Idiopathic Mediastinal Fibrosis Imitating Lung Cancer

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Abstract

Idiopathic mediastinal fibrosis is an occasionally observed disease of mostly unknown etiology characterised by slowly progressive fibrosis and exuberant collagen formation within the mediastinum. This can result in clinical syndromes causing extension to and compression of the mediastinal structures.

In Europe, this disease is exceptionally rare. More cases are seen in USA where the disease may often be associated with fungal infections that can be rarely identified.

We present a young male patient who was at first thought to have pulmonary malignancy but later shown to have idiopathic mediastinal fibrosis after a five-year follow-up.

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Key words: *Idiopathic mediastinal fibrosis, therapy*

Introduction

Idiopathic mediastinal fibrosis is an uncommon disease of uncertain etiology resulting in the deposition of dense fibrous tissue throughout the mediastinum (1,2). The process imitates clinically and radiographically malignant tumors. It can lead to entrapment and compression of the various structures of the mediastinum (2,3). The superior vena cava is most often compromised, but pulmonary arteries and veins, and bronchi can also be involved (1,2). Surgical exploration of the mediastinum will often be necessary to establish the diagnosis and to rule out malignancies (4). We report a case who presented with a mediastinal mass, subsequently confirmed to be idiopathic mediastinal fibrosis.

Case Report

A 29-year-old male patient was admitted to the hospital with complaints of dyspnea, cough, sputum, hemoptysis, weight loss and chest pain in November 1994. He smoked 10 pack/year. His physical examination was normal except for the reduction of the breathing sounds on the right hemithorax.

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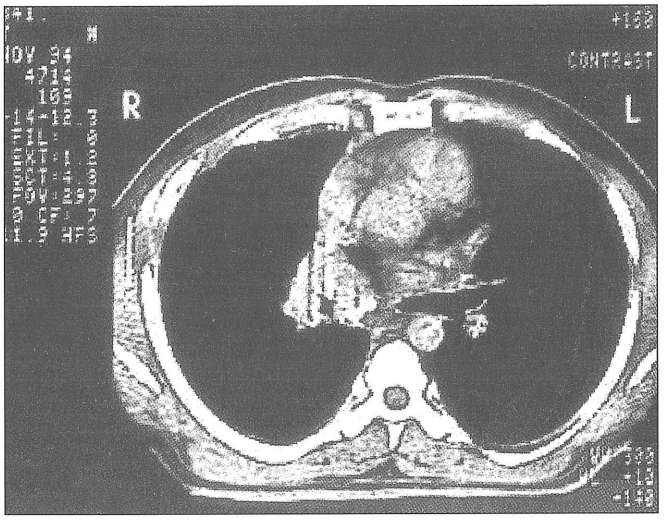

Figure 1. *Chest CT scan in November 1994. A 4.5 cm mass lesion surrounding the right main bronchus*

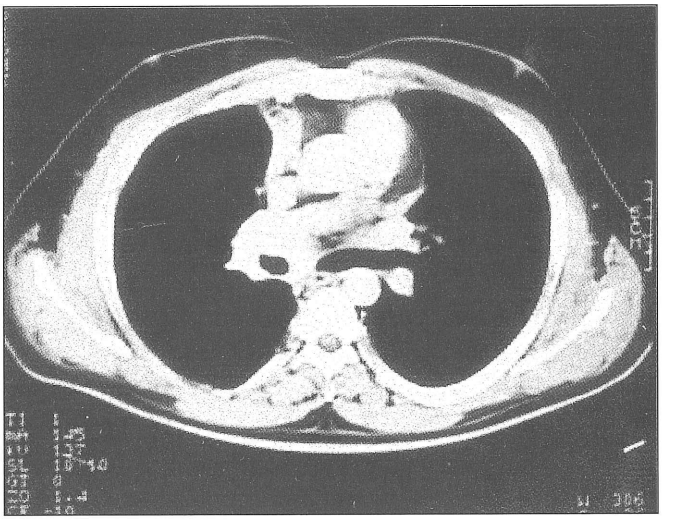

Figure 2. *Stable mass lesion on CT scan after 5 years*

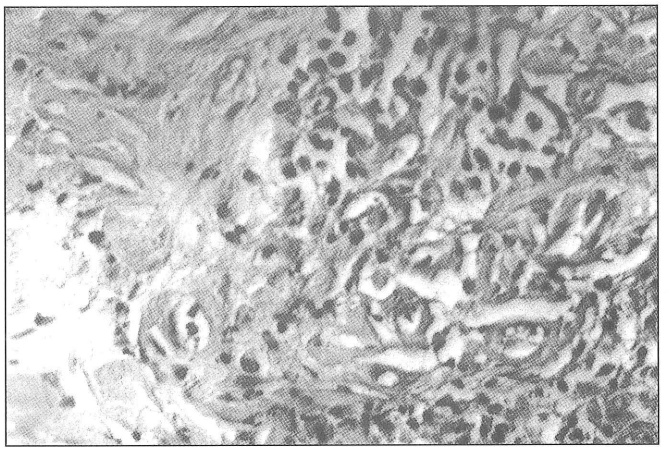

Figure 3. *Hyalinized fibrous tissue obtained by thoracotomy (Hematoxylin-Eosin stain, X 400)*

