Case 15539

Eurorad

Pulmonary epithelioid haemangioendothelioma: a rare clinical entity

Published on 26.03.2018

DOI: 10.1594/EURORAD/CASE.15539
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Thorax Lung Mediastinum
Procedure: Diagnostic procedure
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
Authors: Giuseppe Aquaro1, Vincenza Giorgio2, Manuela Rollo2, Carlo Florio3
Patient: 52 years, female

Clinical History:

A 52-year-old female patient, non-smoker, presented to our chest unit with exertional dyspnoea, weight loss and troublesome dry cough for the previous three months. She had no professional exposure, no clinical signs of connective tissue diseases. Her past medical history and physical examination were unremarkable.

Imaging Findings:

Chest X-ray, performed at a private hospital and not shown by the patient, and chest CT scan, requested by our pulmonologists without intravenous contrast medium, showed multiple, random and bilateral nodules of various size from 3 mm to 14 mm in diameter and predominantly lower lobe thickening of both bronchovascular bundles and interlobular septa with small amount of right-sided pleural effusion. Abdominal ultrasound was unremarkable. Routine laboratory findings, tumour markers and autoantibodies were within normal limits. BAL on bronchoscopy revealed mild lymphocytosis (52%) with CD4/CD8 ratio of 2.2%. A wedge resection by video-thoracoscopic surgery was required and immunostaining of the tissue was positive for CD31 and ERG, but negative for CAMTA1 and a diagnosis was established. After adequate therapy, we observed complete resolution of the nodules on the last CT scan of the chest, abdomen and pelvis with iodine intravenous contrast medium of 2 months before.

Discussion:

Pulmonary epithelioid haemangioendothelioma (PEH) is a rare, vascular tumour of low-to intermediate grade malignancy, described as an intravascular bronchioloalveolar tumour by Dail and Liebow in 1975. Most affected patients are women, ranging in age from 19 to 70 years; many of them are asymptomatic but others show pleuritic pain, cough, dyspnoea or haemoptysis. This tumour may arise simultaneously or sequentially from many organs (lungs, liver, bone and soft tissues); it is difficult to determine if the tumour is multicentric or a primary lesion with metastases to other tissues. Lung involvement is relatively rare with 19% of all cases of epithelioid haemangioendothelioma (EHE). On chest X-ray there are multiple, well or ill-defined small nodules measuring up to 2 cm in both lungs but in less than 10% of the cases reported in the literature a solitary pulmonary nodule was found. There is no hilar or mediastinal lymph node enlargement, but sometimes diffuse infiltrative pleural thickening. Chest CT scan shows multiple nodules with irregular margins, rarely with calcifications, in a perivascular distribution. 20% of PHE patients may have hepatic nodules with peripheral contrast enhancement and some calcifications. [1-4] PHE can also be associated with congenital anomalies of the musculoskeletal system such as hemihypertrophy and scoliosis. [5]
Histologically, the tumour shows oval or round nodules with a central hypocellular zone and a cellular peripheral

one; immunostaining of the tissue is positive for endothelial markers, particularly CD31 and CD34 in about 90% of the cases. The independent risk factors of poor prognoses are weight loss, anaemia, pulmonary symptoms and pleural haemorrhagic effusion. The treatment for PHE can vary from observation in asymptomatic patients, sometimes with spontaneous regression, to surgery in patients with unilateral lung nodule and drugs in patients with disseminated disease using carboplatin, etoposide, interferon 2a, bevacizumab, azathioprine and corticosteroids. Most patients die from respiratory failure due to increasing size and number of tumour nodules, but death can occur from sepsis, myocardial infarction or other malignancy. The 5-year survival is around 60%. [1, 3, 4, 6]
According to some authors the FDG uptake on FDG-PET scan is useful because it reflects the activation of tumour cells, resulting in progression of the disease, but in some patients it was observed that PET findings were negative, probably related to a low proliferation rate of the tumour cells.
So it is difficult for any clinicians to manage PHE because of its variable course and the therapeutic options. [1, 6, 7]
Differential Diagnosis List: Pulmonary epitheliod haemangioendothelioma, Sarcoidosis, Non-small cell lung cancer, Metastases, Vasculitis, Organising pneumonia, Infectious diseases particularly tuberculosis

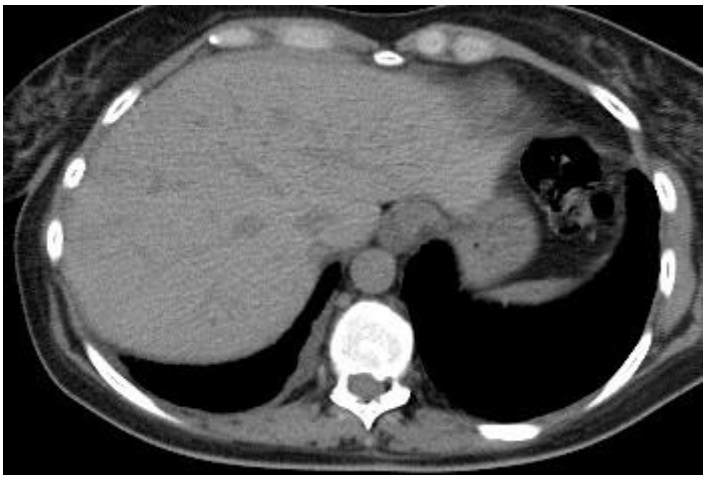
Final Diagnosis: Pulmonary epitheliod haemangioendothelioma

References:

