

## Pulmonary Langerhans cell histiocytosis in an early stage

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**Section:** Chest imaging

**Area of Interest:** Thorax

**Procedure:** Diagnostic procedure

**Imaging Technique:** CT

**Special Focus:** Inflammation Case Type: Clinical Cases

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**Patient:** 50 years, female

### Clinical History:

A 50-year-old female patient with a recently diagnosed left breast cancer, underwent chest CT for cancer staging. She was a smoker, diagnosed with mild chronic obstructive pulmonary disease (COPD) 4 years before. Occasional dyspnoea on exertion. No chest pain. Afebrile. No evidence of cyanosis, lymphadenopathy or clubbing.

### Imaging Findings:

Chest CT (Fig. 1-3) shows innumerable nodules, with ill-defined margins and measuring less than 10 mm, with centrilobular location. They are predominantly distributed in the upper lobes. A few cavitated nodules can be seen posteriorly in the left lower lobe (Fig. 1d).

A chest CT performed 5 months later showed complete resolution of the aforementioned nodules (Fig. 4).

### Discussion:

Langerhans Cell Histiocytosis (LCH) is a disorder characterised by the accumulation of LCs in different organs and tissues [1]. LCH may affect an isolated organ (an entity previously known as eosinophilic granuloma) or may present as a multisystemic disease [2].

Pulmonary involvement of Langerhans Cell Histiocytosis (PLCH) is rare, occurring almost exclusively in smokers and affecting mainly young adults between the ages of 20 and 40 [3].

There is no gender preference.

Pathologically PLCH consists of an accumulation of Langerhans cells (and other inflammatory cells) in small airways, which results in the formation of nodular inflammatory lesions. This may be accompanied by variable lung vascular and interstitial involvement [2]. Whilst cellular inflammation is prominent in early disease, cystic lung destruction, cicatricial scarring of the small airways and vascular remodelling are the hallmark of more advanced stages [2].

Patients with PLCH commonly present with unspecific symptoms such as non-productive cough or dyspnoea [3]. They may, however, be entirely asymptomatic.

Chest radiographs are usually abnormal, showing reticulonodular infiltrates in early disease and cystic lesions in

advances stages [4]. High resolution computed tomography (HRCT) should be obtained in every patient. Nodules (with or without cavitation) measuring 1 to 10mm with a centrilobular location are often seen in early stages [5], as in our case.

Pulmonary cysts, although seen in any stage of disease, are more commonly found in more advanced disease [6]. Their wall may be thin or thick (up to 20 mm).

This pattern of nodular and cystic changes is usually distributed in the upper and middle lobes (with relative sparing of lung bases) [5], although our patient presented with cysts in the left lower lobe.

Typical chest CT features may be sufficient for establishing a presumptive diagnosis. However, when findings are non-diagnostic (predominant lower lobe involvement, cystic changes without nodules or nodular changes without cysts), either transbronchoscopic or surgical lung biopsy may be required [2]. Due to the patient's oncologic history, she was submitted to surgical lung biopsy of the nodules at another institution, which confirmed PLCH.

Prognosis is variable and unpredictable.

Smoking cessation is fundamental and may lead to disease regression [7].

Immunosuppressive agents should be considered for all adults with severe disease [2].

Our patient showed complete spontaneous resolution without immunosuppressive agents or smoking cessation.

**Differential Diagnosis List:** Pulmonary Langerhans cell histiocytosis (early/nodular stage), Miliary tuberculosis, Wegener granulomatosis, Metastases, Sarcoidosis

