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Fibrosing mediastinitis: An unusual cause of superior vena cava symptoms

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Case Report

Abstract

Fibrosing mediastinitis is a rare benign disorder caused by the proliferation of acellular collagen and fibrous tissue within the mediastinum. Although many cases are idiopathic, many (and perhaps most) cases are thought to be caused by an abnormal immunologic response to *Mycobacterium tuberculosis* and *Histoplasma capsulatum* infections. Collagen formation leads to compression of vital structures, resulting in cough, chest pain, and dyspnea. The following case is a former healthy middle-age man who presented with an 8-year history of cough, chest pain, facial swelling, and trouble breathing, and was subsequently found to have fibrosing mediastinitis. Fibrosing mediastinitis should be considered in the differential diagnosis of cough, chest pain, and dyspnea, primarily when findings such as increased venous pressure are present on physical exam, and hilar abnormalities are seen on chest radiograph.

KEYWORDS: Fibrosing Mediastinitis, *Mycobacterium tuberculosis*, Superior Vena Cava Syndrome

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Introduction

Fibrosing mediastinitis also known as granulomatous or sclerosing mediastinitis is an uncommon benign disorder characterized by the proliferation of dense fibrous tissue within the mediastinum.[1] Osler in 1903 published the first major review on mediastinal fibrosis.[2] Since then, a few case series have been published with stray case reports.[3,4] Mediastinal structures are surrounded, constricted and sometimes invaded by the fibrous tissue, which may extend to affect other intrathoracic organs. The pattern of involvement of mediastinum is variable, and so are the clinical features.[5]

Venous structures (superior vena cava and pulmonary veins), due to thin walls and low intraluminal pressure, tend to be compressed earlier than the arteries and tracheobronchial tree and esophagus. For this reason, venous hypertension in the drainage area of superior vena cava is considered the hallmark of the disease. It is a rare cause of superior vena cava obstruction (1-2%).[6] Various hypotheses have been proposed for the fibrous lesions. Radiological, computed tomography (CT), and magnetic resonance imaging features help in diagnosis; however, histological confirmation is difficult. Medical therapy is discouraging; surgery has limitations while stenting of vessels and dilatation of bronchi and esophagus may provide some relief of disease.[3]

Case Report

A 47-year-old male, Iraqian by nationality attended the clinic due to 8-month history of dyspnea on minimal exertion, orthopnea, cough with scanty mucoid-white sputum and

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sometimes bloody expectoration, dull aching, and nausea. He described an 8-year history of progressive puffiness of face, swelling over both upper limbs and dilated neck vein, more after getting up in the morning and decreased over day. There was no history of fever, feet edema, urinary complaints, change in voice, and palpitation. Past history was not contributory. There was no history of drug intake or exposure to radiation. However, patient had a history of active smoking (15 pack/years). Personal and family histories were not contributory.

Clinical examination revealed blood pressure of 120/70 mmHg, pulse of 90/min, respiratory rate of 20/min, and temperature of 36.9° C. He had experience of puffiness of conjunctiva, face, and both upper limbs. The jugular veins were full and non-pulsatile, prominent veins were seen over arms, chest and anterior aspect of neck with flow away from mediastinum (Figure 1). Respiratory system examination revealed bilateral rhonchi and crepitations. Rest of the general physical and systemic examination was unremarkable.

Complete blood count revealed hemoglobin of 11.8 g%, a white blood cell of 4.9×10^9/l with 83% polymorphs, 13% lymphocytes, and normal platelets. Serum biochemical parameters were normal. On investigation, his electrocardiogram and two-dimensional echocardiogram were normal. Sputum for acid-fast bacilli and purified protein derivative test was negative.

The chest X-ray showed mild superior mediastinal widening (Figure 2). Chest CT demonstrated soft tissue density and intermingled calcifications adjacent to the superior vena cava, right pulmonary artery, and ascending aorta. The three-dimensional (3D) venographic reconstruction showed an extensive network of collateral circulation due to obstruction of the right and left brachiocephalic vein that extended toward the chest wall, upper limbs, and abdomen (Figure 3). No thrombi were observed in the vena cava. Based on these findings, a diagnosis of superior vena cava syndrome was made. There was no evidence of mediastinal mass or lymphadenopathy. Mediastinoscopy, mediastinotomy, or histopathological confirmation could not be done due to refusal for consent to undergo invasive interventions/procedure.

