

Imaging Features of a Child with Isolated Scapular Langerhans Cell Histiocytosis Associated with Malignancy: A Case Report

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HUMAN AND ANIMAL RIGHTS

None

ABSTRACT

Background: Langerhans cell histiocytosis (LCH) is a rare clonal proliferative disorder with heterogeneous clinical manifestations. Isolated bone involvement is the most common form, but a solitary lesion in the scapula is unusual and its imaging appearance can mimic a primary malignant bone tumor, posing a diagnostic challenge. This report aims to highlight this rare presentation to improve clinical recognition and management.

Case Report: A 12-year-old boy presented with a two-week history of left shoulder pain. Imaging revealed a solitary, expansile lytic lesion in the scapula with an associated soft tissue mass, initially raising suspicion for Ewing's sarcoma or osteosarcoma. A definitive diagnosis of LCH was confirmed via ultrasound-guided core needle biopsy and immunohistochemical analysis, which showed positivity for CD1a and Langerin (CD207). The patient underwent intralesional curettage and bone grafting and achieved a favorable outcome.

Conclusion: This case illustrates an unusual presentation of LCH as a solitary scapular lesion mimicking a bone sarcoma. It underscores the importance of considering LCH in the differential diagnosis of pediatric osteolytic lesions, even in uncommon locations. Awareness of this presentation can guide appropriate diagnostic biopsy and prevent misdiagnosis and unnecessary aggressive treatment.

Key words: Langerhans Cell Histiocytosis; Child; Scapula; Osteolytic Lesion; Ultrasound-Guided Biopsy; Differential Diagnosis

CASE REPORT

CASE REPORT

A 12-year-old boy presented to our orthopedic outpatient department with a two-week history of left shoulder pain and tenderness without obvious history of trauma. The pain, which worsened with activity, had been initially neglected. Over the preceding week, the pain intensified, particularly at night and during specific movements of the upper limb, interfering with sleep. There were no systemic symptoms such as fever, night sweats, or weight loss. Self-application of a topical analgesic ointment was ineffective. His past medical history and family history were unremarkable, with no history of significant illness or tumors.

On physical examination, there was no redness, swelling, or increased skin temperature around the left shoulder joint. A well-localized tender point and a definite soft tissue mass could be palpated over the body of the left scapula. The mass was of medium firmness and poorly mobile. The active and passive range of motion of the left shoulder joint was largely normal but demonstrated terminal limitation due to pain. The neurovascular examination was unremarkable.

Imaging Findings

Ultrasound examination revealed a $25 \times 30 \times 24$ mm hypoechoic mass in the left shoulder region with an irregular shape, ill-defined boundaries, and a discontinuous posterior cortical line. A small amount of color Doppler flow was seen internally, and the mass was considered highly likely to be malignant.

The plain and contrast-enhanced MRI scan results showed the left scapula and surrounding soft tissues presenting as a mass-like signal. The lesion appeared slightly hypointense on T1-weighted imaging and hyperintense on T2-weighted fat-suppressed imaging. Mild soft tissue edema was visible around the mass, which measured approximately $25 \text{ mm} \times 29 \text{ mm} \times 26 \text{ mm}$. The lesion demonstrated significant enhancement on the post-contrast scan, raising suspicion for a malignant tumor of the scapula.

A whole-body bone scan was performed to rule out additional skeletal lesions, which showed abnormal radiotracer concentration isolated to the left scapula with no other bony abnormalities.

Management and Follow-up

To obtain a definitive diagnosis, the patient underwent an ultrasound-guided biopsy of the mass. The pathological diagnosis was Langerhans cell histiocytosis. Immunohistochemical staining results were as follows: Langerin (+), CD1a (+), CD68 (+), S-100 (+), CD30 (-), and Ki-67 (+, approximately 5%). Following the diagnosis, the patient was discussed at a multidisciplinary team (MDT) meeting and received appropriate treatment, leading to a favorable outcome.

