

Pulmonary inflammatory pseudotumour

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Section: Chest imaging

Area of Interest: Lung Thorax

Procedure: Diagnostic procedure

Imaging Technique: CT

Imaging Technique: Digital radiography

Imaging Technique: PET-CT

Special Focus: Pathology Case Type: Clinical Cases

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Patient: 62 years, female

Clinical History:

A 62-year-old female patient with a diagnosis of SAPHO was hospitalised at the rheumatology department. She presented persistent fever, cough, pain in the left hemithorax, enlarged palpable lymph nodes and weight loss.

Imaging Findings:

The chest radiograph showed multiple coarse nodules, with irregular margins in the apical and medial regions of the lungs.

A chest CT showed multiple nodular parenchymal consolidations, partially confluent with irregular margins. At the periphery of the consolidations, there were ground-glass opacities, while limited pleural thickening and areas of ground were localised in the basal region of the left lung.

Enlarged mediastinal lymph nodes were also present.

PET/CT showed evidence of glucid hyper-metabolism with some of the known pulmonary parenchymal nodularities, the greatest localised in the upper lobe of left lung (SUV max 4.5).

Given the unclear nature of the findings, the patient underwent an atypical superior left lobe resection in VATS. The histological sample showed a nodular mass characterised by a lymphoplasmacellular infiltration, eosinophils and myofibroblasts. An immunohistochemical staining revealed IgG4+ plasmacells (with 100 cells/high power field, IgG4+/IgG+ plasmacell > 40%).

Discussion:

The IgG4-related disease is a chronic fibrotic inflammatory disease [1] characterised by: an infiltration of lymphocytes in the tissue, IgG4-positive plasma cells, a high serum IgG4 concentration and fibrosis [3].

It may affect one or more organs, and the spectrum of symptoms depends on localisations and the grade of fibrosis [1].

The disease also may affect the lung and the pleura. The patient can present cough, haemoptysis, dyspnoea, fever and chest pain [2].

It is possible to recognise four types of pulmonary involvement [2, 5]:

- 1) Solid nodular/mass lesions, sometimes with spiculations;
- 2) Thickening of the bronchovascular bundles and interlobular septa;
- 3) Alveolar interstitial disease, with a diffuse reticular shadow and bronchiectasia;
- 4) Round-shaped, ground-glass opacities.

Plus mediastinal lymphadenopathy, and/or pleural involvement [1].

Comprehensive clinical diagnostic criteria for IgG4-RD are [6]:

(1) Clinical examination reveals a particular diffuse/localised swelling or mass in a single or in multiple organs.

(2) Elevated serum IgG4 concentrations (≥ 135 mg/dL).

(3) Histological examination presents:

(a) Fibrosis and marked lymphocyte and plasmacyte infiltration;

(b) Infiltration of IgG4-positive plasma cells: a ratio of IgG4/IgG positive cells greater than 40% and greater than 10 IgG4-positive plasma cells/HPF.

Definite: condition (1), condition (2) and condition (3) have to be satisfied.

Probable: condition (1) and condition (3) have to be satisfied.

Possible: condition (1) and condition (2) have to be satisfied.

IgG4RD has a very good response to corticosteroid therapy but relapses are common [1, 4, 5].

A lung needle biopsy revealed an inflammatory infiltration rich of lymphocytes.

A clear diagnosis was needed before an immunosuppressive treatment could be started. Consequently, the patient underwent an atypical superior left lobe resection in VATS. This approach was preferred to a lobectomy, because it is less demolitive in a context of multiple lesions. The histological sample showed a nodular mass characterised by a lymphoplasmacellular infiltration, eosinophils and myofibroblasts. An immunohistochemical staining revealed IgG4+ plasmacells (with 100 cells/high power field, IgG4+/IgG+ plasmacell > 40%).