Complete blood count, routine biochemical analysis and urine analysis were normal. Sputum smears and cultures for acid-fast bacilli and fungi were negative for six times.

On admission, the chest x-ray revealed a right pulmonary infiltration extending to the right hilus. Thoracic computerized tomography (CT) scans revealed a 4.5 cm mass lesion surrounding and narrowing the right main bronchus (Figure 1). Brain CT scans and ultrasonographic examination of the abdomen were normal. Fiberoptic bronchoscopy was performed; narrowing and submucosal infiltration was observed in the right middle lobe bronchus. Biopsies were performed. The result of the pathological examination was reported as unsatisfactory material, though it suggested small cell lung carcinoma.

Right thoracotomy was performed. A hard, fixed, nodulary lesion extending to the mediastinum, surrounding oesophagus and the other mediastinal structures was observed. Biopsies were taken from the lesion, paratracheal, supradiaphragmatic lymph nodes and pleura. Pathological examination of the specimens revealed idiopathic mediastinal fibrosis (Figure 3). Subsequently, the patient was examined for multifocal idiopathic sclerosis (mediastinal fibrosis, retroperitoneal fibrosis, orbital pseudotumor, Riedel's thyroiditis). There was no evidence of these disorders on ultrasonography of the abdomen, ophthalmologic examination, and ultrasonography of the thyroid, respectively.

Corticosteroid therapy was initiated. Prednol 40 mg was started and continued in a decremental fashion for 1.5 months.

The patient has been followed for 5 years. New CT scans obtained show no progression of the lesion and the patient has no complaints to date (Figure 3).

Discussion

Idiopathic mediastinal fibrosis (syn: sclerosing mediastinitis, fibrosing mediastinitis, granulomatous mediastinitis, fibrous mediastinitis, chronic fibrosing mediastinitis, mediastinal granulomas, fibroinflammatory lesions) represents a rare chronic inflammatory or inflammatory-like process, resulting in the deposition of dense fibrous tissue typically located in the superior mediastinum near the bifurcation of the trachea as well as in the pulmonary hilar and often extending into the adjacent structures (1).

In Europe, this disease is rare, but in the United States more cases are seen and is often associated with fungal infections, particularly histoplasmosis (1). Histoplasmosis

is almost unknown in Europe and other infectious diseases can rarely be identified (1). Some cases were associated with radiotherapy and radiofrequency radiation exposure (2,3). In our patient there was no prior radiotherapy, thoracotomy, known infection or amyloidosis. He had also no evidence of silicosis, autoimmune factors, familial factors or collagen vascular disease.

The presentation of the disease is variable; many patients present with respiratory symptoms like cough, wheezing, dyspnea, hemoptysis or superior vena cava syndrome. Therefore, asthma, malignancy, chronic bronchitis, superior vena cava syndrome associated with other disease entities must be considered in differential diagnosis (4,5). These aspects were investigated in our patient too. He had symptoms mimicking malignancy.

In some patients retroperitoneal fibrosis, orbital pseudotumor, Riedel's thyroiditis are also seen along with idiopathic mediastinal fibrosis (6). Our patient was also investigated for these aspects, but no associated disorder has been found.

Radiographical findings may be distinctive. The findings include: bronchial narrowing, pulmonary artery obstruction, oesophageal narrowing and superior vena cava obstruction (7). In our patient, thoracic CT scans revealed bronchial narrowing in the right main bronchus.

Currently no definite therapy is present for idiopathic mediastinal fibrosis. Antifungal therapy and if needed, surgical procedures may be performed in cases secondary to histoplasmosis (8). When etiology is unknown, steroid therapy is given, although there is no evidence about its benefits (2,4,5). Our patient has also been given steroid therapy for 1.5 months in a decremental fashion.

Prognosis is variable. In idiopathic mediastinal fibrosis the required surgical procedures may carry a high mortality risk. Our patient has been followed for 5 years and he still has no complaints or signs of progression of the disease.

References

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