

A Curious Case of a Solitary Bone Lesion: Langerhan Cell Histiocytosis

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Introduction

- Langerhan Cell Histiocytosis (LCH): A rare disorder that originates from pathogenic myeloid dendritic cells

- Debate as to whether the disease is a neoplastic versus reactive process.¹ The presence of various cytokine involvement in addition to the possibility for spontaneous remission supported a reactive process. Discovery of the BRAF-V600E mutation in about 50% of cases documented however solidified LCH as a neoplastic process.²

- Affecting both children and adults, the disorder is clinically heterogenous, from self-resolving skin lesions to disseminated multiorgan involvement.

- Though more common in children below the age of 15, this is a rare case of LCH presenting as a solitary bone lesion in an otherwise healthy young man.

Case Presentation

HPI:

- Healthy 34-year-old male presented to ED with persistent and severe right sided rib pain, which he thought started after he hit his chest against a chair
- Pain had been progressively worsening over past several weeks.
- Denied any chest pain, shortness of breath, or aggravating symptoms
- OTC pain medication provided no relief

Past Medical History: unremarkable

Family history: unremarkable

Social history: 8.5 pack year history of tobacco

Pertinent exam findings:

- Mild tenderness to palpation along the chest wall
- No bruising or signs of trauma noted along the anterior or posterior chest wall.

Preliminary work up:

- No acute rib fracture noted on X ray

Leading diagnosis:

- Rib contusion
- Discharged with guidance for supportive care and pain management

Interval history:

- Patient returned to ED 2 weeks later with no resolution of symptoms

Further work up:

- CT chest concerning for an aggressive appearing lytic lesion on the right posterior 6th rib (Figure 2)
- Otherwise clinically stable, the patient was discharged with an oncology referral and close follow up.

Outpatient Oncology work up:

- PET CT scan confirmed a hypermetabolic lytic osseous lesion at the right 6th rib. A small lytic lesion in the medial right scapula was also noted (Figure 3).
- Biopsy of the FDG avid lesion confirmed the diagnosis of Langerhan Cell Histiocytosis (Figures 4a, b, c).
- Immunostaining of the biopsy sample was positive for CD1a. The mutation BRAF-V600E mutation was also detected in the biopsy sample.
- MRI of the brain showed no evidence of metastatic disease
- Baseline labs including CBC, CMP, and LDH were all within normal limits.

Treatment:

- 2,000 CGY of radiation therapy to the right medial scapula as well as the right posterior rib in 10 fractions
- Post treatment PET CT showed no evidence of disease, and the patient was scheduled to be monitored with surveillance scans.

Imaging



Figure 1. No definite acute right rib fracture



Figure 2. Aggressive appearing lytic lesion in right 6th rib posterior-laterally. Mature appearing periosteal reaction and cortical destruction along inter margin of rib.

