Received: 7 July 2023 | Revised: 20 November 2023 | Accepted: 18 December 2023

DOI: 10.1002/ccr3.8391

Clinical Case Reports Open Access WILEY

CASE REPORT

Solitary Langerhans cell histiocytosis of the sternum in a 21-year-old woman

Neda Soleimani[1] | **Massood Hosseinzadeh**[1] | **Armin Amirian**[2] | **Masha Hassani**[1] | **Sahand Mohammadzadeh**[1]

[1]Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran

[2]Thoracic and Vascular Surgery Research Center, Shiraz University of Medical Science, Shiraz, Iran

Correspondence
Sahand Mohammadzadeh, Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran.
Email: mohammads@sums.ac.ir

Key Clinical Message

Children are more likely to develop Langerhans cell histiocytosis (LCH), a rare disorder with an unknown cause. LCH often invades skeletal systems, while it has occasionally been seen in the sternum or ribs. The best course of treatment for single-site, skeletal LCH is yet unknown. We present an instance of sternal LCH with adult onset. By fusing and reconstructing chest computed tomography, it was possible to determine the extent of surrounding soft tissue invasion. Because LCH is so uncommon, it could be challenging to recall when we see a sternal lesion. Adult Patients who arrive with anterior chest discomfort and an osteolytic sternal lesion should include LCH on their differential diagnosis list.

KEYWORDS

bone, Langerhans cell histiocytosis, radiograph, sternum

1 | INTRODUCTION

Infants and children can develop Langerhans cell histiocytosis (LCH), a relatively uncommon illness. Several variables have been proposed as the origin of this illness. LCH is likely a clonal neoplastic illness with extremely varied biologic behavior, according to Willman et al.[1] Because many LCH patients exhibit hypergammaglobulinemia, thymic dysfunction, or reduction in suppressor T lymphocytes in the peripheral blood, Shannon et al. suggested that LCH is Langerhans cell proliferation and accumulation at a focal area by immunoregulatory dysfunctions.[2]

The Histiocytosis Society Writing Group reported LCH to be the preferred word in 1987,[3] replacing Lichtenstein's 1953 term "histiocytosis X."[4] According to their clinical and pathologic characteristics, this condition covers three diseases: Letterer-Siwe syndrome, eosinophilic granuloma (EG), and Hand-Schuller-Christian syndrome.[5] The most frequent form of LCH is EG, which accounts for roughly 70% of patients.[5] Lesions of the sternum are infrequent.[6–15] Using a literature review, we describe one patient with an EG that affected the sternum. This article describes a young woman with sternal LCH and discusses classification, clinical signs and symptoms, pathological results, potential differential diagnoses, and recommended treatments.

2 | CASE PRESENTATION

A 21-year-old woman who had been experiencing increasing, spontaneous chest tightness for 3 months presented to our hospital. During the physical examination, the area over the sternum body and near the manubrium was

quite painful. She did not have any systemic symptoms. The chest computed tomography scan revealed a destructive mass in the right para midline of the sternum measuring about 24×19 mm with mild expansion (Figure 1). Also, near this mass in the retrosternal section, another soft tissue component about 26×15 mm is seen in favor of the enlarged lymph node. Clinical lab data were within normal limits. The patient underwent partial sternotomy with resection of chest wall mass and reconstruction of the upper sternum. The family history was insignificant, and she had no past medical history.

Microscopic analysis (Figure 2) reveals loose clumps of histiocytic-appearing cells against a background of mixed inflammation and localized, obvious eosinophilia. The nuclei of Langerhans cells range from being oval to reniform and have a longitudinal groove. Regarding immunohistochemistry, the Langerhans cells showed diffuse staining strongly positive for S-100 and CD1a (Figure 3).

As a result, LCH was identified in the patient. She is currently attending routine outpatient department follow-up at the first month, 3rdmonth, 6th month, and 1 year after leaving our hospital, and She has shown no signs of a local recurrence.

