

NEAR FATAL DESQUAMATIVE INTERSTITIAL PNEUMONIA WITH BILATERAL RECURRENT TENSION PNEUMOTHORAX

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ABSTRACT. Desquamative interstitial pneumonia (DIP) is an uncommon form of interstitial lung disease demonstrating good response to corticosteroid therapy and favorable prognosis. In rare cases, patients with DIP fail to respond to corticosteroid therapy, and may progress to end-stage fibrotic lung disease with respiratory failure. For such patients, well defined treatment regimen does not exist until now. We report a rare case of near fatal DIP with recurrent bilateral tension pneumothorax despite of corticosteroid maintenance therapy. Clinical and radiological findings improved with surgical intervention in addition to combination therapy with prednisolone and clarithromycin. The patient has remained in an improved state and has been receiving prednisolone and clarithromycin for 9 months. (*Sarcoidosis Vasc Diffuse Lung Dis* 2015; 32: 167-171)

KEY WORDS: desquamative interstitial pneumonia; tension pneumothorax; macrolide

CASE REPORT

A 34-year-old man with a 4-month history of dry cough and progressive dyspnea on exertion was admitted. He was a 15-pack-year smoker and stopped smoking as symptoms occurred. There was no history of exposure to industrial toxic gases or drugs. Laboratory findings including peripheral blood cell counts, liver, and renal function tests were

within normal limits. Serologic tests for specific autoantibodies to nuclear antigens, and rheumatoid factor were all negative. Pulmonary function tests revealed a restrictive disease of moderate severity (forced vital capacity decreased to 35.1% of predicted value; diffusing capacity of carbon monoxide decreased to 20.6% of predicted value). Chest X-ray was normal; however, chest computed tomography (CT) showed diffuse ground-glass opacities underlying mildly emphysematous lungs, without nodules or honeycomb pattern (Figure 1). A wedge resection was performed via video-assisted thoracic surgery (VATS), and microscopy revealed prominent accumulation of alveolar macrophages in alveolar spaces, with mildly fibrotic and thickened alveolar septa (Figure 2). The patient was diagnosed with DIP and was encouraged to continue to refrain from smoking. Prednisolone (60 mg/day) was administered for 14

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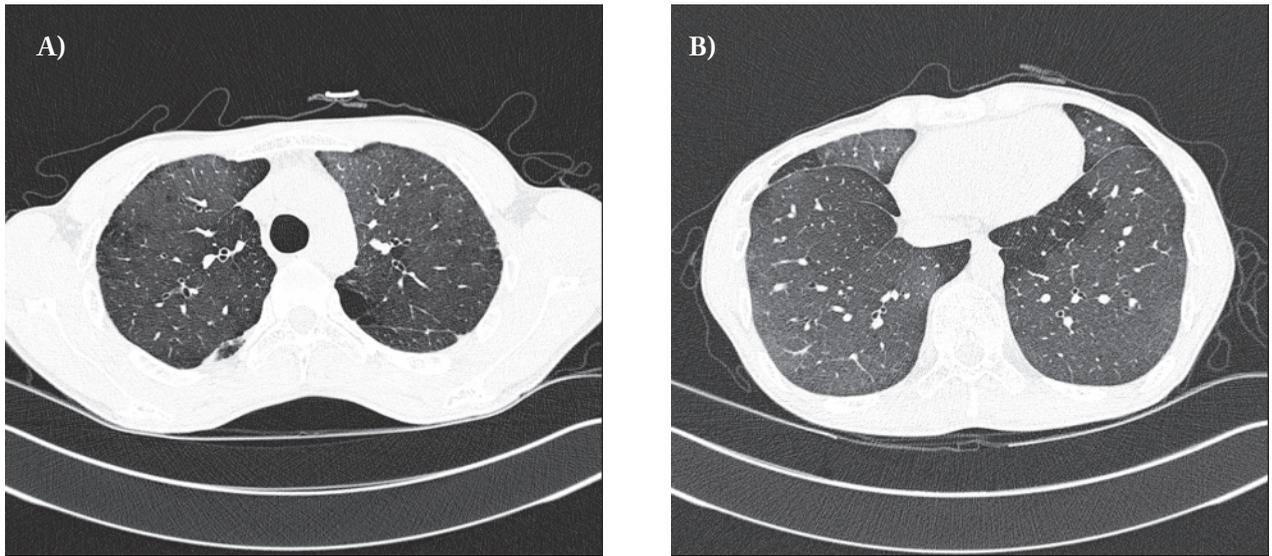