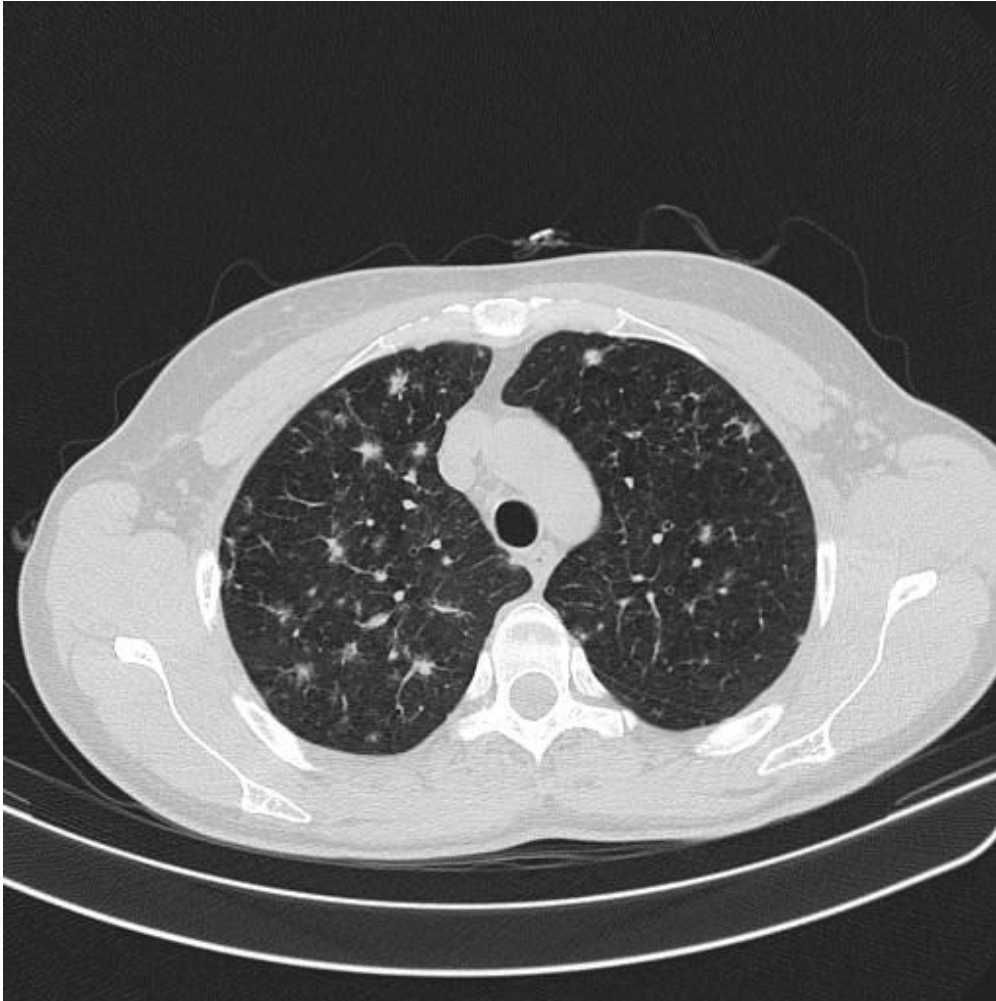
**Final Diagnosis:** Pulmonary Langerhans cell histiocytosis (early/nodular stage)

## References:

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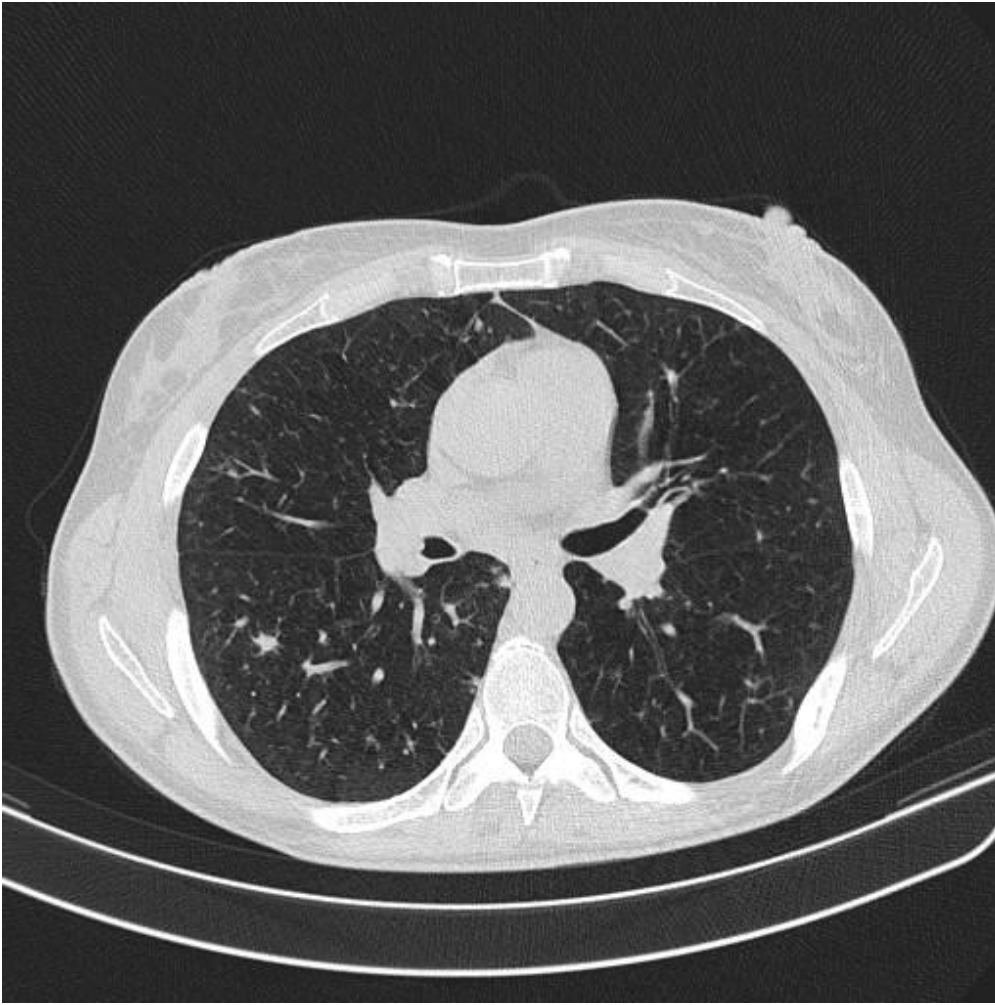
**Figure 1**