Differential Diagnosis List: Pulmonary inflammatory pseudotumour IgG4-related disease., Adenocarcinoma or bronchioloalveolar carcinoma, Inflammatory-myofibroblastic tumour

Final Diagnosis: Pulmonary inflammatory pseudotumour IgG4-related disease.

References:

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Figure 1

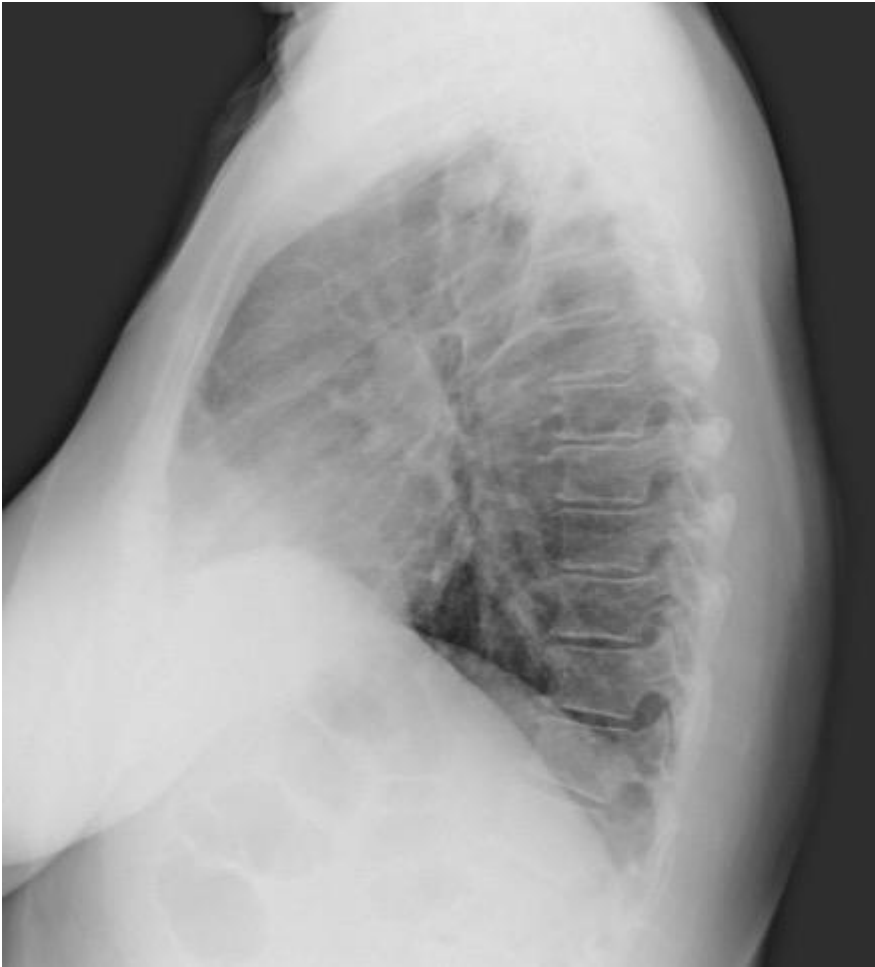
a



Description: PA and LL plain film shows nodular opacities in the upper lobe of the right and left lung