Okamura K, Ohshima T, Nakano R (2010) A Case of Pulmonary Epitheliod Hemangioendothelioma Surviving 10 years without treatment. Ann Thoracic Cardiovasc Surg 16: 432-435 (PMID: 21263426)
Sakamoto N, Adachi S, Monzawa S (2005) High resolution of pulmonary epitheliod hemangioendothelioma. J Thoracic Imaging 20: 236-238 (PMID: 16077343)
Kim M, Chang J, Choi H (2015) Pulmonary hepitheliod hemangioendothelioma misdiagnosed as a benign nodule. World Journal of Surgical Oncology 13: 107 (PMID: 25889253)
Rishi Mehta S, Das A (2012) Metastatic pulmonary epithelioid hemangioendothelioma: a case report and review of the literature. Respiratory Medicine Case Reports 7: 17-20 (PMID: 26029602)
Dahabreh M, I.Hmeideen N (2012) Multicentric epithelioid hemangioendothelioma involving the lungs, trachea, liver and the skeletal muscles. Respiratory Medicine Case Reports 5: 20-22 (PMID: 26057597)
Rosengarten D, R.Kramer M (2011) Pulmonary epithelioid hemangioendothelioma. Israel Medical Association Journal 13(11): 676-9 (PMID: 22279701)
Cazzuffi R, Calia N, Ravenna F (2011) Primary pulmonary epithelioid hemangioendothelioma: a rare cause of PET-negative pulmonary nodules. Case reports in medicine 2011:262674 (PMID: 21869893)

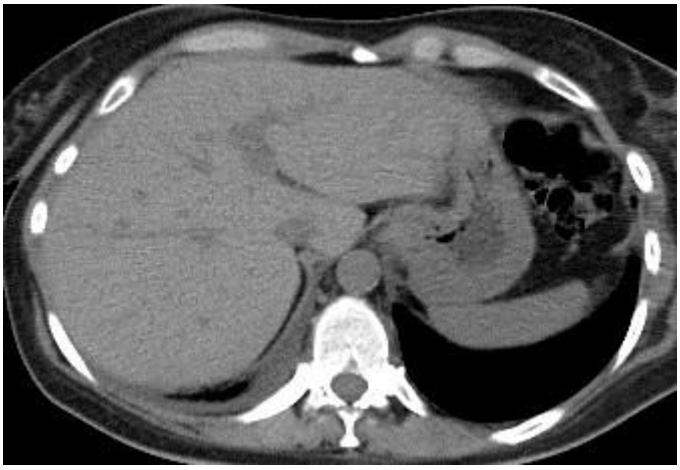
Figure 1

a



Description: Chest CT scan with mediastinal window setting, requested without intravenous iodine contrast medium, shows a small amount of right-sided pleural effusion. **Origin:** G.Aquaro, Department of Radiology, Di Venere Hospital - Bari (BA) - Italy

b



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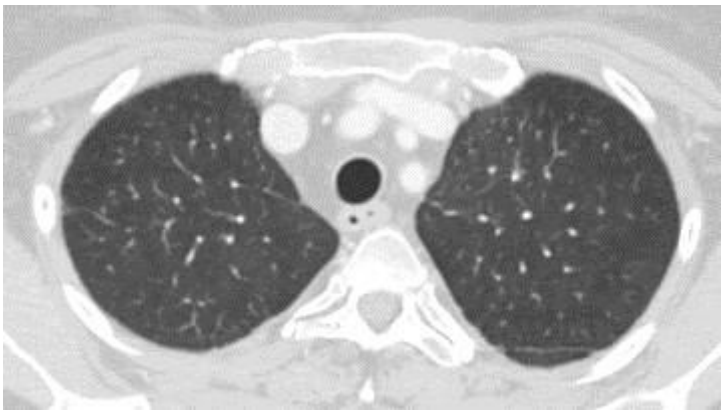

Description: Chest CT scan – with lung window setting - shows complete resolution of the pulmonary nodules after adequate therapy. **Origin:** G.Aquaro, Department of Radiology, Di Venere Hospital - Bari (BA) - Italy

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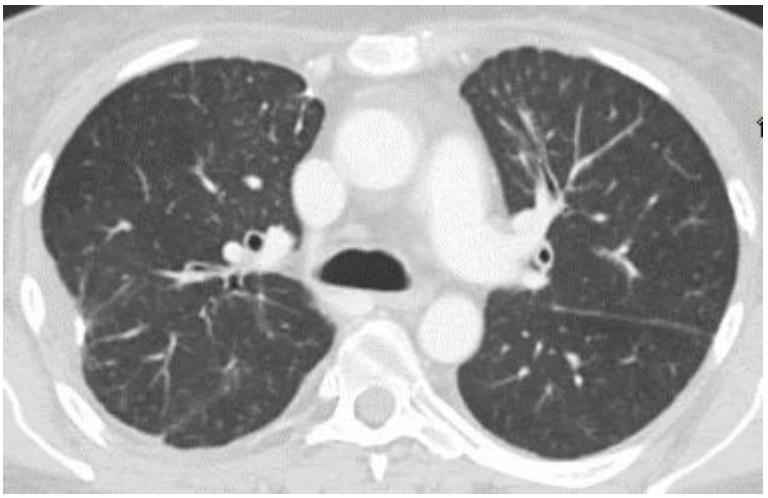

