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# Desquamative interstitial pneumonia complicated by diffuse alveolar haemorrhage

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## Abstract

We report a rare case of desquamative interstitial pneumonia (DIP) with diffuse alveolar haemorrhage (DAH). A 56-year-old man diagnosed with DIP by surgical lung biopsy 2 years ago was admitted to our hospital because of severe acute respiratory failure. The DIP had progressed despite smoking cessation. On admission, the patient appeared extremely ill, and physical examination revealed respiratory distress. The patient required mechanical ventilation. High-resolution computed tomography showed diffuse ground glass opacity in both lungs. The bronchoalveolar lavage fluid was bloody, and numerous haemosiderin-laden alveolar macrophages were detected. Pulse steroid therapy followed by oral prednisolone immediately relieved the respiratory failure and improved the long-term control of the DIP. Paired sera tests confirmed the diagnosis of influenza A/H3N2 virus infection, which was the cause of the DAH. Chronically progressive DIP with acute respiratory failure due to DAH was successfully treated by steroid therapy.

## Introduction

Desquamative interstitial pneumonia (DIP) was first described in 1965 by Liebow et al. [1]. The aetiology of DIP is unknown, but it is widely associated with cigarette smoking and passive exposure to cigarette smoke. Although smoking cessation and systemic corticosteroids are important components of disease management, such treatments do not always produce clinical effects. In a minority of cases, the outcome is poor [2]. We report a case of chronic progressive DIP despite smoking cessation, in which the patient exhibited acute respiratory failure due to diffuse alveolar haemorrhage (DAH) that was successfully treated by steroid therapy.

## Case Report

A 56-year-old man was admitted to our hospital with severe acute respiratory failure. The patient had been well until 2 years before this admission, when he noted general fatigue and shortness of breath during exertion. At that time, he was a 30-pack-year current smoker. He was admitted to hospital at that time, and high-resolution computed tomography (HRCT) of the chest revealed bilateral ground glass opacities (GGOs), predominantly in the lower lobes, and no honeycombing. Surgical lung biopsy showed numerous aggregates of alveolar macrophages within the alveolar space and dense thickening of the alveolar septa. Mild interstitial fibrosis was noted (Fig. 1A, B). He was diagnosed with DIP, and he quit smoking completely. No corticosteroids or other immunosuppressive agents were administered. The patient's exercise tolerance, vital capacity, and features of the chest radiographs deteriorated gradually over the next 2 years.

Five days before the current admission, he experienced progressive dyspnoea and high fever with chilling. On admission, his body temperature was 37.9°C, blood pressure was 121/67 mmHg, heart rate was 106 beats/min, and respiratory rate was 40 breaths/min. Cobblestone appearance in posterior oropharynx, crackles, and finger clubbing were not found. The arterial blood gas values were

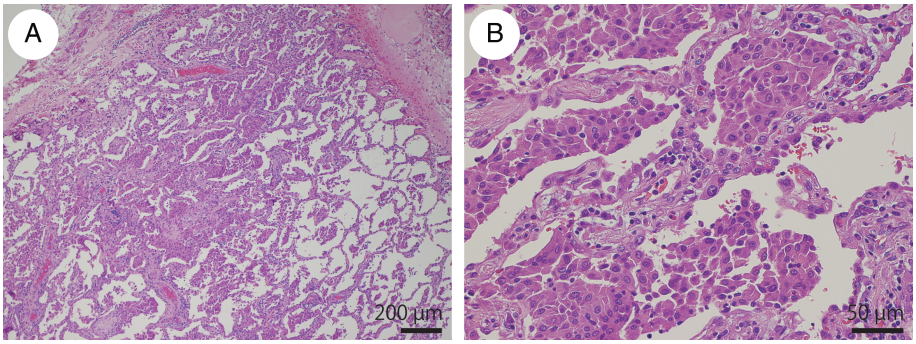


**Figure 1.** Surgical lung biopsy specimen showed numerous aggregates of macrophages in the alveoli and mild interstitial fibrosis without fibroblastic foci (haematoxylin and eosin (HE) staining). (A) 40× and (B) 200×.

