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

Serhat FINDIK[1] MD, M. Levent ERKAN[1] MD, Levent YILDIZ[2] MD, Davut UĞURLU[1] MD, Mahmut ÇAKIR[3] MD, Fatma Aras DORU[1] MD, Bedri KANDEMİR[2] MD,

Ondokuz Mayıs Üniversitesi, Tıp Fakültesi, Göğüs Hastalıkları[1], Patoloji[2] ve Çocuk Sağlığı ve Hastalıkları[3] Anabilim Dalı, SAMSUN

✔ 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*

✔ **İdiopatik Bronşiolitis Obliterans Organize Pnömoni: Hastanemizde Tanı Konan İlk Vaka**

Altı ay önce başlayan ve giderek artan dispne, kuru öksürük ve halsizlik, kilo kaybı, iştahsızlık gibi konstitüsyonel yakınmalar ile başvuran 68 yaşında bir kadın hastayı sunuyoruz. Fizik muayenede her iki akciğerde inspiryum sonu ince ralleri mevcuttu. Akciğer grafisinde ve toraks bilgisayarlı tomografisinde (BT) her iki akciğerde yaygın multipl alveoler opasiteler ve hava bronkogramları mevcuttu. Transbronkoskopik akciğer biopsisinin patolojik inceleme sonucu bronşiolitis obliterans organize pnömoni (BOOP) ile uyumlu geldi. Ayrıntılı değerlendirmede BOOP' a neden olabilecek altta yatan bir hadise olmadığından idiopatik olarak kabul edildi. Steroid tedavisine başlandı ve ilk ay içinde hastada belirgin klinik ve radyolojik düzelme gözlendi. Hastanemizde tanı konan ilk idiopatik BOOP vakası olması bakımından yayınlanmaya değer olduğunu düşündük.

Anahtar kelimeler: *İdiopatik BOOP, hava bronkogramı, alveoler opasite*

INTRODUCTION

BOOP is a rare immune disorder charecterized 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 charecteristic 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/mm^3, 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, PaO_2: 57 mmHg, $PaCO_2$: 29 mmHg, HCO3: 23 mmol/L. Pulmonary function tests (PFTs): FVC: 1.85 L (76% of predicted), FEV_1: 1.48 L (73% of predicted), FEV_1/FVC: 95% of predicted, that was consistent with mild restrictive defect. Alveolearterial oxygen gradient (D A-a O_2): 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.

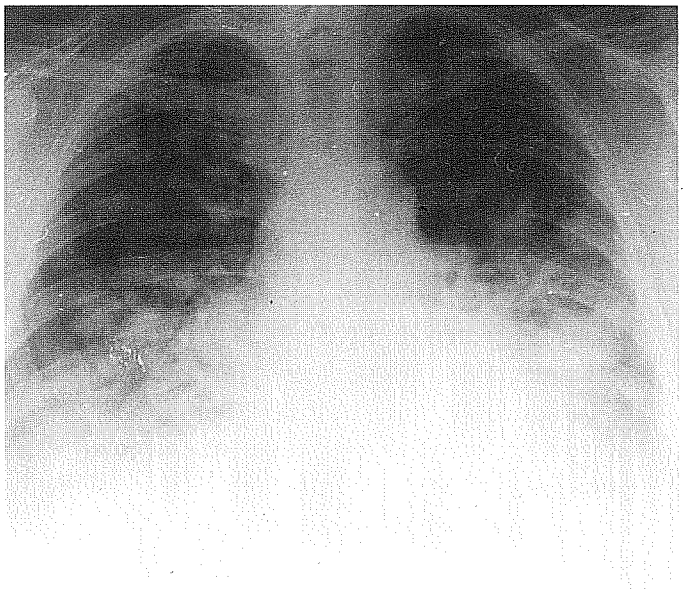

Figure 1. Initial chest x-ray demonstrated bilateral multiple alveolar opacities

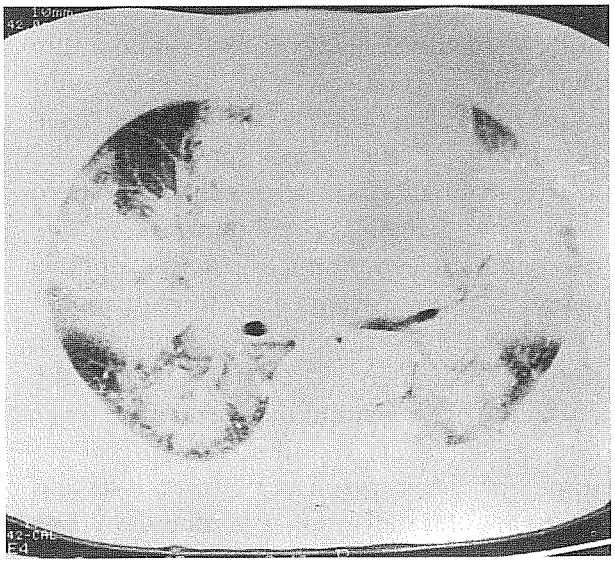

Figure 2. Initial thorax CT revealed multiple alveolar opacities with air bronchograms involving both lung fields.