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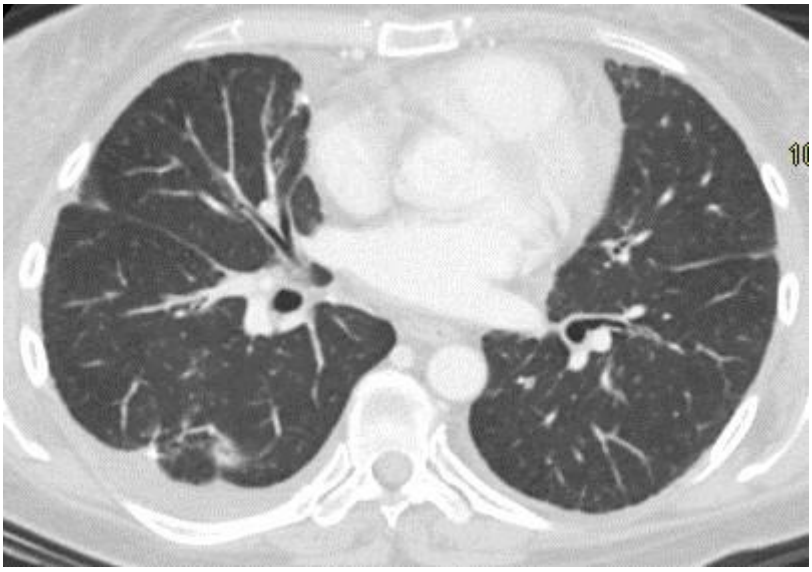

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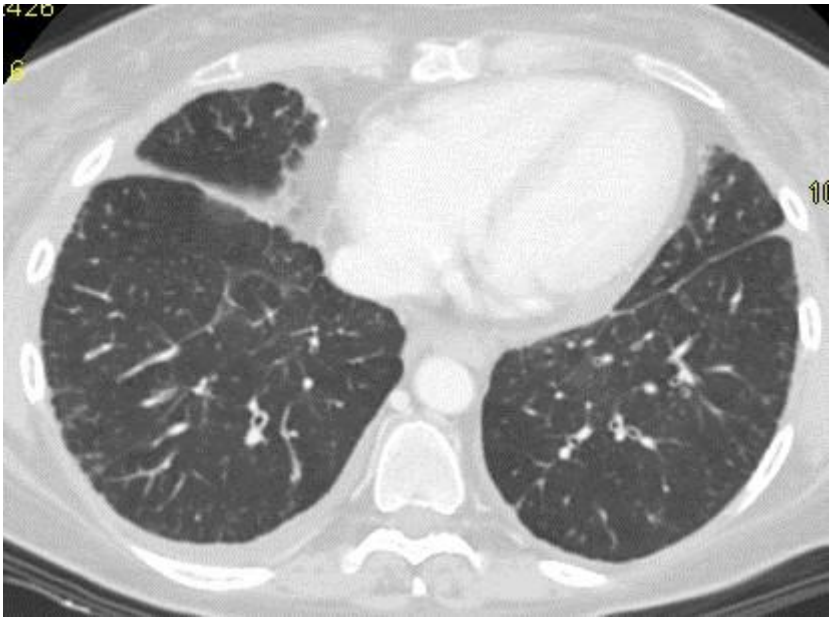

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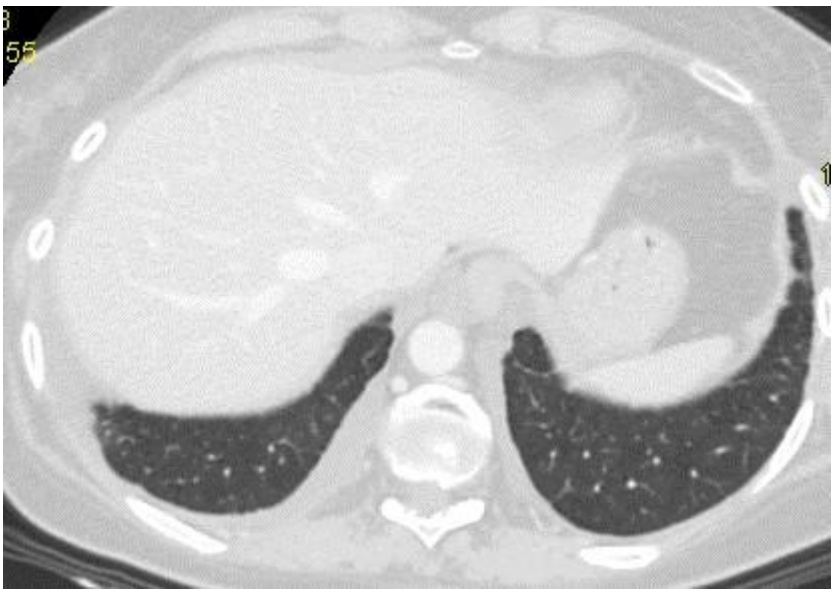

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Figure 3

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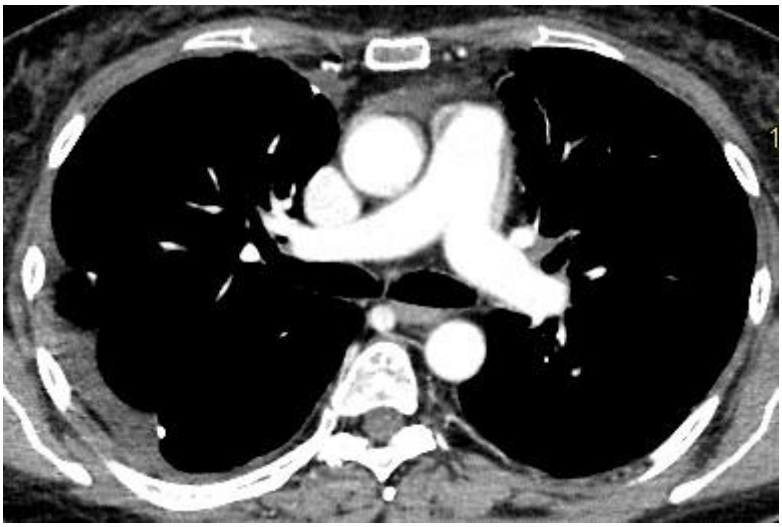