pH 7.46, partial pressure of carbon dioxide ($PaCO_2$) 37.6 Torr, and partial pressure of oxygen ($PaO_2$) 98.6 Torr while receiving 10 L/min of oxygen. Chest HRCT showed new GGO in both lungs, in addition to worsening GGO of the bilateral lower lobes (Fig. 2A, B). The patient required mechanical ventilation due to severe respiratory distress. Total cell count in the bronchoalveolar lavage fluid was $4.3 \times 10^5$/mL, with 52.0% alveolar macrophages, 45.0% neutrophils, 1.0% lymphocytes, and 2.0% eosinophils. The recovered bronchoalveolar lavage fluid was bloody, and many haemosiderin-laden alveolar macrophages were detected. Serum levels of both lactate dehydrogenase (LDH) and Krebs von den Lungen-6 (KL-6) were elevated. Autoantibodies associated with DAH were within normal limits. Blood and sputum cultures were negative for fungi, acid-fast bacteria, and other pathogenic bacteria. Throat swabs were negative for the influenza A and B virus antigen. Pulse steroid therapy followed by 1 mg/kg/day oral prednisolone, 1.5 g/day meropenem hydrate, and 500 mg/day levofloxacin hydrate were administered for 3 days. The patient was successfully weaned from mechanical ventilation 3 days after intubation. Chest X-ray and HRCT revealed remarkable improvement of the GGO. After discharge, a paired sera test revealed an eightfold increase in the antibody titre against influenza A/H3N2 virus. Oral prednisolone was gradually tapered over a period of 2 years and then discontinued. This patient has remained smoke-free and has no respiratory symptoms or GGOs on chest HRCT.

## Discussion

Here, we report a case of chronically progressive DIP with acute respiratory failure due to DAH, which was successfully treated with steroid therapy. The DIP is a rare form of interstitial lung disease, characterized by the presence of numerous macrophages within the alveolar air spaces. The aetiology of this disease is unknown, but approximately 90% of patients with DIP are smokers. Smoking cessation is important in the management of the DIP, but the effects of smoking cessation on the clinical course are not fully understood. Most patients become stable or improve with steroid treatment [1]. Carrington et al. [2] divided DIP into three groups according to the extent of the fibrosis. The prognosis does not differ in patients with severe fibrosis, regardless of whether they have idiopathic pulmonary fibrosis or DIP [2]. In patients with mild or moderate fibrosis, the prognosis is better in DIP than in idiopathic pulmonary fibrosis. Our patient did not have severe fibrosis of the lung.

The cause of acute respiratory failure in this case was DAH. Usual causes of DAH are diffuse alveolar damage due to vasculitis, infection, or drug reaction. Influenza virus infection causes diffuse alveolar damage. As the antibody titre against influenza A/H3N2 was significantly

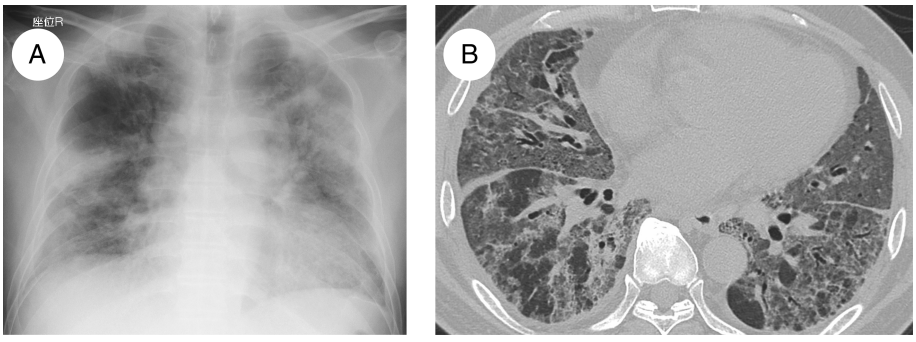


**Figure 2.** (A) Chest X-ray on admission showed bilateral ground glass opacity and consolidation. (B) Chest high-resolution computed tomography (HRCT) on admission showed diffuse ground glass opacity and a small amount of pleural effusion bilaterally. Traction bronchiectasis was also found, to a degree similar to that obtained before admission.

elevated in the present case, the cause of DAH was thought to be an influenza infection. The patient fully recovered following pulse steroid therapy without any anti-influenza agents, although the infection was sufficiently fulminant to cause the DAH.

In this case, despite smoking cessation, the respiratory symptoms and chest CT features indicated disease progression. The clinical course of the patient would suggest a poor prognosis. It is not clear whether the diffuse alveolar damage affected the clinical course of the DIP. Of three previous case reports of patients with DIP and acute respiratory failure, steroid therapy produced a good response in two [3,4], and because steroid treatment was not effective in the third case, combination therapy with immunosuppressants was necessary to control the disease [5]. In our case, both the DAH and DIP responded to pulse steroid therapy followed by long-term oral prednisolone. After discontinuation of the steroid therapy, no recurrence was detected, and the long-term prognosis is considered good. As Carrington et al. [2] previously reported, the extent of lung fibrosis is thought to be important in the prognosis of DIP. Further accumulation of cases and investigation are needed.

## Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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