Origin: © Department of Radiology, UNIVERSITY OF PISA, Pisa, Italy

b

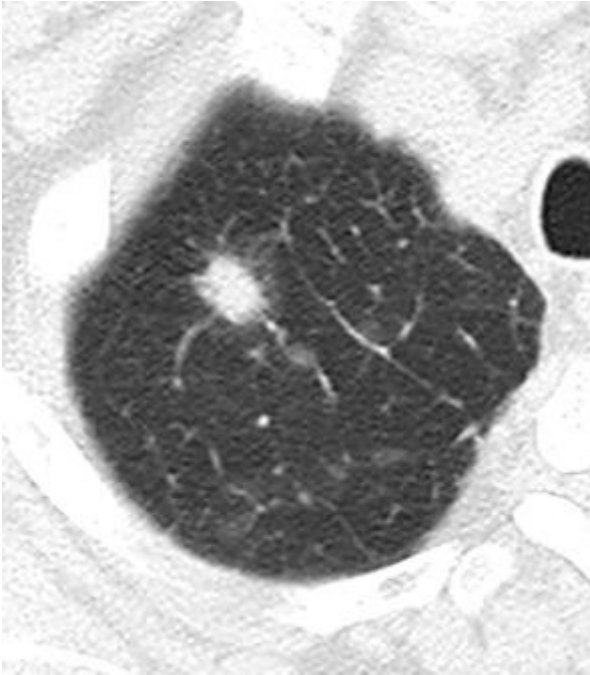


Description: PA and LL plain film shows nodular opacities in the upper lobe of the right and left lung

Origin: © Department of Radiology, UNIVERSITY OF PISA, Pisa, Italy

Figure 2

a

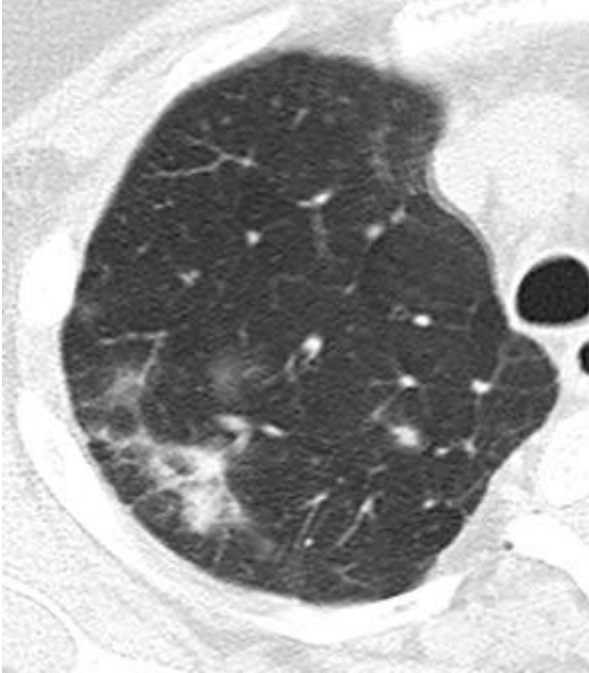


Description: CT image shows solid nodule in right upper lobe accompanied by peripheral ground glass opacity.

Diffuse pseudo-nodular area of ground glass **Origin:** © Department of Radiology, UNIVERSITY OF PISA, Pisa, Italy

Figure 3

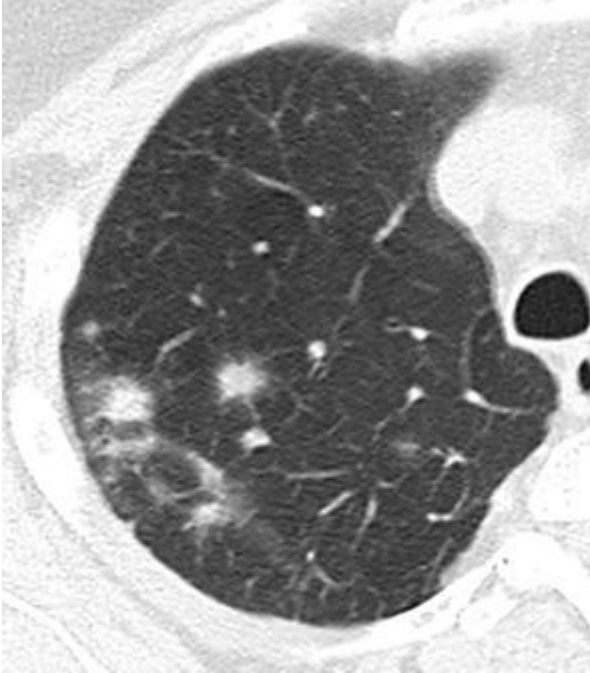
a



Description: CT image shows peribronchial spread of inflammation **Origin:** © Department of Radiology, UNIVERSITY OF PISA, Pisa, Italy

