Spontaneous Remission of Desquamative Interstitial Pneumonia

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We report a case of spontaneous remission of desquamative interstitial pneumonia (DIP) in a 50-year-old male. The histological diagnosis of DIP was based on open lung biopsy. A chest X-ray revealed reticulo-nodular shadows in the bilateral lung fields, and the patient had mild dyspnea on exertion. Without treatment, these shadows decreased gradually and disappeared after several months. The patient recovered completely within one year, and recurrence of the disease has not been observed for 4 years. Recently, DIP has rarely been described, and the spontaneous remission of DIP has not been reported since Carrington et al in 1978 (1).

Key words: bronchoalveolar lavage (BAL), open lung biopsy (OLB), respiratory bronchiolitis-associated interstitial lung disease (RB/ILD)

Introduction

Desquamative interstitial pneumonia (DIP) is one of the chronic progressive interstitial pulmonary diseases described in the histological classification proposed by Liebow et al in 1968 (2). The histological findings of DIP are characterized by the presence of numerous macrophages filling the alveolar spaces, without marked fibrosis. The prognosis of DIP is generally considered to be good compared with usual interstitial pneumonia (UIP), because of its good response to corticosteroid treatment. Additionally, the incidence of DIP is much lower than that of UIP (1, 3–5). Furthermore, it is a matter of controversy whether or not DIP is an independent clinical entity. We describe herein a patient who displayed reticulo-nodular shadows on chest X-rays, was diagnosed as having DIP by open lung biopsy and showed spontaneous remission during several months of follow-up.

Case Report

A 50-year-old male was referred to our hospital in September 1991 because diffuse reticulo-nodular shadows in the bilateral lung fields were detected on chest X-rays during a regular checkup. He complained of mild exertional dyspnea (Hugh-Jones grade II) though his general condition was good, without fever, cough or sputum. Though he quit smoking upon admission, he had had a smoking habit (30 cigarettes a day) for 30 years without history of any dust exposure. His family history and past history were not contributory. Physical examination on admission did not reveal any abnormal findings such as skin rash, clubbed finger, joint swelling or deformity, muscle weakness, respiratory rales and heart murmur.

Laboratory findings including peripheral blood cell counts, liver and renal function tests were all within normal limits. In serological tests, C-reactive protein, rheumatoid factor, antinuclear antibody and anti-Jo-1 antibody were negative. Pulmonary function tests revealed that forced vital capacity (FVC), forced expiratory volume in one second/FVC (FEV1/0%), and diffusing capacity of carbon monoxide (DLCO) were within the normal range except for a slightly increased residual volume. However, arterial blood gas analysis at rest showed moderate hypoxemia (PaO2: 71.1 Torr). Bronchoalveolar lavage (BAL) was performed through the right middle lobe (B3a) as previously described (6). Total cell count was 39.8 x 10³/ml and differential cell counts of BAL fluid were 75% macrophages, 18.3% lymphocytes, 0.9% neutrophils and 5.8% eosinophils. A CD4/CD8 ratio was elevated at 5.61.

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A chest X-ray revealed diffusely increased reticulo-nodular shadows in the bilateral lung fields as shown in Fig. 1A. Chest computed tomography (CT) scan showed diffusely increased density of the lungs and multiple small bullous changes just beneath the pleura bilaterally (Fig. 1B). Honeycombing was not noted. There was no enlargement of hilar or mediastinal lymph nodes. For differential diagnosis of interstitial pulmonary diseases, lung biopsy under thoracotomy was performed. Microscopically, the alveolar wall was slightly thickened and the alveolar spaces were filled with macrophages (Fig. 2). These changes were observed diffusely in the biopsied specimen, while centriacinar tendency of alveolar infiltration was not noticed. Immunohistochemically, macrophages in the alveolar spaces were positive for the PG-M1 antibody (CD68) which was used as a specific marker for macrophages, and they were negative for anti-S-100 protein antibody which is usually

Figure 1. A) Chest X-ray, taken on admission, which revealed reticulo-nodular shadows distributed diffusely in the bilateral lung fields. B) Chest CT scan showing diffusely increased density of the lungs and multiple small bullous changes just beneath the pleura bilaterally.

Figure 2. Low magnification view of lung specimen stained with hematoxylin-eosin showing mild diffuse thickening of the alveolar walls (×40A, ×100B). Higher magnification showed a marked accumulation of alveolar macrophages filling the alveolar spaces (×400, C).
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Figure 3. A) Within several months of follow-up, the reticulo-nodular shadows in bilateral lung fields disappeared gradually without any treatment. B) Multiple small bullous changes just beneath the pleura remained, as shown in Fig. 1b.

Discussion

Differences in clinical manifestations and histological findings between UIP and DIP have been the subject of debate since the 1970’s (1, 3–5). Carrington et al (1) reported that UIP was characterized by onset at an older age, decreased pulmonary function and severe fibrotic changes of the alveolar wall when compared with DIP, but there were no differences in relation to sex and smoking history between them. In general, UIP is less responsive to steroid treatment than DIP, and the prognosis of UIP is worse than that of DIP (1, 3, 4). UIP is characterized by the temporal heterogeneity of histological findings, including various structural changes from normal alveolar architecture to terminal fibrotic changes in the same specimen (1–4). On the other hand, DIP shows relatively uniform lesions in almost the entire specimen with different degrees of infiltration of macrophages in the alveolar spaces (1–4). Moreover, some investigators claim that DIP may represent an early stage of UIP with marked cell infiltration (3, 4). In this case, the histological findings obtained from lung biopsy specimens clearly excluded the existence of advanced UIP lesions. Furthermore, spontaneous remission of UIP has never been reported and a moderate increase of eosinophils in BAL fluid, as in the case of the present patient, has been shown to indicate generally slow but steady progression with episodes of exacerbation and poor prognosis in UIP (3, 6). These histological findings, such as marked cell infiltration and uniform distribution of alveolitis, might be interpreted as an early or cellular stage of UIP. However, our patient's spontaneous remission and good prognosis even with eosinophilia in his BAL fluid deny the possibility of UIP.

Some studies have shown that approximately 90% of DIP patients are smokers while 70% of UIP patients are smokers (1, 3, 4). Myers et al (7) have recently stressed that DIP should be histologically differentiated from respiratory bronchiolitis-associated interstitial lung disease (RB/ILD) that occurs frequently in smokers. RB/ILD is reported to be terminal bronchiolar alveolitis with an increased number of pigmented macrophages phagocytizing dusts in the adjacent air space (7–9). It should be emphasized that these lesions, resembling DIP, are confined to the peribronchiolar parenchyma rather than having a diffuse distribution in the lobuli. Thus, uniform lesions of the alveolar spaces in the present patient are characteristic of DIP and are different from RB/ILD. Carrington et al reported in 1978 that 7 (21.9%) of 32 untreated patients with DIP improved spontaneously (1). However, there has been no report concerning the spontaneous remission of DIP after their documentation. Patients with RB/ILD have shown improvements in their symptoms and physical findings after the cessation of smoking (7–9). Accordingly, some cases of RB/ILD might have previously been misdiagnosed as DIP, and the spontaneous remission of DIP could be exceptional. The patient in this report is certainly a rare case of the spontaneous remission of DIP. Thus, the diagnosis of DIP should be done based on the precise histological examination of an adequate specimen obtained by open lung biopsy or thoracoscopic lung biopsy at the appropriate timing in its clinical course.
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References