DISCUSSION

Etiology & Demographics

Langerhans cell histiocytosis (LCH) is an uncommon clonal proliferative disorder of dendritic-cell lineage Langerhans cells [1]. It primarily affects children, with an estimated annual incidence of three to five cases per million [2]. The disease exhibits a broad clinical spectrum, ranging from isolated, self-resolving bone lesions to life-threatening multi-system involvement. Skeletal lesions are the most common manifestation, occurring in approximately 80% of cases; however, LCH confined solely to the scapula is unusual [3].

Clinical & Imaging Findings

The diagnosis process of this case highlights the non-specific nature of LCH clinical manifestations. The patient only presented with local pain, which was easily mistaken for muscle strain or shoulder joint inflammation. For persistent bone pain in adolescents, especially when it worsens at night, even without a history of trauma, sufficient vigilance should be exercised.

Despite its typically indolent biological behavior, its imaging appearance can be aggressively osteolytic, often mimicking malignancy and leading to potential diagnostic errors and inappropriate management [4,5]. In this case, the soft tissue masses shown on MRI plain scans and ultrasound eroded the bone and stimulated surrounding tissues, causing peripheral bone marrow edema, which are aggressive imaging manifestations [6-8]. The heterogeneous enhancement of the lesion on enhanced scans and the internal blood flow signals observed by ultrasound suggest the vascular-rich nature of the lesion [9,10]. The contradictory characteristics of "good and bad interweaving" in imaging are typical manifestations of LCH.

Differential Diagnoses

Given the patient's age and lesion location, imaging findings of a solitary lytic lesion need to be differentiated from other entities such as Ewing's sarcoma, osteosarcoma, eosinophilic granuloma (another form of LCH), and Brodie abscess. Isolated scapular Langerhans cell histiocytosis is a localized bone lesion with a favorable prognosis. However, due to its insidious clinical onset and nonspecific imaging features that can mimic malignancy, the diagnosis is often challenging [11]. (See Table 1 for a detailed summary of imaging differential diagnoses).

Treatment & Prognosis

Isolated bone LCH is the mildest form within the LCH spectrum and has an excellent prognosis. Confirmatory diagnosis relies on histopathological examination, specifically positive immunohistochemical staining for CD1a and Langerin (CD207) [6]. The gold standard for diagnosing Langerhans cell histiocytosis relies on immunohistochemical staining, typically showing positivity for both CD1a and Langerin. A Ki-67 proliferation index of approximately 5% suggests relatively low

tumor cell proliferative activity, which aligns with its biological behavior [11]. When imaging findings suggest malignancy but clinical manifestations are atypical, ultrasound-guided core needle biopsy is decisive in obtaining a tissue sample for accurate diagnosis. This case highlights the significant advantages of ultrasound-guided biopsy: it is convenient, minimally invasive, and provides precise targeting, offering critical evidence for final diagnosis. The entire diagnostic and therapeutic process was conducted within a multidisciplinary framework, integrating imaging, pathology, and clinical evaluation to formulate a scientific and rational treatment strategy, thereby avoiding unnecessary overtreatment. (See Table 2 for a summary of LCH characteristics).

TEACHING POINT

Langerhans cell histiocytosis (LCH) should be considered in the differential diagnosis of pediatric osteolytic bone lesions, even in unusual locations like the scapula, as its imaging features can closely mimic a primary malignant bone tumor. When clinical symptoms are atypical, a minimally invasive ultrasound-guided core needle biopsy is crucial for obtaining a definitive histological diagnosis and avoiding overtreatment.

QUESTIONS

Question 1: A 12-year-old boy presents with left shoulder pain and an osteolytic lesion in the scapula on imaging. Which of the following features from this case report would raise the highest suspicion for a malignant bone tumor rather than a benign process?

- The patient's age of 12 years.
- Presence of nocturnal pain.
- MRI showing a mass with heterogeneous enhancement and surrounding soft tissue edema. (applies)
- Absence of systemic symptoms like fever or weight loss.
- The lesion being solitary on bone scan.

