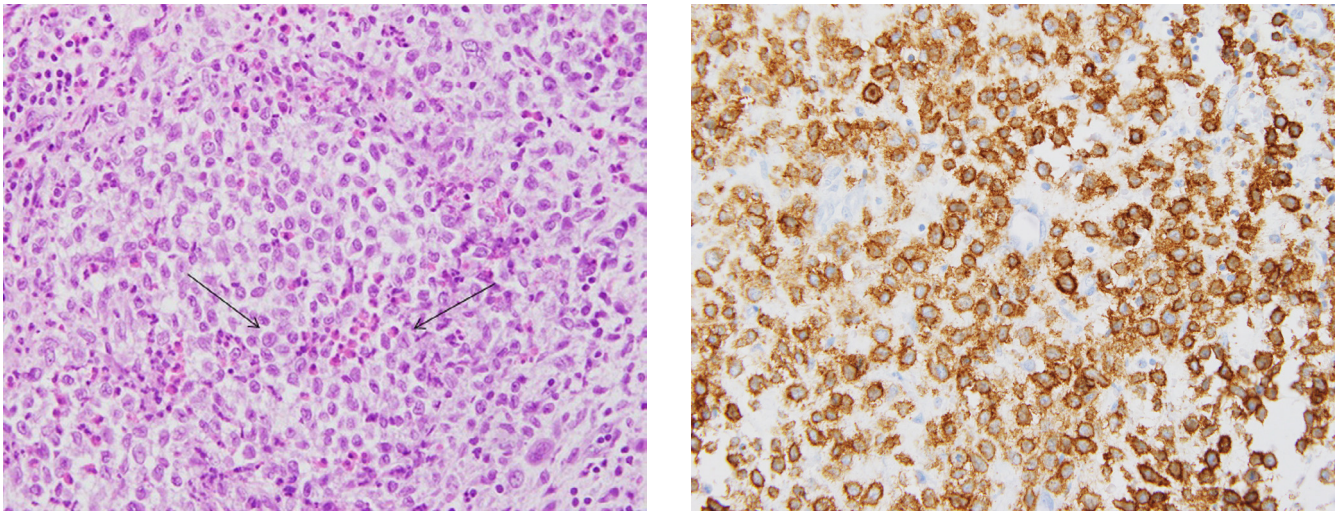


Fig. 1. Forty-year-old female patient with LCH on the left 8th rib. (a) Axial CT image of the chest shows an expansile osteolytic lesion (arrow) with cortical thinning and disruption at the left 8th rib. (b) Axial T2-weighted image with fat saturation shows an intramedullary high-signal intensity mass (arrow) with surrounding soft tissue edema. (c) Axial T1-weighted image shows a low-signal intensity mass (arrow). (d) Axial T1-weighted image with fat suppression after gadolinium administration shows infiltrative enhancement (arrow). Diffusion restrictionis not clearly visible on the (e) axial diffusion-weighted image (DWI) and (f) axial apparent diffusion coefficient map (ADC map) (arrows).

PET CT revealed another hypermetabolic lesions (SUVmax = 15.1) in the left distal femur but no extraskeletal involvement (Fig. 3).

