Computed tomography manifestations of fibrosing mediastenitis, an unusual cause of pulmonary symptoms: A case series

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MRK conceived idea, MRK KN drafted the study, NK collected data, NK did statistical analysis & interpretation of data, MRK NK ARK critical reviewed manuscript, All approved final version to be published.

Declaration of conflicting interests

The Authors declares that there is no conflict of interest.

Abstract

Background: Fibrosing mediastenitis is a rare benign condition in which fibrous tissue proliferates invasively and progressively in mediastinum. It has two types: focal granulomatous and diffuse non-granulomatous form. It presents with diverse clinical presentations depending upon its complications. Contrast enhanced computed tomography is useful modality for diagnosis and assessing the extent and severity of involvement.

Objective: The purpose of this study is to describe computed tomographic manifestations of fibrosing mediastenitis in eight pathologically proven cases of fibrosing mediastenitis.

Methodology: In this study we describe the computed tomography findings in eight cases of pathologically proven fribrosing mediastinitis. Clinical data regarding the presentation and suspected etiology were correlated with location of mediastinal disease, presence of calcification, contrast enhancement, effect on structures of mediastinum and additional associated pulmonary findings on computed tomography.

Results: The mean age of patients was 39 years, with two female and six male patients. Two patients had diffuse involvement of mediastinum and six patients presented with local mass. Calcification was present in five cases. There was no contrast enhancement in all eight cases. Six of eight cases revealed narrowing of mediastenal structures, with two cases showing pulmonary artery narrowing, three with superior vena cava obstruction, two with tracheal narrowing and one with pulmonary vein narrowing.

Conclusion: Fibrosing mediastenitis is an unusual cause for common complaints like cough, shortness of breath and chest pain. Familiarity with its different imaging features is crucial not only for accurate diagnosis as well as for planning noninvasive and surgical procedures. Computed tomography can play a vital role in its diagnosis, work up and follow up.

Key words: Fibrosing Mediastenitis; Computed Tomography; Mediastinum

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Introduction

Fibrosing Mediastenitis is also known as sclerosing mediastenitis. It is a rare benign condition in which fibrous tissue proliferates invasively and progressively.[1] Age range for fibrosing mediastenitis is between 13 and 65 years with female predominance.[2]

It has two types: focal granulomatous and diffuse non-granulomatous form. Non granulomatous is result of autoimmune reaction to autoimmune

syndromes, drugs like methysergide, and post radiation. Frequent association is established with fibrosing conditions like retroperitoneal fibrosis, Behcet disease, primary sclerosing cholangitis etc.[3] Granulomatous fibrosing mediastenitis is due to idiosyncratic fibrous tissue proliferation in response to antigenic stimulation to histoplasma capsulatum.[4] Other causes include tuberculosis, aspergillosis, mucormycosis, sarcoidosis etc.[5-7]

Fibrosing mediastenitis presents with diverse clinical presentation depending upon complications due to compression of vascular structures, esophagus and pulmonary structures.[8] Chest X-ray may show nonspecific findings of mediastenal widening, lymphadenopathy and tracheal narrowing.[9] Contrast enhanced CT is a useful modality for diagnosis and assessing the extent and severity of involvement. Mulitiplanner reformatted images help in assessing airway and vascular stenosis and in planning therapeutic intervention. Localized fibrotic areas with calcification is seen in majority of patients.[10] Some cases show diffuse soft tissue mass without calcification.[11] Post contrast study may show variable hetrogenous enhancement.[12] Biopsy is mandatory to exclude any malignant pathology.[13]

Superior vena cava syndrome is considered most common complication of granulomatous type. Resultant venous collaterals may appear as downhill varices around esophagus and stomach. Collateral flow through vein of Sapey may appear as pseudo lesion on CECT in segment 4 of liver.[14] Pulmonary artery encasement may lead to pulmonary oligemia and pulmonary infarction. Pulmonary vein narrowing may lead to pulmonary edema. Long standing pulmonary vessel obstruction may mimic chronic pulmonary thromboembolic disease and ultimately lead to pulmonary hypertension and corpulmonale.[15] Bronchial encasement can lead to collapse and pneumonia secondary to obstruction.[10] Other less common complications include chylothorax, constrictive pericarditis, coronary artery narrowing, narrowing of aorta and its branches, entrapment of recurrent laryngeal nerve, phrenic nerve, and esophageal narrowing.[16-20]

Fibrosing mediastenitis is a rare disease with very few cases reported in Asian countries. With no specific histological criteria available, diagnosis is usually based on exclusion.[21] Till date, only two large case series regarding fibrosing mediastenitis have been reported.[22-23] No local study regarding fibrosing mediastenitis are available. In this case series we highlight different imaging features of this rare disease on CT scan and its associated complications which can assist in better diagnosis.

Objective

The purpose of this study is to describe computed tomographic manifestations of fibrosing mediastenitis in eight pathological proven cases of fibrosing mediastenitis.