Explanation: While LCH can occur in children and cause pain [Case Report], the imaging findings in this case were initially concerning for malignancy. The MRI revealed an aggressive-appearing mass with significant heterogeneous enhancement and surrounding soft tissue edema, which are features that can mimic malignant bone tumors such as Ewing's sarcoma or osteosarcoma [Discussion, Paragraph 2, citing 4,5,7,8]. Nocturnal pain (choice 2) can occur in both LCH and malignancy. The absence of systemic symptoms (choice 4) and a solitary lesion (choice 5) do not rule out malignancy.

Question 2: According to the case report, what was the definitive method used to diagnose Langerhans cell histiocytosis (LCH) and differentiate it from other malignant bone tumors?

- Plain radiography showing a lytic lesion.
- Whole-body bone scan showing isolated uptake.
- Contrast-enhanced MRI characteristics.
- Ultrasound-guided core needle biopsy with immunohistochemical staining. (applies)
- Clinical presentation of shoulder pain and tenderness.

Explanation: The case report states that while imaging

raised suspicion for a malignant tumor, a definitive diagnosis was obtained via ultrasound-guided biopsy of the mass [Case Report, Management and Follow-up]. The confirmatory diagnosis of LCH relies on histopathological examination, specifically positive immunohistochemical staining for CD1a and Langerin [Discussion, Treatment & Prognosis, citing 6]. The other imaging modalities (choices 1, 2, 3) and clinical presentation (choice 5) are useful for raising suspicion but cannot provide a definitive diagnosis.

Question 3: Which of the following immunohistochemical markers are typically positive in a confirmed case of Langerhans cell histiocytosis (LCH), as highlighted in this report?

- CD20 and CD3.
- Cytokeratin and EMA.
- CD1a and Langerin (CD207). (applies)
- Desmin and MyoD1.
- HMB-45 and Melan-A.

Explanation: The case report specifies that the immunohistochemical staining results for the patient were positive for Langerin and CD1a [Case Report, Management and Follow-up]. The discussion reinforces that the gold standard for diagnosing LCH relies on immunohistochemical staining, typically showing positivity for both CD1a and Langerin [Discussion, Treatment & Prognosis, citing 6]. The other options are markers for lymphoid (1), epithelial (2), muscle (4), and melanocytic (5) lineages, respectively.

Question 4: Based on the discussion in this article, what is a key reason why a solitary LCH lesion of the bone can be diagnostically challenging?

- It always presents with systemic symptoms such as fever and night sweats.
- It is exclusively found in the long bones of the lower extremities.
- Its imaging appearance can be aggressively osteolytic and mimic a primary malignant bone tumor. (applies)
- Biopsy is never required for diagnosis, as imaging is pathognomonic.
- It does not cause any pain, leading to delayed presentation.

Explanation: The article's discussion emphasizes that despite LCH's typically indolent biological behavior, its imaging appearance can be aggressively osteolytic, often mimicking malignancy and leading to potential diagnostic errors [Discussion, Clinical & Imaging Findings, citing 4,5]. This is further supported by the case where the MRI findings initially raised suspicion for a malignant tumor [Case Report, Imaging Findings]. The other options are incorrect; LCH can occur in flat bones like the scapula, may or may not have systemic symptoms, requires biopsy for diagnosis, and frequently presents with pain.

Question 5: A 10-year-old patient is diagnosed with a solitary Langerhans cell histiocytosis (LCH) lesion of the scapula after biopsy. According to the principles discussed in this case report, what is the most appropriate next step in management?

- Immediate radical surgical resection of the entire scapula.
- High-dose multi-agent chemotherapy.
- Radiation therapy to the entire hemithorax.

Observation only, as all LCH lesions resolve spontaneously. Multidisciplinary team (MDT) discussion to formulate a treatment plan, considering options like curettage or intralesional steroids. (applies)

Explanation: The case report concludes by emphasizing the importance of a multidisciplinary collaborative mechanism that leverages the expertise of various specialties to improve the overall management of complex diseases. This approach helps in formulating a scientific and rational treatment strategy, thereby avoiding unnecessary overtreatment [Discussion, Treatment & Prognosis, citing 12]. For a solitary bone lesion, treatment options might include local curettage or intralesional steroids, not aggressive resection (1), high-dose chemotherapy (2), or radiation (3). Observation alone (4) is not standard for a symptomatic, biopsy-proven lesion.