Description: Chest CT scan – with mediastinal window setting - shows post-surgical wedge resection on the right lung and bilateral pleural effusion. There are no mediastinal or hilar lymphadenopathies. Abdominal findings are unremarkable. **Origin:** G.Aquaro, Department of Radiology, Di Venere Hospital - Bari (BA) - Italy

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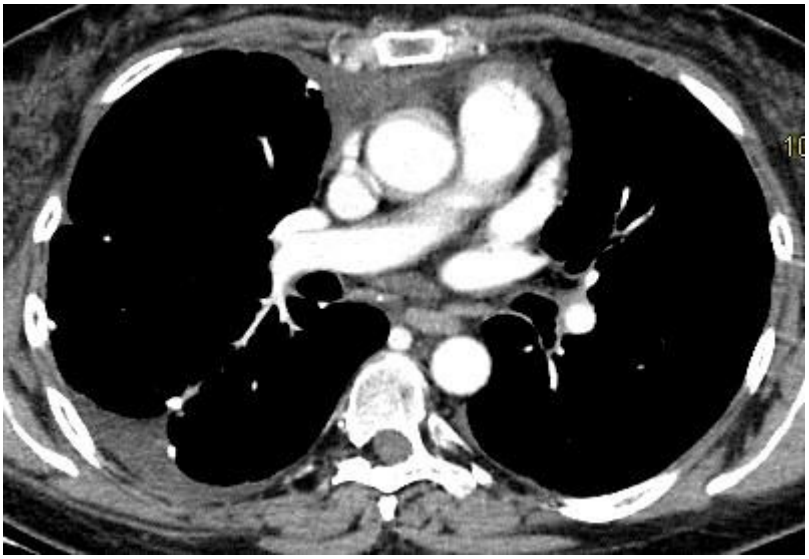

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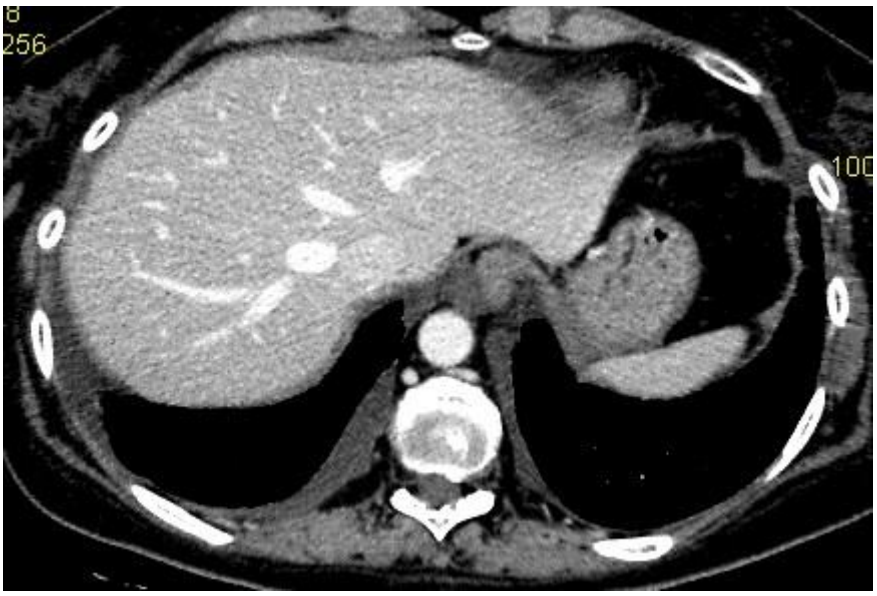

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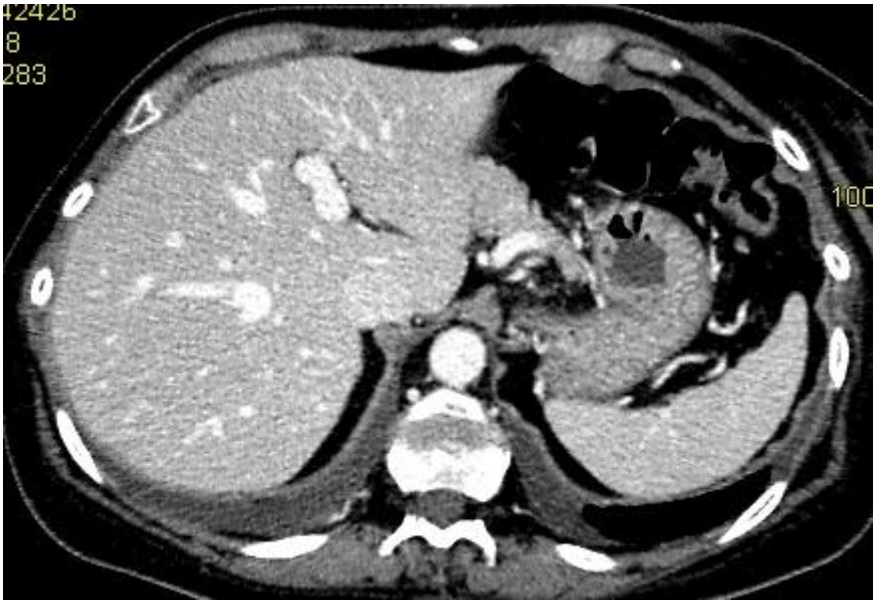

