Case 11988

Eurorad

Pulmonary amyloidosis

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ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Lung Thoracic wall Thorax
Procedure: Screening
Procedure: Contrast agent-intravenous
Procedure: Sampling
Imaging Technique: Conventional radiography
Imaging Technique: CT
Imaging Technique: PET-CT
Special Focus: Cavitation Case Type: Clinical Cases
Authors: Carpentier K, De Beuckeleer L, Van Mieghem F, De Foer B, Pouillon M, Tavares MDC.
Patient: 72 years, female

Clinical History:

A 72-year-old woman underwent chest X-ray in the preoperative setting of a total knee replacement (Fig. 1). For this patient there were no significant data regarding her medical history.

Imaging Findings:

As a coincidental finding, multiple pulmonary nodules were seen on the preoperative chest X-ray. A subsequent CT of the chest, performed after administration of intravenous contrast, confirmed the presence of multiple nodules in both lungs with a predominantly peripheral distribution (Fig. 2). Some of these nodules were cavitated. There was no perilymphatic or centrilobular distribution pattern. There were no signs of interstitial disease and there were no associated lymphadenopathies. In order to rule out malignancy she underwent a FDG-PET examination (Fig. 2) demonstrating FDG-uptake in these nodules (SUVmax 6.1). Because malignancy could not be excluded, a wedge resection of the right lower lobe was performed for histological sampling purposes. Staining with Congo red under crossed polarization light demonstrates a yellow-green birefringence confirming the diagnosis of amyloidosis (Fig. 3). The affinity of amyloid for Congo red did not alter after incubation with potassium permanganate, specifying this type as a non-AA type.

Discussion:

Amyloidosis represents a heterogeneous group of disorders characterized by overexpression of specific subunit proteins with an extracellular accumulation [1]. There are two forms of amyloidosis, the systemic and the organ-limited form. Besides the form in which amyloidosis can occur, different categories of amyloidosis are known in a classification system based on the different subunit proteins [2]:

- AL: Primary or immunoglobulin light-chain disease
- AA: Secondary or amyloid protein disease
- ATTR: Hereditary or mutant transthyretin disease
- A?2M: Dialysis-associated or ?2-microglobulin disease

Pulmonary involvement is most commonly found in the primary form (AL) of amyloidosis in which it can be a part of a widespread process involving many organs or in which it may be localized in the lung parenchyma [1]. Regardless

of the form, three distribution patterns of lung involvement are described: tracheobronchial, nodular parenchymal and diffuse alveolar septal distribution [3].

In the tracheobronchial distribution pattern, submucosal depositions of amyloid are found mainly in the trachea and main bronchi, most frequently located in the subglottic region. This form can be nodular or diffuse with narrowing of the lumen. Radiologic signs include nodular and irregular narrowing of airway lumen, airway wall thickening and calcified amyloid deposits [2].

The presence of nodular parenchymal involvement is most frequently found in the localized form of the disease [2, 4]. These nodules tend to be located at the periphery of the lung and are most often multifocal, well-defined and lobulated. They are variable in size ranging from 0.6 to 9 cm [2]. Large nodules may, as in our case, cavitate.

Diffuse parenchymal involvement is suspected when fine reticular or reticulonodular opacities are found. They may even be accompanied with alveolar infiltrates [2].

Cyst formation is rare and has been reported in associating with Sjögren syndrome accompanied with pulmonary nodules, parenchymal opacities and/or bronchiectasis [4]. Other rare manifestations of pulmonary amyloidosis are lymph node involvement [5] and pleural effusion (mostly due to cardiac amyloidosis)[2].

A chest X-ray normally yields sufficient information about the extent of the pulmonary involvement. A (high-resolution) CT is more sensitive in detecting tracheobronchial involvement and subtle parenchymal lesions. FDG-PET imaging has been used to exclude malignancy in cases of pulmonary nodules in which it has a higher specificity than CT imaging [6]. However, false positive results can occur due to infections, inflammatory disorders and, as in our case, amyloidosis [7]. This makes histologic sampling necessary in establishing a definite diagnosis.
Differential Diagnosis List: Localized pulmonary amyloidosis - nodular form, Cavitated lung metastases, Granulomatous disease, Rheumatoid nodules, Septic pulmonary embolism

Final Diagnosis: Localized pulmonary amyloidosis - nodular form

References:

