Case Report

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The case of idiopathic pleuroparenchymal fibroelastosis diagnosed by transbronchial biopsy

Huseyin Arpag, Muhammet Sayan[1], Nurhan Atilla, Fulsen Bozkus, Hasan Kahraman, Abdulkadir Yasir Bahar[2], Mahmut Tokur[1]

Abstract:

Pleuroparenchymal fibroelastosis (PPFE) is claasified in idiopathic interstitial pneumonitis group is rarely seen. Etiology and pathophysiology of this entity are not completely understood. The prognosis is poor, and there is no effective treatment except for lung transplantation. Here, we presented a case of PPFE diagnosed by transbronchial biopsy through fiberoptic bronchoscopy and its histopathological and clinicoradiological features.

Keywords:

Idiopathic, pleuroparenchymal fibroelastosis, transbronchial biopsy

Introduction

Pleuroparenchymal fibroelastosis (PPFE) is an idiopathic interstitial pneumonitis. This rare entity characterized by fibroelastic the pleural and subpleural lung parenchymal thickening particularly located in the upper lobes of lungs. The prognosis is poor and there is no effective treatment except for lung transplantation. There is a very few information about etiology and pathophysiology of PPFE.[1-3] Here, we presents a case of PPFE.

Case Report

A 54-year-old, nonsmoker farmer male admitted to clinic with cough and for dyspnea complaints. He had no known respiratory, rheumatologic disease, or tuberculosis. There was general decreased respiratory sound on thorax auscultation. Clubbing and pretibial edema were not detected. The oxygen saturation was measured as 98% with finger probe in the room air. His body mass index was 18.1 kg/m^2. Right tracheal deviation, bilateral apical fibrotic areas, increment in bilateral reticular trails, and blunting in bilateral costophrenic angle were seen on chest X-ray and thorax computerized tomography [Figure 1]. Mix pattern was detected on pulmonary function test (forced vital capacity; 2.25 L [60%], forced expiratory volume1; 1.23 L [41%], and Tiffeneau index; 55%). There was no hypercapnia in the arterial blood gas and the oxygen pressure was 82 mmHg. He had no known occupational-environmental exposure and familial lung diseases and his rhematological markers were negative. Informed consent form included intervention and publishability of his medical records was obtained from the patient then fiberoptic bronchoscopy and transbronchial biopsy was performed. The results of acid-fast bacilli tests and tuberculosis culture (Löwenstein–Jensen medium) from sputum samples were negative. Final pathology report revealed that alveolar collapse, thickening of alveolar septum, and peribronchial hyalinization were observed. Elastic fiber accumulation was detected in the alveolar septum and

Departments of Chest Diseases, [1]Thoracic Surgery and [2]Pathology, Faculty of Medicine, Kahramanmaras Sutcu Imam University, Kahramanmaras, Turkey

Address for correspondence:
Prof. Muhammet Sayan, Department of Thoracic Surgery, Faculty of Medicine, Kahramanmaras Sutcu Imam University, Avsar Campus, Onikisubat 46100 Kahramanmaras, Turkey.
E-mail: drsayann@gmail.com

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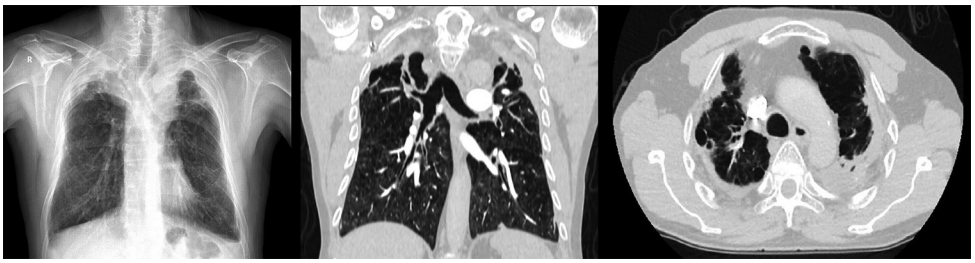

Figure 1: Chest X-ray and computed tomography revealed; right tracheal deviation, bilateral apical fibrotic areas, increment in bilateral reticular trails, and blunting in bilateral costophrenic angles

air spaces with the histochemically applied orcein stain [Figures 2 and 3]. The patient was diagnosed as idiopathic PPFE according the clinicoradiological and histopathological findings.

