

Case Report

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Pulmonary multinodular amyloidosis: A case report

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Abstract

Nodular pulmonary amyloidosis is rare in our environment with a low incidence. A 77-year-old female patient, who began her illness 20 days before hospital admission with sporadic non-productive cough without attack to the general condition. Computed tomography scan of the chest showed multiple nodular images of heterogeneous density, some of them hyperdense in calcific range, poorly defined borders, patchy in both hemithoraxes with confluence in basal regions. The histopathological study reported areas of amyloid with positive Congo red staining and upon exposure to polarized light, apple green birefringence was observed. Generally the presentation is asymptomatic, and the diagnosis is incidental, some clinical manifestations may be dyspnea, cough, hemoptysis. It is important to have a high degree of clinical suspicion to make timely diagnosis.

Keywords: Case report; Amyloidosis; Pulmonary multinodular.

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Introduction

Amyloidosis comprises a group of disorders characterized by deposition of abnormally folded proteins in tissues. The deposits are formed from soluble globular proteins, which misfold and aggregate into insoluble fibrils, causing progressive organ damage [1].

At the pulmonary level, these deposits envelop the interstitium, affecting gas exchange and presenting progressive interstitial lung disease [2].

In addition to protein production, there is a failure of the elimination mechanisms for this type of misfolded proteins. Amyloid deposits are metabolically inert, but physically interfere with organ structure and function; some prefibrillar oligomers of amyloidogenic proteins have direct cellular toxicity, being an important component of the pathogenesis of the disease [3].

Amyloid deposits can be localized (10%-20%) or systemic (80-90%). At pulmonary level there are 4 clinical forms: diffuse

alveolar septal, pulmonary nodule, bronchial tracheobronchial and pleural amyloid [4]. It predominates in the male gender. The most frequent symptoms are fatigue and weight loss; physical findings are not characteristic, being macroglossia one of the most specific signs [5].

In the USA, 1% of patients with chronic inflammatory conditions such as rheumatic disease, familiar mediterranean fever will develop secondary amyloidosis, while the incidence of primary amyloidosis is 4.5 per 100,000 inhabitants. In Mexico, pulmonary amyloidosis is rare and its incidence is unknown.

Case report

A 77-year-old female patient, who began her illness 20 days before hospital admission with sporadic non-productive cough without attack to the general condition, and she went for medical evaluation. Physical examination revealed pulmonary consolidation syndrome in the right hemithorax.

A Computed Tomography (CT) scan of the chest showed multiple nodular images of heterogeneous density, some of them

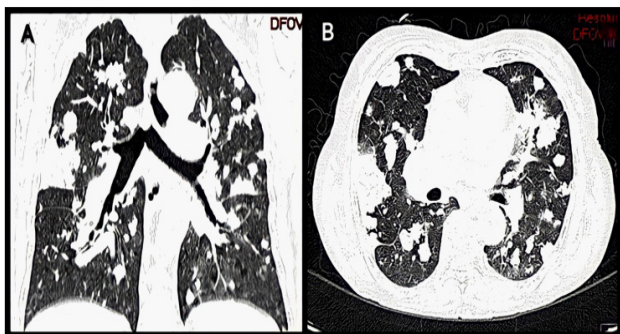


Figure 1: A) Coronal section: multiple bilateral irregular nodular lesions. B) Axial section: multinodular lesions, the largest in the right lower lobe.

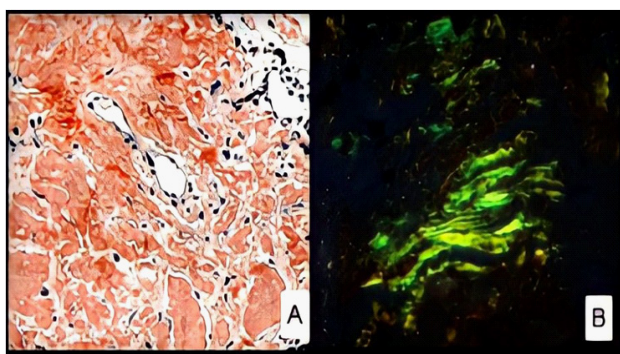


Figure 2: Microscopic photographs: A) Lung histological section at 10x with positive Congo Red staining, where abundant extracellular homogeneous material stained in “brick red” color is observed. B) Lung histological section exposed to polarized light where the apple-green birefringence is observed.

hyperdense in calcific range, poorly defined borders, patchy in both hemithoraxes with confluence in basal regions (Figure 1). A lung biopsy was performed by posterolateral thoracotomy. The histopathological study reported areas of amyloid with positive Congo red staining and upon exposure to polarized light, apple green birefringence was observed (Figure 2).

Laboratory results showed serum kappa levels of 3.3 mg/dl, lambda 26.3 mg/dl, kappa/lambda ratio of 0.125, respiratory function tests reported severe restrictive pattern with a vital capacity of 40%. In our patient there was no evidence of systemic amyloidosis.

Discussion

Nodular pulmonary amyloidosis is rare in our environment with a low incidence, frequent in men aged 50-60 years, with a 3:2 ratio and a history of chronic inflammatory conditions such as rheumatic disease, Sjögren’s syndrome, MALT lymphoma or multiple myeloma [6].

At pulmonary level it shows isolated lesions that can be confused with malignant processes. It usually presents as interstitial disease of tracheobronchial location, due to diffuse amyloid deposition in the alveolar septa with a reticular pattern on chest X-ray. Rarely, intraparenchymal lesions appear in a multinodular form, as in the present case.

Generally, the presentation is asymptomatic, and the diagnosis is incidental, some clinical manifestations may be dyspnea,

cough, hemoptysis [7]. In the chest X-ray peripheral nodules can be found that can be bilateral of variable size with cystic changes of slow growth. Contrast computed axial tomography is an ideal technique to establish the characteristics and location of the pulmonary nodule. In our case, chest CT showed multiple nodules in both hemithoraxes, with confluence in basal regions. Bronchoscopy is of little use in the diagnosis of peripherally located pulmonary nodules, although it can help in central lesions. Radiologically guided percutaneous fine needle puncture of a peripheral pulmonary nodule has high diagnostic capability and lower risk than a tracheobronchial puncture [8]. To establish a diagnosis of certainty it is essential that a biopsy be stained with Congo red and show apple-green birefringence in polarized light. Histopathological studies reveal eosinophilic density and aggregates of plasma cells and lymphocytes [9].

Systemic presentation requires chemotherapy treatment or autologous stem cell transplantation; treatment for localized forms is symptom oriented. Nodular localized amyloidosis usually progresses and shows recurrence after resection, there is no specific treatment [4,9]. Patients with nodular pulmonary amyloidosis have a good prognosis with timely diagnosis, unfortunately amyloidosis with myeloma has a poor prognosis, with a median life expectancy of 12 to 15 months after diagnosis.

The case presented is relevant for being female, absence of specific clinical data and no history of chronic inflammatory disease, presenting a benign evolution and adequate survival.

Declarations

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Statement of ethics: Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Ethical review board: The authors declare that they have followed the protocols of their work center on the publication patient data.

Right to privacy and informed consent: The authors have obtained the written informed consent from the patient for publication of this case report and any accompanying images. The author is in possession of this document.

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