CASE REPORT

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Recurrent bronchiolitis obliterans organizing pneumonia
in a patient with limited cutaneous systemic sclerosis

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Abstract
Systemic sclerosis (SSc) is a generalized disorder characterized by fibrosis and vascular obliteration in the skin, lung, gastrointestinal tract, and kidney. One of its two subsets is a stable, limited cutaneous group (ISSc). Pulmonary involvement in scleroderma is common, and several types of pulmonary disorders are associated with SSc. Bronchiolitis obliterans organizing pneumonia (BOOP) is a rare finding in lung disorders associated with SSc. We describe a case of ISSc with BOOP that was responsive to steroid therapy. Of interest is that the lung disorders appeared in different periods and areas. It might be important to diagnose abnormal shadows in lung fields before treatment of patients with SSc.

Keywords
Bronchiolitis obliterans organizing pneumonia · Computed tomography · Prednisolone · Systemic sclerosis · Video-assisted thoracoscopic surgery

Introduction
Systemic sclerosis (SSc) is a generalized disorder characterized by fibrosis and vascular obliteration in the skin, lung, gastrointestinal tract, and kidney. It is divided into two subsets: a stable, limited cutaneous group (ISSc) and a more rapidly advancing, diffuse cutaneous group (dSSc) [1]. Pulmonary involvement in scleroderma is common, observed in about 70% of patients at autopsy [2]. Several types of pulmonary disorders are associated with SSc, including interstitial pneumonia, pulmonary hypertension, pulmonary vascular diseases, and pleural disease [3]. Bronchiolitis obliterans organizing pneumonia (BOOP) is a rare finding in lung disorders associated with SSc [4].

In this report, we describe a case of ISSc with BOOP that was responsive to steroid therapy. Interesting in this case is that the lung disorders appeared in different periods and areas. It might be important to diagnose abnormal shadows in lung fields before treatment of patients with SSc.

Case report
A 79-year-old woman was admitted to our hospital because of cough and fever. She had also suffered from Raynaud’s phenomenon for several years, and sclerodactyly was observed on both her fingers. The nailfold capillary loops were slightly enlarged. Her physical examination showed no abnormalities other than finger disorders. The serum lactic dehydrogenase (LDH) level was 373 IU/l (normal range 180–450) and C-reactive protein (CRP) 3.5 mg/dl. The serum level of KL-6, a mucin-like high-molecular-weight glycoprotein that is elevated in sera of patients with interstitial pneumonia [5] was 457 U/ml (normal range <500). Blood gas analysis revealed slight hypoxia (pH 7.42, PaO₂ 74.7 Torr, PaCO₂ 38.4 Torr). Pulmonary function tests appeared normal, with vital capacity (VC) at 85.4%, relation of forced expiratory volume to forced vital capacity (FEV₁/FVC) 88.7, and diffusion capacity (DLCO) 89.8%.

The patient had received steroid therapy twice because of pulmonary disorder. At her first visit 3 years ago, chest radiography and computed tomography (CT) (Fig. 1, parts A and B, respectively) showed an infiltrative shadow at the left lower lung. Although video-assisted thoracoscopic surgery (VATS) was not done, since informed consent was not obtained, transbronchial lung biopsy (TBLB) was performed, and the histological findings were consistent with BOOP. It was responsive to steroid therapy (30 mg prednisolone, or PSL, for 2 weeks, then tapered off), and chest radiography and CT (Fig. 1, parts C and D, respectively) showed...
little abnormal shadow after the cessation of PSL administration (Fig. 1). At the second admission a year ago, new infiltrative shadows were visible in the anterior segment of the right upper lung and left upper lobe (Fig. 1, parts E and F) and were also diagnosed as BOOP by TBLB. Prednisolone was administered similarly, and the abnormal shadow disappeared on chest radiography and CT (Fig. 1, parts G and H, respectively).

At the present, third admission, the major part of the lung disorder was localized in the anterior segment of the left upper lung as determined by chest radiography (Fig. 1, part I) and chest CT (Fig. 1, part J). This time, VATS was performed, and histological findings of the lung tissue showed filled granulomatous plugs in bronchioles, alveolar ducts, and alveoli. Macrophages had accumulated in alveolar spaces, and infiltration of lymphocytes, plasma cells, and eosinophils and fibrotic change were observed in alveolar walls. These findings are consistent with BOOP (Fig. 2) [6]. Steroid therapy (30 mg/day of PSL) was effective for the lung disorder, and little abnormal shadow remained on chest radiography and CT after the cessation of PSL (Fig. 1, parts K and L). However, the severities of Raynaud’s phenomenon and sclerodactyly have not improved with steroid therapy.

Fig. 1. Chest radiography (A, C, E, G, I, and K) and CT (B, D, F, H, J, and L). A and B at first visit, C and D after improvement with prednisolone, E and F at second visit, G and H after improvement with prednisolone, I and J final visit, K and L after improvement with prednisolone.

Fig. 2. Histological finding of lung specimen obtained by video-assisted thoracoscopic surgery (H&E, original magnification ×100)