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FIGURES

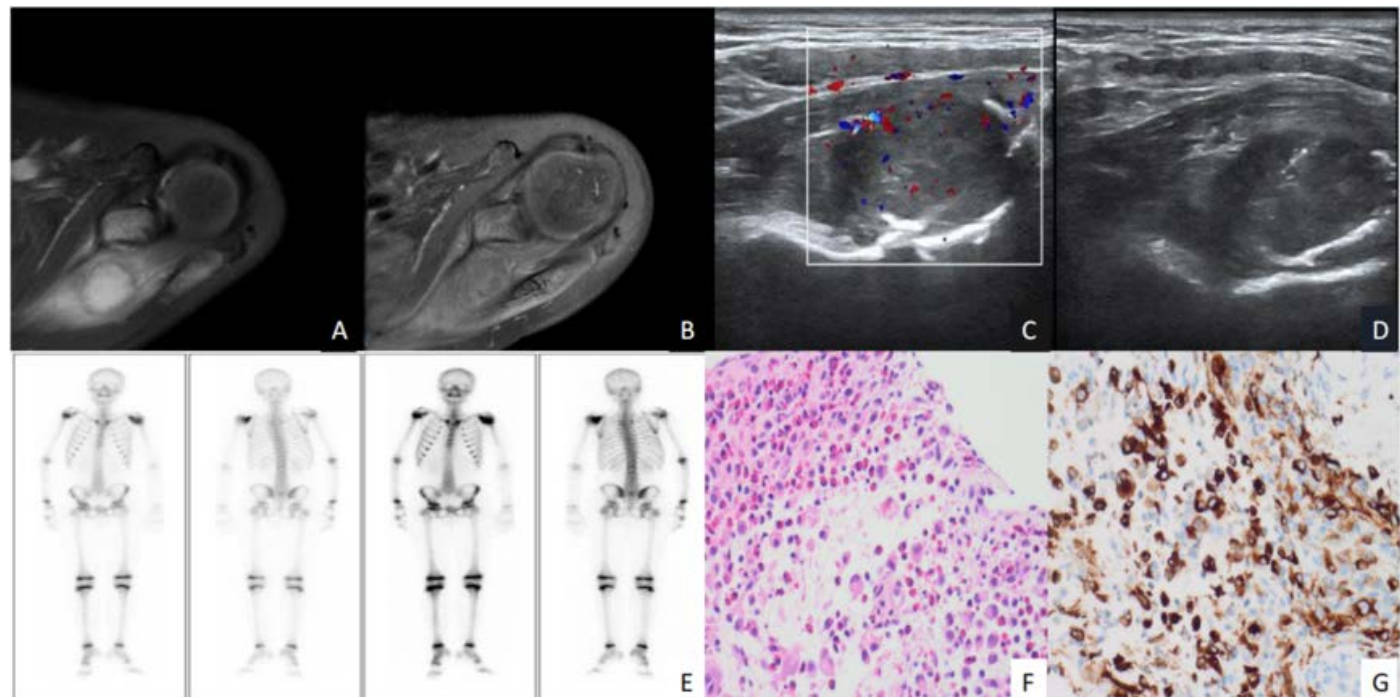


Figure 1: 12-year-old male with solitary Langerhans cell histiocytosis (LCH) of the left scapula.