Discussion

PPFE is a rare condition and characterized by fibroelastic thickening in the pleura and subpleural lung parenchyma especially in the upper lobes.[1] It was first reported in 2004 by Frankel *et al*.[2] In recent years, PPFE was included into idiopathic interstitial pneumonias (IIP) group by the international multidisciplinary classification.[1-3] There is a very few information about etiology and pathophysiology of PPFE. Although PPFE is been usually idiopathic, it may be associated connective tissue diseases, lung transplantation, bone marrow transplantation, or genetic condition in some cases. Our case was considered as idiopathic. Clinical manifestations are vary but the most common complaints of patients with PPFE are dypsnea and nonproductive cough.[4] Our patient had exertional dyspnea and dry cough symptoms. Thorax computerized tomography findings of PPFE are usually characterized by bilateral fibrosis, bronchiectasis, and subpleural reticular pattern located especially upper zones.[1,2] Similar radiological findings were detected in our case. Usually, restrictive pattern is observed in pulmonary functions tests.[5] However, the combination of restriction and obstruction (mix pattern) was seen in ours. Histopathologically, it is characterized by collagen fibrosis, septal elastosis, and elastin accumulation, especially in subpleural space and intraalveolar space.[3,6-8] Elastin accumulation was shown through the orcein stain in tissue sample taken through transbronchial biopsy in our case. Although the definite diagnosis of PPFE is surgical lung biopsy, it is not preferred by physicians due to the bad general condition of these patients and complications of operation, especially pneumothorax and prolonged air leak.[6] Therefore, less invasive diagnostic procedures such as transthoracic biopsy, bronchoscopic transbronchial lung biopsy, or transbronchial crio biopsy which is used in increasingly day-by-day

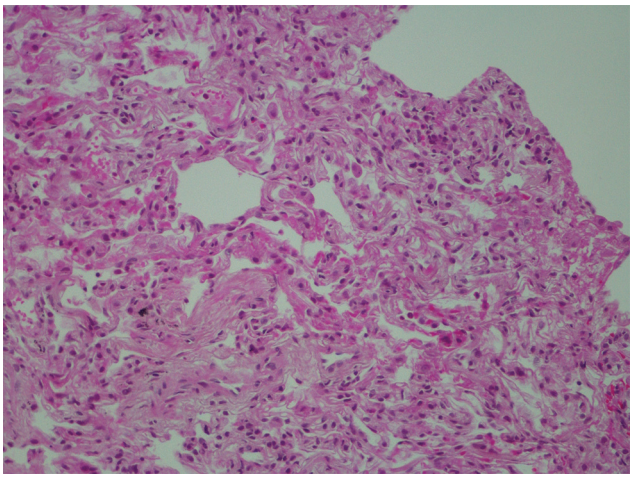

Figure 2: Markedly fibrosis and increased chronic inflammatory cells in the interstitial area (H and E, ×200)

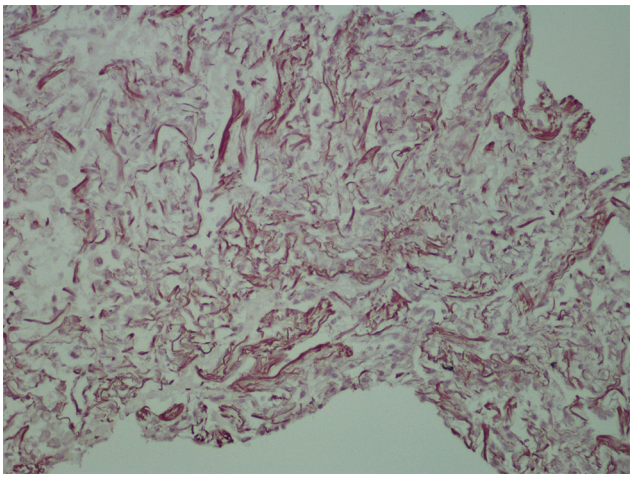

Figure 3: Positive reaction with orcein stain in elastic fibers (dark red)

are recommended for diagnosis of diseases.[1-3,6] Despite immunosuppressive therapy is used in some patients, definitive treatment currently known is lung transplantation.[7] Bronchodilator therapy is started for airway obstruction to our case and he referred to lung transplantation center.

The less invasive procedures should be preferred for diagnosis of PPFE which is classified in IIP group so possible complication can be prevented.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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