Fig. 1. A and B: A high resolution CT chest scan revealed ground-glass opacity bilaterally that predominantly involved peripheral lung region

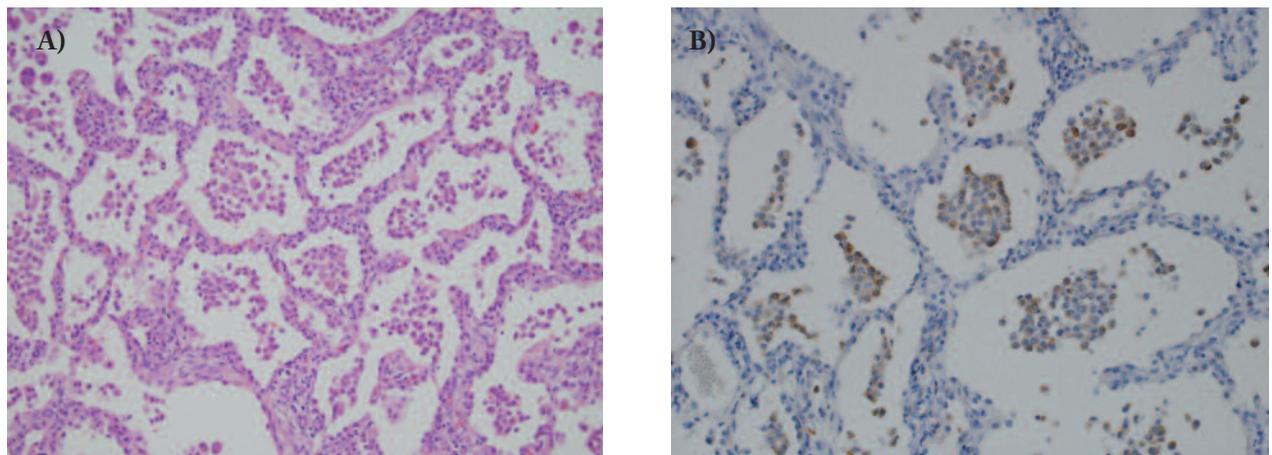


Fig. 2. Lung biopsy specimen. A: Mild fibrotic thickening of alveolar septa (hematoxylin-eosin, original 200x). B: prominent accumulation of alveolar macrophages in alveolar spaces (glycoprotein CD 68, 200x).

days, resulting in improvement of symptoms. The patient was discharged from the hospital with a maintenance dose of prednisolone (30 mg/day), and was followed up in the outpatient clinic.

Four months later, he returned complaining of sudden dyspnea and left sided chest wall pain. A chest radiograph revealed left massive pneumothorax with mediastinal shift to the right (Figure 3, A). The left lung failed to fully expand despite tube thoracostomy by the third hospital day, and a wedge resec-

tion was performed via VATS. Moreover, right massive pneumothorax with collapsed lung, which newly developed on the seventh hospital day, failed to completely recover even after tube thoracostomy (Figure 3, B). Therefore, a right upper lobectomy was performed. Histological analysis of the excised lung specimens confirmed findings that were very similar to those of the initial biopsy. Persistent ground-glass opacities on chest CT were regarded as a sign that the DIP was unresponsive to corticoids.