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: 97mmHg, $satO_2$: 97%, $PaCO_2$: 28 mmHg, HCO3: 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.

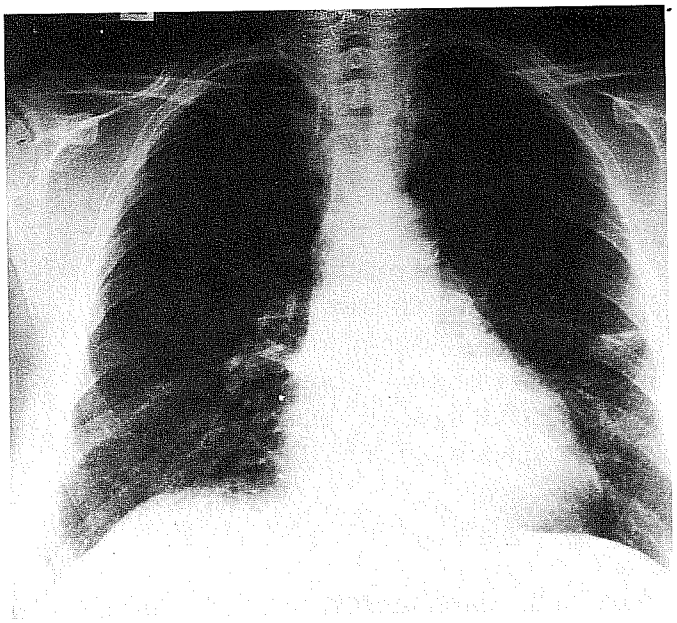

Figure 3. Control chest x-ray taken at first month of treatment showed prominent decrease in alveolar opacities.

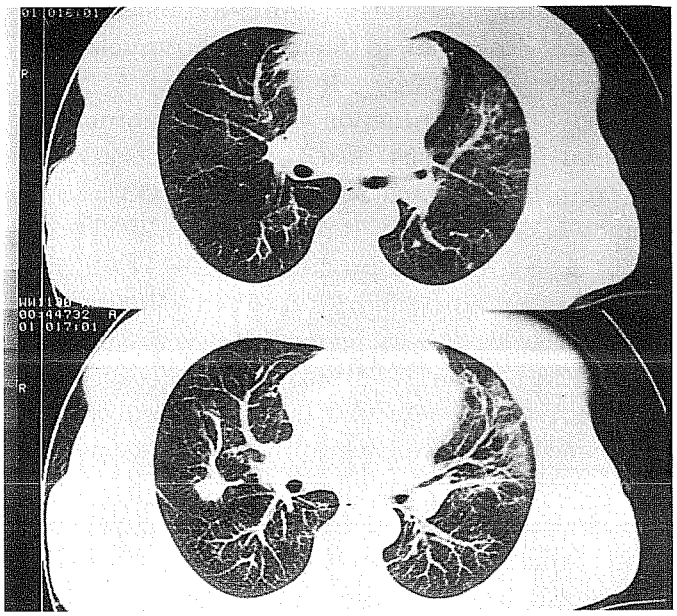

Figure 4. Control thorax CT taken at first month of treatment showed prominent decrease in alveolar opacities

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 [2]. 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, weihgt 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 rhonci in 40% of patients. İnspiratory rales may be present. In our case, inspiratory rales were heard over both lungs. Sedimentation is elevated in 80% of cases, but was 10mm/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[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 ia a clue to diagnosis. The alveolar architecture is preserved and fibrosis is notably absent[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 transbronchocopic 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 trasplants, 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[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.

Geliş tarihi : 28.09.1998
Yayına kabul tarihi : 07.05.1999
Yazışma adresi:
Dr. Serhat FINDIK
Ondokuz Mayıs Üniversitesi, Tıp Fakültesi,
Göğüs Hastalıkları Anabilim Dalı
55139 Kurupelit, SAMSUN

KAYNAKLAR

1. Epler GR, Colby TV, McLoud TC, et al. Bronchiolitis obliterans organising pneumonia. N Engl J Med 1985; 312: 152-158
2. Alasaly K, Muller N, Ostrow DN, et al. Cryptogenic organising pneumonia: A report of 25 cases and a review of the literature. Medicine 1995; 74: 201-211
3. Cordier JF, Loire R, Brune J. Idiopathic BOOP: Definition of charecteristic clinical profiles in a series of 16 patients. Chest 1989; 96: 999-1004
4. Bouchardy LM, Kuhlman JE, Ball WC Jr, et al. CT findings in BOOP with radiographic, clinical and histologic correlation. J Comput Asist Tomogr 1993; 17: 352-357
5. Lee KS, Kullnig P, Hatman TE, et al. Cryptogenic organising pneumonia: CT findings in 43 patients. AJR 1994; 162: 543-546
6. Colby TV, Myers JL. The clinical and histologic spectrum of bronchiolitis obliterans including BOOP. Semin Respir Med 1992; 13: 119-126
7. Karnak D, Kayacan O, Beder S, ve ark. Tüberküloz ve Toraks 1998; 46: 180-185