**a**



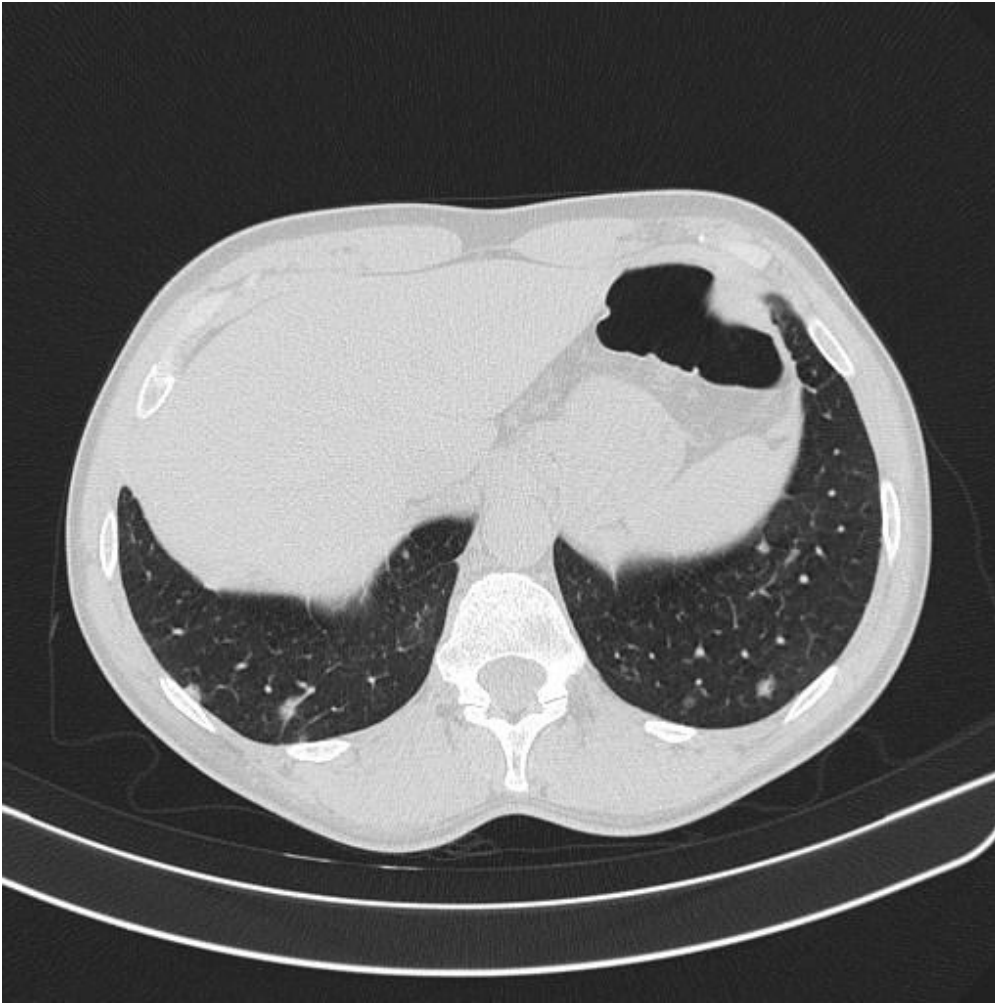
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**b**



