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Case report

Direct hemoperfusion with polymyxin B-immobilized fiber for the treatment of the acute exacerbation of idiopathic pulmonary fibrosis in patients requiring invasive mechanical ventilation

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ABSTRACT

The prognosis of acute exacerbation of idiopathic pulmonary fibrosis (AE-IPF) in individuals undergoing invasive mechanical ventilation is known to be poor.

We describe the cases of three men, who were former smokers and required mechanical ventilation, whose AE-IPF was treated with direct hemoperfusion with polymyxin B-immobilized fiber column (PMX-DHP). In all cases, we successfully weaned the patients from mechanical ventilation. Two of the patients survived for more than 180 days after development of AE-IPF. PMX-DHP may improve the prognosis of severe respiratory failure in patients with AE-IPF.

Abbreviations: AE-IPF, acute exacerbation of idiopathic pulmonary fibrosis; ALAT, Latin American Thoracic Association; ATS, American Thoracic Society; BAL, bronchoalveolar lavage; CT, computed tomography; ERS, European Respiratory Society; FVC, forced vital capacity; IPF, idiopathic pulmonary fibrosis; MV, mechanical ventilation; NPPV, non-invasive positive pressure ventilation; PMX-DHP, direct hemoperfusion with polymyxin B-immobilized fiber column.

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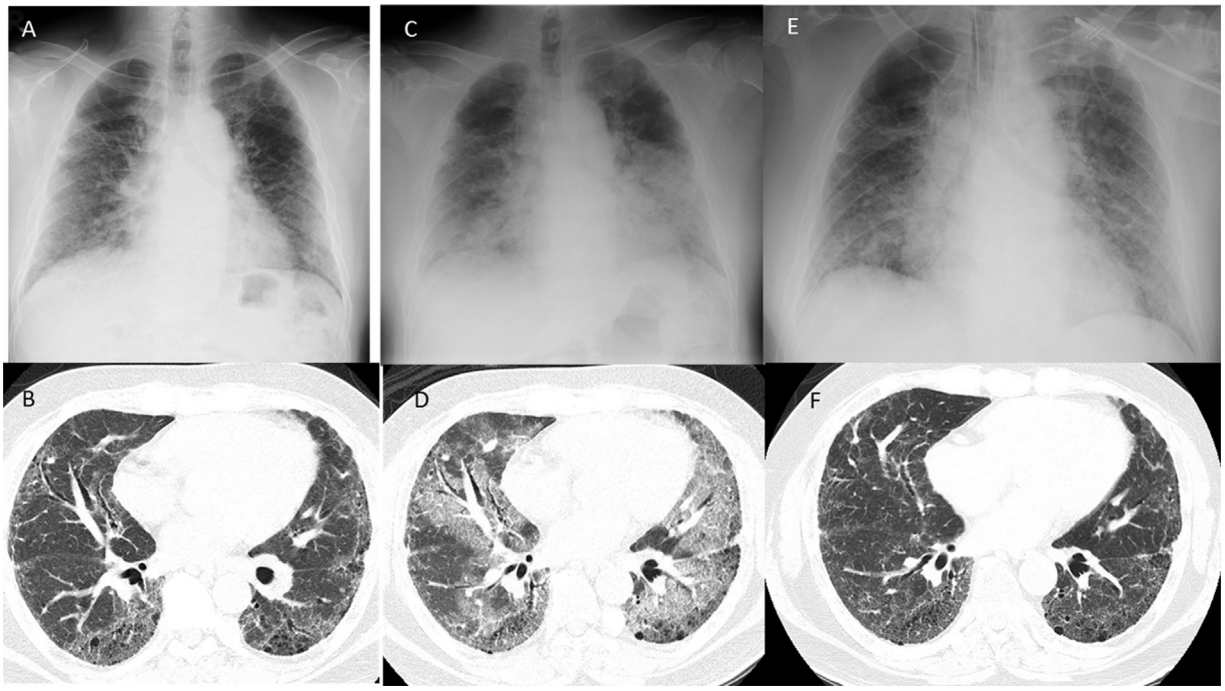

Fig. 1 – Sixty-three-year-old man (case 2). Chest radiograph (A) and chest CT (B) before acute exacerbation showed a reticular shadow and honeycombing with traction bronchiectasis. Chest radiograph (C) and chest CT (D) upon admission revealed bilateral ground-glass opacification. Chest radiograph 2 days after PMX-DHP treatment (E) and chest CT 25 days after admission (F) revealed improvement in the bilateral ground-glass opacification. Abbreviations: CT, computed tomography; PMX-DHP, direct hemoperfusion with polymyxin B-immobilized fiber column.

1. Introduction

Acute exacerbation of idiopathic pulmonary fibrosis (AE-IPF) requiring mechanical ventilation (MV) has a devastating prognosis [1,2]. There are no proven effective therapies for AE-IPF. Recently, several studies reported the effectiveness of direct hemoperfusion with polymyxin B-immobilized fiber column (PMX-DHP) for the treatment of AE-IPF [3–6]. Here we describe the cases of three patients with AE-IPF requiring MV treated with PMX-DHP.

2. Case report

2.1. *Case 1*

A 68-year old man presented to our former hospital in May 2011 with an abnormal reticular shadow, predominantly in the lower lobes with pleural honeycombing, on chest computed tomography (CT). Risk factors for interstitial pneumonia, including environmental exposure, connective tissue disease, and drug toxicity, were excluded. Thus, the patient was diagnosed with idiopathic pulmonary fibrosis (IPF) according to the 2011 consensus criteria of the American Thoracic Society (ATS), European Respiratory Society (ERS), Japanese Respiratory Society, and Latin American Thoracic Association (ALAT) [7]. Pirfenidone was administered from 2013. The patient developed dyspnea in January 2015, and a chest CT revealed newly bilateral ground-glass opacification. He was diagnosed with AE-IPF and treated with corticosteroids. His clinical condition and imaging findings improved, although he had hypoxemia upon exertion. The patient was discharged with prescriptions for prednisolone (22.5 mg/day) and oxygen therapy.

The patient presented in the emergency room with exertional dyspnea and disturbance of consciousness. The patient's partial pressure of oxygen was 73.6 Torr, although he had an oxygen supply at 15 L/min. Fine crackles were audible in both lungs. Laboratory findings upon admission revealed an increased white blood cell count (21,810/μL), elevated C-reactive protein level (6.88 mg/dL), and high serum levels of Krebs von de Lungen-6 (2201 U/mL). Chest CT revealed bilateral ground-glass opacification with reticular shadows and honeycomb.

The patient's respiratory state soon deteriorated, and mechanical ventilation was initiated. We performed bronchoalveolar lavage (BAL) for differential diagnosis and found a 70% increase in the number of neutrophils. The possibility of infection was eliminated based on the results of the BAL fluid culture, serum β-D glucan, and other tests. Cardiac failure and pulmonary embolism were also excluded. We diagnosed the patient with AE-IPF for the second time.

The patient was treated with methyl prednisolone pulse therapy (1 g for 3 days), antibiotic therapy with ampicillin/sulbactam and azithromycin, sivelestat sodium hydrate, and direct hemoperfusion with polymyxin B-immobilized fiber column (PMX-DHP). PMX-DHP was performed for 48 h. Subsequently, the condition and respiratory state of the patient improved. The PaO_2/FiO_2 ratio improved from 73.6 to 255.4 after 48 h of PMX-DHP. On day 10 after admission, the patient was successfully weaned from MV. His condition was good and stable, but his respiratory state worsened suddenly again

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