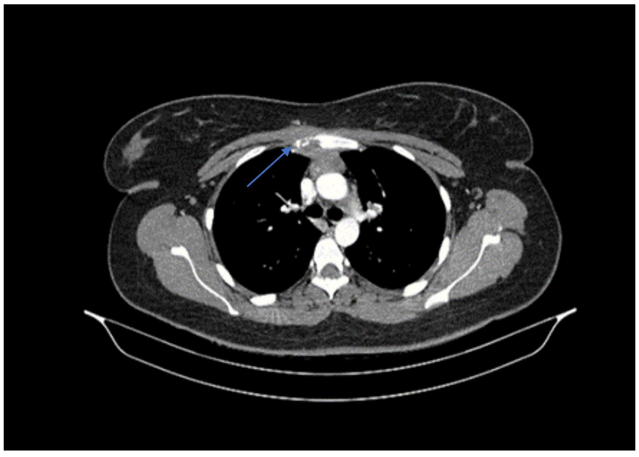

FIGURE 1 An axial CT image of the chest shows an expansile osteolytic lesion (arrow) with cortical thinning and disruption at the right side of the sternum.

3 | DISCUSSION

Depending on the place afflicted, LCH has different clinical signs. The skeleton is the most often affected organ, accounting for >57%–75% of LCHs. As a result, local bone pain is LCH's most prevalent symptom. Additional recorded LCH symptoms included dyspnea, lethargy, and a painful lump on the head, spontaneous pneumothorax, and diabetes insipidus.[7]

The patient's physical examination, medical history, imaging tests, histology, immunohistochemistry, and electron microscopy all play a role in diagnosing LCH. Given the rare nature of LCH of bone and its resemblance to radiographic osteolytic lesions, it can be challenging to distinguish benign lesions from other primary or metastatic bone cancers. Thus, a biopsy is required for these osteolytic lesions. Clinical and pathological evidence in our case led to a definitive diagnosis based on Langerin (CD1a) positivity.[4]

The abnormal growth of tissue macrophages known as Langerhans cells in one or more organs, such as the liver, bone, skin, lung, lymph nodes, spleen, and bone marrow, is known as Langerhans cell histiocytosis. 90% of patients range in age from 5 to 15 years, with slightly more men than women.[9] Less than 1% of bone tumor-like lesions are LCH, and most (79%) are solitary lesions.[8] The skull is the most common place, and the jaw, femur, pelvis, spine, ribs, humerus, scapula, and clavicle are next in decreasing order of incidence.[10] Lesions of the sternum are incredibly uncommon, accounting for fewer than 1% of LCH's bone lesions.[11]

To our knowledge, there are only 10 cases with LCH of the sternum (Table 1).[6–15] Pain, mass, and soreness were the three most frequent symptoms. Without any focal recurrence, all of these patients fully recovered.[12]

In the described cases, the sternum typically presented radiographically as a pure osteolytic lesion without

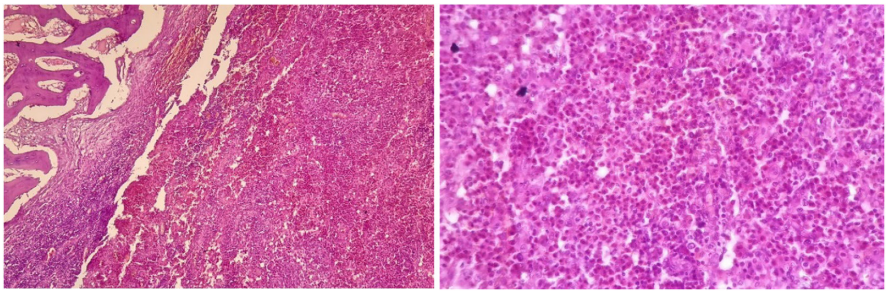

FIGURE 2 A histopathological examination of a sternum tissue biopsy sample. Hematoxylin and eosin (H&E) stains reveal Langerhans cells with ovoid nuclei and occasional nuclear grooves in a mixed inflammatory background with prominent eosinophilia. (Left 100x), (Right 400x).