Methodology

Eight patients who were diagnosed with fibrosing mediastenitis in the study center from 2011 to 2018 were included in our study. Clinical record, radiological and biopsy findings were reviewed. Due to absence of universally accepted diagnostic criteria for fibrosing mediastenitis, diagnosis was made on the bases of radiological findings, exclusion of other diseases and repeated CT examinations. Clinical data regarding the presentation and suspected etiology were correlated with location of mediastinal disease, presence of calcification, contrast enhancement, effect on structures of mediastinum and additional associated pulmonary findings on computed tomography.

All eight patients underwent contrast enhanced CT chest in Khyber Teaching Hospital using Aestion VP single slice Toshiba CT machine. 5mm cuts were taken from root of neck to upper abdomen with images acquired 20seconds after intravenous injection of 150ml of contrast at rate of 5ml/sec. The

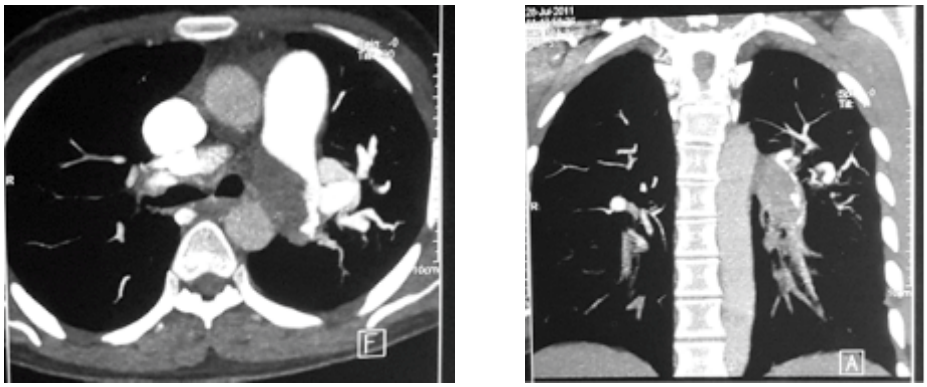

Figure 1: Focal non enhancing mediastenal fibrosis narrowing left pulmonary artery.

CT images were evaluated by experienced radiologists. Diagnosis of fibrosing mediastenitis was established when CT revealed infiltrative process in mediastinum with associated compressive effect on mediastenal structures. Diseases like active pulmonary tuberculosis, sarcoidosis and mediastenal malignancies were ruled out by sputum smear, bronchoscopy, repeated CT examination and biopsy. Patients with history of pulmonary or mediastenal malignancy and previous mediastenal radiotherapy were excluded from our study. Patients were followed by their clinical symptoms and repeated CT.

Results

Eight patients were included in the present study including two female and six men. The mean age of patients was 39 years. The common clinical symptoms in all patients were dull chest pain and shortness of breath. Three out of eight patients had a past history of tuberculosis. None of the patients had a history of malignant tumour. No underlying cause was identified in five cases and were labelled as idiopathic. In all eight patients BAL was negative for any infection. Serology for immune and connective tissue disorders were negative in all eight cases. Biopsy samples through mediastenoscopy from the mediastenal lesion revealed no malignant or granulomatous tissues, though it revealed fibrous tissues. On CT two patients had diffuse involvement of mediastinum and six patients presented with local mass. Calcification was present in five cases. There was no contrast enhancement in all eight cases. Six of eight cases revealed narrowing of mediastenal structures, with two cases showing pulmonary artery narrowing, three with superior vena cava obstruction, two with tracheal narrowing and one with pulmonary vein narrowing. Right ventricular dilatation was seen in two cases. Three out of eight cases had additional pulmonary findings. One patient had localized interstitial pulmonary edema due to pulmonary venous compression and additional findings of pulmonary arteriovenous malformation in left lung. Segmental atelectasis with pneumonia was present in three cases. All eight patients were followed by follow up CT which revealed stable findings with slight progression of findings due to slow disease process

Discussion

Fibrosing mediastenitis is a rare disease presenting with different pulmonary manifestations. Many case reports regarding fibrosing mediastenitis are found in geographical regions endemic for Histoplasmosis, specifically regions in Midwest like Ohio, Indiana etc.[24] The Mayo clinic electronic chart review found the average age of patients to be 42 years.[22] A case series of twenty Asian patients over a period of 10years in a single center study shows female predominance and tuberculosis as the major associated factor.[25] Three patients in our case series had previous history of tuberculosis.

Fibrosing mediastenitis usually presents on CT as soft tissue density infiltrative mass that cause obliteration of mediastenal fat planes.[26] Middle mediastinum is most frequently involved including paratracheal, subcarinal region and hila.[27] In a study by Sherrick et al focal pattern of mediastenal fibrosis was seen in 82% of cases which appear as soft tissue density mass. Calcification was present in 63% cases.[2] Diffuse pattern was observed in 18% cases. These findings are similar to our study in which two out eight patients had diffuse mediastenal involvement (25%) and 6 out of eight patients(75%) had local form of involvement. Calcification was present in 63% cases.