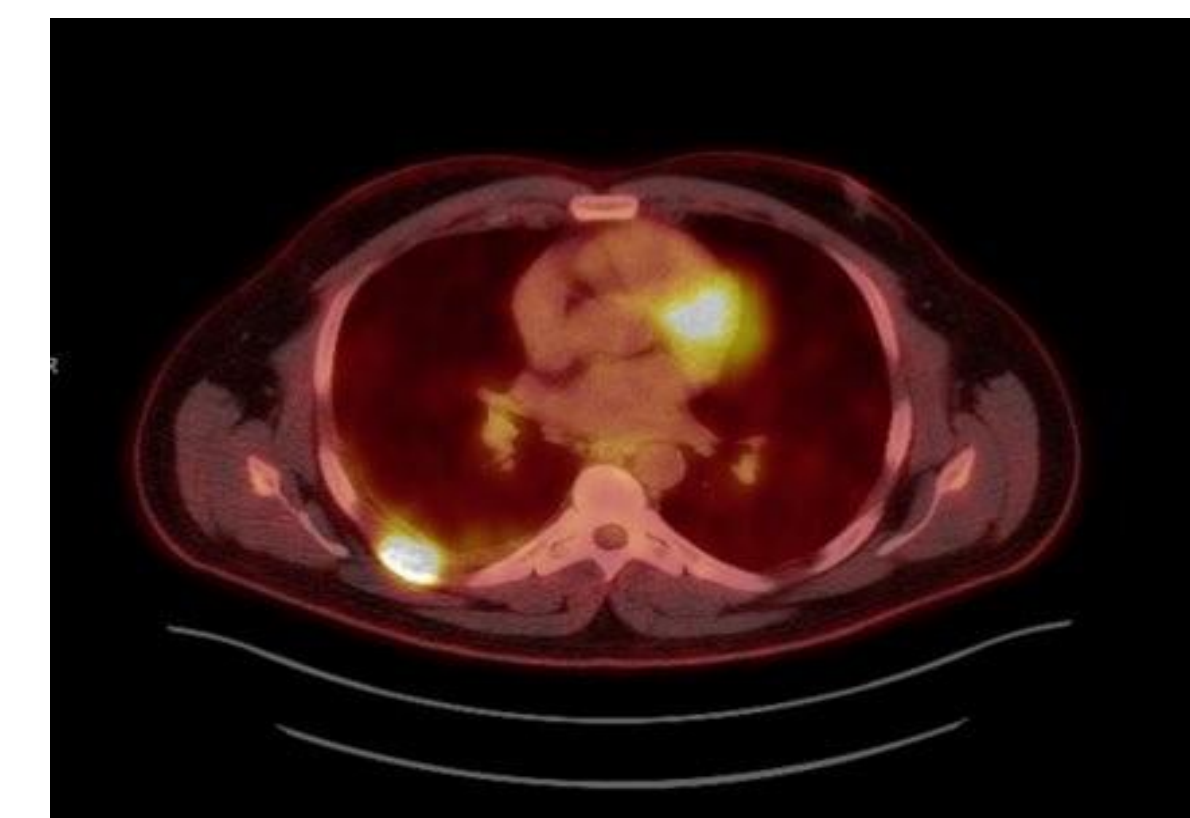


Figure 3. Hypermetabolic lytic osseous lesion in the posterior right 6th rib with a second small lytic lesion in the medial right scapula.

Pathology

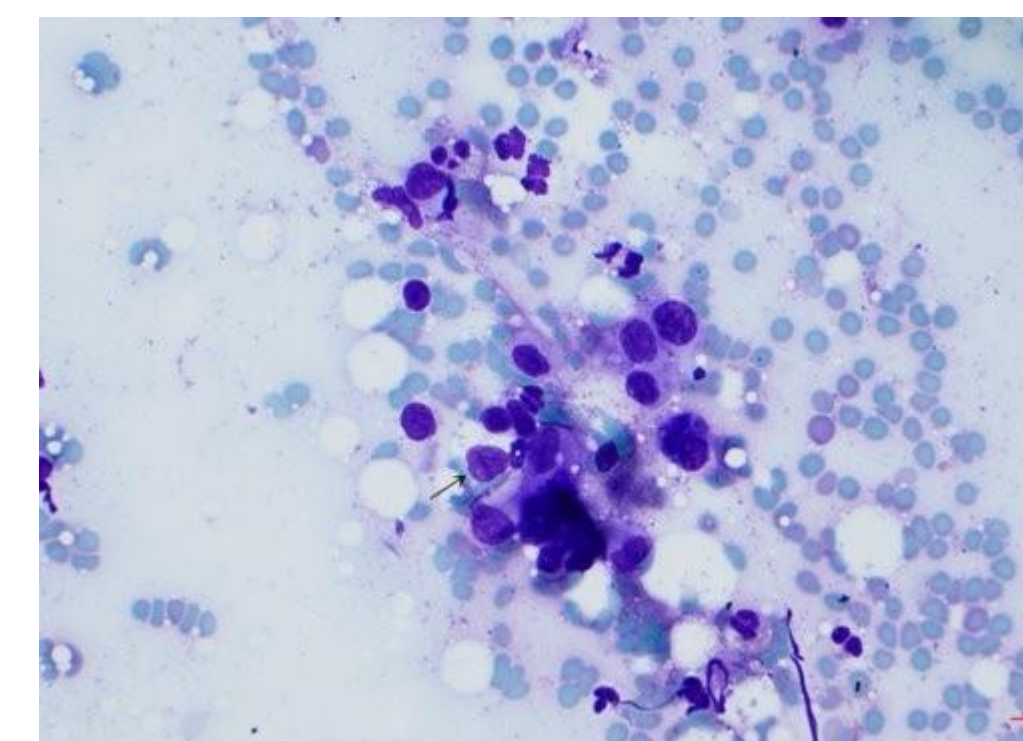


Figure 4a. Neoplastic Langerhans cell with irregular and folded nucleus (arrow, Papanicolaou stain).