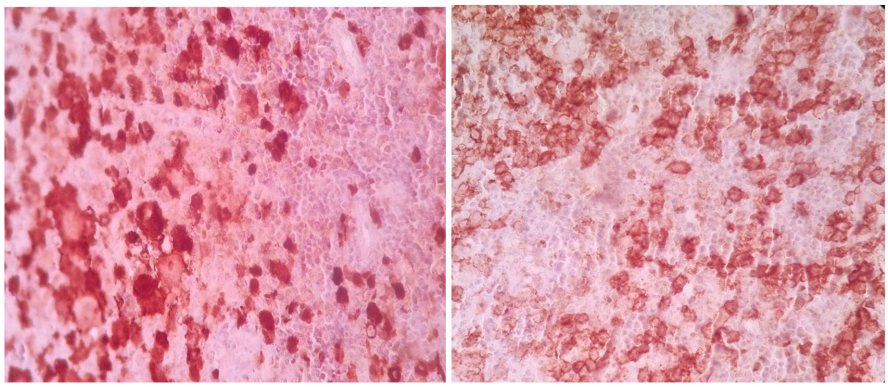

FIGURE 3 (Left) S100-positive and (Right) CD1a-positive immunohistochemical staining of Langerhans cells (×400).

TABLE 1 Literature regarding LCH of the sternum.

Reference	Year	Age (y)	Treatment	Recurrence	Follow-up
Eroglu et al.[6]	2004	30	PS	-	1 year
Mansour et al.[7]	1993	6	PS	-	NS
Chiau et al.[8]	1990	5	Curettage	-	3 year
Peer et al.[9]	1985	30	Radiation	-	NS
Gugliantini et al.[10]	1982	2	Chemotherapy	-	9 month
Taillefer et al.[11]	1981	24	Radiation	-	6 month
Fazio et al.[12]	2005	38	PS	NS	NS
Sai et al.[13]	2005	25	Curettage	-	2 year
Wilson et al.[14]	2005	6	Intralesional steroid injection	-	1 year
Bayram et al.[15]	2008	13	PS, Mesh plate	-	5 year

Abbreviations: NS, not stated; PS, partial sternotomy.

sclerotic borders. CT scans confirmed cortex disruption and the lytic lesion extension from the parasternal to the retrosternal region, better defining the disease's extent than radiography.[6,7,13]

Osteomyelitis and several malignant bone disorders with severe radiological characteristics, such as cortical disruption and soft tissue extension, are included in the differential diagnosis. Multifocality of the bone lesions, age, and a rise in the incidence of eosinophils all pointed to LCH in the current instance. However, it is important to emphasize that only a biopsy's histopathologic results can help diagnose LCH definitively. The histiocytosis arrangement in loose mesh-works or clusters[15] and positivity for CD1a and S-100 antigens can help to diagnose LCH and roll out differential diagnoses like Ewing's sarcoma, plasma cell neoplasm, chronic infections, lymphomas, or other primary bone lesions.

The severity of LCH will determine how it is treated. A single lytic lesion that affects a lengthy bone has been treated in various ways. The treatment of solitary lesions is always successful in the end (100% of isolated, solitary bone lesions), albeit recurrence happens in some cases (11%) using curettage, radiation, local steroid injection, and chemotherapy alone or in combination.[15] In contrast, chemotherapy is frequently used in conjunction with other therapeutic modalities to treat multifocal and multisystem kinds of LCH. Local treatments, like curettage, biopsy, or intralesional steroid injection, showed more prospective advantages than total excision of the afflicted bone for LCH with bone lesions. It is advised to take oral methotrexate, azathioprine, or thalidomide for LCH without the danger of organ involvement. Systemic therapy with etoposide, cytarabine, or vinblastin/prednisolone can be advised for the treatment of LCH.

4 | CONCLUSIONS

Because LCH is so uncommon, it could be challenging to recall when we see a sternal lesion. Adult patients who arrive with anterior chest discomfort and an osteolytic sternal lesion should include LCH on their differential diagnosis list.

AUTHOR CONTRIBUTIONS

Neda Soleimani: Formal analysis; methodology; software; validation; writing – review and editing. **Massood Hosseinzadeh:** Conceptualization; software; validation; visualization. **Armin Amirian:** Investigation; methodology; software; visualization. **Masha Hassani:** Investigation; methodology. **Sahand Mohammadzadeh:** Conceptualization; data curation; methodology; supervision; writing – original draft.

