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Langerhans Cell Histiocytosis of the Rib in an Adult: A Case Report

Sung Hyun Kim Moon Young Choi

Department of Internal Medicine, Busan Paik Hospital, Inje University College of Medicine, Busan, South Korea

Key Words
Langerhans cell histiocytosis · Rib · Adult

Abstract
Single-site, single-system Langerhans cell histiocytosis (LCH) of the rib is one of the rarest causes of bone tumor in adults. Herein, we report a case of a healthy 35-year-old male who presented with upper back pain that was attributed to a solitary osteolytic lesion at the posterolateral aspect of his sixth rib. For diagnostic confirmation and treatment, partial resection of the sixth rib was performed and pathologic finding was consistent with LCH. At the final follow-up after 2 years, no local recurrence or metastasis was observed.

Introduction

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by proliferation and infiltration of histiocytes at various organs, causing local or systemic effects. The most common sites of involvement include the bone, lung, central nervous system, liver, thymus, skin, and lymph node [1]. Although it is often manifested as a solitary bone lesion in the pediatric age group, single-site, single-system LCH of the bone, especially the rib, is one of the rarest causes of bone tumor in adults. We report a case of a healthy 35-year-old male who presented with upper back pain that was attributed to a single osteolytic lesion at the posterolateral aspect of his sixth rib, consistent with single-site, single-system LCH of the rib.

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Moon Young Choi
Division of Hematology-Oncology, Department of Internal Medicine
Busan Paik Hospital, Inje University College of Medicine
75, Bokji-ro, Busanjin-gu, Busan 614-735 (Korea)
E-Mail medmoon@hanmail.net

Case Presentation

A 35-year-old male was hospitalized because of left-sided upper back pain radiating to the rib cage. The pain had begun 3 weeks earlier and had been progressively getting worse. For the previous 2 days, the pain was exacerbated by deep inspiration and movement and was unresponsive to over-the-counter analgesics. The patient denied any history of trauma. The patient did not have any significant medical history. He denied any personal or family history of cancer. He smoked one pack of cigarette per day for the last 15 years. A review of systems was negative for fever, chills, shortness of breath, weight loss, abdominal pain, extremity weakness or paresthesias. Physical examination revealed a temperature of 36.5°C, blood pressure of 120/80 mm Hg, heart rate of 70 beats per minute, and respiratory rate of 18 breaths per minute. An examination of the thorax showed focal tenderness on the posterolateral arc of the left sixth rib. No lymphadenopathy or hepatosplenomegaly was noted. The remainder of the examination was unremarkable. Complete blood count and other laboratory tests were within normal range. Radiography revealed a focal osteolytic lesion in the posterior arc of the left sixth rib (fig. 1a). Computed tomography (CT) of the chest showed an osteolytic lesion of about 1.6 × 1.1 cm in size with perilesional sclerotic change and inner cortical destruction at the posterior arc of the left sixth rib (fig. 1b). There was no abnormal finding on both lung and mediastinum. A skeletal survey was negative for any other bone involvement. Given this finding, our differential diagnosis included malignant or benign lesions originating from the bone and metastatic disease from unknown origin. An infectious disease was ruled out because the patient lacked any evidence of infection such as local inflammation, fever or leukocytosis. For the further differential diagnosis, positron emission tomography (PET) was conducted. The maximum standardized uptake value (SUVmax) of the lesion at the left sixth rib was 7.1, suggesting malignancy (fig. 1c, d). There were no other abnormal hypermetabolic lesions on PET. For diagnostic confirmation and treatment, partial resection of the sixth rib was performed. The histologic findings revealed proliferation of histiocytes and eosinophil infiltration. No malignant cells were detected in the lesion. Immunohistochemical stains were positive for S100 and CD1a (fig. 2), compatible with a diagnosis of LCH. Thereafter, during the follow-up for 2 years, the patient had no local recurrence or distant metastasis.

Discussion

LCH is pathologically characterized by proliferation of myeloid dendritic cells reminiscent, as far as morphology and surface markers are concerned, of skin Langerhans cells. The etiology and pathogenesis for the expansion of the cells responsible for LCH are still under investigation, and many theories have been proposed so far [2]. LCH can affect patient of any age, but is most common in children from 1 to 3 years old. The incidence appears to be 3–5 cases per million children, and 1–2 cases per million adults [3].

LCH may affect a single or a multitude of different organs. Most commonly it involves the bone, lung, central nervous system, liver, thymus, skin, and lymph node. The extent and severity of the disease may vary widely, and it could range from benign and self-limiting to lethal [1].