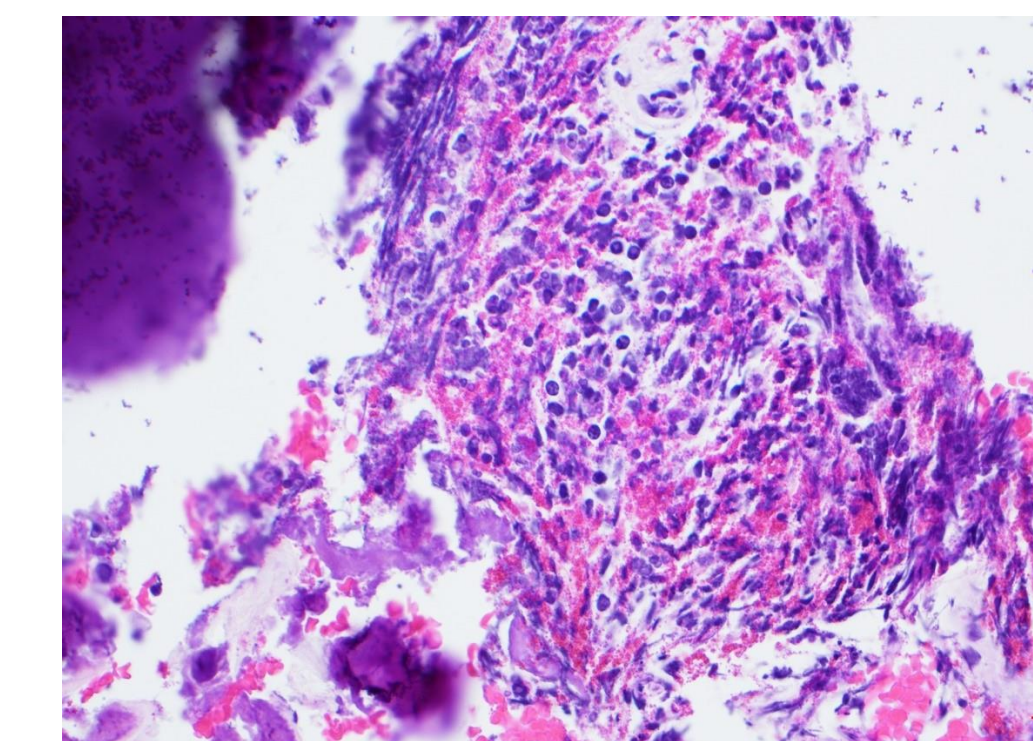


Figure 4b. Clusters of Langerhans cells with bone invasion. Background is rich in eosinophilic granular debris (core biopsy, hematoxylin and eosin stain).

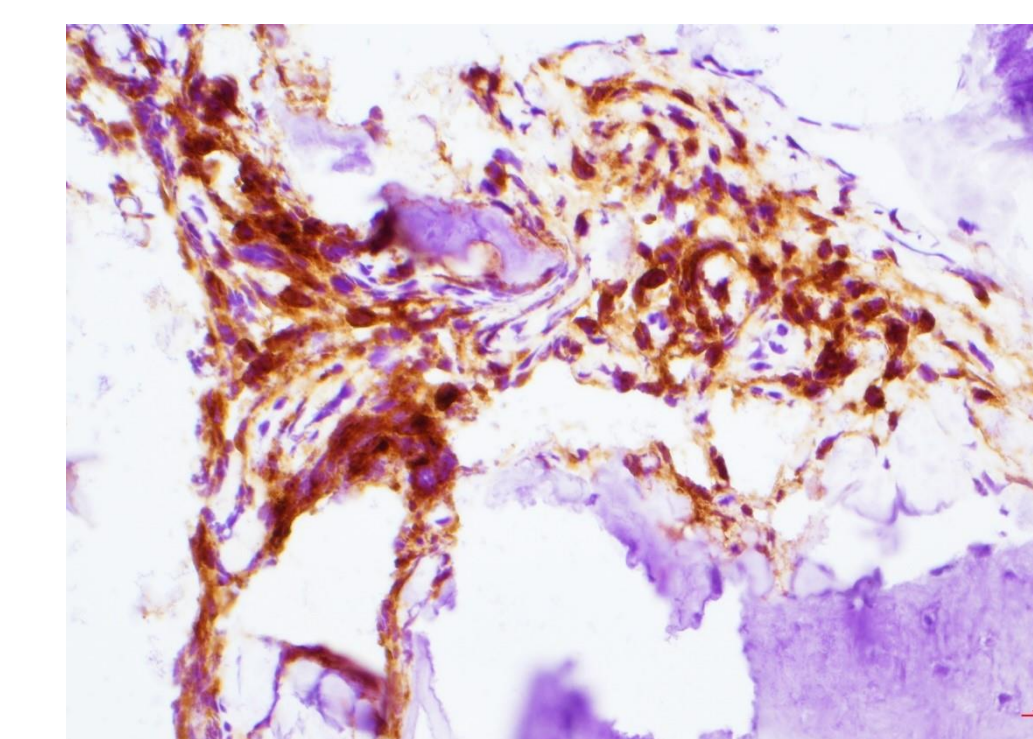


Figure 4c. Langerhans cells are positive for Langerin immunostain (core biopsy)

Discussion

- Langerhan Cell Histiocytosis is classified as the clonal proliferation or aggregation of abnormal CD207+ dendritic cells that can affect single or whole-body tissues.³

- Langerhan cell histiocytosis is rare in the adult population, thus increasing the likelihood of misdiagnosis as it initially occurred in this case.

- There are reported 1-2 newborns diagnosed/ million each year, and 4-5 cases/ million each year in children under the age of 15. In adults, it is even rarer as there are about 1-2 cases/ million.¹

- Bone involvement is seen in almost 80% of cases, though studies have reported that primary rib involvement in adults is extremely rare, about 6%.¹

- Differential diagnosis for osteolytic lesions of the bone includes metastatic disease from carcinoma, multiple myeloma, lymphoma, primary bone malignancies, as well as osteomyelitis.

Conclusion

- In an otherwise healthy patient with no signs of overt trauma to the site, this patient had pain out of proportion to the nature of the physical exam and imaging findings, thus warranting a more detailed work up including CT imaging earlier on.

- This case particularly highlights that a patient's age, social history, and medical comorbidities must always be closely considered when working up patients.

References

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Objectives

- Highlight the importance of a broad differential, specifically in a patient whose subjective story does not align with objective data

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