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Case Report

Life after acute fibrinous and organizing pneumonia: a case report of a patient 30 months after diagnosis and review of the literature

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ABSTRACT

Acute fibrinous and organizing pneumonia (AFOP) is a rare histologic interstitial pneumonia pattern recently described in the literature with fewer than 120 cases published. AFOP is often difficult to diagnose and may be mistaken for other pulmonary disorders such as interstitial pneumonias or pneumonitides. Patients often present with vague symptoms of cough, dyspnea, hemoptysis, fatigue, and occasionally respiratory failure. Radiological findings show diffuse patchy opacities and ground glass appearance of the lungs. On histologic examination, intra-alveolar fibrin balls are observed. We discuss a case of a man who presented with hemoptysis and dyspnea and whose open lung biopsy revealed AFOP. We will describe the presentation, diagnosis, and post-discharge course, and review the current literature. There are only 4 cases which have reported the patients' course of disease after 1 year, the longest being 2 years. To our knowledge, this is the only case of AFOP in the literature that describes the course of a patient more than 2 years after the diagnosis of AFOP, and is the most comprehensive review of the current literature.

1. Introduction

Acute fibrinous and organizing pneumonia (AFOP) is a rare histologic interstitial pneumonia pattern that is histologically characterized by intra-alveolar fibrin "balls" and organizing pneumonia with a patchy distribution. It is associated with viral, bacterial, and fungal infections, connective tissue disorders, autoimmune diseases, certain drugs including amiodarone, everolimus, decitabine, and abacavir, and occupational/environmental exposures such as asbestos, various dusts, exotic animals, aerosol products, and coal; it may also be idiopathic in nature. Patients present with cough, dyspnea, or acute respiratory distress syndrome. Radiological findings demonstrate diffuse patchy opacities. The diagnosis requires a lung biopsy which can be obtained surgically, via bronchoscopy, or under ultrasound- or computed tomographic (CT) scan-guidance. The definitive management of this disease is still unknown; however, there is a role for steroids and other immunosuppressive agents [1].

We present the case of a man diagnosed with AFOP and describe his 30-month post-discharge course, the longest follow-up in the literature to our knowledge. In addition, we will also present the most comprehensive review of the AFOP literature. Patient characteristics, suspected etiology, diagnostic modality, treatments, and outcomes will be reported.

2. Case report

A 60-year-old male smoker was transferred from an outside hospital presenting with progressively worsening dyspnea and blood-tinged sputum over the course of 2 months. He was otherwise healthy and reported a history of working with asbestos and fiberglass from 1975 to 1988, and recent occupational dust exposure. He complained of low grade fevers and sweats and denied weight loss, other significant exposures, recent travel, pets, and sick contacts. At an outside hospital, a chest x-ray showed bilateral diffuse opacities and small bilateral pleural effusions (Fig. 1A). A CT scan of the chest demonstrated diffuse bilateral ground glass densities (Fig. 1B). Bronchoscopy was performed and biopsies were negative for malignancy, granulomas, *Pneumocystis jirovecii* cysts, and trophozoites. Gram stain and culture, periodic acid–Schiff stain, and acid fast-bacilli culture were negative. Bronchoalveolar lavage (BAL) showed many red blood cells, few white blood cells, and no organisms. The BAL culture grew few *Candida albicans*. Routine laboratory work up was normal, except for an elevated white blood cell count. Laboratory work for rheumatoid factor, cold agglutinins, antinuclear antibodies, Scl-70 scleroderma antibody, and histone antibody were negative. In addition, HIV-1 and HIV-2, influenza A and B, urine *Legionella* antigen, and *Mycoplasma pneumoniae* IgG and IgM were negative. He received a 7-day course of antibiotics (ceftriaxone, azithromycin, and doxycycline) and methylprednisolone 125 mg IV every 6 hours for 6 days. Despite therapy, his symptoms did not improve and he was transferred to our hospital for further work up.

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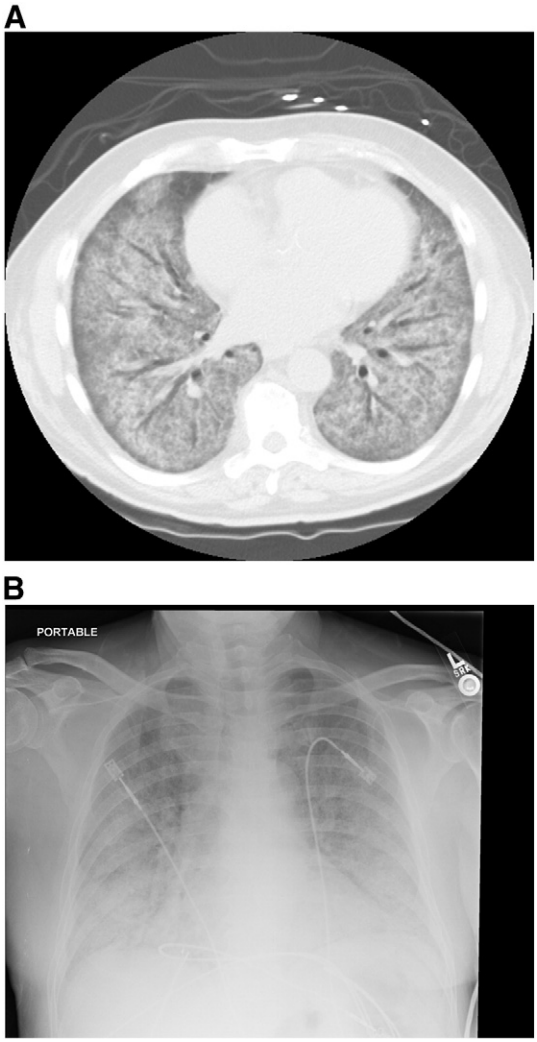

Fig. 1. A, Chest x-ray showing bilateral patchy infiltrates. B, Chest CT scan showing diffuse ground glass opacities.