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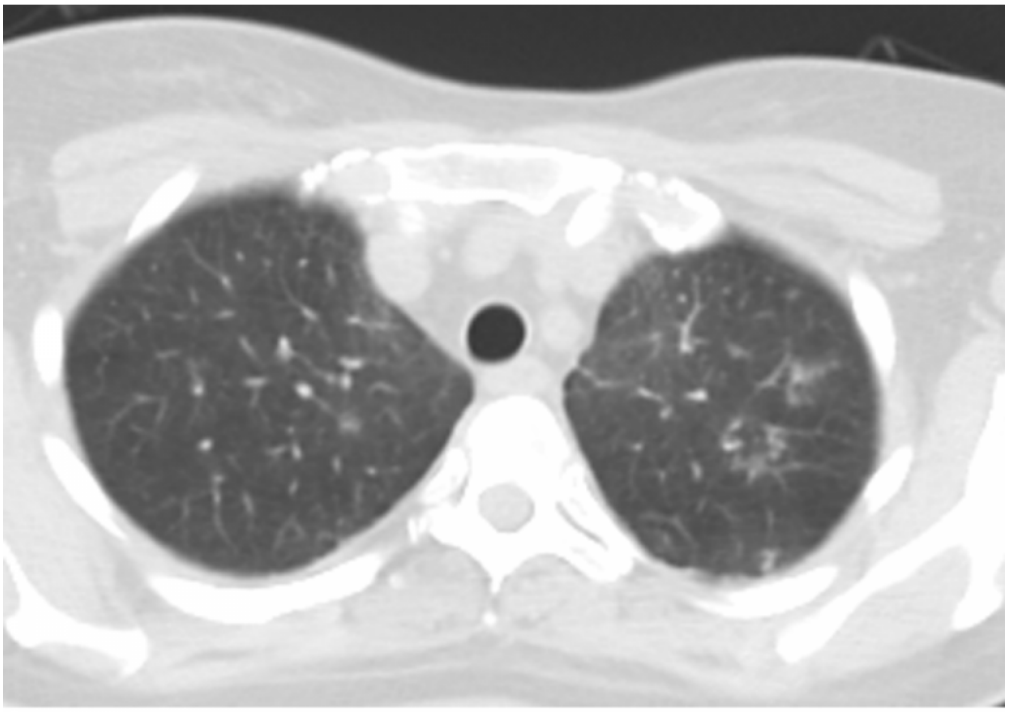

Description: Chest CT scan with lung window setting, requested without intravenous iodine contrast medium, shows multiple, random nodules of various size, lower lobe thickening of both bronchovascular bundles and interlobular septa. **Origin:** G.Aquaro, Department of Radiology, Di Venere Hospital - Bari (BA) – Italy

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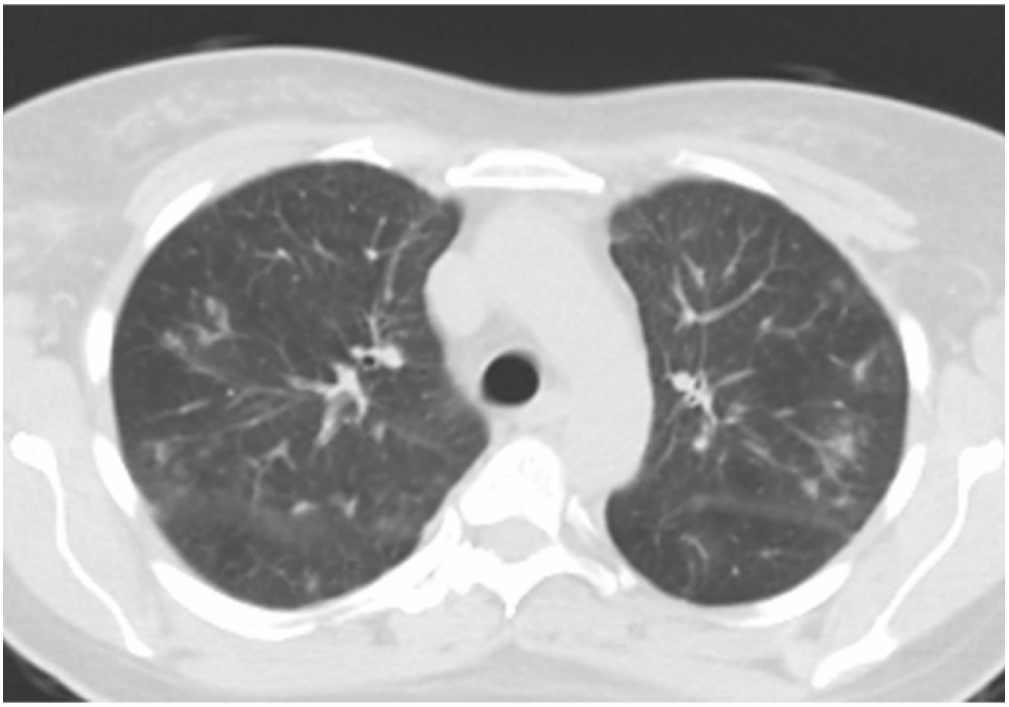

