Pulmonary Alveolar Proteinosis in a Patient with Systemic Lupus Erythematosus

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Secondary pulmonary alveolar proteinosis (PAP) associated with treatment is relatively frequent^{1,2}. However, PAP associated with autoimmune diseases is extremely rare; we have found only 2 cases reported in the literature^{1,3}.

We present a 34-year-old woman diagnosed with systemic lupus erythematosus (SLE) in 2001 based on rash malar, arthritis, leukopenia, positive antinuclear antibody, positive anti-DNA, and hypocomplementemia. She had received immunosuppressive drugs (methotrexate, belimumab, mycophenolate, and leflunomide).

She was admitted to a hospital in September 2014 for the study of respiratory insufficiency. An interstitial pattern could be observed in the thorax radiography and it was interpreted as lupus pneumonitis. She received treatment with methylprednisolone bolus and rituximab.

The patient's disease course declined and in December 2014 she was referred to our hospital to assess the need for lung transplant. Serological and hematological activity of SLE was discarded. Thoracic computerized tomography showed a crazy-paving pattern (Figure 1). After ruling out lymphoma and infections (including pneumocystis), our main diagnostic suspicion was PAP.

A bronchoscopy with transbronchial biopsy showed a

liquid of milky appearance. Histologic studies revealed an amorphous material by positive periodic acid-Schiff staining in the alveolar space compatible with PAP (Figure 2). Serum granulocyte-macrophage colony-stimulating factor antibody was negative.

According to our results, this patient presents with secondary PAP. Treatment of PAP is whole lung lavage to remove the accumulation surfactant on the air space⁴. With this therapeutic approach, our patient presented clinical and radiological improvement. She now has good quality of life, and a lung transplant was not necessary.

REFERENCES

1. Shah SK, Phan NB, Goyal G, Sharma G. Pulmonary alveolar proteinosis in a 67-year-old woman with Wegener's granulomatosis. *J Gen Intern Med* 2010;10:1105–8.
2. Wardwell NR, Miller R, Ware LB. Pulmonary alveolar proteinosis associated with a disease-modifying antirheumatoid arthritis drug. *Respirology* 2006;5:663–5.
3. Compa DR, Judson MA, Beegle SH. Granulomatosis and polyangitis followed by alveolar proteinosis in a 32-year-old woman. *Chest* 2012;5:1359–60.
4. Seymour JF, Presneill JJ. Pulmonary alveolar proteinosis: progress in the first 44 years. *Am J Respir Crit Care Med* 2002;166:215–35.

Figure 1. Extensive areas of ground-glass opacity are associated with thickening of interlobular septa, diffuse distribution, and a mosaic pattern.

Figure 2. Transbronchial biopsy: alveolar spaces occupied with a PAS-positive amorphous acellular material. PAS: periodic acid-Schiff.