LCH has been stratified by the Histiocyte Society expert panel to single-system and multisystem LCH. Single-system LCH can be further subdivided into single site (such as monostotic disease or single lymph node) or multiple site (such as polyostotic disease or multiple lymph nodes). Multisystem LCH outlines the involvement of two or more organ systems with

or without organ dysfunction and can further be divided into low-risk and high-risk according to involvement of high-risk organs such as liver, spleen, lung and hematopoietic system that predisposes to higher mortality [4].

Single-system LCH of the bone, formerly known as eosinophilic granuloma, is a benign form of LCH that is localized to one or multiple bone structures. The most common sites differ depending on the patient's age. In children, the most frequent sites of involvement are the skull (40%), femur, rib, vertebra, and humerus [5]. In contrast, the primary sites of bone involvement in adults in one study were jaw (30%), skull (21%), vertebra (13%), pelvis (13%), extremity (17%), and rib (6%) [3].

Single-site, single-system LCH of the rib in adults can be difficult to diagnose since it is an uncommon disease. The differential diagnosis of osteolytic lesion of the rib includes metastasis form carcinoma, multiple myeloma, lymphoma, primary bone malignancy as well as osteomyelitis. Radiologic examination alone is difficult to differentiate these lesions, and a biopsy of suspicious osteolytic bone lesion is needed to confirm the diagnosis. LCH must be confirmed either by positive immunohistochemical staining for CD1a and CD207 or by the identification of Birbeck granules by electron microscopy [1].

Given the rarity of the condition and the relatively good prognosis, treatment of single-system LCH of the bone has not been standardized. Different approaches such as observation only, surgical curettage or excision, intralesional corticosteroid injection, radiotherapy, and chemotherapy have been implemented according to localization and severity of the disease [1, 6–9]. Single-site, single-system LCH of the bone has been treated successfully with curettage with or without bone graft. One study reported a 4-year event-free survival of 90 percent for patients with solitary bone LCH [10], and another study reported only 2 recurrences among 46 cases with solitary bone LCH [11]. Because solitary small lesions tend to be self-limited, with spontaneous regression noted in multiple studies, aggressive surgical excision resulting in skeletal deformities is not recommended [12]. In our case, partial resection of the involved rib was performed for the diagnosis and treatment because severe pain persisted for weeks and did not respond to analgesics. Thereafter, during the follow-up for 2 years, the patient had no local recurrence or distant metastasis.

In conclusion, although single-site, single-system LCH of the rib is one of the rarest causes of bone tumor in adults, it can be treated successfully with surgical intervention such as curettage or partial resection. Therefore, it should be considered in the differential diagnosis of solitary osteolytic lesion of the rib.

Statement of Ethics

Informed consent was obtained from the patient for publication of this case report and any accompanying images.

Disclosure Statement

The authors have no conflict of interest to declare.

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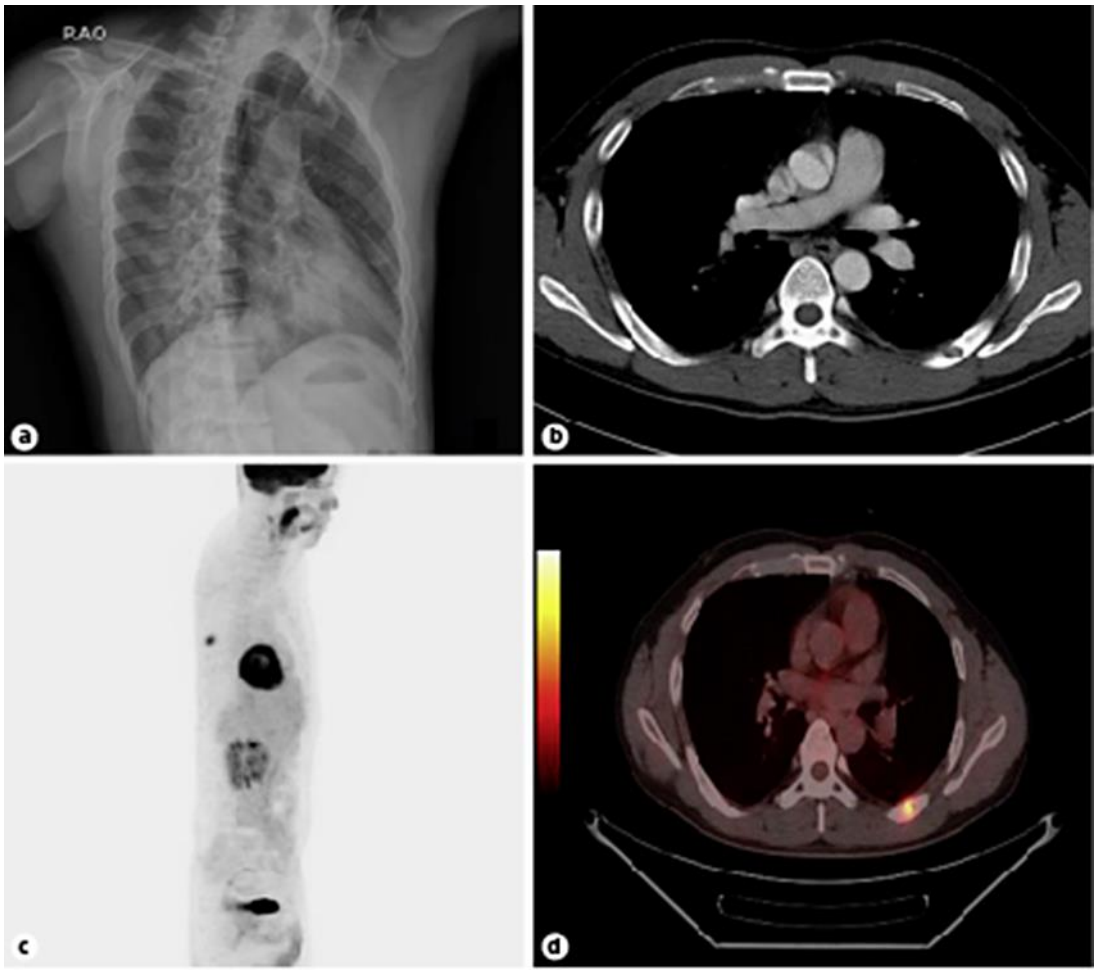

