Unilateral Acute Exacerbation of Pulmonary Fibrosis in Association with Sjögren’s Syndrome

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A 70-year-old female had an abnormal chest roentgenogram. Infiltrative shadows were recognized in the right lung, and an open lung biopsy (OLB) specimen revealed usual interstitial pneumonia (UIP). Xerostomia, keratoconjunctivitis sicca, and lymphocyte infiltration in salivary glands were consistent with Sjögren’s syndrome; she was diagnosed as having pulmonary fibrosis in association with Sjögren’s syndrome (SjS-IP). Acute exacerbation occurred and she was successfully treated with corticosteroids. Unilateral exacerbation and UIP in SjS-IP are rare. OLB is useful for diagnosis and to select a pertinent therapy if lung involvement is unilateral in Sjögren’s syndrome.

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Figure 1. Chest roentgenogram on admission (1991.4.23) shows infiltrative shadows and small nodular shadows localized in the right lower lung field.

Figure 2. Chest CT scan on admission (1991.4.24) shows infiltrative shadows and subpleural curvilinear shadows in the right lower lung field.

Figure 3. Chest CT scan on acute exacerbation (1991.9.18) shows widespread infiltrative shadows and ground-glass appearance localized in the right lung.

Figure 4. Open lung biopsy specimen from both right S8 and S10 shows coexistence of thickness of alveolar septa, inflammatory cell infiltration in alveoli, and destructive changes of the alveolar structures (honeycomb formation) (HE stain, x40).

1,670 ml (84%); FEV1/FEV75.9%; diffusing coefficient (DLco) 11.7 ml/min/mmHg (83%). Arterial blood gas analysis during room air breathing was: pH 7.41; Po2 71.0 Torr; Pco2 42.7 Torr; HCO3 27.1 mM; AaDo2 27.8 Torr.

Biopsy specimens from minor salivary glands revealed lymphoid cell infiltration. Transbronchial brushing and washing cytology were performed at the right basal segment (S10) and no neoplastic cells were recognized. Transbronchial lung biopsy (TBLB) was performed at right S10 and microscopic examination indicated nonspecific interstitial pneumonia with lymphoid cell infiltration in alveolar septa, thickness of the alveolar walls and no foreign body deposition.

After several months, she complained of shortness of breath and her chest roentgenogram and chest CT scan showed in-
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Fig. 5. Chest CT scan after corticosteroid therapy (1991.12.19) shows mild infiltrative shadows remain in the right lower lung field.

creased infiltrative shadows and ground-glass appearances in the right lung field (Fig. 3). She complained of a severe cough, therefore TBLB and bronchoalveolar lavage could not be attempted. We performed an open lung biopsy from right S8 and S10, so as to determine whether the lung shadows were acute exacerbation of interstitial pneumonia or aspiration pneumonitis. Gross examination of the specimen revealed tight adhesion to parietal pleurae. Microscopic findings (Fig. 4) varied from almost normal alveolar structures to extensive interstitial pneumonia with focal alveolar fibrosis and microhoneycomb lung. Furthermore, muscular cirrhosis with peripheral smooth muscle proliferation and organization within the bronchiolar lumen or alveolar cavities were widespread. Severe inflammatory changes were recognized both in the visceral and parietal pleurae and intra-lobular pleurae. No foreign body depositions, giant cells or macrophages, as seen in aspiration pneumonitis, could be recognized.

In conclusion the aggravation of the symptoms and chest roentgenogram findings of this patient indicated acute exacerbation in pulmonary fibrosis, not aspiration pneumonitis. She was treated successfully with corticosteroids and healed, though some honeycombing and slight infiltrative shadows remained (Fig. 5).

Discussion

Sjögren’s syndrome is a chronic inflammatory autoimmune disease characterized by dryness of the mouth, eye and other mucous membranes. The basic pathologic lesions are lymphoid infiltrative processes of the salivary glands and extrasalivary lymphoproliferative disorders. The pleuro-pulmonary manifestations of Sjögren’s syndrome are reported to be pleurisy and/or effusion, interstitial fibrosis, desiccation of the tracheobronchial tree and lymphoid interstitial disease (1). The histopathologic appearance of pulmonary lesions, lymphocytic interstitial pneumonitis, pseudolymphoma and malignant lymphoma are commonly described in Sjögren’s syndrome, as one of the expressions of an autoimmune disorder. These lesions occur frequently in Sjögren’s syndrome (5). An animal study by Hoffman and colleagues (11) suggested that the dryness influences the lower respiratory tract with the lymphocytic infiltrates around bronchioles and pulmonary arteries.

Microscopically, lymphoid interstitial pneumonia is characterized by diffuse, interstitial lymphoid hyperplasia in the lung, and the infiltrate may be nodular and principally around the bronchi and vessels, though it invariably involves the alveolar septum (10). The summary of pathologic findings in this case was varied with focal alveolitis, microhoneycomb lung, smooth muscle hypertrophy in peripheral bronchiole, intra-alveolar organization and pleural thickness. Muscle changes, vasculitis and pleural changes may agree with the diagnosis of autoimmunity, concerning interstitial pneumonia in association with collagen vascular disease (12).

The pathologic finding of UIP in association with Sjögren’s syndrome has been rare, as only two cases, that of Kawaguchi and colleagues (13) and that of Fujita and colleagues (14), have been reported. The former case might be a profile of interstitial fibrosis in association with systemic lupus erythematosus (SLE). By contrast, like the latter case and our case, UIP, not only LIP, might be frequently in association with primary Sjögren’s syndrome. However, we cannot confirm this possibility because of the few reported cases, of the existence of collagen vascular disease preceded by interstitial pneumonia (15), and of the fact that chest CT scan and lung biopsy are not always performed for patients with Sjögren’s syndrome. Many cases should be summarized and an active lung biopsy should be performed for patients with interstitial fibrosis to determine the possible contribution of Sjögren’s syndrome if lung involvement is unclear (9).

Unilaterally throughout the clinical course is very rare in interstitial fibrosis in association with Sjögren’s syndrome, though it may be present in the early stage. In this case, abnormal pulmonary shadows were located only on the right side of her lung throughout her admission, including during exacerbated periods, and that was confirmed by chest CT scan even when she was scanned in the prone position. Because of the facts that her abnormal shadows were localized in the right lower lung field, she had false teeth, and had a history of cerebral arteriosclerosis, we first considered the possibility of aspiration pneumonitis, but pathologic findings revealed pulmonary fibrosis due to Sjögren’s syndrome. Chest roentgenographic features of Sjögren’s syndrome are reticular or nodular infiltrates, or dense patchy infiltrates (16). The infiltration is usually bilateral and most prominent in the bases of the lungs (5). Furthermore, neither severe interstitial fibrosis without malignant lymphoma nor rapidly progressing lung manifestation has been reported. This suggests that we should consider the possibility of sub-
clinical Sjögren’s syndrome, if symptoms are not marked, in cases of acute exacerbation in patients diagnosed as having idiopathic interstitial pneumonia or idiopathic pulmonary fibrosis.

Though the present patient experienced exacerbation of the interstitial fibrosis, corticosteroid treatment was effective to reduce the lung shadows. Corticosteroid was tapered off gradually without exacerbation. But, apparently due to arteriosclerosis, she appeared to have a psychosomatic disorder and entered a psychiatric hospital. Up to at least three years after corticosteroid therapy she had experienced no exacerbation and had complained of no other collagen-vascular diseases.

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References