Idiopathic Bronchiolitis Obliterans Organising: Pneumonia: The First Case That Was Diagnosed in Our Hospital

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We report the case of a woman who presented with progressive dyspnea, nonproductive cough and constitutional symptoms including weight loss, anorexia and malaise for six months. Physical examination revealed end-inspiratory crackles over both lungs. Chest-X-ray (CXR) and thorax computerised tomography (CT) revealed bilateral multiple alveolar opacities with air bronchograms. With transbronchoscopic lung biopsy, bronchiolitis obliterans organising pneumonia (BOOP) was the pathologic diagnosis. We could not find any underlying condition, that can cause BOOP, so classified as idiopathic. The patient rapidly responded to, both clinically and radiologically, steroid treatment within one month. This is the first case of idiopathic BOOP that was diagnosed in our hospital, and was considered worth reporting.

Key words: Idiopathic BOOP, air bronchogram, alveolar opacity

Idiopatik Bronşiolitisi Obliterans Organize Pnömoni: Hastanemizde Tani Konan İlk Vaka


Anahtar kelimeler: Idiopatik BOOP, hava bronkogramı, alveoler opasite

INTRODUCTION

BOOP is a rare immune disorder characterized by granulation tissue that obstructs terminal and respiratory bronchioles and extends into the distal alveolar ducts and alveoli. Cryptogenic organising pneumonia (COP) is a synonymous term. Clinical features include dry cough, fever, dyspnea. Radiologically, bilateral alveolar infiltrates with air bronchograms is the most characteric pattern. BOOP may complicate collagen vascular disease, lower respiratory tract infections, drugs, toxic fume inhalation and
organ transplants, or idiopathic, in which no specific etiology can be established.

**CASE**

A 68 years-old female who was previously healthy, presented to our department with a history of progressive dyspnea, nonproductive cough and constitutional symptoms including malaise, weight loss (about 10 kg) and anorexia. CXR revealed bilateral multiple alveolar opacities (figure 1). The patient was admitted to our ward for further evaluation. The patient was centrally cyanotic, dyspneic and orthopneic. There were end-inspiratory crackles over both lung fields on auscultation. Laboratory findings were as follows: Complete blood count (CBC): White blood cell: 5400/mm³, hemoglobin: 13 gr/dL, sedimentation: 10mm/hour. Serology: Antistreptolysin-O (ASO): -, Rheumatoid factor: -, Antinuclear antibody: -, antiDNA: -, Ig G, Ig A and Ig M: Normal. Thyroid function tests were normal. Arterial blood gases (ABG): pH: 7.52. PaO₂: 57 mmHg, PaCO₂: 29 mmHg, HCO3: 23 mmol/L. Pulmonary function tests (PFTs): FVC: 1.85 L (76% of predicted), FEV₁: 1.48 L (73% of predicted), FEV₁/FVC: 95% of predicted, that was consistent with mild restrictive defect. Alveolararterial oxygen gradient (D A-a O₂): 56 (N: <20), that was increased. Echocardiography was normal. Thorax CT revealed bilateral multiple alveolar opacities with air bronchograms involving both lung fields (figure 2). We performed fiberoptic bronchoscopy, that was normal, but we obtained transbronchoscopic lung biopsy through lateral segment of the right middle lobe bronchus. The result of pathologic examination of biopsy specimen was consistent with BOOP. We could not find any underlying condition that can cause

**BOOP including collagen vascular diseases, drug usage, lower respiratory tract infections, inflammatory bowel diseases, exposure to organic or inorganic dust, or thyroid disease.**
We started steroid treatment, as 60 mg prednisolone/day orally. Within one month, the patient responded to treatment, clinically and radiologically. At first month of treatment: ABG: pH: 7.39, PaO$_2$: 97 mmHg, satO$_2$: 97%, PaCO$_2$: 28 mmHg. HCO$_3$: 17 mmol/L. D A-a O$_2$: 18. Pulmonary function tests: FVC: 2.35 L (111% of predicted), FEV1: 2.17 L (113% of predicted). FEV$_1$/FVC: 112% of predicted. CXR (figure 3) and thorax CT (figure 4) showed prominent decrease in alveolar opacities at first month of treatment. Then, the patient was discharged with a prescription of 50 mg prednisolone per day orally.