Fig. 1. Simple X-ray (**a**) and chest CT (**b**) showed a focal osteolytic lesion in the posterior arc of the left sixth rib. PET (**c**, **d**) revealed a solitary hypermetabolic lesion (SUVmax 7.1) at that site.

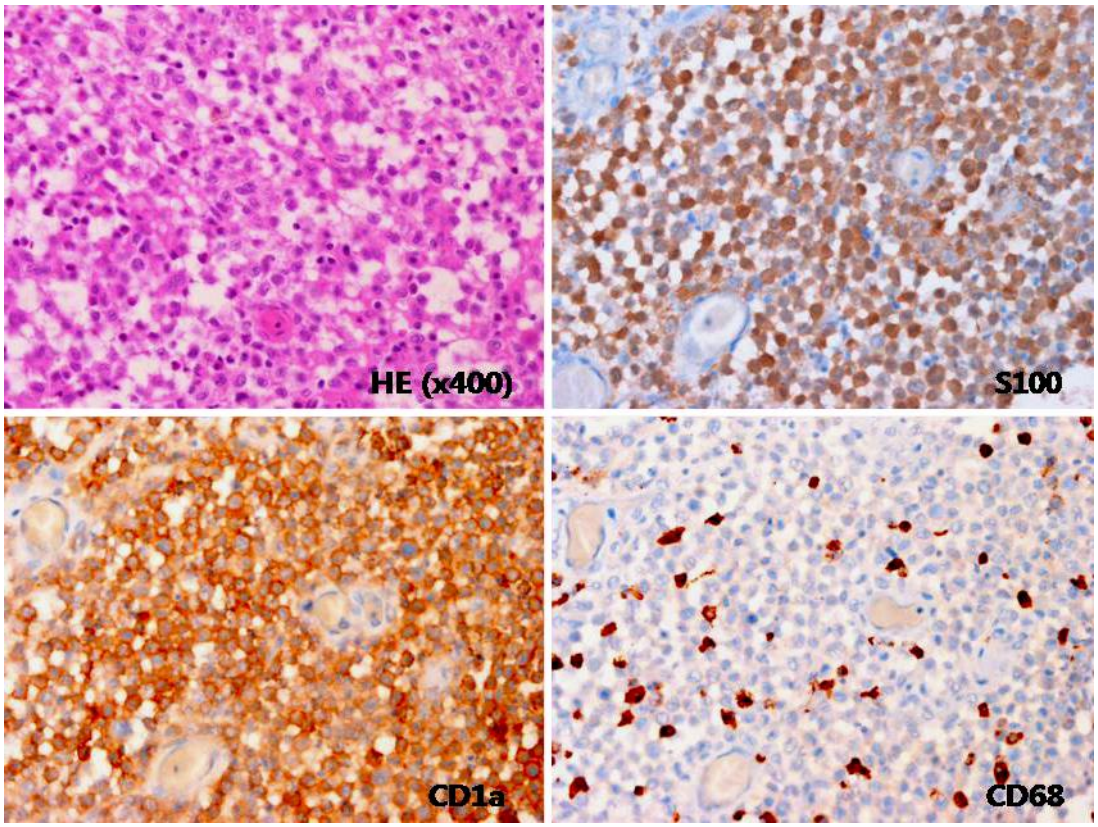

Fig. 2. Histologic findings showed proliferation of histiocytes and eosinophil infiltration. Immunohistochemical stains were positive for S100 and CD1a, and negative for CD68.