

A rare case of pulmonary amyloidosis with lymphocytic interstitial pneumonia in a patient with systemic lupus erythematosus

Sirs,

We report the case of a patient with systemic lupus erythematosus (SLE) presenting with concomitant nodular pulmonary amyloidosis (NPA) and lymphoid interstitial pneumonia (LIP). NPA is a rare condition associated with lymphomas and inflammatory conditions such as SLE (1-3). Lymphoid interstitial pneumonia (LIP) is usually associated with SS and is rarely reported in patients with SLE. The co-existence of pulmonary amyloidosis (PA) and LIP in the same SLE patient has not been reported before.

A 54-year-old woman who has been followed for SLE for 21 years presented with dyspnea and dry cough that had started 4 months earlier. The patient was diagnosed in 2006 with SLE with leucopenia, lymphopenia, positive anti-nuclear antibody (ANA), anti-double-stranded DNA antibody (anti-dsDNA), arthritis, and malar rash, with no major organ involvement. At the time of diagnosis a labial salivary gland biopsy was performed which resulted negative. 19 years ago, she suffered a second-trimester miscarriage at the 20th week of gestation. The antiphospholipid antibodies (aPL) were negative that time and she did not experience another miscarriage. She has been taking oral hydroxychloroquine 400 mg/day since her diagnosis with an uneventful disease course and was considered to be in remission. Her laboratory showed positive ANA at 1/320 titre with a homogeneous pattern, extracted nuclear antigen (ENA) results were anti-histone (+), anti-SSA (++) , complement 3 (C3): 0.58 g/dl (0.9-1.8), complement 4 (C4): 0.087 g/dl (0.1-0.4), anti-dsDNA: 418 IU/ml (<20), erythrocyte sedimentation rate (ESR):25 mm/h (0-18), C-reactive protein (CRP): 8 mg/dl (0-5). Her CXR showed a nodular opacity in the posterior segment of the left upper lobe (Fig. 1). Her thoracic CT with contrast revealed paraseptal emphysema in both lungs, more prominent in the upper lobes; several air cysts compatible with lymphoid interstitial pneumonia (LIP) (Fig. 2); and an irregularly contoured nodular lesion measuring 18 mm in the posterior segment of the left upper lobe (Fig. 3), two nodular subpleural opacities in the lingula, adjacent to the mediastinum the largest measuring 12 mm, consistent with amyloid foci (Fig. 4). After consultation with the Department of Chest Diseases, a PET/CT was performed for presumed malignancy. PET/CT showed mildly increased FDG uptake in the lesion, with a standard uptake value (SUV) max of 2.55 (Fig. 5). Pulmonary function tests (PFTs) were performed before bronchoscopy to assess respiratory

compromise, but spirometry and lung volumes were unaffected (Fig. 6). A bronchoscopy showed all lobar and bronchial orifices were patent with normal-appearing mucosa (Fig. 7). Cytological results from bronchoalveolar lavage were benign. A biopsy was deemed appropriate and demonstrated amyloid deposition on histochemical examination with Congo red and crystal violet staining (Fig. 8 and 9). The Department of Cardiology was consulted regarding cardiac amyloidosis, which resulted in a normal transthoracic echocardiogram (TTE) with an ejection fraction (EF) of 60% and no pulmonary hypertension. Serum and urine protein electrophoresis, immunofixation electrophoresis, serum free light chain assay, bone marrow examination, and skeletal imaging were negative. Systemic amyloidosis was excluded following analysis of abdominal subcutaneous fat, which showed no binding to Congo red, and no green birefringence was seen under polarised light. The Division of Nephrology was consulted regarding a renal biopsy, but it was deemed unnecessary. Oral methylprednisolone 0.5 mg/kg/day, oral azathioprine 150 mg/day, and oral colchicine 1 mg/day were initiated, resulting in symptom improvement.