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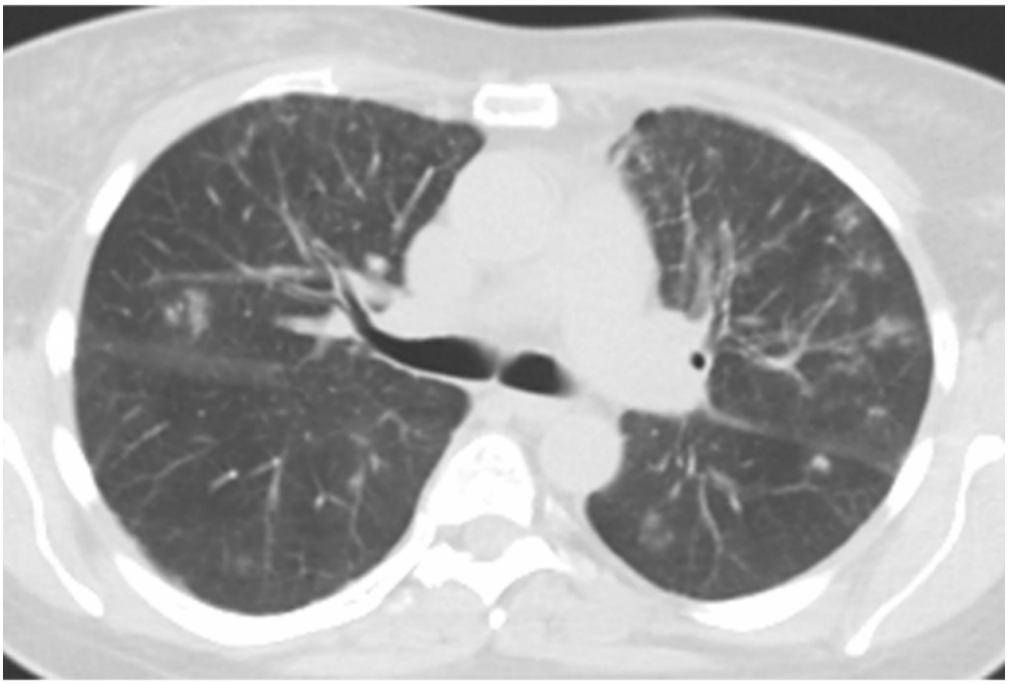

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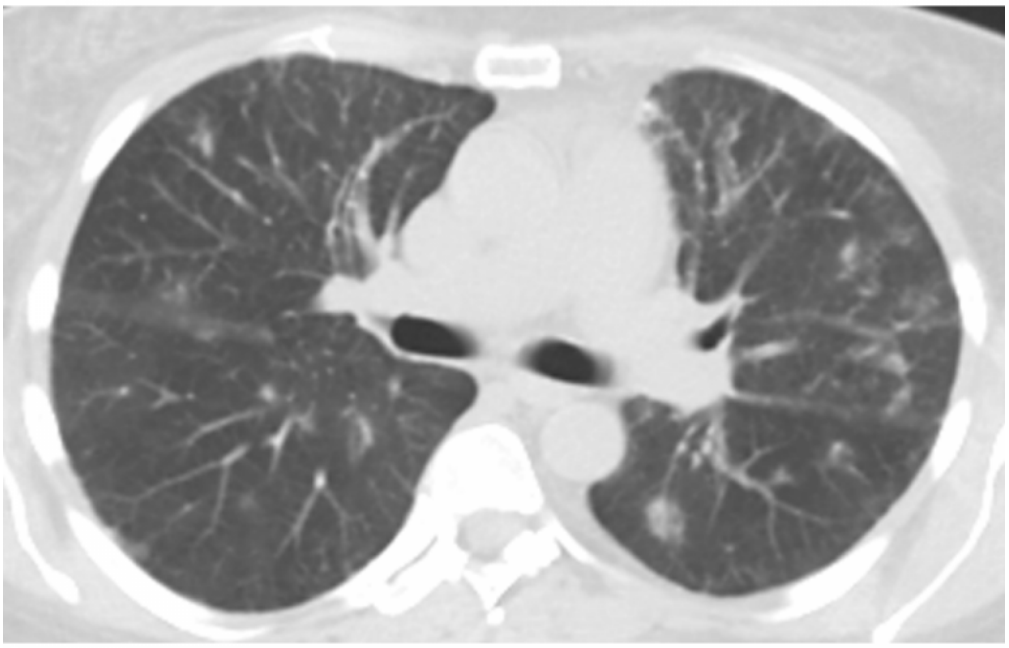

