Two Cases of Idiopathic Mediastinal Fibrosis Associated With Obstructive Lung Disease

Halil Yanardag 1, Tetikkurt Cuneyt2*, and Muammer Bilir1

1Department of Internal Medicine, Cerrahpasa Medical Faculty, Istanbul University
2Department of Pulmonary Medicine, Cerrahpasa Medical Faculty, Istanbul University

Abstract

Idiopathic mediastinal fibrosis is a rare benign disease of unknown etiology. The disease is characterized by abnormal proliferation of fibrous tissue leading to obliteration of mediastinal structures. Abnormal immunologic response to granulomatous infections like mycobacterium tuberculosis and histoplasma capsulatum is the most common etiologic factor leading to fibrosing mediastinitis.

We present two cases of idiopathic mediastinal fibrosis with atypical clinical features. The first patient had chronic obstructive pulmonary disease and the second patient had asthma in addition to idiopathic mediastinal fibrosis. Both patients presented with dyspnea due to past airway disease that resolved with bronchodilator treatment. Comorbid diseases may predominate the clinical picture in patients with fibrosing mediastinitis causing a crucial predicament in the absence of specific features relevant to the existence of a significant mediastinal fibrous tissue load. Symptoms of comorbid airway disease may overlap with symptoms of mediastinal fibrosis leading to a diagnostic impasse. Normal chest X-ray does not rule out fibrosing mediastinitis in such cases. In patients without a pathognomic clinical profile fibrosing mediastinitis is diagnostic dilemma for the clinician.

Keywords: Mediastinal fibrosis; Idiopathic mediastinal fibrosis; Fibrosing mediastinitis

Introduction

Fibrosing mediastinitis is a disorder characterized by the proliferation of dense fibrous tissue in the mediastinum. Fibrosis may compromise the vital mediastinal structures leading to symptoms pathognomonic for the syndrome. The morbidity is precisely associated with the location and the bulk of fibrous tissue. The pattern of vital mediastinal involvement by fibrous tissue is variable. Consequently, the clinical features and the outcome of fibrosing mediastinitis is relevant with the load of fibrous tissue that compromises the mediastinal structures [1-4].

We present two cases of idiopathic fibrosing mediastinitis presenting with dyspnea due to their previous obstructive airway disease. Fibrosing mediastinitis has not been described simultaneous to chronic obstructive lung disease and asthma. Clinicians should bear in mind that the symptoms of previous comorbid disease in fibrosing mediastinitis may predominate the clinical picture and cause a diagnostic impasse.

Case 1

A 57 year old male with a 40 pack/year smoking history was admitted for dyspnea in exertion for three months. Past medical history included chronic obstructive pulmonary disease, type II diabetes mellitus, bacterial pleurisy, and hypertension. His father had died of myocardial infarction, his mother had hypertension, and his sister had type II diabetes. Blood pressure was 110/65 mm Hg. Physical examination did not reveal any pathologic findings. Complete blood count and biochemistry were within normal limits.

Urine analysis and rheumatologic markers were unremarkable. Tuberculine test was negative. ECG was showed a sinus rhythm of 84/minute. Chest X-ray showed fibrotic pleural changes in the right costal pleura, blunting of the right costodiaphragmatic sinus and enlargement of the right upper mediastinum (Figure 1). Pulmonary function tests demonstrated moderate obstructive lung function with FEV1: 62% and FEV1/FVC: 68% of the predicted. Following bronchodilator and steroid inhaler treatment, the patient had resolution of symptoms. Computed tomography revealed abnormal fibrous tissue in the mediastinum and pleural thickening (Figure 2,3). Magnetic resonance imaging showed compact fibrotic tissue in the mediastinum (Figure 4). Sputum culture for mycobacterium
tuberculosis and fungi were negative. Bronchoscopic examination revealed normal airways and culture of bronchial lavage was negative for bacteria, tuberculosis and fungus. Histopathologic examination of the mediastinoscopic biopsy specimen was compatible with idiopathic mediastinal fibrosis revealing cellular fibrous reaction with plasma cells and eosinophils. There was no history of mycobacterial or fungal infection, fibrosing conditions, radiotherapy and drug intake that might be associated with the histopathologic findings of excessive fibrotic reaction. The final diagnosis was idiopathic mediastinal fibrosis.