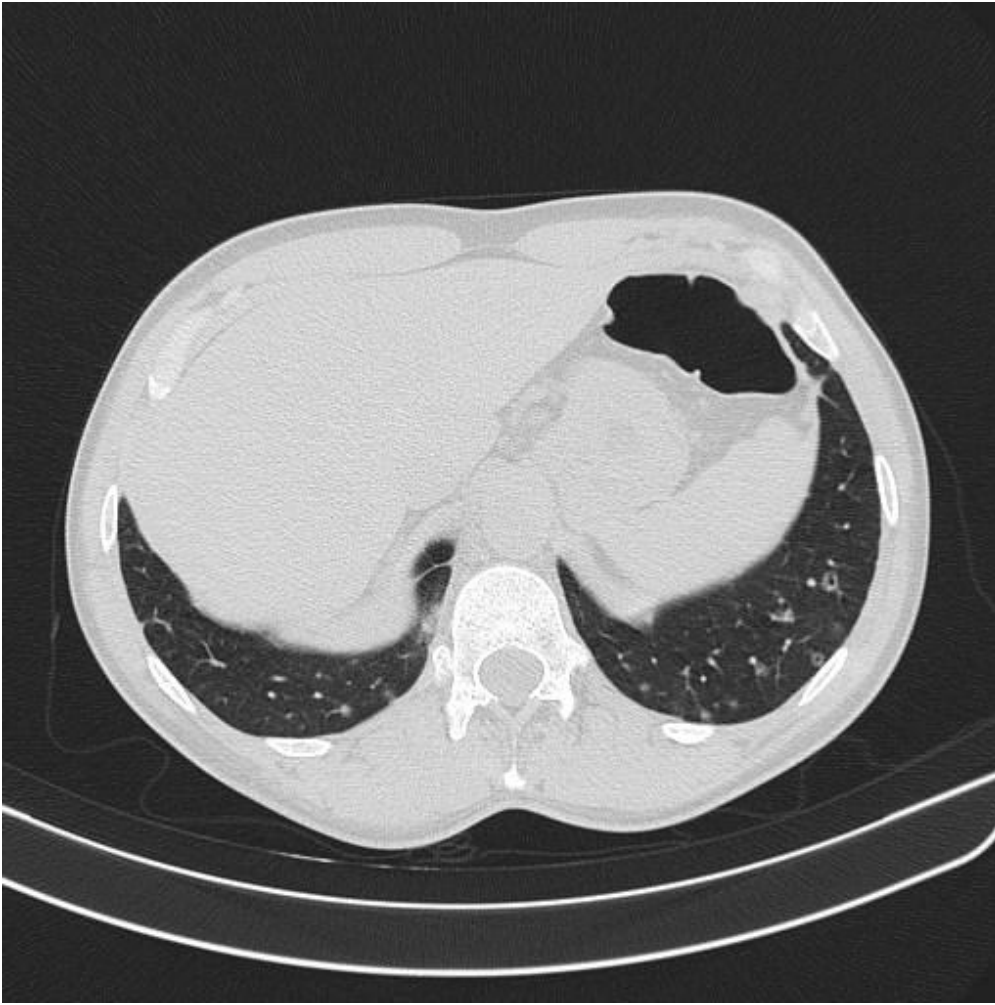
**Description:** Middle segments **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