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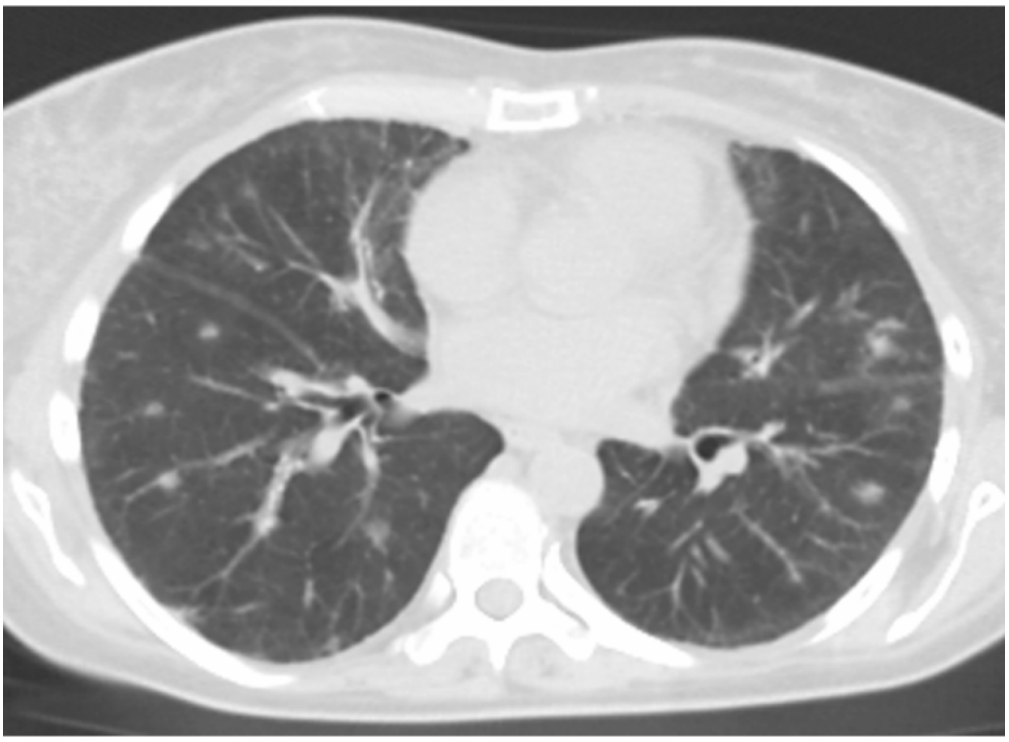

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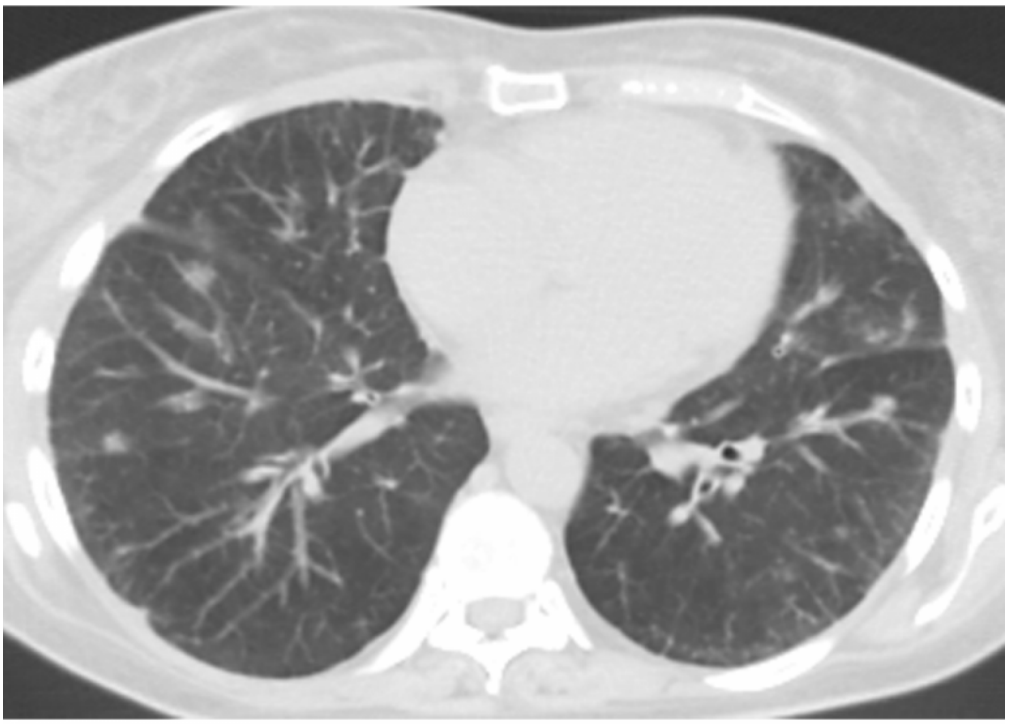

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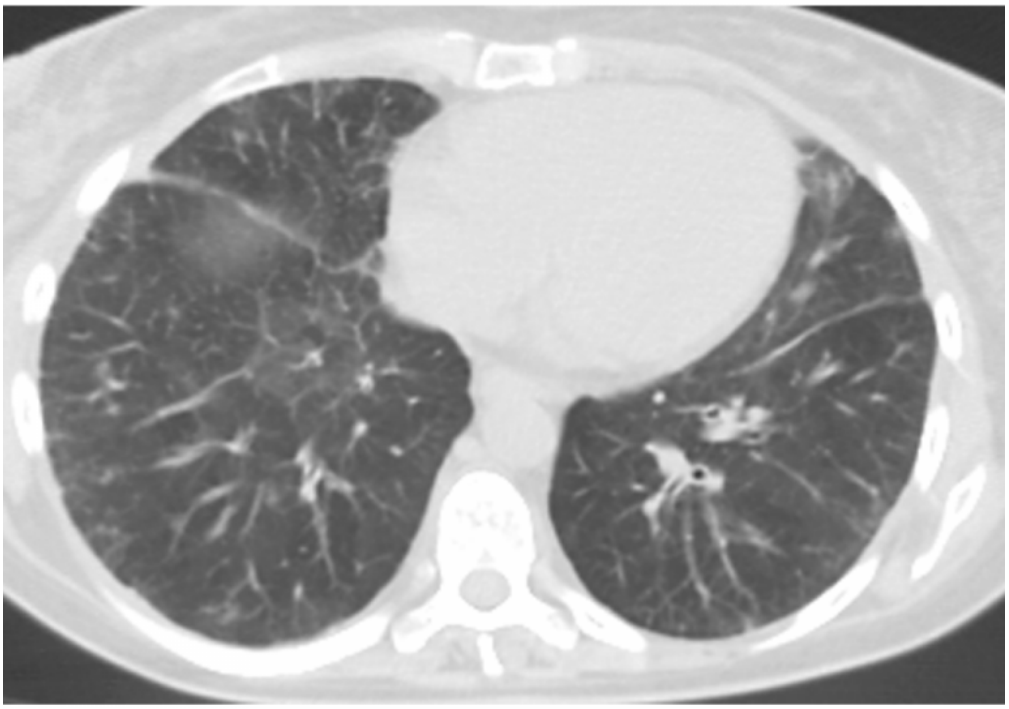