Figure 4

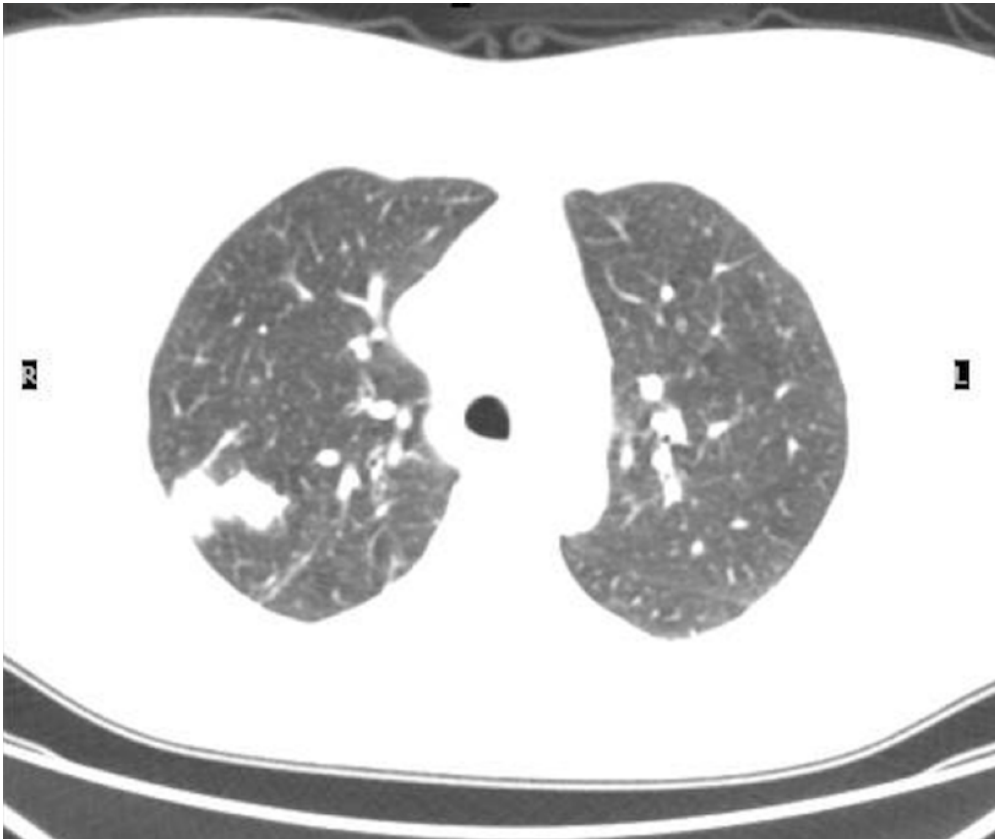
a



Description: Multiple small nodules, with non defined margin and ground glass opacities **Origin:**
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Figure 5