**c**



**Description:** Lower segments **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

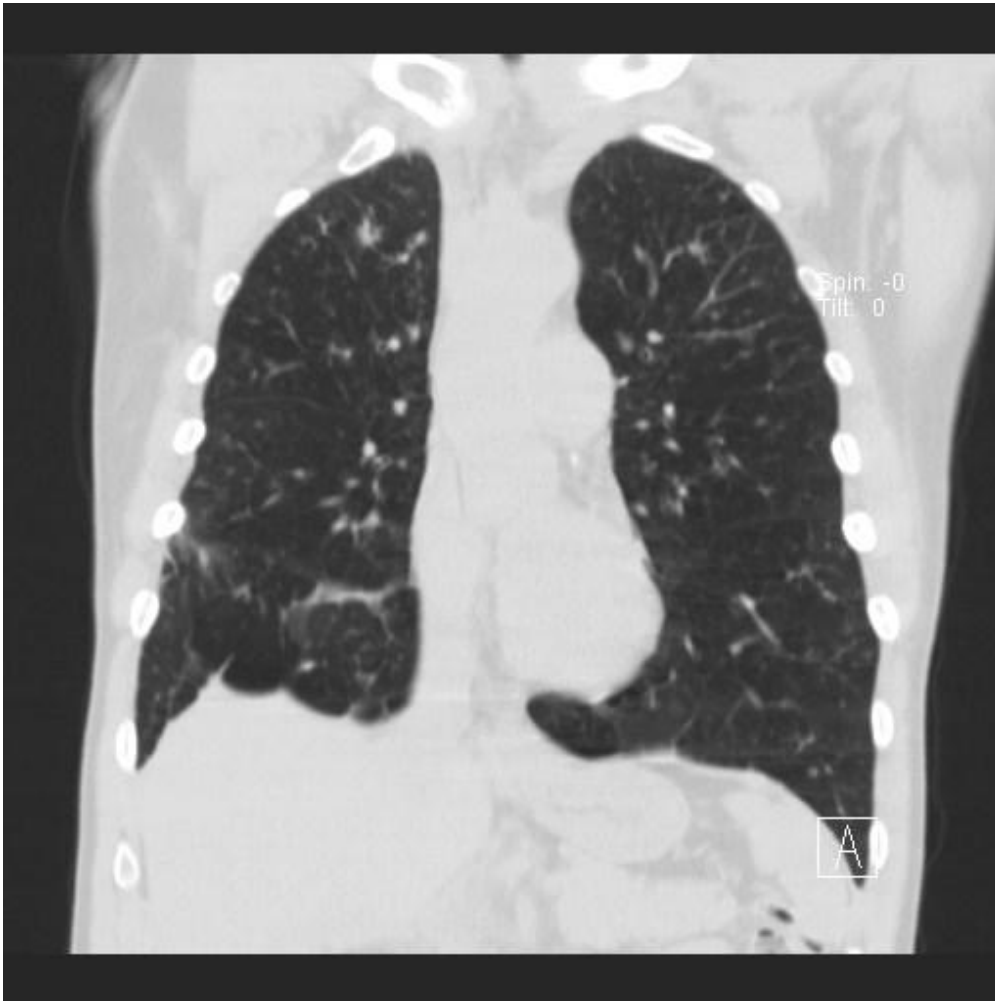
d



**Description:** Cavitated nodules in the left lower lobe **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

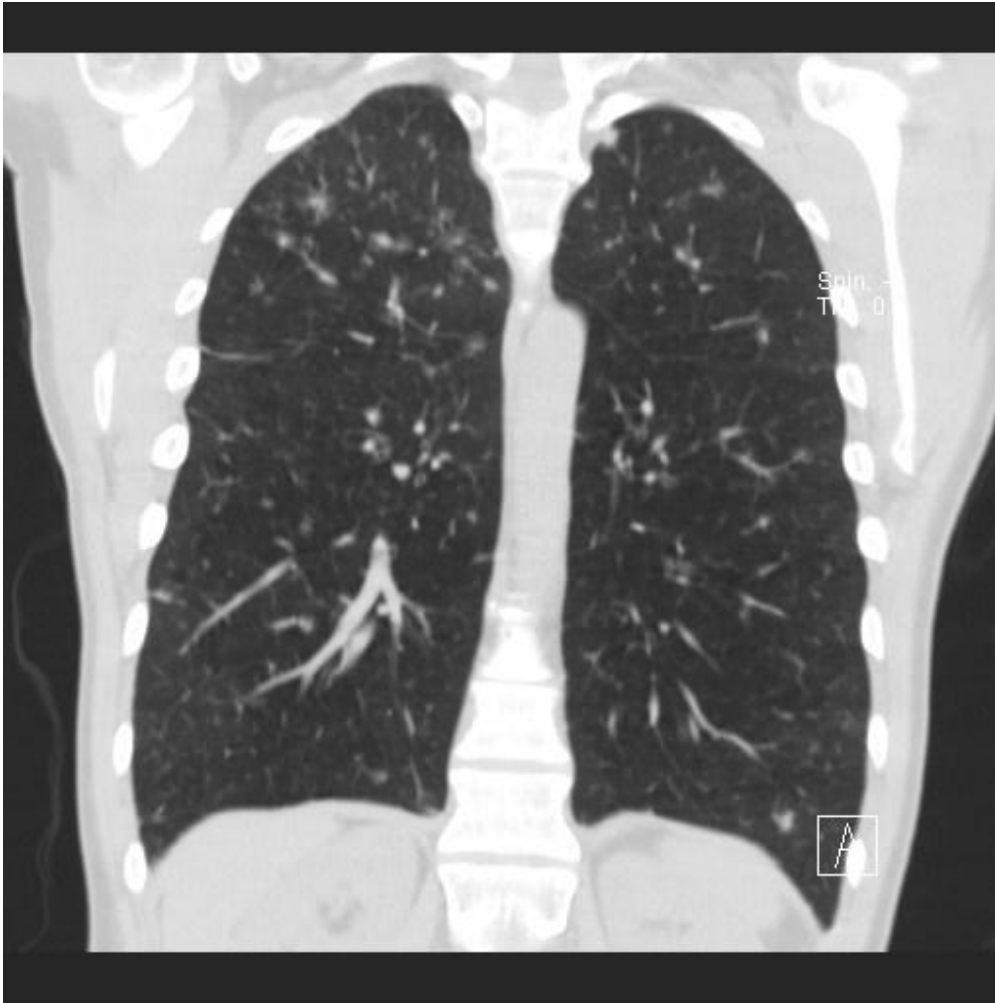
**Figure 2**

a