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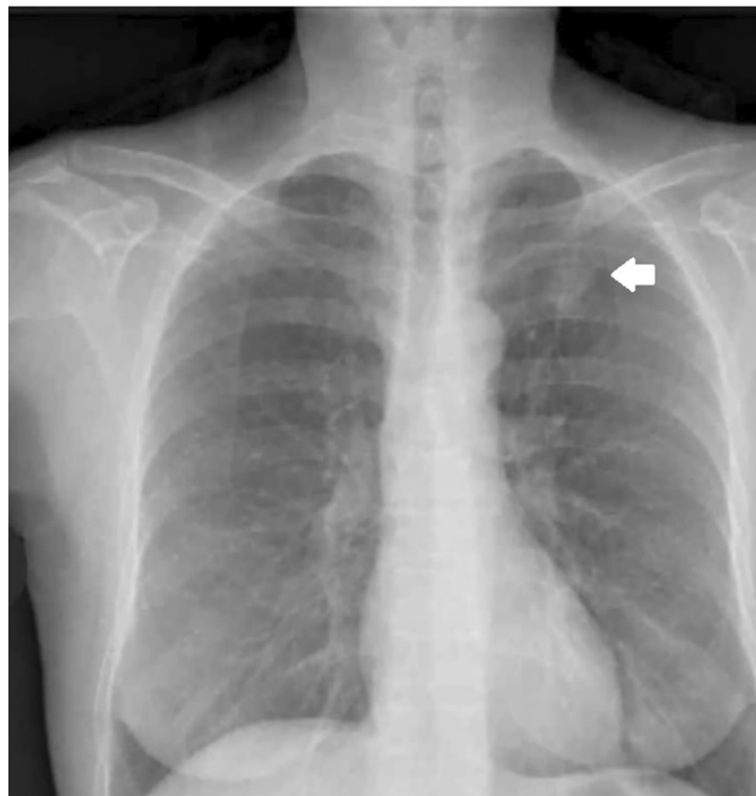


Fig. 1. Nodular opacity in the posterior segment of the left upper lobe (white arrow).

Letters to the Editors



Fig. 2. Irregularly contoured nodular lesion measuring 18 mm in the posterior segment of the left upper lobe (white arrow).

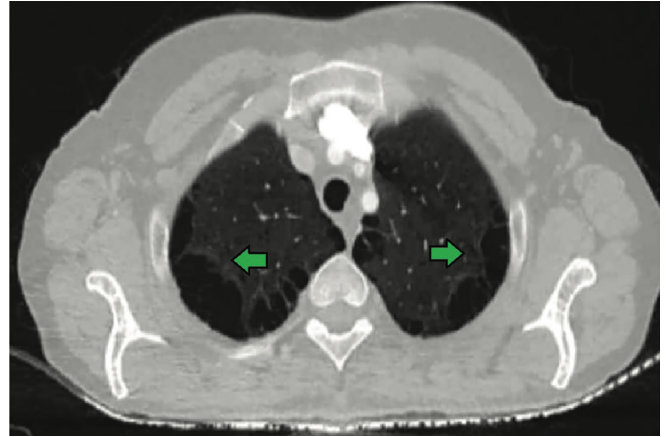


Fig. 3. Several air cysts are compatible with LIP (green arrow).

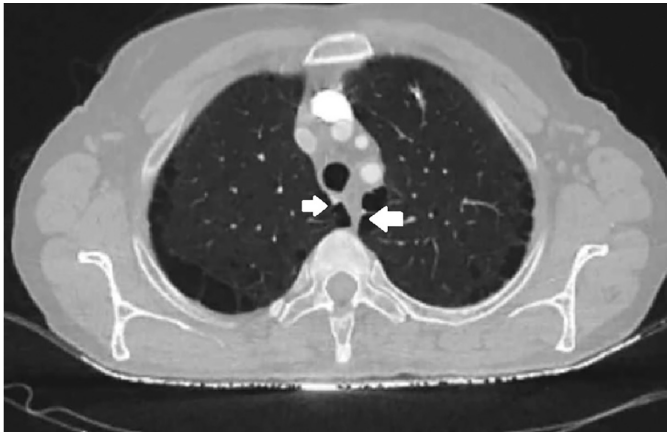


Fig. 4. Two amyloid foci in lingula (white arrows).

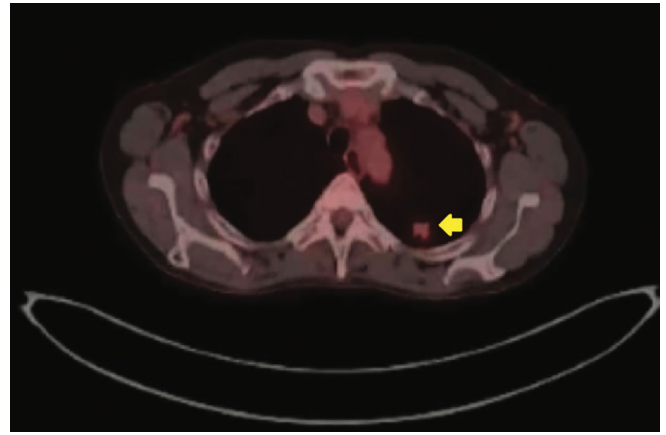


Fig. 5. A mildly increased FDG uptake was observed in an irregularly margined, spiculated mass.