FUNDING INFORMATION

No institutional funding.

CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

The research has been carried out by the World Medical Association Declaration of Helsinki. The study was approved by the Ethics Committee of Shiraz University of medical science.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy on the title page of the manuscript.

ORCID

Neda Soleimani https://orcid.org/0000-0001-5769-8452
Massood Hosseinzadeh https://orcid.org/0000-0003-0531-0457
Armin Amirian https://orcid.org/0000-0002-0780-2175
Sahand Mohammadzadeh https://orcid.org/0000-0001-6974-1323

REFERENCES

1. Willman CL, Busque L, Griffith BB, et al. Langerhans'-cell histiocytosis (histiocytosis X)–a clonal proliferative disease. *N Engl J Med*. 1994;331(3):154-160.
2. Héritier S, Emile JF, Hélias-Rodzewicz Z, Donadieu J. Progress towards molecular-based management of childhood Langerhans cell histiocytosis. *Arch Pediatr*. 2019;26(5):301-307.
3. Fang J, Chen J, Deng C, et al. Langerhans cell histiocytosis in a pediatric patient with simultaneous gastrointestinal and skin involvement: a case report and literature review. *Fetal Pediatr Pathol*. 2023;42(3):483-487.
4. Radzikowska E. Update on pulmonary Langerhans cell histiocytosis. *Front Med*. 2021;8(7):582581.
5. Papo M, Cohen-Aubart F, Trefond L, et al. Systemic histiocytosis (Langerhans cell histiocytosis, Erdheim–Chester disease, Destombes–Rosai–Dorfman disease): from oncogenic mutations to inflammatory disorders. *Curr Oncol Rep*. 2019;21:1.
6. Eroglu A, Kürkçüoglu IC, Karaoglanoglu N. Solitary eosinophilic granuloma of sternum. *Ann Thorac Surg*. 2004;77(1):329-331.
7. Mansour KA, Anderson TM, Hester TR. Sternal resection and reconstruction. *Ann Thorac Surg*. 1993;55(4):838-843.
8. Chiau JH, Chang TK, Hwang B, Tiu CM. Eosinophilic granuloma of sternum. Report of one case. Zhonghua Minguo Xiao er ke yi xue hui za zhi. *Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi*. 1990;31(1):35-39.
9. Peer A, Witz E, Yona E, Manor H, Reif R. Solitary eosinophilic granuloma of sternum: case report with review of the literature. *Br J Radiol*. 1985;58(696):1173-1176.
10. Gugliantini P, Barbuti D, Rosati D, Donfrancesco A, Miano C. Histiocytosis X: solitary localization in the sternum of a 2-year-old child. *Pediatr Radiol*. 1982;12:102-104.
11. Taillefer R, Levasseur A, Robillard R. Ga-67 imaging in eosinophilic granuloma. *Clin Nucl Med*. 1981;6(6):270-271.
12. Fazio N, Spaggiari L, Pelosi G, Presicci F, Preda L. Langerhans' cell histiocytosis. *Lancet*. 2005;365(9459):598.
13. Sai S, Fujii K, Masui F, Kida Y. Solitary eosinophilic granuloma of the sternum. *J Orthop Sci*. 2005;10:108-111.
14. Wilson GJ, Versluys AB, Bax KN. Solitary Langerhans cell histiocytosis of the sternum in a 6-year-old girl: how should it be treated? *J Pediatr Surg*. 2005;40(7):e13-e15.
15. Bayram AS, Köprücüoglu M, Filiz G, Gebitekin C. Case of solitary eosinophilic granuloma of the sternum. *Thorac Cardiovasc Surg*. 2008;56(2):117-118.

How to cite this article: Soleimani N, Hosseinzadeh M, Amirian A, Hassani M, Mohammadzadeh S. Solitary Langerhans cell histiocytosis of the sternum in a 21-year-old woman. *Clin Case Rep*. 2024;12:e8391. doi:10.1002/ccr3.8391