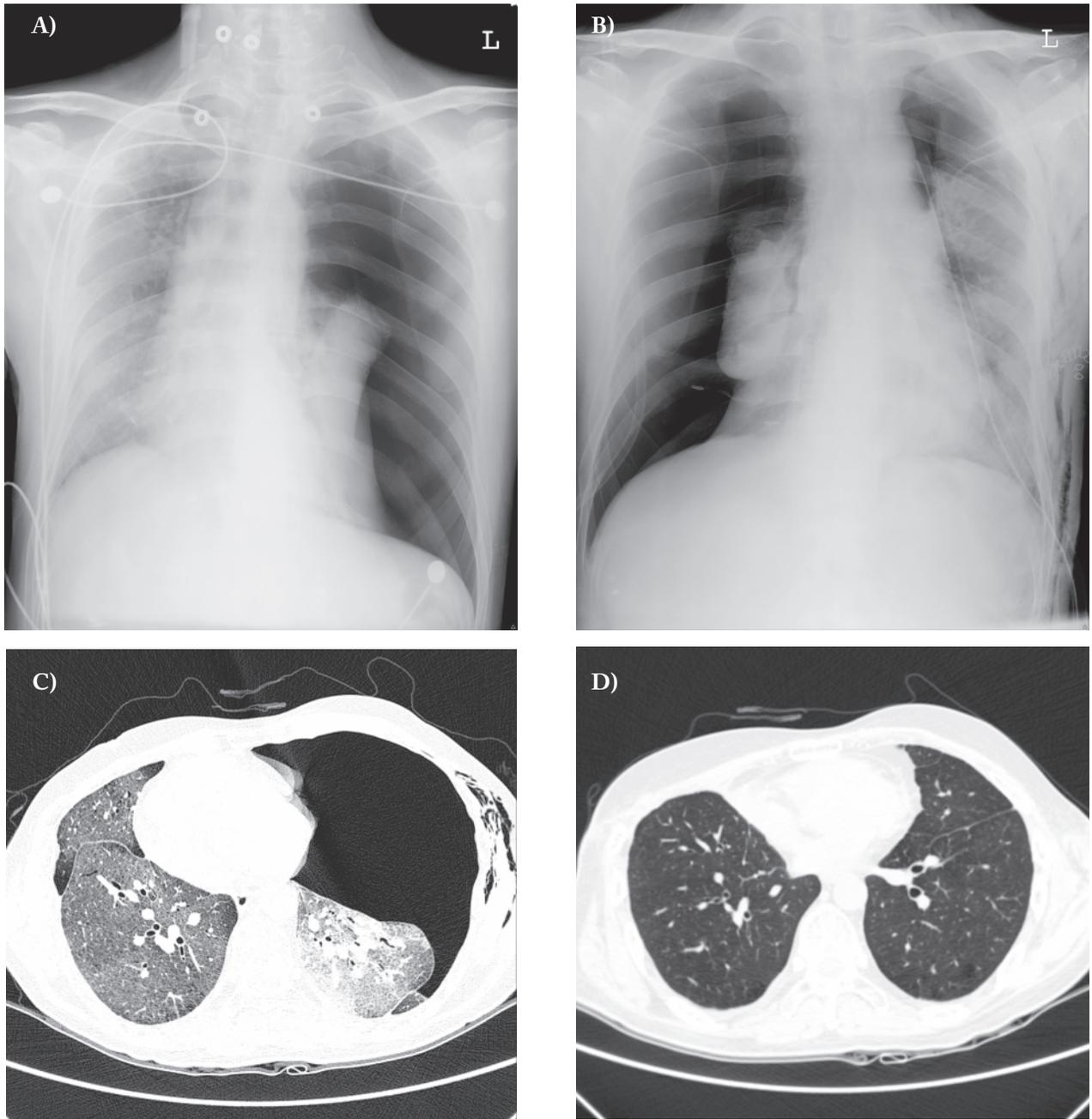


Fig. 3. A: Chest radiograph showed left tension pneumothorax on admission. B: chest radiograph on 7th day of hospitalization showed right tension pneumothorax. C: chest CT scan revealed left tension pneumothorax and ground-glass opacities on right lung. D: Chest CT scan, obtained after surgical intervention in addition to one month of combination therapy with clarithromycin and corticosteroid, showed a remarkable decrease in the degree and extent of ground-glass opacities.