On initial physical examination he was afebrile, tachypneic, and hypoxic, requiring 80% oxygen to achieve a SpO_2 of 92%. Lung auscultation revealed diffuse rhonchi and rales. Additional tests performed at our institution included blood and sputum cultures, ANCA, Jo-1 antibody, glomerular basement membrane antibody, cyclic citrullinated peptide IgG and IgA, and creatine phosphokinase, which were negative. On admission to our hospital, methylprednisolone 60 mg IV every 6 hours was given for 2 days, and the dose was later increased to 125 mg IV every 6 hours. Two days later a steroid taper was initiated, decreasing the methylprednisolone dose to 60 mg IV every 6 hours for 5 days, and then prednisone 60 mg PO daily was started. He required oxygen with a high flow nasal cannula. On the fourth day of hospitalization, he underwent an anterior thoracotomy, right wedge lung biopsies, and right chest tube placement. He was brought intubated from the operating room for ventilator weaning in the intensive care unit and he was extubated on postoperative day 1. Specimens of the lung biopsy were sent for pathology, microbiology, and virology. The pathology demonstrated AFOP with a background of chronic interstitial pneumonia (Fig. 2A and B). A trichrome stain revealed fibrosis. Tissue specimens were negative for acid fast-bacilli, mycobacteria, fungi and cytomegalovirus. Gram stain and culture were also negative. However, an anaerobic and aerobic culture grew a coagulase negative *Staphylococci*. Sulfamethoxazole/trimethoprim was initiated on postoperative day 3. On repeat chest x-ray, the lungs appeared unchanged. The chest tube was eventually removed and he was discharged to pulmonary rehabilitation on

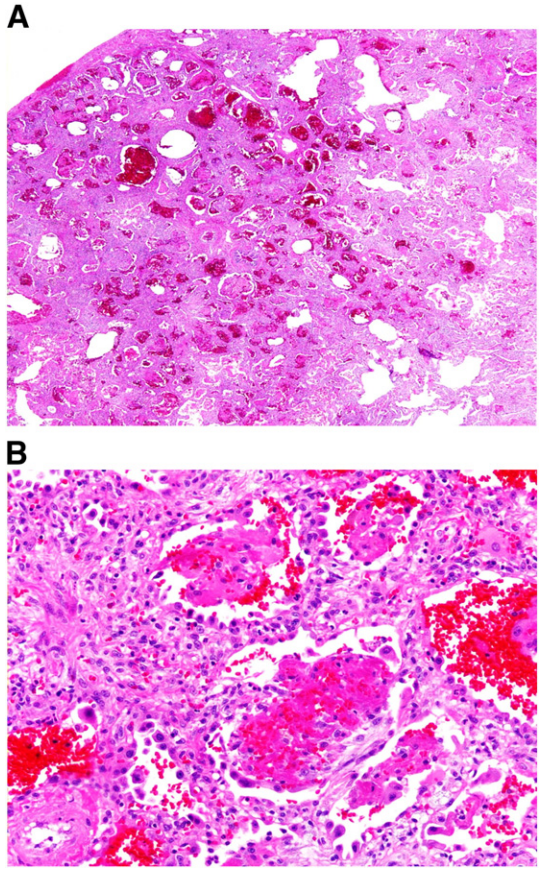

Fig. 2. A, Histology of AFOP with diffuse and patchy involvement. B, Histology of AFOP with intraalveolar fibrin balls.

prednisone 60 mg PO daily and continuous oxygen via nasal cannula at 3 L/min.

Upon discharge, the patient was followed by the pulmonology team. Eight months after his hospital discharge, he experienced arthralgias and Raynaud's syndrome, and developed telangiectasias. On further evaluation, blood work was positive for antinuclear antibodies and Sjogren's syndrome antibody and he was diagnosed with an unspecified connective tissue disease (CTD). He was started on azathioprine, but because of an adverse reaction, his therapy was changed to mycophenolate mofetil 500 mg PO 3 tablets in the morning and 2 tablets at night. Over 2 years, several steroid tapers were attempted unsuccessfully. His symptoms worsened at lower doses. Thirty months after discharge he remains dependent on mycophenolate mofetil, prednisone 10 mg PO daily, and oxygen, requiring 4 L/min via nasal cannula at rest and during sleep, and 8 to 10 L/min via an oxymizer pendant with activity. He is currently on disability. Although his exercise tolerance improved after completing pulmonary rehabilitation, his functional status has not returned to his pre-illness baseline. He has been evaluated and listed for lung transplantation.

3. Discussion

Acute fibrinous and organizing pneumonia (AFOP) is a rare histologic interstitial pneumonia pattern; as of 2015, there have been 111 cases reported in the literature (Table 1). Of these cases, 43 were male, 35 were female; gender was not specified in 33 cases. It most commonly occurred in patients in their fifth and sixth decade of life; however, there were 5 reports in children. In most cases, the etiology was unknown. Possible causes of AFOP include connective tissue disease, infections, environmental or occupational exposure, drug reactions, autoimmune disease, after organ transplantation, and cancers