CASE REPORT

Localised Nodular Pulmonary Amyloidosis in a Patient with Sicca Syndrome

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Summary

A 52 year old Chinese woman with a 25 year history of sicca syndrome (primary Sjogrens syndrome) was investigated for 3 episodes of haemoptysis. Clinical examination was unremarkable except for the presence of dry eyes and xerostomia. Computed tomography of the chest revealed a lobulated mass in the posterior basal segment of the left lower lobe. Histopathological examination of this resected nodule confirmed the diagnosis of nodular amyloidosis. The normal radiolabelled serum amyloid P component scintigraphy and the absence of monoclonal plasma cell dyscrasia in the bone marrow strongly support the diagnosis of localised nodular pulmonary AL amyloidosis in this patient. Nodular pulmonary amyloidosis can be associated with sicca syndrome and often simulates bronchogenic carcinoma, bronchiectasis or pulmonary tuberculosis.

Key Words: Sicca syndrome (primary Sjogrens syndrome), Haemoptysis, Localised nodular pulmonary amyloidosis

Introduction

Amyloidosis is characterised by extracellular deposition of the substance amyloid which appears as a typical amorphous eosinophilic extracellular material with green birefringence under polarising microscope after staining with congo red dye. The lower respiratory tract is unique because it can be involved in systemic amyloidosis or by localised nodular amyloid lesions[1]. We report a case of localised nodular pulmonary amyloidosis in a 52 year old women with primary Sjogrens syndrome (sicca syndrome).

Case Report

A 52 year old woman was referred to our Chest Clinic with a history of three episodes of haemoptysis over the previous week. She did not have any other respiratory or constitutional symptoms. She had been suffering from primary Sjogrens syndrome which was diagnosed 25 years earlier on the basis of the presence of keratoconjunctivitis sicca and xerostomia without evidence of a connective tissue disease. Findings on physical examination were remarkable only by the presence of dry eyes and xerostomia. Chest-X ray examination showed a round mass in the left lower lobe behind the heart shadow. Computed tomography of the chest revealed a lobulated mass measuring 3cm in diameter in the posterior basal segment of the left lower lobe and a smaller mass in the right middle lobe (Figure 1). The full blood count was normal with an ESR of 58mm/1hr. Blood biochemistry was normal. Serum and urine electrophoresis were essentially normal. Anti-nuclear antibody was positive at 1:2560 dilution (speckled), and the rheumatoid factor positive at 1:219. However, the complement levels were normal and double stranded DNA antibody was negative. She had polyclonal hyperglobulinaemia (IgG 26g/l, IgA 7.48g/l, IgM 0.8g/l). SSA (Ro) and SSB (La) (antibodies to extractable nuclear antigens) were positive. A

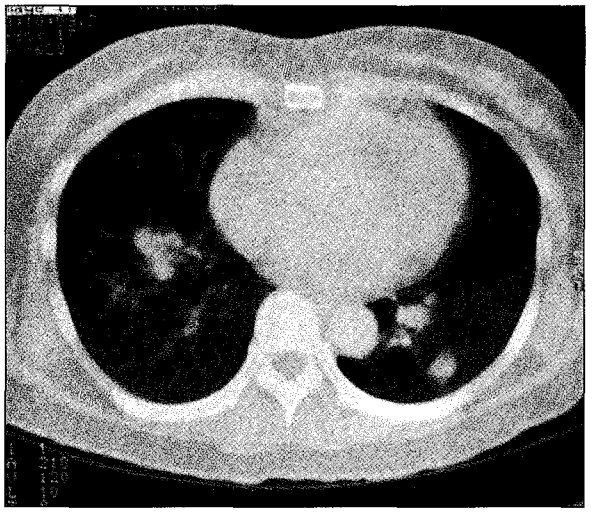

Fig.1: Computed tomography of the chest showing a lobulated mass in the left posteriorbasal segment of left lower lobe of the lung and a smaller mass in the right middle lobe.

