Case 18103

Eurorad

Pulmonary Langerhans' Cell Histiocytosis and solitary eosinophilic granuloma in an adult

Published on 18.04.2023

DOI: 10.35100/eurorad/case.18103
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Lung Musculoskeletal bone
Musculoskeletal soft tissue
Imaging Technique: CT
Imaging Technique: SPECT
Case Type: Clinical Cases
Authors: Arnaud Roger Goossens1,2, Jan Hendrickx3, Jan Vandereycken3, Filip M. Vanhoenacker1,2,4
Patient: 32 years, female

Clinical History:

A 32-year-old female presented at the emergency department with acute right-sided thoracic pain. She also complained of a chronic dry cough. She was an active smoker with 10 packyears. Clinical examination and laboratory results were unremarkable. The patient was hemodynamically stable and afebrile.

Imaging Findings:

Computed tomography (CT) reveals multiple thick-walled cystic lesions (<10 mm) as well as some micronodules, predominantly in the upper- and middle lung fields (Figure 1). An osteolytic lesion with non-sclerotic margins, cortical breakthrough and adjacent soft tissue swelling is seen on the lateral aspect of the right seventh rib (Figure 2A). Subsequent bone scintigraphy confirms its unique location (Figure 2B). Immunohistochemical diagnosis of Langerhans' Cell Histiocytosis was made after a surgical lung biopsy.

Discussion:

Langerhans' Cell Histiocytosis (LCH) is a rare systemic disorder caused by (idiopathic) infiltration and proliferation of Langerhans cells or histiocytes in various organs. Manifestations occur most commonly in bones, lungs, liver–spleen, central nervous system, thymus and lymph nodes.

Pulmonary LCH (PLCH) presents in young adults aged 20 to 40 years old, independent of gender. There is an important association with current (or previous) cigarette smoking in 90 to 100%[1]. Clinical presentation includes chronic cough, dyspnoea and constitutional symptoms (asthenia, weight loss). Only a minority of lesions present with pneumothorax. CT is the modality of choice for pulmonary evaluation. The typical manifestation pattern of PLCH is centrilobular lesions that predominate in the upper and middle lung fields. Basal fields are typically spared. The morphology of the lesions differs depending on the stage of disease. In the early or "florid" stage, (cavitated) nodules are found, followed by cavitation and formation of thick-walled cysts which progressively transform into thin-walled cysts. End-stage is characterized by fibrotic and cystic changes. The key features for diagnosis are patient age, a history of smoking, and the distribution pattern and morphology of the lesions. Cysts with a round shape in random distribution (no basal sparing) and absence of parenchymal nodules favors the diagnosis of lymphangioleiomyomatosis (LAM), while cysts predominate in the lower lobes favors lymphocytic interstitial pneumonitis (LIP). In centrilobular emphysema, destructed parenchyma lacks a visible wall. Overall, pulmonary findings can be confounding, thus biopsy is mandatory especially in young females with a cystic pattern at onset[1-3].

LCH in a solitary bone was previously referred to as eosinophilic granuloma. It is more commonly seen in children aged 3 to 5 years old, with predilection site of the skull, pelvis and femur. It is rarer in adults, in whom the jaw, skull and vertebra are the preferred locations [4]. Patients present with pain, tenderness and swelling around the lesion. CT is the preferred modality to evaluate matrix and adjacent structures. The osteolytic defect has non-sclerotic margins and is often beveled due to uneven destruction of the inner and outer cortex [5]. Adjacent soft tissue swelling can also be appreciated on CT and ultrasound, which is a good site for biopsy. Bone scintigraphy and PET-CT are useful to evaluate multiplicity [4, 6]. The differential diagnosis for an osteolytic rib lesion with non-sclerotic margins includes metastasis, multiple myeloma, primary bone malignancy, osteomyelitis and lymphoma.

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Pulmonary Langerhans' Cell Histiocytosis, Lymphangioleiomyomatosis (LAM), Emphysema, Lymphocytic interstitial pneumonitis (LIP), Usual interstitial pneumonia (UIP), Wegener granulomatosis, Septic emboli, Pneumatocoeles secondary to pulmonary infections, Metastasis, Osteomyelitis, Multiple myeloma, Primary bone malignancy, Lymphoma

