A Case Of Bronchiolitis Obliterans with Organizing Pneumonia

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Abstract

Bronchiolitis obliterans with organizing pneumonia is a very special disorder. The lesions nearly completely resolve with steroid therapy. Clinical improvement is often dramatic sometimes within two days of the therapy. Because its therapy is so steroid-related, early diagnosis is very important lest the irreversible changes occur. Our patient is a 50 year old man, complaining of cough and progressive dyspnea, that did not respond to antibiotherapy. After our clinical and cytological examinations, we diagnosed him as BOOP. Our diagnosis was supported with the steroid therapy response in the first month.

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Key words: *BOOP, bronchiolitis obliterans, cryptogenic organizing pneumonia, cough, progressive dyspnea, steroid therapy*

Abbreviations: *BOOP: Bronchiolitis obliterans with organizing pneumonia, BAL: Bronchoalveolar lavage, DLCO: Carbonmonoxide diffusion capacity, IPF: Interstitial pulmonary fibrosis, PNL: Polymorphonuclear leucocyte*

Introduction

Bronchiolitis obliterans with organizing pneumonia (BOOP) was first described by Lange in 1901 (1,2). It is generally idiopathic and the most common symptoms on presentation are, progressive dyspnea and nonproductive persistent cough. The characteristic histopathological lesion is excessive proliferation of granulation tissue within terminal bronchioles and alveolar ducts along with a fibroblastic growth through the alveoli. The lesion dramatically resolves with steroid therapy (3,4).

Commonly, patchy and/or reticular infiltration is seen in bilateral middle and inferior zones. The lesions may be migratory. The ground glass opacities seen on computerized tomography are also typical (5).

The patients have a restrictive type defect in the respiratory functions and a decrease in the carbonmonoxide diffussion capacity (DLCO) is also seen (6).

The diagnosis is mostly made by great biopsy materials taken by open lung biopsy. Rarely, by the help of bronchoalveolar lavage and transbronchial forceps biopsy, the diagnosis may also be made.

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The pathological definition is very important for the diagnosis to be confirmed. In tissue biopsy materials, the mononuclear cell infiltration and obliteration by fibroblasts and organizing connective tissue in the distal bronchioles, alveolar ducts and alveoli must be seen.

In bronchoalveolar lavage (BAL) the CD4/CD8 ratio and the proportion of macrophages decrease while the proportion of lymphocytes, eosinophils, neutrophils, mast cells and plasma cells increase. The other BAL abnormalities include foamy macrophages, mast cells and plasma cells.

Before calling it as BOOP, bronchiolitis obliterans, atipic pneumonia, idiopathic pulmonary fibrosis, (IPF) eosinophilic pneumonia, bronchioloalveolar carcinoma and extrinsic allergic alveolitis must be differentiated. In bronchiolitis obliterans pathological report is different and there's an obstructive ventilatory defect. It can be differentiated from atipic pneumonia by its being nonresponsive to the antibiotherapy, from IPF by high lymphocyte proportion in BAL (in IPF lymphocyte proportion is low), from eosinophilic pneumonia, by a lower level of eosinophilia, and from bronchioloalveolar carcinoma by not having any malignant cell.

Appropriate therapy is 1mg/kg of prednisone per day for 1-3 months and decreasing the dose by 10 mg per month, to 10-20 mg per day and complete the therapy to 9-12 months. Radiological improvement is seen earlier than the functional, and BAL findings are the last to recover. Early cessation of the therapy makes the patients susceptible to recurrences.

Case Report

Our patient is a 50 years old male, living in Adapazari. At the time of his admission, he complained of cough with white sputum, and exercise dyspnea of a two months' time. He had no hemoptysis or chest pain. Before attending our center, he had taken antibiotherapy for 20 days regularly and as no improvement in the clinical status and in the chest radiographs is seen by the doctors he was reffered to our hospital. He was a driver before his retirement. He had no other illness and operations or accidents. His mother had died from lung cancer. He used no cigarettes or alchohol or any drugs. He did not have any other systemic disorders.

At the time of his referral the vital signs were normal. He had no fever. Edema, icterus, cyanosis, clubbing were not seen and none of the peripherical lymh nodes were palpable.The cardiovascular examination was normal. The end inspiratory fine crackles were the only respiratory system findings. The other systems were found normal. His chest roentgenogram revealed an acinar pattern, utmost at the inferior zones of each lung. In his thorax computerized tomography (CT), bilateral basilar alveolar infiltration was seen.

His hemogram and routine biochemical results were normal. The erithrocyte sedimentation rate was 115 mm/hr.