**Description:** Anterior segments **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

**b**



**Description:** Middle segments **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal



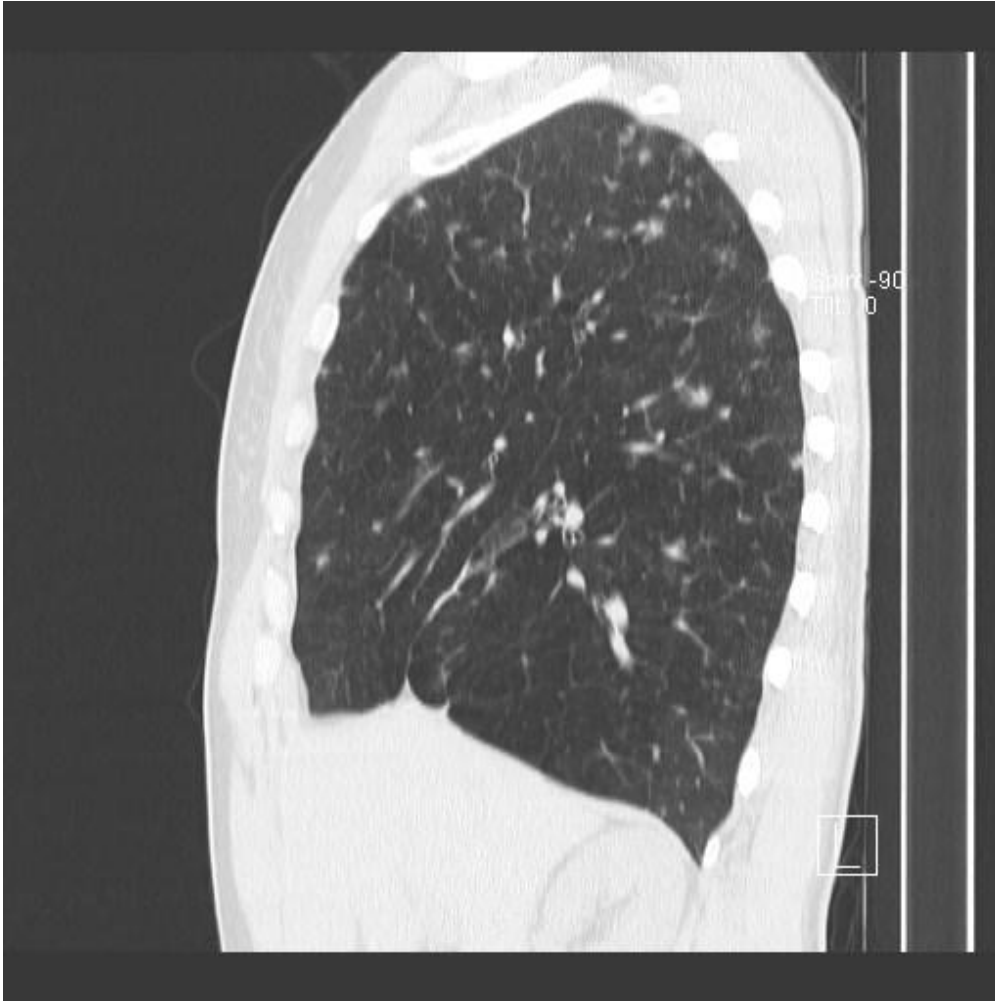
**c**



**Description:** Posterior segments **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

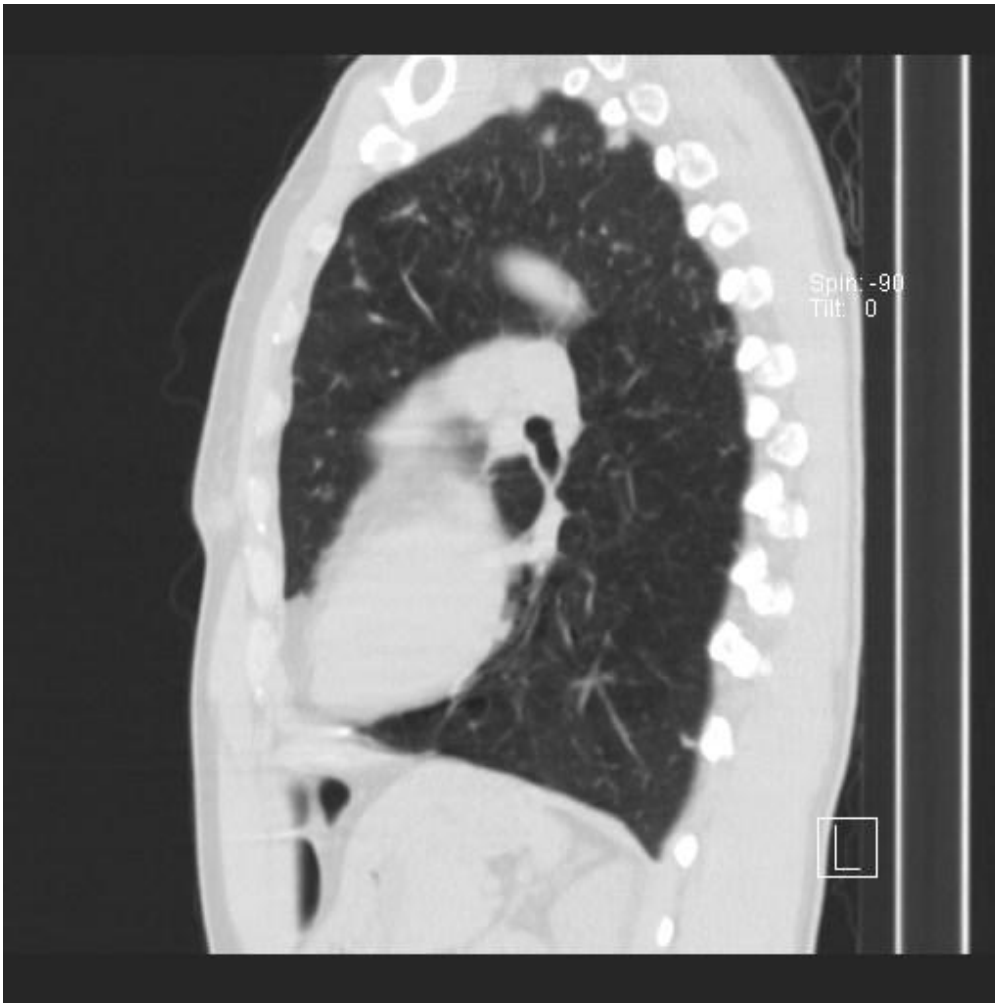
**Figure 3**

a