**DISCUSSION**

In 1985, idiopathic BOOP was described as a distinct entity with clinical, radiologic and histopathologic features$^{[1]}$. Idiopathic BOOP is a relatively rare disorder and most referral centers see two to five cases per year$^{[3]}$. Most cases present in the fourth through sixth decades of life. There is no gender predominance. The predominant symptoms are nonproductive cough and dyspnea. Constitutional symptoms of fever, weight loss, and malaise may be prominent. The course is usually subacute with symptoms developing over two weeks to six months. An antecedent upper or lower respiratory tract infection precedes the onset of symptoms in one third of patients. The diagnosis is often not suspected until repetetive courses of antibiotics have failed$^{[3]}$. Our patient's course, including onset and symptoms, were actually compatible with BOOP.

Physical examination reveals mid-inspiratory squeaks or rhonchi in 40% of patients. Inspiratory rales may be present. In our case, inspiratory rales were heard over both lungs. Sedimentation is elevated in 80% of cases, but was 10 mm/hour in our patient. Roentgenologically, bilateral alveolar
infiltrates, often peripherally and with air bronchograms, are the cardinal features, observed in 60-80% of patients. A more diffuse pattern, associated with reticulonodular infiltrates, occurs in 20-30% of patients. The other pattern is solitary focal pneumonia which usually occurs in upper lobes. Thorax CT scans depict the salient features of BOOP more clearly\(^\text{[4,5]}\). Radiologic appearance, both CXR and thorax CT, were consistent with the bilateral multiple alveolar infiltrates with air bronchograms, the most typical pattern of BOOP, in our case.

PFTs in BOOP characteristically demonstrate reductions in lung volumes and carbon monoxide diffusion capacity (DLCO), mild hypoxemia with widened alveolar-arterial oxygen gradient. These deficits usually reverse with corticosteroid therapy. In our case, PFTs gave similar results. There were reductions in both FVC and FEV1 proportionally, no obstruction was there, and also mild hypoxemia with increased alveolar-arterial oxygen gradient, and these parameters improved with steroid treatment. We could not make DLCO measurement due to technical problem.

The cardinal histologic feature of BOOP is an exuberant inflammatory and fibrotic process involving terminal and respiratory bronchioles. Tufts of granulation tissue, aggregates of neutrophils, fibrin, connective tissue plug the terminal bronchioles. Inflammatory cells, mostly mononuclear cells, extend into the peribronchiolar regions, alveolar ducts and alveolar spaces. The disease is patchy, but the peribronchiolar distribution is a clue to diagnosis. The alveolar architecture is preserved and fibrosis is notably absent\(^\text{[6]}\). The diagnosis can be confirmed with transbronchoscopic lung biopsy. The absence of bronchitis or purulent secretions within airways makes infection less likely. In our case, bronchoscopic appearance was normal and there was no secretion. We made transbronchoscopic lung biopsy and result of pathologic examination of biopsy specimens was consistent with the findings of BOOP, including intraluminal granulation tissue in terminal and respiratory bronchioles extending into alveolar ducts and alveolar spaces and also there was peribronchial inflammation.

BOOP may complicate collagen vascular diseases, inflammatory bowel diseases, lower respiratory tract infections, thyroid diseases, drugs, and organ transplants, and also can be seen after exposure to organic and/or inorganic dust. But, we could not find any underlying condition that can cause BOOP in our case. Thus, we diagnosed the patient as idiopathic BOOP.

Corticosteroids are the cornerstone of treatment for BOOP. Response to treatment is usually rapid and complete\(^\text{[6,7]}\). We started corticosteroid treatment, as prednisone 60 mg per day orally, that resulted in symptomatic and clinical improvement including PFTs and arterial blood gases, and also control CXR and thorax CT, within one month, showed clearing of alveolar opacities.

In conclusion, in patients with a history of subacute onset of nonproductive cough, dyspnea, and constitutional symptoms including malaise, weight loss and anorexia, and also with bilateral multiple alveolar opacities with air bronchograms on CXR and thorax CT, and if there is failure of multiple antibiotic regimens, idiopathic BOOP should be considered in differential diagnosis.

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