The diseases resulting in bilateral acinar pattern was considered and further evaluation was done. Angiotensin converting enzyme was 26. Serum Ca:10.4 mg/dl urine Ca:0.13 mg/24h (N:0.1-0.4). Differential leucocyte count: segment 76%, lymphocyte 14%, eosinophil 10%. PPD (-). Anti HIV (-), anti HbsAg (-). EKG was normal. In the spirometer FEV1: 3.11 (87%), FVC: 3.66 (81%),

FEV1/FVC: 85%. Arterial blood gases were: pH:7.46, pO2: 83 mmHg, pCO2: 34 mmHg, sat.:96%

Fiberoptic bronchoscopy was performed. Hyperemia and the increase of capillarity was seen especially on the basal segments' mucosa. There were no endobronchial lesions. BAL was performed in the left basal segments because there were more lesions there than the other sites on CT. The cells were counted and CD4/CD8 ratio was calculated. Transbronchial biopsy was performed from the left 9th and the 10th segments and sent to the cytologic examination, just like the biopsy taken from the left common bronchial mucosa. The catheter biopsy material was sent to mycobacterial and routine bacterial cultures.

The BAL results were 83% alveolar macrophage, 12% lymphocyte 3% PNL, 2% monocyte. No pathogen bacteria was seen in the culture media and in the direct miroscopic and cultural investigation of mycobacterium tuberculosis. From the cytologic examination of transbronchial biopsy, thickenning of the alveolar walls, connective tissue proliferation filling some alveolar lumens, the infiltration of lymphocytes, histiocytes and few polymorphonuclear leucocytes in the alveolar walls and some foamy histiocytes were recorded.

As the patient's cough and dyspnea and the radiologic patterns did not respond to the antibiotherapy and as the cytologic result confirmed BOOP, we began to give 1mg/kg/day oral methylprednisolone to the patient and discharged him with the plan of monthly control by chest x-ray and clinical examination. After eight weeks at this dose, we gradually decreased the dose to a continuous

