Sjögren's syndrome and pulmonary alveolar proteinosis co-existence

Sirs,

Sjögren's syndrome (SS) is a systemic autoimmune disease characterised by lymphocytic infiltration of the exocrine glands and by a wide number of systemic manifestations (1). SS is classified as primary SS (pSS) when alone or as secondary SS (sSS) when accompanied by another systemic autoimmune disease (2). The pulmonary manifestations of SS are polymorphic. It can affect all compartments of the lung, including the lung parenchyma, airways, vascular structures, and pleura (3).

Pulmonary alveolar proteinosis (PAP) is a syndrome characterised by surfactant accumulation in alveoli and resulting in hypoxemic respiratory failure/insufficiency. According to its aetiology, three different clinical forms have been defined as primary, secondary and congenital (4).

The high aetiological diversity in PAP and the increased levels of lipid (5) and protein (6), which are the subunits of surfactant, suggest that its aetiopathogenesis may be variable. In the aetiology of secondary PAP, haematological disorders, infectious diseases, autoimmune diseases, post-organ transplantation and non-haematological malignancies have been identified. In a study examining patients with secondary PAP, autoimmune diseases Behçet's disease, Wegener's granulomatosis and microscopic polyangiitis were reported (7). Case studies and literature review involving 164 patients with secondary PAP using the PubMed database showed the rate of autoimmune disease to be 6.7% (8).

There are several case reports of the coexistence of PAP and SS (8-10). A 61-year-old female patient who presented with chest pain that had persisted for the past two years had peripheral, diffuse, subpleural ground-glass opacity on high-resolution computed tomography (HRCT) (Fig. 1). SS was diagnosed by salivary gland biopsy and PAP was diagnosed by open lung biopsy.

Diseases that cause radiological ground glass opacity in patients with pSS, cellular non-specific interstitial pneumonia, lymphocytic interstitial pneumonia, lymphoma, drug-induced lung disease, infection, hypersensitivity pneumonitis (11). We think that PAP disease should also be kept in mind, although it is rare, when ground glass opacity is found in the thoracic radiological imaging of PSS patients.

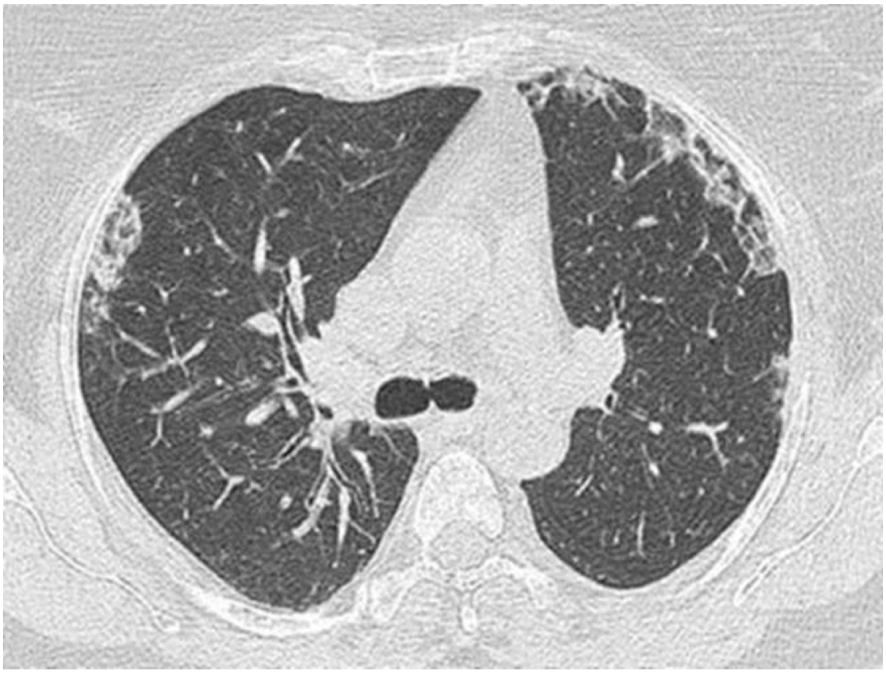

Fig. 1. High-resolution computed tomography.

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