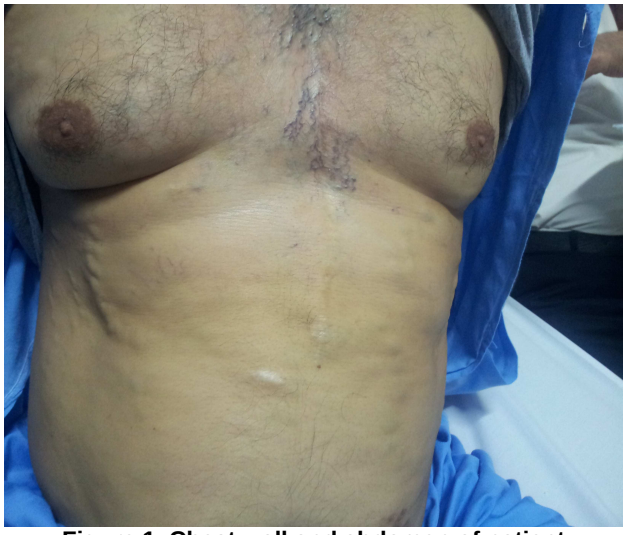


Figure 1. Chest wall and abdomen of patient demonstrated extensive engorgement of vessels

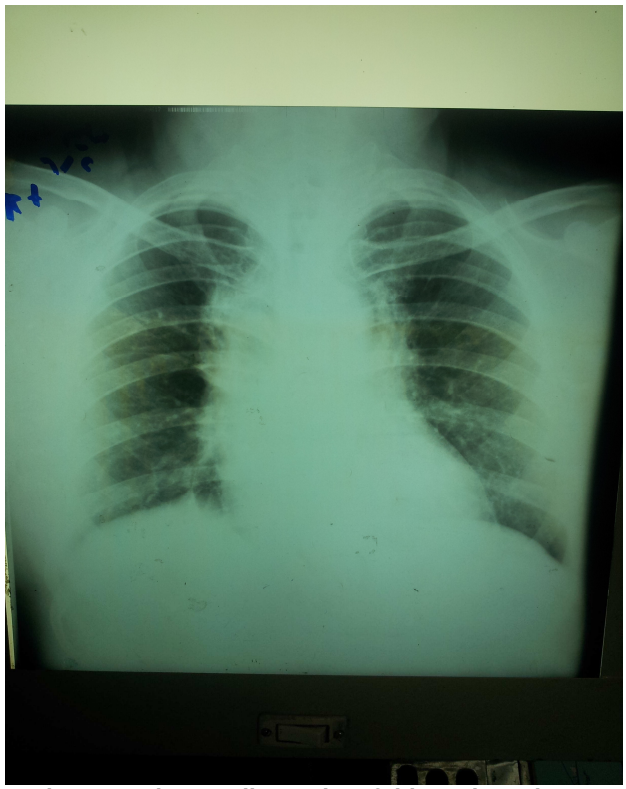
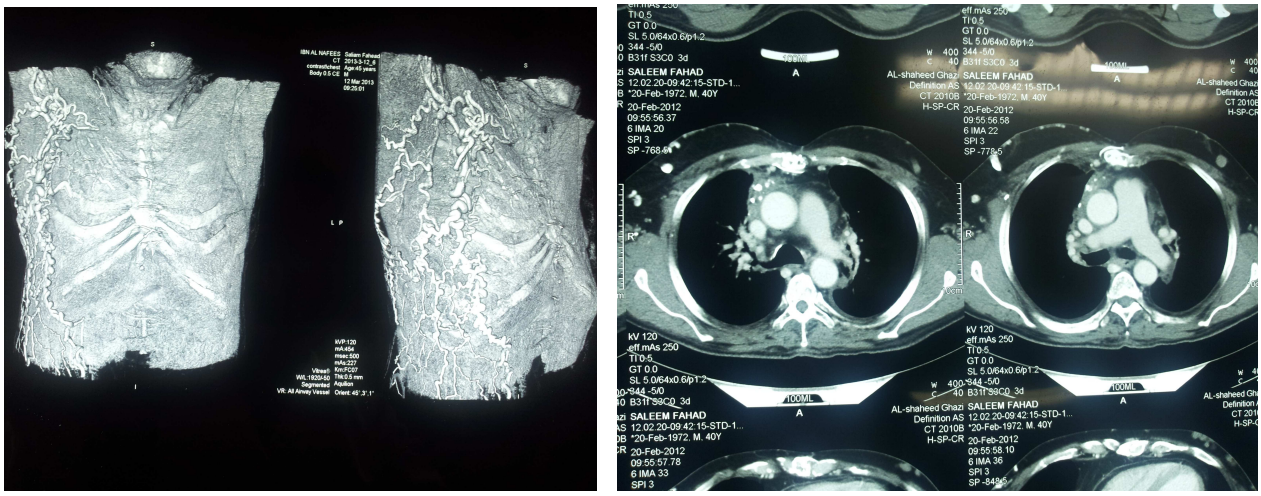