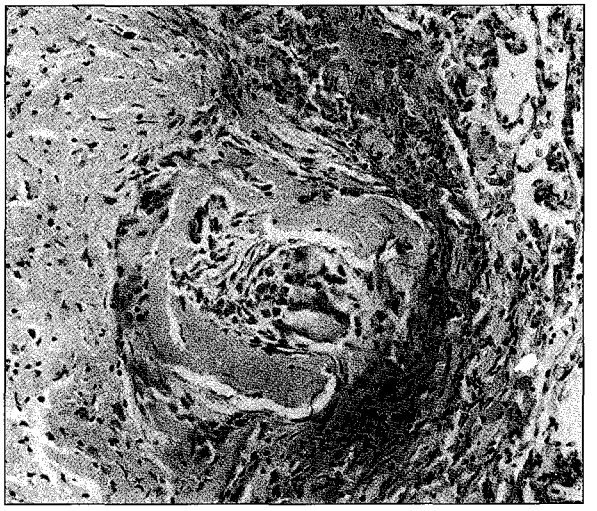

Fig.2: Histopathological section of the resected pulmonary nodule shows the lung tissue extensively replaced by masses of pink acellular amyloid deposits surrounded by plasma cells, foreign body giant cells and interpersed with lymphoid cells.

radiolabelled serum amyloid P component scintigraphy (SAP) showed a normal blood pool picture without any extra-pulmonary uptake of tracer. A bone marrow examination was normal and a minor labial gland biopsy showed glandular atrophy on histopathological examination.

Fibreoptic bronchoscopy did not reveal any endobronchial lesion and there was no evidence of bleeding at the time of bronchoscopy. Bronchial washings from the posterior basal segment of the left lower lobe were negative for acid fast bacilli and malignant cells. At thoracotomy through a median sternotomy, two nodules were palpated on the right middle lobe and one nodule each on the right and left lower lobes. The nodules which measured 1 to 2cm in their greatest diameters, were yellowish brown in colour and were hard to firm in inconsistency. The right middle lobe and the right lower lobe nodules were resected and submitted for histopathological examination. Multiple histopathological sections studied showed that the lung tissue was extensively replaced by masses of pink acellular amyloid deposits surrounded by plasma cells and foreign body giant cells (Figure 2). Lymphoid cells were interpersed. These deposits which stained red with Congo-red stain, and showed a yellow green birefringence under polarizing light, were at places perivascular in distribution. Some of them were surrounded by fibro-collagenous tissue and there were foci of calcification and ossification. The amyloid masses did not express immuno-cytochemical reactivity to amyloid A protein.

Discussion

The normal SAP scan and the absence of a monoclonal plasma cell dyscrasia in the bone marrow strongly support the diagnosis of localised nodular pulmonary AL amyloidosis in this patient. Localised pulmonary amyloidosis is defined as amyloid deposition isolated to the respiratory tract and does not include amyloidosis associated with systemic deposition (primary,secondary or familial)[2]. Localised pulmonary amyloidosis may involve the tracheobronchial tree or the pulmonary parenchyma in a nodular or diffuse distribution. In patients with tracheobronchial deposition, cough, dyspnea, wheezing and haemoptysis are common complaints. In contrast patients with nodular amyloidosis may have cough but dyspnea and haemoptysis are uncommon[1]. Haemoptysis is often caused by either amyloid infiltration into small blood vessels or compression of a bronchus by the nodular amyloid and resultant bronchiectasis[3]. Nodular pulmonary amyloid has a more benign natural history and a better prognosis than other forms of pulmonary amyloid[4]. They are frequently seen in older patients with an average age in the sixth decade. Although rare Sjogrens syndrome has been previously associated with nodular pulmonary amyloidosis of the lung[5].

Diagnosis may be made on the basis of transbronchial biopsy or transcutaneous needle aspiration. However surgical resection is often needed to obtain a satisfactory biopsy specimen. Thoracotomy and resection are unnecessary because many patients will show little or no progression when followed up for years.

At three years from the date of diagnosis our patient continues to remain well, with no chest symptoms and no radiological progression of the amyloidosis. Localised pulmonary amyloidosis is probably more common than is generally recognised. Pulmonary amyloidosis often simulates carcinoma, bronchiectasis and pulmonary tuberculosis.

As it can mimic various other diseases, it should be thought of and investigated for when in doubt regarding the diagnosis.

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