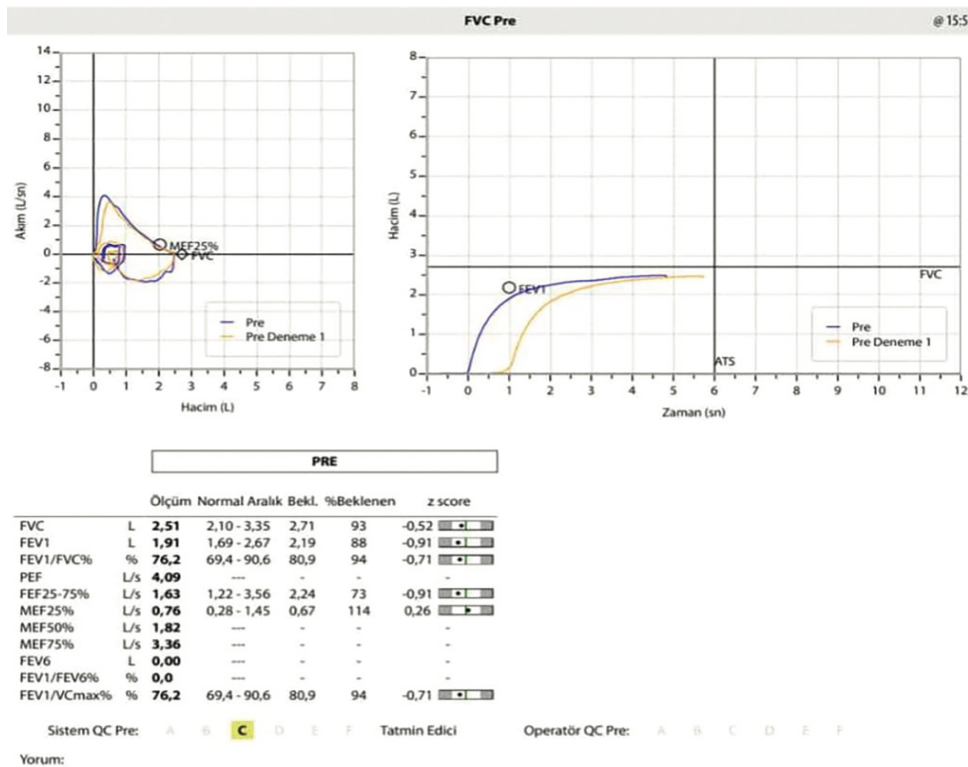


Fig. 6. Normal PFTs of the patient.

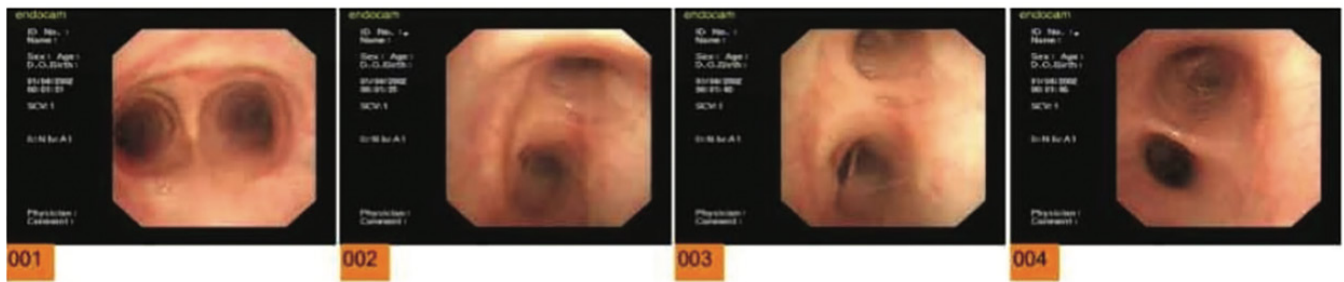


Fig. 7. All lobar and bronchial orifices are patent with normal-appearing mucosa.