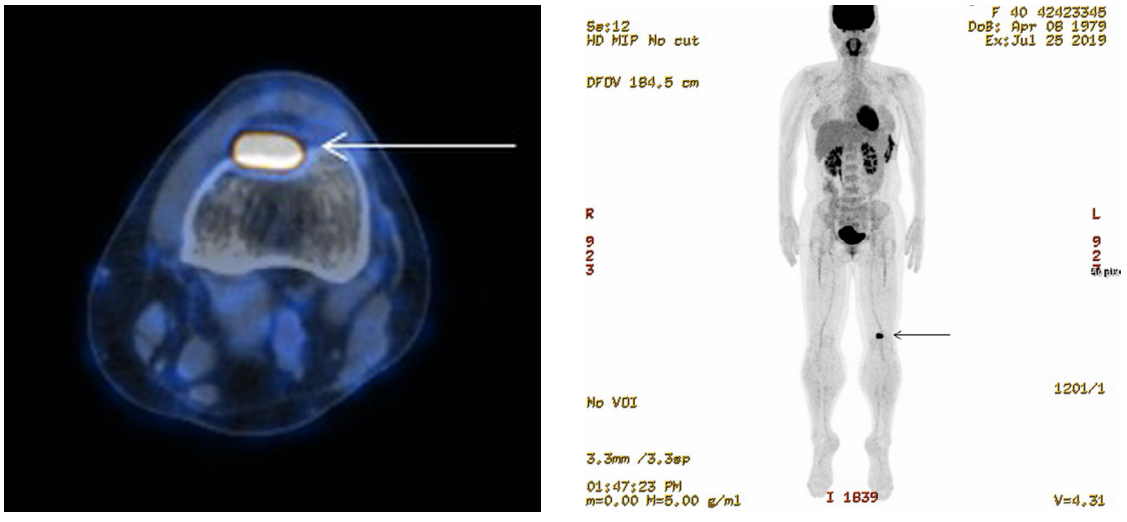
DISCUSSION

Langerhans cell histiocytosis (LCH) is an abnormal accumulation of histiocytes derived from dendritic cells that exhibit the same antigens (i.e., CD1a, S100, and CD207) (2, 3). The etiology and pathogenesis of this disease remain unclear. LCH occurs predominantly in children and can also occur in adults, with an annual incidence of 3-5 cases per million in children and 1-2 cases per million in adults (4). Although LCH can affect any organ, bone involvement is the most common among adults, with monostotic (single site) involvement more common than polyostotic (multiple sites) involvement. LCH has a predilection for the axial skeleton,



a b

Fig. 2. Histopathological examination of the rib mass. (a) Hematoxylin and Eosin (× 400) staining reveals Langerhans cells with ovoid nuclei and occasional nuclear grooves in a mixed inflammatory background with prominent eosinophilia (arrows). (b) Immunohistochemical staining (× 400) is positive for CD1a.



a b

Fig. 3. (a, b) PET CT demonstrates focal hypermetabolic bone lesion (SUVmax = 15.1) in the left distal femur (arrows).

with more than 50% of cases occurring in the skull (5, 6). Long tubular bones (17%), vertebrae (13%), pelvis (13%), and ribs (6%) are less frequently affected (7). Long bone involvement is more common in children, while isolated flat bone involvement, such as of the rib or mandible, is more frequently encountered in adults (3). A study by Islinger et al. (8) reviewed 211 adult cases and 330 pediatric cases of LCH and found that skull lesions were present in 28% of the cases, rib lesions in 25%, pelvic lesions in 8%, and spine lesions in 3%. Rib involvement accounted for 25% of the lesions in adults compared to 8% of the lesions in the children.

Although radiography is usually the first step for evaluating bone tumors and tumor-like lesions, bony abnormalities can also be identified using other imaging modalities such as CT, MRI, and bone scintigraphy. Among these, MRI is considered the best technique for detecting and characterizing bone lesions because bone marrow and soft tissues are shown intricately. A few studies have reported MRI findings of LCH in the skull, axial skeleton, and long bones (9). However, to the best of our knowledge, there have been no reports focusing on the MRI features of rib lesions. CT is helpful for further evaluation of a rib lesion. MRI examination of the thoracic region is not routine, and poses a challenge for radiologists due to physiological movements and susceptible artifacts between the air and tissues. In recent years, the rapid development of MRI techniques such as motion correction technique, very short echo times, breath-hold imaging, and projection reconstruction technique have improved its effectiveness for evaluating rib lesions. The MRI protocol used in this study comprised advanced techniques for motion correction and fast imaging (e.g., MultiVane, fat-suppressed three-dimensional gradient echo).

The imaging features of osseous LCH vary considerably depending on the site of involvement and phase of the disease. Bone lesions usually appear as a single or multiple round or oval-shaped lytic lesions. They may exhibit either well- or ill-defined margins. Rib lesions may be expansile or aggressive and are sometimes accompanied by soft tissue mass. In addition, they may be associated with a pathological fracture. LCH usuallyshows intramedullary lesions with increased T2 signal intensity, decreased T1 signal intensity, and extensive edema in the surrounding soft tissue on MRI, which is concordant with the MRI findings in our case. The most common radiological finding in adults (53% of cases) is cortical destruction (9). Skull or vertebral body lesions may have specific appearances such as punch-out defects or vertebral plana. The differential diagnosis of aggressive bone lesions should include Ewing's sarcoma in children and metastasis or multiple myeloma in adults. Infection should be considered in all patients.

Until recently, universally accepted guidelines for the management of adult patients with LCH were not available. Treatment often focused on the most evidently affected organ without sufficient assessment of other systems, which often led to underdiagnosis or incomplete staging. Recently, Girschikofsky et al. (10) and the Histiocytosis Association have developed a set of recommendations for managing adult LCH patients with bone involvement. For LCH patients with unifocal lesions in a single system, local treatment such as biopsy, curettage, or intralesional steroid injection may be more beneficial than complete excision of the affected bone. For patients with multifocal lesions in a single system or multi-system involvement (i.e., two or more organ systems), systemic therapy should be considered.

In conclusion, although LCH with rib involvement is rare in the adult population and the clinical and radiological findings of LCH overlap with other pathological conditions, LCH should be considered in an adult patient who presents with a rib lesion with aggressive features as evaluated through imaging studies.

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