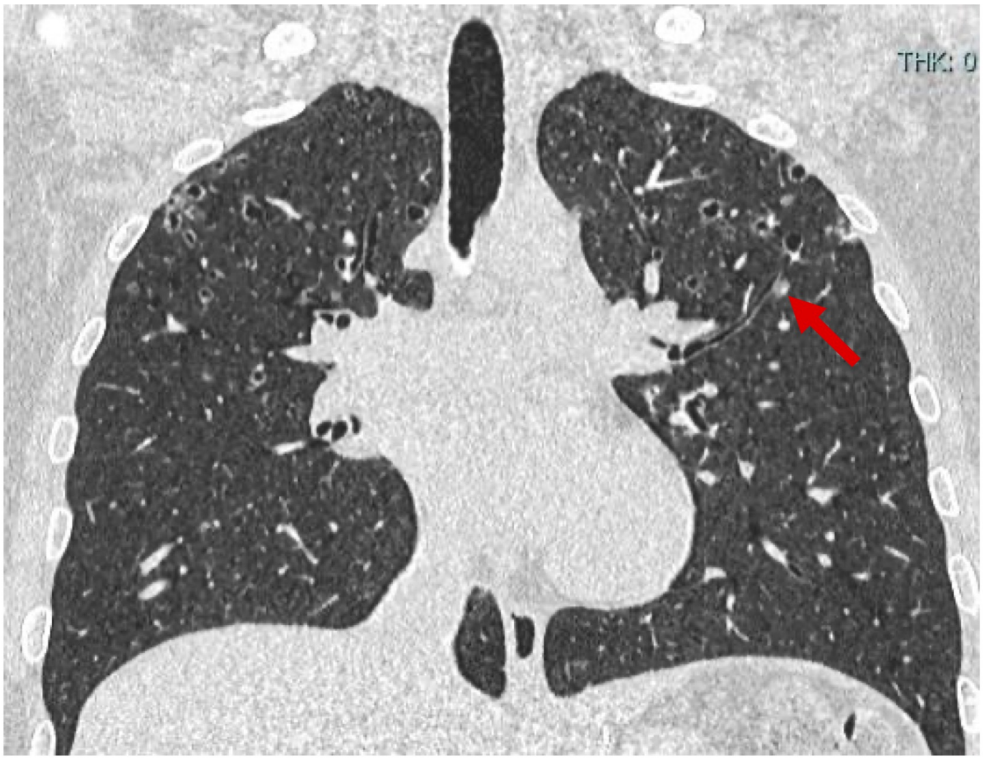
Final Diagnosis: Pulmonary Langerhans' Cell Histiocytosis

References:

Tazi A (2006) Adult pulmonary Langerhans' cell histiocytosis. European Respiratory Journal 27:1272–1285 (PMID: 16772390)
Castoldi MC, Verrioli A, de Juli E, Vanzulli A (2014) Pulmonary Langerhans cell histiocytosis: The many faces of presentation at initial CT scan. Insights Imaging 5:483–492 (PMID: 24996395)
Brauner MW, Grenier P, Tijani K, et al (1997) Pulmonary Langerhans cell histiocytosis: Evolution of lesions on CT scans. Radiology 204:497–502 (PMID: 9240543)
Kim SH and Choi MY (2016) Langerhans cell histiocytosis of the rib in an adult: a case report. Case Reports in Oncology 9(1):83-88 (PMID: 26933424)
Azouz EM, Saigal G, Rodriguez MM, Podda A (2005) Langerhans' cell histiocytosis: Pathology, imaging and treatment of skeletal involvement. Pediatric Radiology 35:103–115 (PMID: 15289942)
Zhao S, Luo C, Tang B, et al (2022) Langerhans cell histiocytosis of the rib in an adult: A case report and review of the literature. International Journal of Surgery Case Reports 97:107432 (PMID: 35939864)