**Description:** Left lung **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

**b**



**Description:** Right lung - medial **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

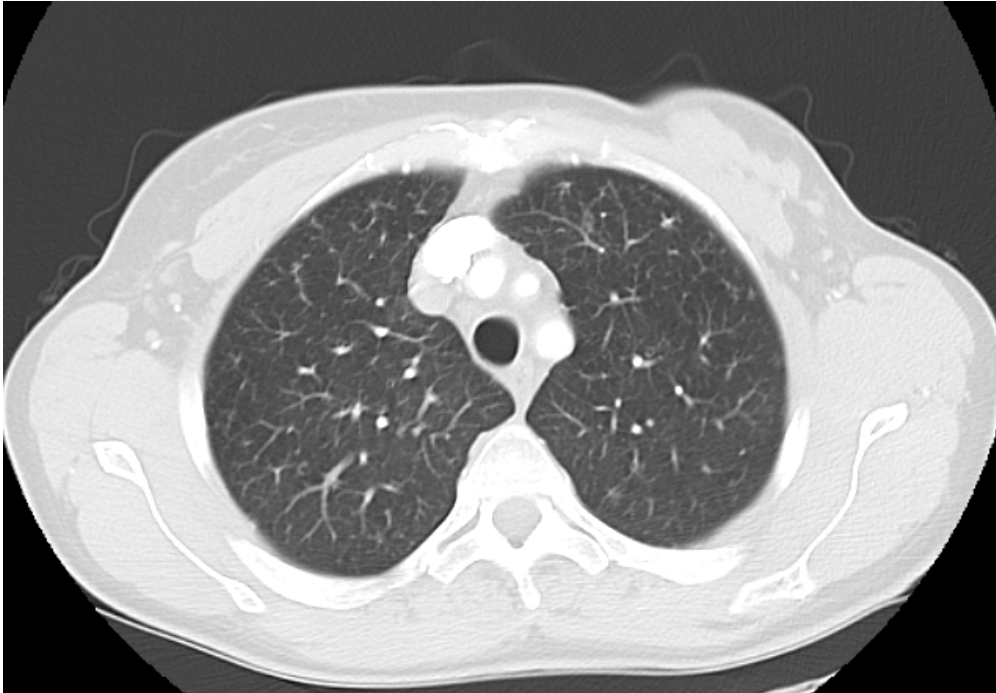
**c**



**Description:** Right lung - lateral **Origin:** Department of Radiology, Centro Hospital de Lisboa Ocidental, Lisboa, Portugal