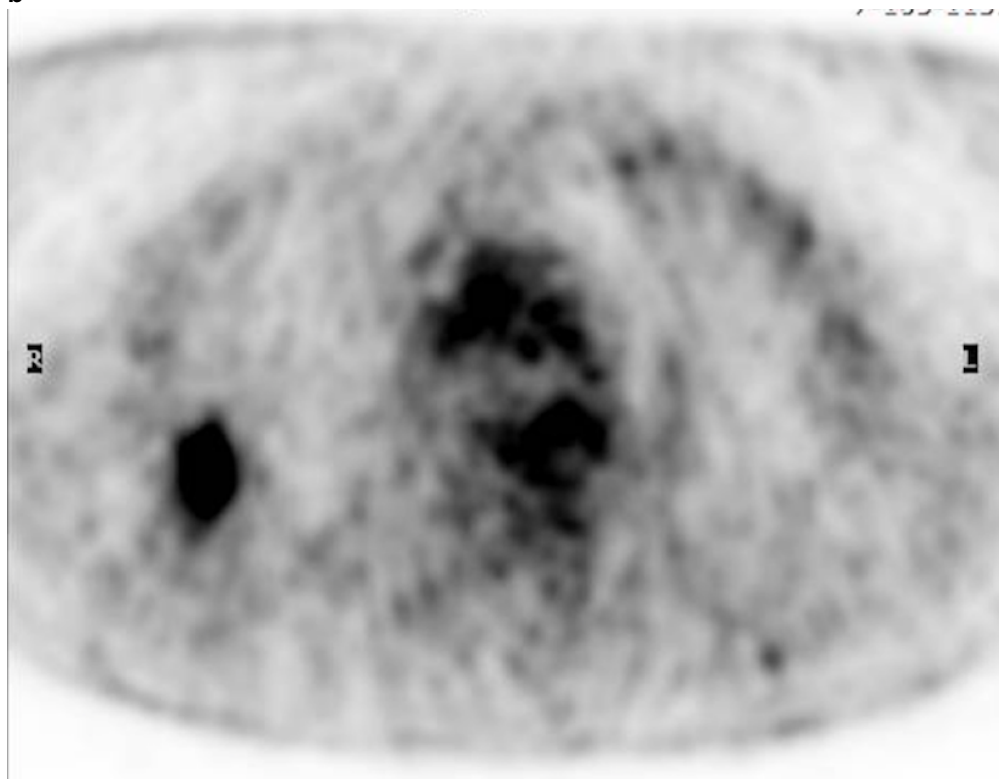
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Description: Images show increased FDG uptake in lung nodule of the upper lobe of the right lung

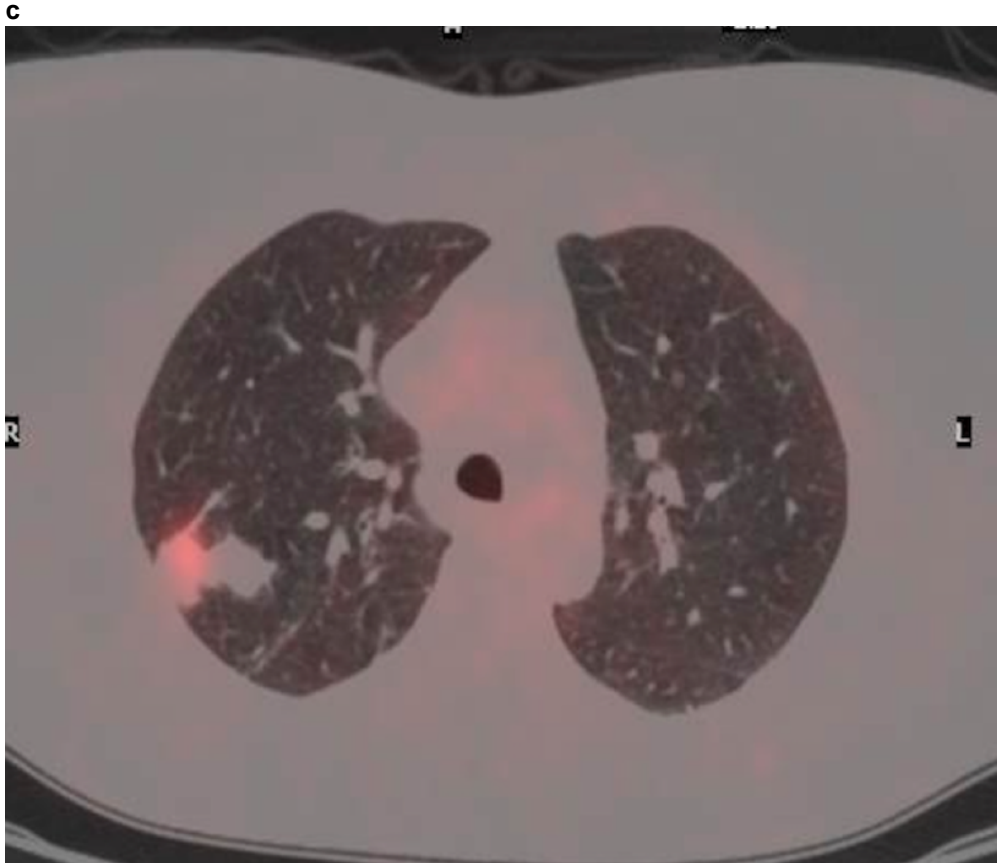
Origin: © Department of Nuclear Medicine, UNIVERSITY OF PISA, Pisa, Italy