**Case 2**

A 41 year old non-smoker was admitted for headache and dyspnea. Past medical history was significant for asthma from fifteen years. Her mother had hypertension, her father died of lung cancer, and her aunt had sarcoidosis. Physical examination did not reveal any pathologic findings. Blood pressure was 120/70 mm Hg. Complete blood count, biochemistry, urine analysis, and rheumatologic markers were within normal limits. ECG was showed a sinus rythm of 74/minute. Tuberculine test was negative. Chest X-ray was normal (Figure 5). Pulmonary function tests showed reversible moderate obstructive defect with FEV1: 58% and FEV1/FVC: 64% of the predicted. The symptoms improved with bronchodilator treatment. EMG was normal and did not reveal any pathologic findings. Sputum culture for mycobacterium tuberculosis and fungi were negative. Computed tomography revealed abnormal fibrous tissue with calcifications in the right paratracheal and subcarenal area surrounding the trachea, both main bronchi, and vena cava superior (Figure 6). Bronchoscopic examination showed normal airways. Culture of bronchial lavage was negative for bacteria, tuberculosis and fungus. Histopathologic examination of the mediastinoscopic biopsy samples revealed chronic inflammatory infiltrate and cellular fibrous reaction rich in plasma cells. The final diagnosis was compatible with idiopathic mediastinal fibrosis because there was no history of granulomatous infection, fibrosing diseases, drug treatment, and radiotherapy.

**Discussion**

Fibrosing mediastinitis is a rare disorder characterized by proliferation of abnormal fibrous tissue in the mediastinum. The majority of the cases are associated with granulomatous infection including tuberculosis and histoplasmosis [1,2]. Idiopathic fibrosing mediastinitis develops as an autoimmune reaction and is commonly associated with other fibrosing conditions such as retroperitoneal fibrosis, primary sclerosing cholangitis, orbital pseudotumors, and Riedel’s thyroiditis [5,6]. Dense fibrous tissue is usually located in the upper mediastinum but diffuse fibrosis may also occur extending from brachiocephalic veins to the lung bases [7]. Mediastinal fibrosis may be self-limiting or lead to serious complications by compromising the airways, vessels, and other structures. Clinical manifestations develop as the fibrotic process compresses or obliterates mediastinal structures.
Both of our patients presented with clinical features associated with their past obstructive airway diseases while there was not any specific symptom associated with the fibrosing mediastinitis. Symptoms of fibrosing mediastinitis arise from the compression of airways or vessels while 40% of the patients are asymptomatic at presentation [8–10]. This silent clinical picture of fibrosing mediastinitis may be explained by the fact that the patients tend to be asymptomatic in the exudative stage of the inflammation. Location and the load of fibrotic tissue are other crucial components of symptom arousal. The manifestations of fibrosing mediastinitis may become apparent following invasion of mediastinal structures by dense fibrotic tissue as related to the involvement of vessels, airways, esophagus or mediastinal nerves [11–12]. It may bring forward that the patients with fibrosing mediastinitis may be asymptomatic at the initial exudative stage of the inflammation. Chest X-ray was abnormal in one of our patients revealing fibrotic lesions associated with previous disease and completely normal in the other. Computed tomography images identified a significant load of compact fibrotic tissue while compression or obliteration of vital mediastinal structures were not observed in our patients revealing that the clinical profile of both patients was due to previous obstructive airway disease.

Approximately half of the patients with fibrosing mediastinitis are asymptomatic and the disease is discovered incidentally by chest radiography. In the reminder, a variety of symptoms may occur due to the compression of vital mediastinal structures by dense fibrous tissue reaction [8,9,11]. Another clinical profile is the presence of predominant symptoms of previous comorbid disease that is not associated with mediastinal fibrosis as it is the case in our patients. The asymptomatic presentation in fibrosing mediastinitis is probably associated with the exudative initial stage of inflammation, location of fibrotic tissue, or a low burden of fibrotic tissue sequela on the mediastinal structures. Computed tomography and pathologic examination identified dense fibrotic tissue in both cases thereby eliminating the active inflammation stage in these patients. Response to bronchodilator treatment indicates that the symptoms in our patients were relevant to past obstructive airway disease and were not due to fibrosing mediastinitis.

In both patients, the presenting clinical profile was relevant to antecedent obstructive lung disease and responded well to bronchodilator treatment. The chest X-ray was normal in one patient and did not point out to fibrosing mediastinitis while the computed tomography and magnetic resonance imaging identified the fibrotic tissue in the mediastinum. There may be an overlap of symptoms of mediastinal fibrosis and obstructive airway diseases. CT with MRI may provide useful data revealing the extent and the load of mediastinal fibrotic tissue comprising the vital structures in such cases. Resolution of dyspnea with bronchodilator treatment indicates that the clinical symptom profile was associated with past obstrucive lung disease since CT did not reveal any compression of airways that may be the cause of dyspnea in both patients. Clinicians should bear in mind that the symptoms of past comorbid conditions may prevail the clinical picture or overlap with mediastinal fibrosis thereby hindering the diagnosis. Specific symptoms of vital organ compression may not be apparent due to the initial exudative inflammatory stage of mediastinal fibrosis or to the absence of excessive fibrotic tissue in fibrosing mediastinitis. Idiopathic mediastinal fibrosis may exist simultaneous to obstructive airway disease.

Conflict of Interest

Authors declared no conflict of interest to disclose

References