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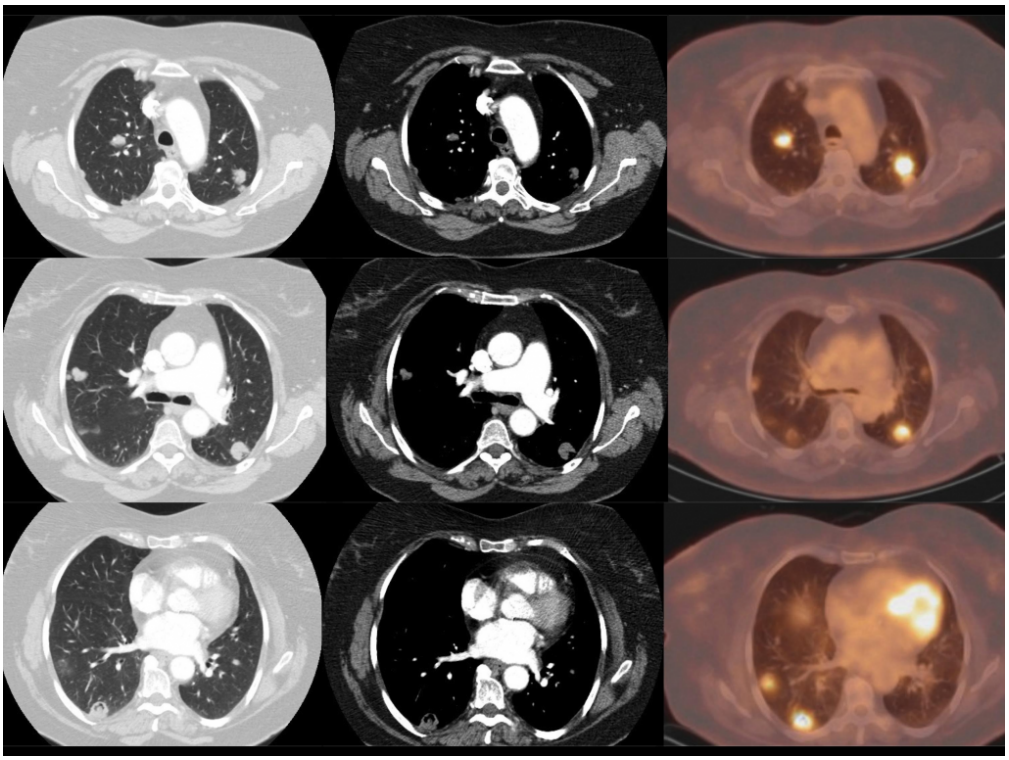
Figure 1

a



Description: Chest CT images showing multiple peripherally distributed nodules. Some are cavitated. Comparative PET shows FDG-uptake in these nodules. Since there are no pathognomonic radiographic features of pulmonary amyloidosis, a wedge resection was performed. **Origin:** Carpentier K, Department of Radiology, GZA Sint-Augustinus, Wilrijk, Belgium