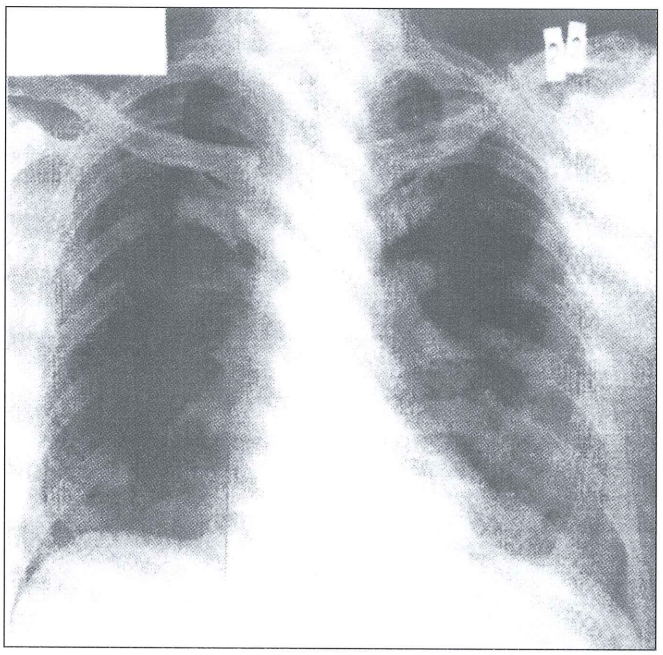

Fig. 1. The posteroanterior graphy before therapy

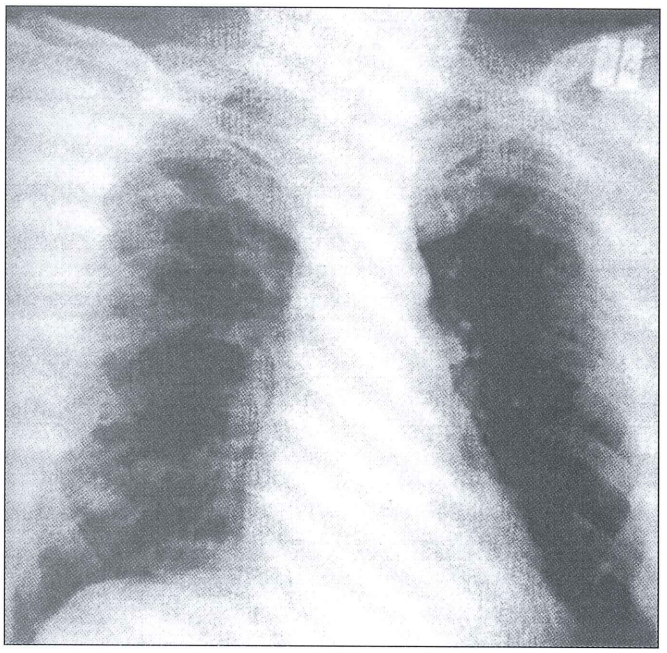

Fig. 3. The posteroanterior graphy after seven months of therapy

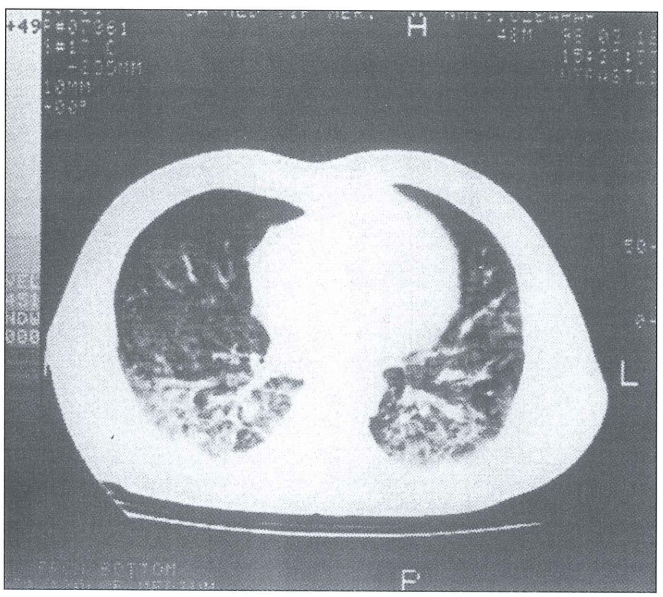

Fig. 2. The computed tomography scan before therapy

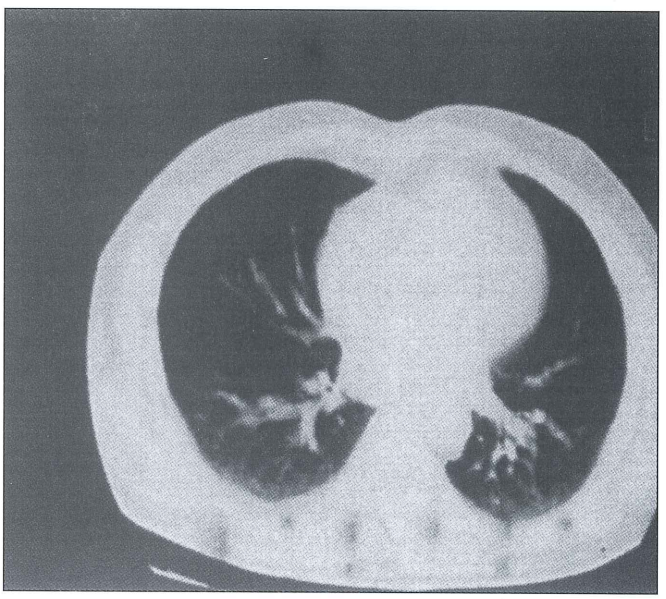

Fig. 4. The computed tomography scan after seven months of therapy

dose of 16 mg/day. Six months later, the dose was slowy reduced and in twelveth month the therapy was stopped.

Discussion

Our case can be a typical example of BOOP. For the patients with persistant cough and alveolar infitration in the radiograms and who do not respond to any antibacterial therapy, BOOP must be one of the first diseases we must think of. Most of them have a story of slowly resolving viral pneumonia in weeks or months (7). In our case, our patient's complaints did begin two months before.

Epler and associates (3) described bilateral patchy infiltration and ground glass pattern in 81% of the patients. King and associates (8) reported bilateral diffuse alveolar opacities in 79% of the patients. Other radiologic imaging patterns are miliar and miliar-nodular pattern.Our case had bilateral inferior lobe infiltration.

All of the laboratory results including spirometry and DLCO did correlate with the ones in the literature (9,10,11).

As in almost all of the patients with BOOP, clinical and

physiological improvement is seen with steroid therapy, which is another confirmation for the diagnosis. Generally, open lung biopsy is needed to distinguish BOOP from irreversible interstitial lung diseases. Transbronchial lung biopsies are generally inadequate in confirming BOOP and ruling out the other disorders. Small biopsies can miss the definite diagnosis, stepsectioning of transbronchial biopsies is useful in identifying the lesions of BOOP (2). But in our case the bronchoscopic material was helpful for the pathologist to confirm our diagnosis.

We conclude that the early diagnosis of BOOP is very important for its dramatic response to steroid therapy. We must treat it, before the irreversible changes occur in the lung. For that reason, we must always keep in mind this situation while differentiating the etiology of cough and dyspnea.

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