Pulmonary fibrosis and lymphocytic alveolitis associated with triple antiphospholipid antibody positivity: a diagnostic puzzle

Sirs.

To our knowledge, only two cases of fibrosing alveolitis associated with antiphospholipid antibody syndrome have been described so far (1, 2). Pulmonary fibrosis is a clinical, pathophysiological entity difficult to characterise that includes a heterogeneous group of disorders all causing the progressive destruction of lung architecture, leading to respiratory failure (3). On the other hand, primary antiphospholipid syndrome (PAPS) identifies a well defined condition at increased risk of vascular occlusion and/or pregnancy complications (4). In the spectrum of pulmonary diseases that may occur in PAPS (5), the most common are thromboembolism and pulmonary hypertension, while isolated fibrosing alveolitis is an extremely rare presentation of PAPS (1, 2).

We report the case of a 74-year-old woman, former smoker, hospitalised for worsening dyspnea, non-productive cough in the absence of fever. Her past medical history was notable for obesity (BMI 32), dyslipidemia, arterial hypertension and a triple positive history for clear thrombotic events or pregnancy complications (4). The physical examination revealed crackles of DLCO. The six minute walking test (6MWT) was interrupted after 220 meters. Association between PAPS and interstitial lung disease is a still debated entity (5).

Spontaneous pneumothorax in a patient with known PAPS (6MWT) was interrupted after 220 meters.

Association between PAPS and interstitial lung disease is a still debated entity (5). Negative history for clear thrombotic events or pregnancy morbidity and the presence of a CD4+ T cells alveolitis led us to diagnose an interstitial lung disease with antiphospholipid antibody positivity. Furthermore high levels of TGF-beta found in BAL could be related to diffuse lung fibrosis. We decided to treat fibrosing alveolitis with methylprednisolone pulse therapy (125 mg/die) followed by azathioprine (75 mg/die) combined with a maintenance dose of prednisone; due to the triple aPL positivity associated with cardiovascular risk factors we started also a primary prophylaxis with low dose acetylsalicylic acid (100 mg/die). After one month of therapy the patient respiratory condition was improved, arterial blood gas analysis showed an increased PaO2 (74 mmHg) and a normal PaCO2 (36 mmHg); a new 6MWT was interrupted after 320 meters.

In conclusion, association of fibrosing alveolitis and antiphospholipid antibodies may simply reflect a stochastic coexistence of two different diseases at the same time or the antibodies may have had a role in the evolution of this particular condition. We describe a case with uncertain results, where perfusion defects are not easily ascribable to fibrosis or microthrombosis; in the light of this rare association, and recent findings of an inflammation role of antiphospholipid antibodies (6, 7), could it be worthy to start a retrospective study on fibrosing alveolitis searching for antiphospholipid antibodies.

References