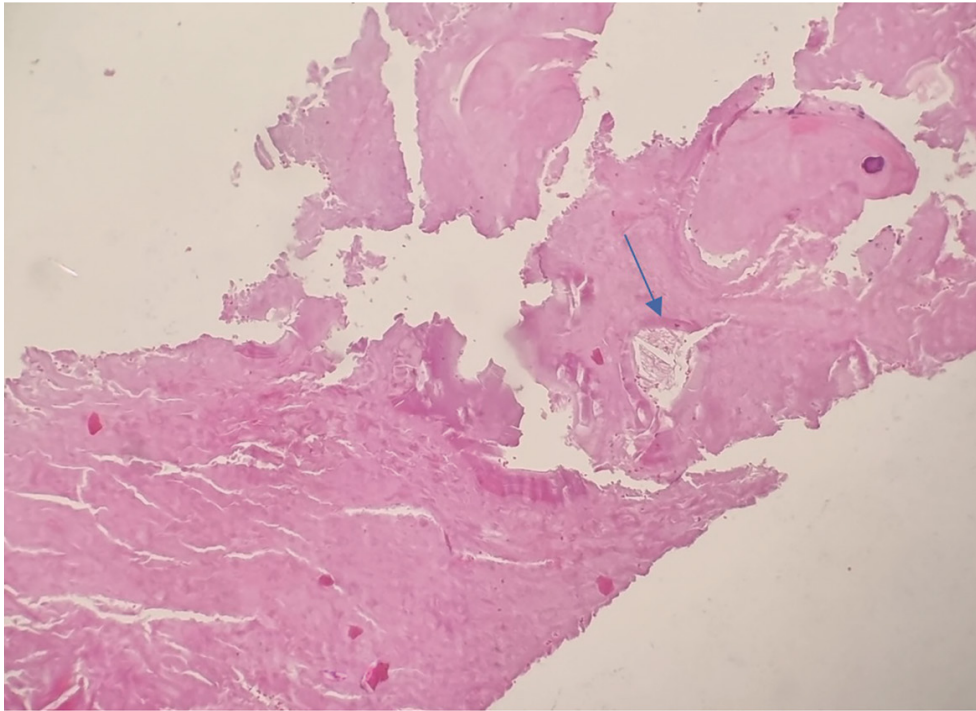


Fig. 8. Accumulation of acellular proteinaceous material with a hyalinised appearance (blue arrow).

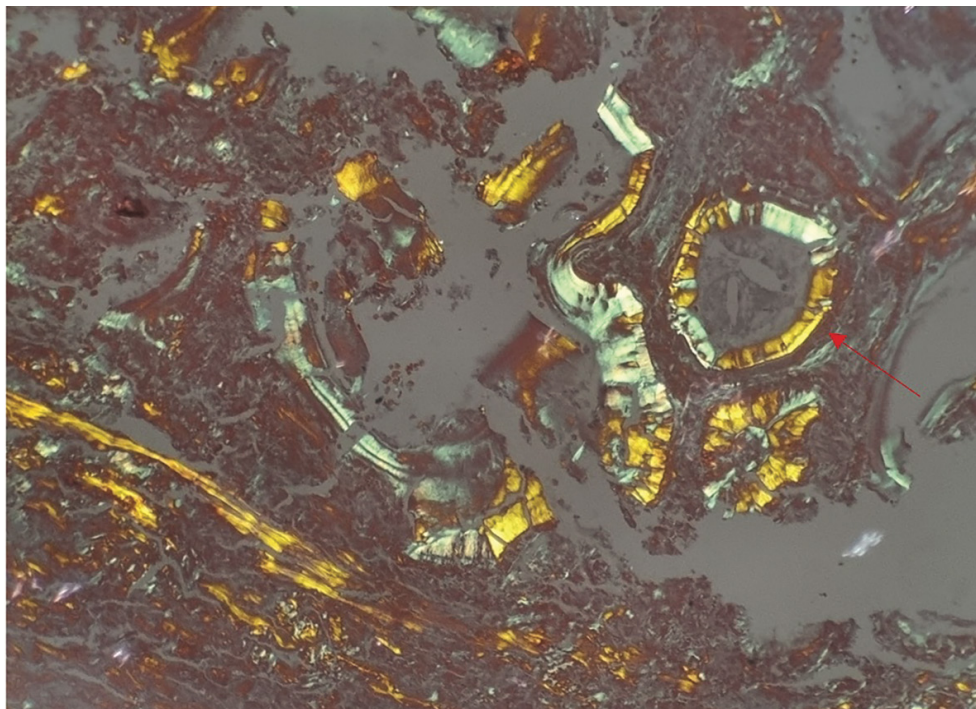


Fig. 9. Congo red staining shows apple-green birefringence under polarised light (red arrow).