FINDINGS: (A) Axial T2-weighted fat-suppressed MRI shows a hyperintense mass (arrow) arising from the scapula with surrounding soft tissue edema. (B) Axial contrast-enhanced T1-weighted fat-suppressed MRI demonstrates heterogeneous avid enhancement of the lesion (arrow). (C) Ultrasound shows a 25 × 30 × 24 mm hypoechoic mass with irregular borders and internal vascularity on color Doppler. (D) Ultrasound-guided core needle biopsy (arrow indicates needle path) was performed to obtain a tissue sample. (E) Whole-body bone scan (posterior view) reveals isolated increased radiotracer uptake in the left scapula (arrow). (F) Photomicrograph (original magnification 400x) shows positive immunohistochemical staining for Langerin (CD207) (brown), confirming the diagnosis of LCH. (G) High-power view (original magnification 400x) of Hematoxylin and Eosin (HE) staining shows the characteristic Langerhans cells.

TECHNIQUE: (A) MRI, 3T, T2-weighted fat-suppressed, axial plane, TR/TE: 4000/85 ms. (B) MRI, 3T, T1-weighted fat-suppressed, axial plane, post-contrast (0.1 mmol/kg gadolinium-based agent). (C) Ultrasound, linear array transducer (5-12 MHz), grayscale and color Doppler. (D) Ultrasound-guided biopsy, linear array transducer (5-12 MHz), 14-gauge core needle. (E) Whole-body bone scan, 3 hours post-injection of 99mTc-MDP (10.0 mCi). (F) Immunohistochemistry, anti-Langerin antibody, 400x magnification. (G) Hematoxylin and Eosin stain, 400x magnification.

Summary table:

Table 1: Summary of Langerhans Cell Histiocytosis (LCH)

Feature	Description
Etiology	Clonal proliferative disorder of dendritic-lineage Langerhans cells [1].
Incidence	Rare; 3-5 cases per million children annually [2].
Gender Ratio	Slight male predominance.
Age Predilection	Primarily children (peak 1-3 years).
Risk Factors	Unknown; possible association with family history of cancer.
Treatment	Varies by extent: local curettage, intralesional steroids, low-dose radiation, or chemotherapy for multifocal/systemic disease [12].
Prognosis	Excellent for isolated bone lesions; variable for multisystem disease [2].
Findings on Imaging	Aggressive-appearing lytic bone lesion that can mimic malignancy; "vertebra plana" in spine; soft tissue component and surrounding edema are common [7, 9].

Table 1 Legend: Summary of key clinical, epidemiological, and imaging characteristics of Langerhans cell histiocytosis (LCH), based on current literature [1, 2, 7, 9, 12].

Differential table:

Table 2: Differential Diagnoses for a Lytic Scapular Lesion in a Child

Diagnosis	X-Ray	US	CT	MRI	PET
Langerhans Cell Histiocytosis (LCH)	Lytic lesion, "beveled edge" appearance possible.	Hypoechoic mass with internal vascularity. Cortical disruption.	Lytic lesion with soft tissue component, aggressive periosteal reaction possible.	Hypointense	Increased FDG uptake.
Ewing's Sarcoma	Permeative, moth-eaten lytic lesion with lamellated periosteal reaction (onion-skin).	Large, heterogeneous soft tissue mass.	Large soft tissue mass, bone destruction.	Hypointense	Intense FDG uptake.
Osteosarcoma	Mixed lytic/sclerotic lesion, sunburst periosteal reaction, Codman triangle.	Soft tissue mass, possible osteoid matrix (shadowing).	Osteoid matrix, bone destruction, soft tissue mass.	Hypointense	Intense FDG uptake.
Brodie's Abscess (Subacute Osteomyelitis)	Lytic lesion with sclerotic rim, often in metaphysis.	Fluid collection, may show hyperemia on Doppler.			

Table 2 Legend: Imaging characteristics of Langerhans cell histiocytosis and its main differential diagnoses for a lytic scapular lesion in a child, based on current literature and typical imaging findings [3, 4 5, 7, 9].

KEYWORDS

Langerhans Cell Histiocytosis; Child; Scapula; Osteolytic Lesion; Ultrasound-Guided Biopsy; Differential Diagnosis

ABBREVIATIONS

LCH = Langerhans Cell Histiocytosis

MRI = Magnetic Resonance Imaging

MDT = Multi Disciplinary Team

HE = Hematoxylin and Eosin

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