Figure 1

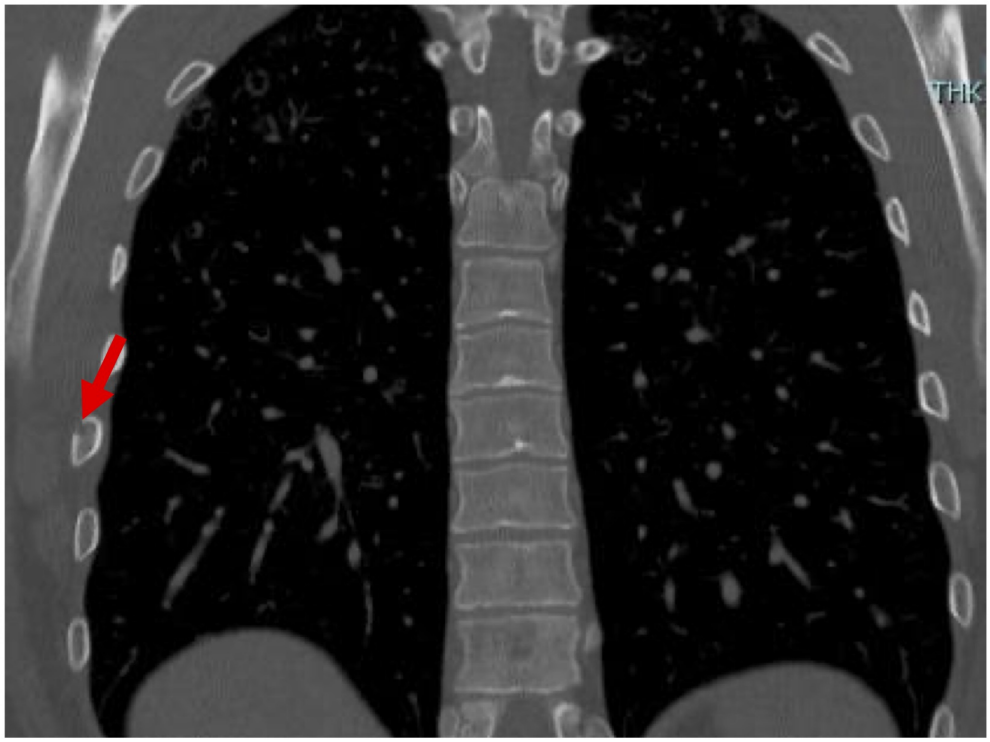
a



Description: Coronal CT scan of the thorax in lung window. Description: Multiple thick-walled cystic lesions (< 10 mm) in the upper- and middle lung fields, with some micronodules (red arrow). Notice sparing of the lower fields **Origin:** © Department of Radiology, AZ Jan Palfijn Ghent, East-Flanders, Belgium, 2022

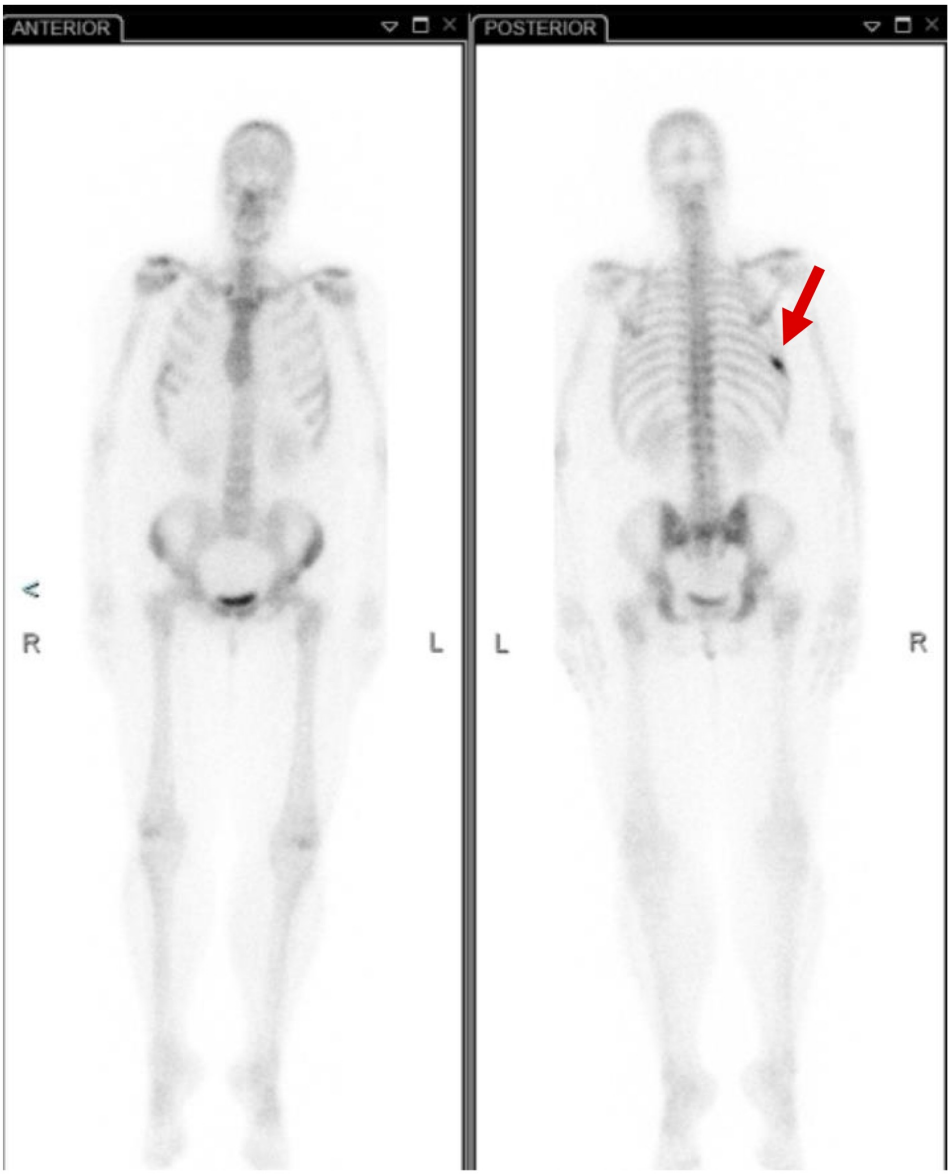
Figure 2

a



Description: Coronal CT scan of the thorax in bone window. Description: Ill-defined osteolytic lesion with cortical breakthrough and adjacent soft tissue swelling is seen on the lateral aspect of the seventh rib on the right (red arrow) **Origin:** © Department of Radiology, AZ Jan Palfijn Ghent, East-Flanders, Belgium, 2022

b



Description: Bone scintigraphy in the early osteoblastic phase. Description: Bone scintigraphy was used to exclude multifocality of musculoskeletal LCH. Osteoblastic activity is seen on the lateral aspect of the seventh rib on the right (red arrow), correlating with a solitary eosinophilic granuloma **Origin:**