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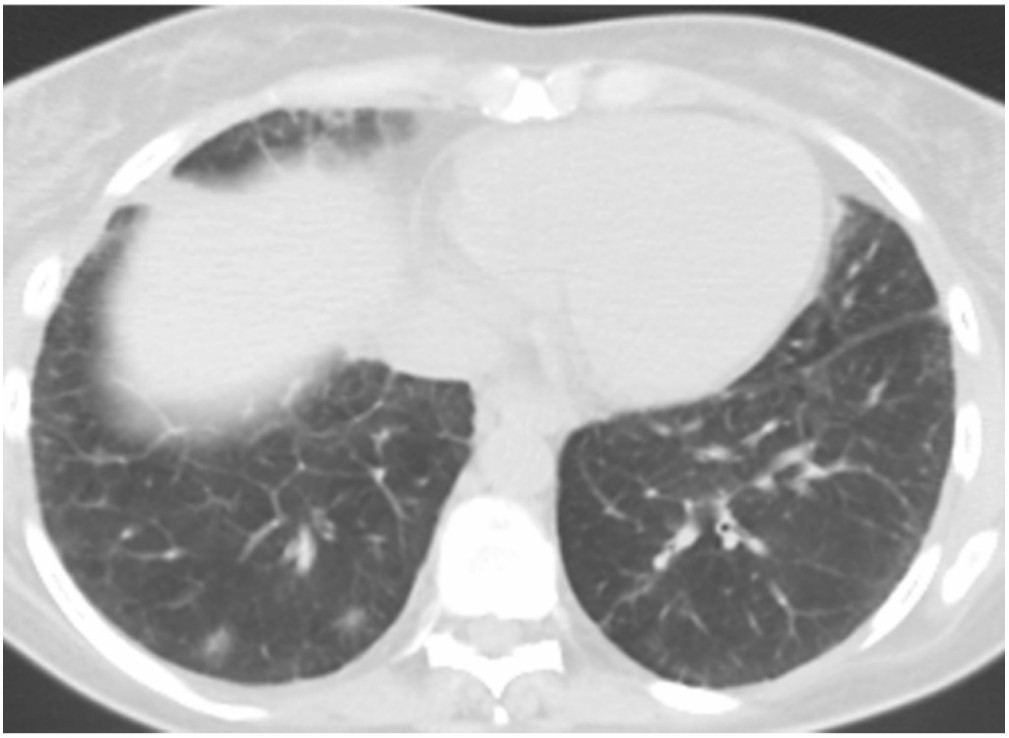

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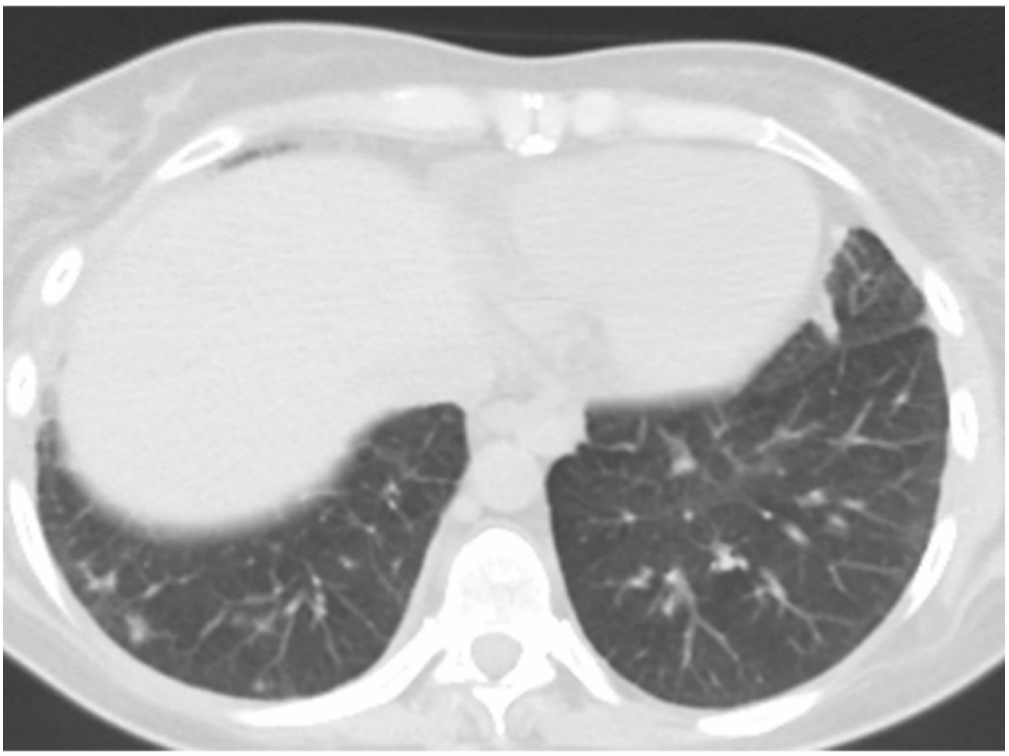

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