teroid maintenance therapy (Figure 3, C). Prednisolone (60 mg/day) was administered for 14 days, and then rapidly tapered to 30 mg/day because of the possibility of delayed wound healing. Because

macrolide antibiotics are known to be an efficient therapeutic option for other chronic inflammatory lung diseases, the patient was treated with combination therapy consisting of clarithromycin (500 mg,

twice daily) and prednisolone (30 mg/day). His exertional dyspnea improved within two weeks. After one month of treatment, the ground-glass opacities disappeared in the chest CT (Figure 3, D). He has maintained this improved state and has been receiving prednisolone (20 mg daily) and clarithromycin (500 mg twice daily) in our outpatient clinic for 9 months.

DISCUSSION

DIP is a rare form of idiopathic interstitial pneumonia characterized by a large number of pigmented macrophages in the alveoli with interstitial fibrosis and/or inflammation (1). Most patients with DIP show a favorable response to corticosteroid therapy with smoking cessation, which results in a good prognosis (2,3). In some cases of DIP, even spontaneous remission has been described (3,4). The literature contains a few cases that were refractory to corticosteroid therapy, and showed fulminant or rapidly progressive clinical course (5, 6). In our case, bilateral recurrent tension pneumothorax developed during the course of corticosteroid tapering. Although pneumothorax is a possible complication of DIP (3), this recurrent bilateral tension pneumothorax has not been described in the literature as a near fatal complication of DIP. In addition, this case is the first report of the administration of a macrolide as an alternative to immunosuppressive agents in the treatment of a near fatal course of DIP.

Interestingly, recurrent bilateral tension pneumothorax had developed despite of corticosteroid maintenance therapy with smoking cessation. These findings appear to suggest that the response to treatment could be dependent on the dose of corticosteroid. Previous studies have shown variable results with regard to the response of DIP to corticosteroid (2,3,7). Earlier studies reported that approximately one-quarter of patients with DIP may continue to worsen despite therapy (3,7). On the other hand, a recent study indicated that 97% of patients with DIP show a favorable response to corticosteroid therapy (2). This discrepancy may be related to the differences in definitions before and after the international classification of idiopathic interstitial pneumonia (1). However, Individualization of treatment with corticosteroid could influence the outcome. Kim et

al. (8) reported the improvement of DIP after initiation of corticosteroid treatment and pneumatocele formation during the course of corticosteroid tapering. For patients with DIP, a standard guideline regarding the dose and duration of corticosteroid has not been suggested yet. Therefore, corticosteroid therapy needs to be tailored according to individual circumstances and the severity of disease.

In this case, we could taper the dose of corticosteroid to 20 mg/day with the use of clarithromycin. Although cytotoxic and other immunosuppressive agents have been considered in the treatment of patients with DIP who are poorly responsive to corticosteroid treatment, their efficacy remain undefined until now. Macrolide antibiotics are known to have anti-inflammatory properties and a beneficial effect in the treatment of chronic airway diseases, including cystic fibrosis and diffuse pan-bronchiolitis (9). Recent case report with clarithromycin monotherapy showed a beneficial response in a patient with slowly progressive DIP, refractory to corticosteroid therapy (10). We found the possible role of clarithromycin as an adjunctive therapy in a near fatal DIP with bilateral recurrent tension pneumothorax. However, an increased dose of corticosteroid could have mainly related to the improvement of DIP. Therefore, additional clinical research is needed to define the role of macrolide in the treatment of DIP.

In conclusion, we presented a case of near fatal DIP with recurrent bilateral tension pneumothorax, treated with appropriate surgical intervention in addition to combination therapy with corticosteroid and macrolide.

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