Figure 2. Chest radiography of this patient shows mediastinal widening



A **B**

Figure 3. Three-dimensional venographic and reconstruction computed tomography scan of the same patient; (A) Widespread arrangement of collateral circulation due to obstruction of the right and left brachiocephalic vein that extended toward the chest wall; (B) Soft tissue density and calcifications adjacent to the superior vena cava and great vessels

Discussion

Mediastinal fibrosis is a rare disease characterized by the proliferation of collagen tissue and establishment of fibrous tissue in the mediastinum.[1] In most cases, the cause of this process is unknown, although in endemic zones it has been related with *Histoplasma capsulatum* infection, specifically with an abnormal inflammatory response to the antigens of this fungus and, with other granulomatous diseases such as tuberculosis.[7] There is an idiopathic form with a possible autoimmune component that may be associated with fibrosing processes in other locations, such as retroperitoneal fibrosis, pseudo-tumor of the orbit and Riedel's thyroiditis. It affects young patients, with a slight predominance in males, and its symptoms are insidious and progressive, with a variable natural history.[8] The inflammatory process usually sets in the upper half of the mediastinum, in the paratracheal region. It is commonly located to the right, anteriorly to the trachea and close to the pulmonary hilum; however, it may also develop as a diffuse fibrosis in the mediastinum, extending from the brachiocephalic veins to the pulmonary bases. The symptoms are generally caused by obstruction of the superior vena cava, esophagus, trachea, bronchi, or pulmonary veins, also causing pulmonary arterial hypertension by direct compression of pulmonary arteries, or secondary to pulmonary venous compression.[9]

Investigations showed suggestive of idiopathic mediastinal fibrosis are mediastinal widening on chest X-ray and lung CT. Barium swallow and bronchoscopy may show areas of compression or distortion of the esophagus and bronchus. Venogram showing collaterals and mediastinal veins are diagnostic. Mediastinoscopy, mediastinotomy may be difficult, and histopathology may be confirmative and requires large sample of tissue if obtained surgically though rarely CT guided automated needle biopsy may be possible.[10]

Our patient presented with features of superior vena cava obstruction for which no clinical or radiological cause could be demonstrated; hence, a possibility of idiopathic mediastinal fibrosis was entertained as a diagnosis on exclusion of common causes, which was supported by the 3D angiographic study of the upper extremity and chest wall.

There is no curative treatment for this disease. Anti-fungal agents have been used in cases that may be related with histoplasmosis, although they

have not been effective.[11] The use of corticoids does not provide any benefit except in cases of autoimmune etiology, in which there may be a response. Therefore, therapeutic measures will be aimed at relieving obstructive symptoms in the airways, major vessels, and esophagus. When there is involvement of the vena cava, the placement of endovascular stents to the vessel is an option that produces a symptomatic improvement. Other techniques have been described, such as bypass surgery with saphenous vein grafts or bioprostheses.[12] We recommended palliation therapy such as endovascular stent in brachiocephalic vein to the patient. However, he did not submit this alternative management.

Conflict of Interests

Authors have no conflict of interests.

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