Acute fibrinous and organizing pneumonia

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A 45-year-old man presented with malaise, arthralgia, and dyspnea. The chest CT scan showed bilateral patchy consolidation in the lower lobes. A lung biopsy revealed intra-alveolar fibrin “balls” deposits and focal features of organizing pneumonia, both of which are typical pathological features of acute fibrinous and organizing pneumonia (AFOP). The patient had a good clinical course after treatment with prednisone. We report this case of idiopathic AFOP and review the published studies on this newly recognized clinicopathological entity that is still underdiagnosed and underreported.

Acute fibrinous and organizing pneumonia (AFOP) was first described by Beasley et al in 2002, as a new pattern of lung injury, with histological similarities to organizing pneumonia (OP), diffuse alveolar damage (DAD), and eosinophilic pneumonia (EP). However, it has a distinct overall histological pattern, characterized by intra-alveolar fibrin associated with OP in a patchy distribution. Since Beasley's initial description, 14 individual case reports of AFOP have been published in English research papers. Although the histopathological features are well described, the clinical manifestations, course, and treatment of AFOP are not characterized. We report a case of a male with bilateral patchy lower lobes consolidation, in which lung biopsy revealed the classical histopathological features seen in AFOP.

CASE

A 45-year-old man, without significant past medical history, presented with a history of malaise and arthralgia of 8 weeks duration preceded by a flu-like illness. Two weeks prior to his presentation to the emergency room, he complained of left-sided chest pain with dyspnea and nonproductive cough. There was no history of fever, chills, sweats, hemoptysis, or weight loss. Neither a history suggestive of connective tissue diseases was available nor significant occupational or environmental exposure was reported.

On examination the patient was alert with temperature of 37.2°C, pulse rate 100/min, respiratory rate 18/min, and blood pressure 130/60 mm Hg. He appeared lethargic but without obvious signs of respiratory discomfort. Auscultation of the lungs revealed bilateral inspiratory crackles. The rest of his physical examination was unremarkable.

Laboratory data revealed a white blood count of 9.8 k/cu mm, white blood count of 9.8×10⁹ (4-10×10⁹) hemoglobin 11.9 g/dL, and platelet count of 546k/mm³. The ESR was 34 mm/h. Electrolytes, creatinine, liver function tests, and urine analysis were within normal limits. A sputum gram smear showed scant neutrophils, and a sputum culture detected a growth of normal respiratory flora. Arterial blood gas at room air revealed pH 7.4, PaO₂ 63 mm Hg and a PaCO₂ 35 mm Hg. Serologic tests for ANA, RF, and C-ANCA and P-ANCA were negative. The pulmonary function test revealed a moderate restrictive ventilatory impairment and a moderate decrease in diffusing capacity. A chest CT scan revealed bilateral patchy peripherally located consolidations, mainly of the lower lobes. Some consolidations appear nodular with thickening of the interlobular septae (Figure 1).

An open lung biopsy from the left lower lobe revealed a patchy consolidation of the lung parenchyma with interstitial thickening associated with a mixed inflammatory infiltrate of mononuclear cells and a few neutrophils (Figure 2). Some alveoli contained fibrin deposits as “fibrin balls” admixed with macrophages (Figure 3). Focal features of OP were present and characterized by intra-alveolar fibroblastic plugs (Figure 4). Some alveoli were lined by reactive and hyperplastic pneumocytes. The intervening parenchyma between the consolidated areas showed mild interstitial thickening, sparse inflammatory
Afop

Ann Saudi Med 2013 May-June

www.annsaudimed.net

302

case report

infiltrate, and focal emphysematous changes. Subpleural fibrosis and chronic inflammation were present. No hyaline membranes were seen. The special stains for fungal and mycobacterial organisms were negative. These findings were consistent with AFOP.

After establishing the diagnosis of AFOP, the patient was started on methylprednisone 60 mg every 6 hours, followed by 50 mg of oral prednisone daily. He showed substantial clinical and radiological improvements after 12 weeks of corticosteroid treatment. The chest scan revealed almost normal findings after 3 months (Figure 5).

DISCUSSION

From the 17 cases that comprised Beasley’s initial report of AFOP, which were identified by a retrospective review of case material at the Armed Forces Institute of Pathology, 9 cases had fulminating illness and died of the disease, and 8 cases had a subacute disease with recovery. The most common symptoms reported were dyspnea in 11 patients, fever in 6 patients, and cough in 3 patients. Bilateral basilar infiltrates were the most common radiographic pattern. Seven of the 17 patients were treated with corticosteroids. Fourteen individual case reports of AFOP have been published in English,2-15 after Beasley’s initial description. All patients had symptoms for less than 2 months, with cough and dyspnea as the most common. Radiographic abnormalities included bilateral basilar infiltrates in 11 patients,2,3,5-15 while unilateral complete lung consolidation solitary nodule,4 miliary nodules, and patchy consolidations6 were noted in the remaining 3 patients. Identified exposures or associated conditions included medications (abacavir and decitabine),5,10 connective tissue disease,8,11 hematopoietic stem cell transplant,6 acute lymphoblastic leukemia,13 HIV,1 juvenile dermatomyositis,1 and Whipple disease.15 No underlying cause or association were identified in
5 cases.2,4,9,12,14 Patients were treated with medications including corticosteroids, mycophenolate mofetil, cyclophosphamide, and cyclosporine. Among the 14 case reports, 3 patients required mechanical ventilation,3,5,9 and 2 patients died.1,4 In addition to these individual case reports, Hwang et al16 reported autopsy finding from the lungs of 20 patients who died from severe acute respiratory syndrome. The histological findings of these 20 patients were AFOP pattern in 6 cases, DAD in 8 cases, and both patterns in the remaining cases. In conclusion, AFOP is a newly recognized clinicopathological entity that is still underdiagnosed and underreported. Although the histopathological features are well described, identification of precipitating factors, clinical course, and elements affecting prognosis and treatment need to be fully established.

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