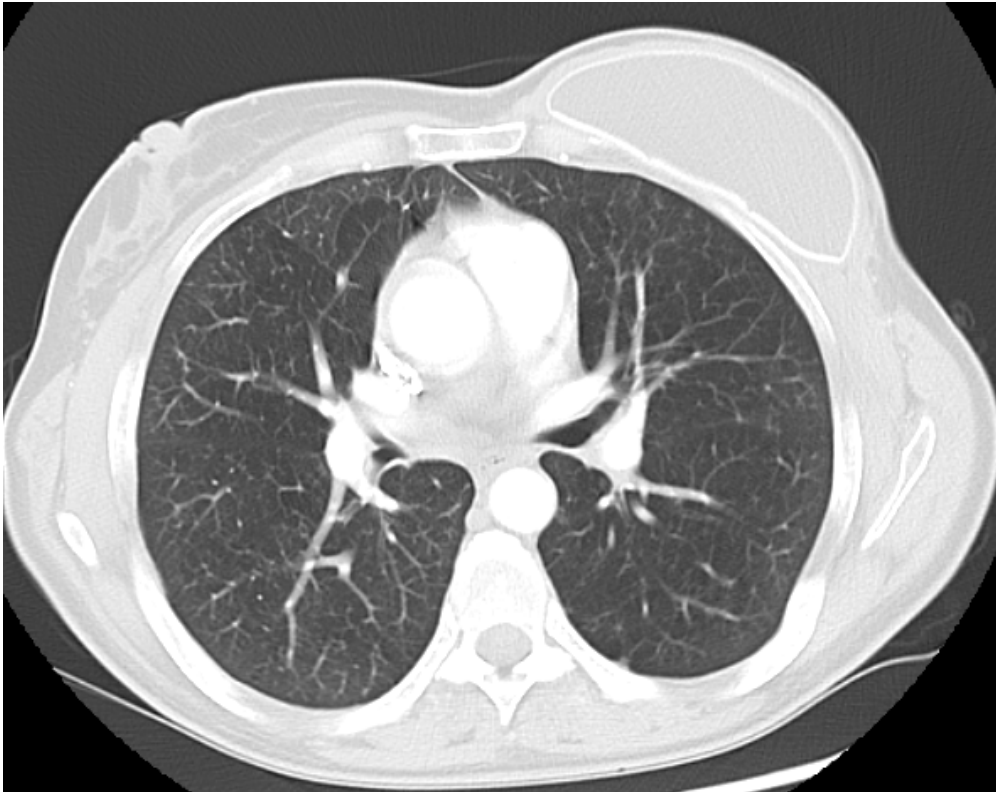
**Figure 4**

**a**



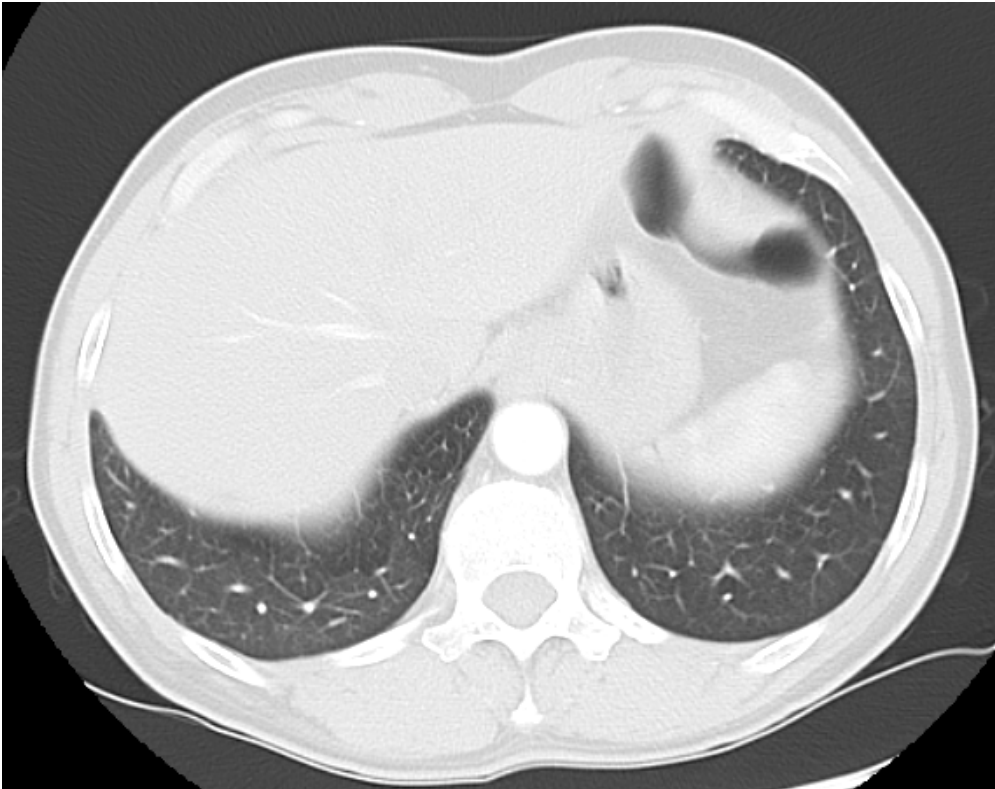
**Description:** Upper segments **Origin:** Centro Hospitalar de Lisboa Ocidental, Serviço de Radiologia, Lisboa, Portugal

**b**



**Description:** Middle segments **Origin:** Centro Hospitalar de Lisboa Ocidental, Serviço de Radiologia, Lisboa, Portugal

**c**



**Description:** Lower segments **Origin:** Centro Hospitalar de Lisboa Ocidental, Serviço de Radiologia, Lisboa, Portugal