Figure 2

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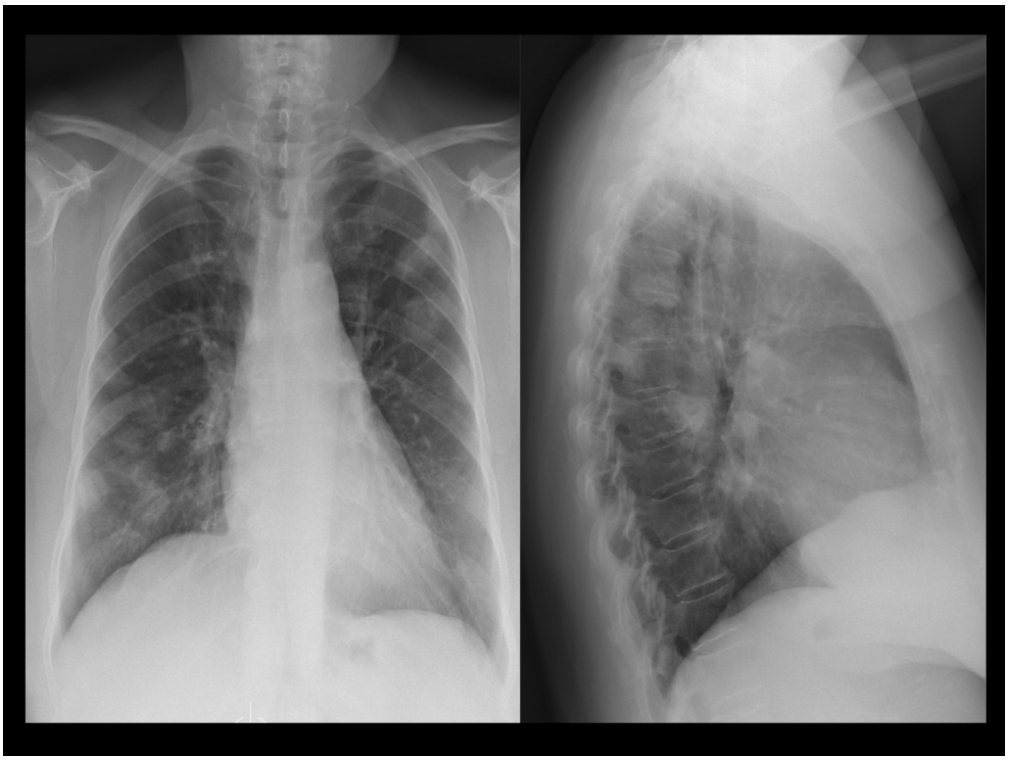

Description: Preoperative chest X-ray (AP and lateral view) showing multiple nodular opacities distributed throughout both lungs. **Origin:** Carpentier K, Department of Radiology, GZA Sint-Augustinus, Wilrijk, Belgium

Figure 3

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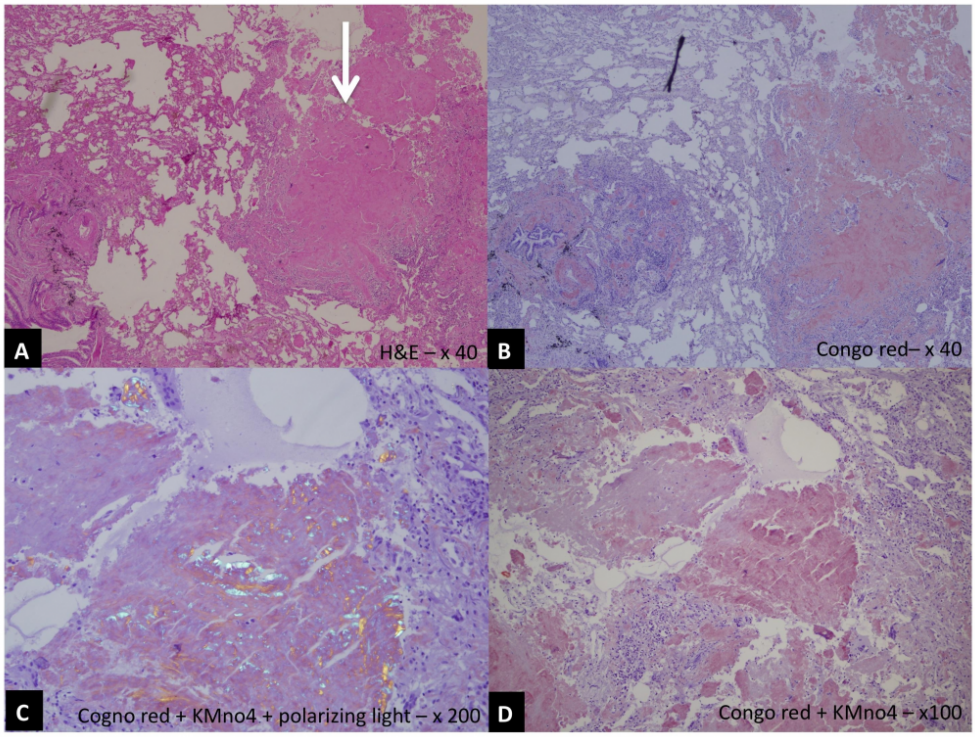

Description: H&E staining (A) shows an amorphous component (arrow) within the normal alveolar morphology. Congo red staining (B) under crossed polarization light (C) demonstrates a yellow-green birefringence. No effect of potassium permanganate (C-D). **Origin:** Carpentier K, Department of Radiology, GZA Sint-Augustinus, Antwerp, Belgium