b



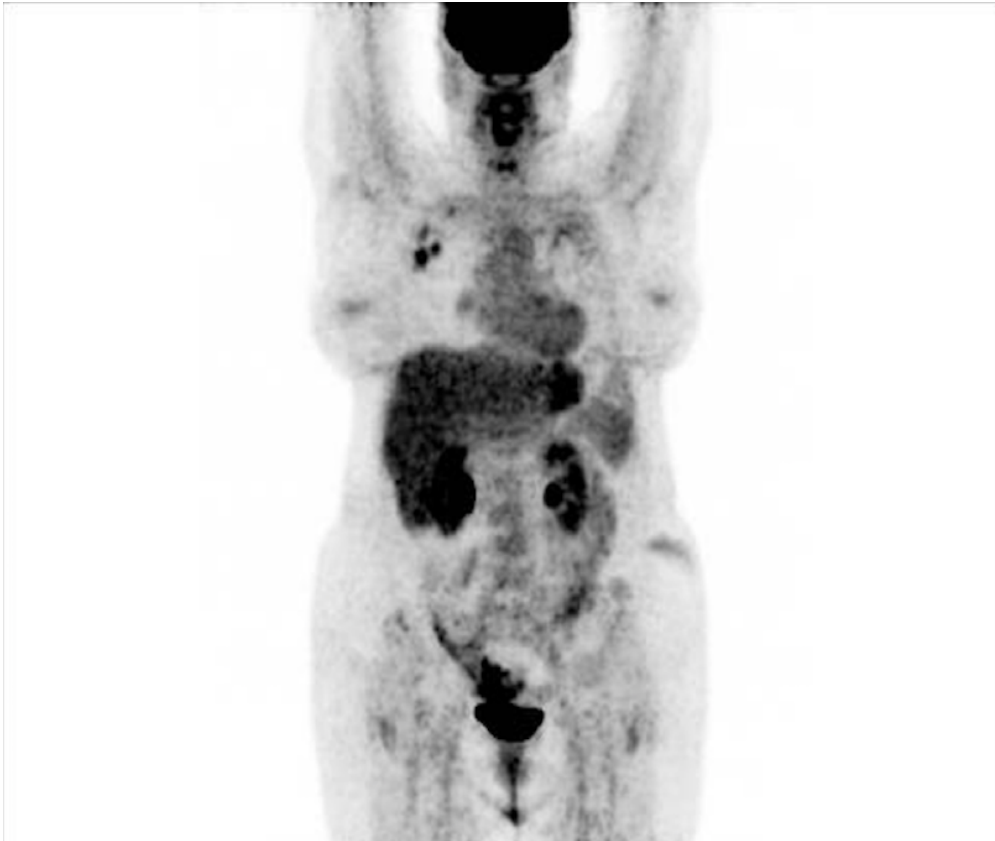
Description: Images show increased FDG uptake in lung nodule of the upper lobe of the right lung

Origin: © Department of Nuclear Medicine, UNIVERSITY OF PISA, Pisa, Italy



Description: Images show increased FDG uptake in lung nodule of the upper lobe of the right lung
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d



Description: Images show increased FDG uptake in lung nodule of the upper lobe of the right lung
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