Jain et al in their study on fibrosing mediastenitis reported non enhancing soft tissue attenuation mass on contrast enhanced CT in superior mediastinum causing encasement of vascular structures. These included SVC, aortic arch and its branches. Mole et al in his study of 18 cases of fibrosing mediastenitis reported 33% patients had SVC obstruction.[28] Three out of eight patients in our study presented with SVC obstruction.

Extension of mediastenal fibrosis to encase bronchi may result in atelectasis (lobar or segmental) and pneumonia. Hevroni et al reported a case of recurrent pneumonia due to mediastenal fibrosis in which

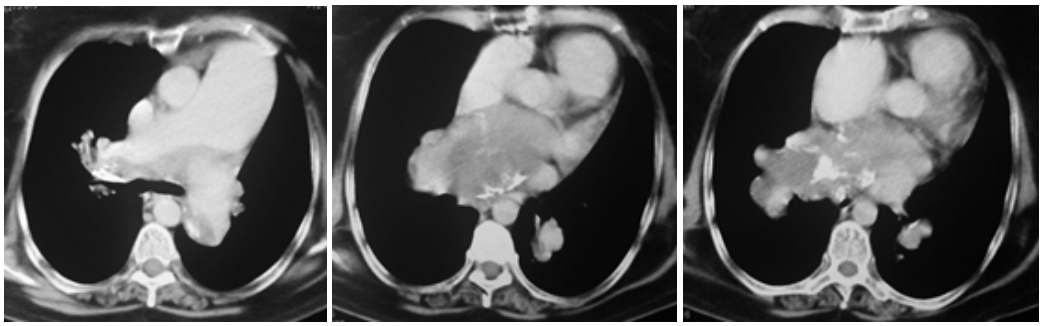

Figure 2: Diffuse fibrosing mediastenitis encasing middle mediastenal and right hilar structures.

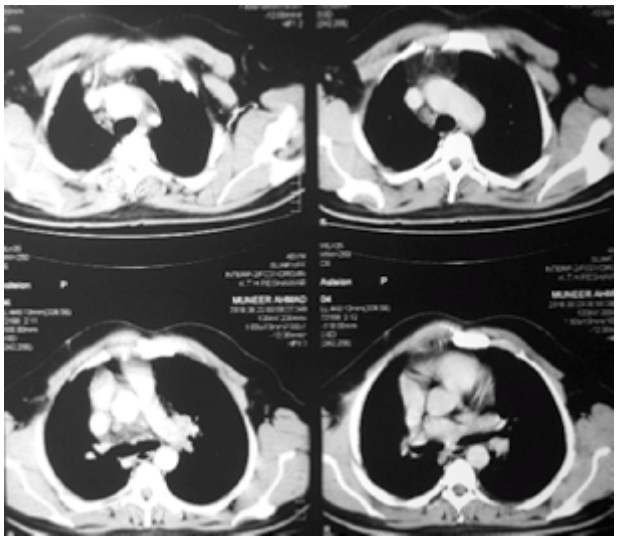

Figure 3: Focal form of fibrosing mediastenitis in right paratracheal and pre carinal region.

contrast enhanced CT chest reveal subcarinal and right hilar soft tissue masses compressing the right bronchial tree with right middle lobe atelectasis.[29] Sherrick et al reported 36% of his patients presented with airway narrowing on imaging studies with pulmonary parenchymal opacities seen in 33% cases. In our study 2 out eight cases showed tracheal narrowing on contrast enhanced CT chest.

Fibrosing mediastenitis leading to pulmonary arterial constriction can lead to pulmonary oligemia. Pulmonary venous narrowing leads to congestion and edema. Long standing pulmonary vascular obstruction leads to pulmonary hypertension and cor pulmonale. Wu et al reported a case of mediastenal fibrosis with pulmonary hypertension.[5] Contrast enhanced CT chest showed soft tissue mass in mediastinum and hilar region causing proximal pulmonary artery stenosis and pulmonary veins narrowing. Two cases in our study revealed pulmonary artery narrowing, one pulmonary vein narrowing and two cases revealed right ventricular dilatation due to pulmonary vascular encasement. One patient with pulmonary vein narrowing had additional findings of arteriovenous malformation in left lung. Two patients had changes of obstructive pneumonia in right lung.

In the presence of characteristic imaging features and appropriate clinical findings, diagnosis of fibrosing mediastenitis becomes certain. In endemic areas of histoplasmosis and tuberculosis patients presenting with calcified mediastinal mass with associated calcified granulomas in lung, spleen or liver suffice for diagnosis. Differential diagnosis for fibrosing mediastenitis include lung cancer, hilar or mediastinal metastasis and lymphoma. Therefore histopathology and culture is required for definite diagnosis.[10,28]

Conclusion

Fibrosing mediastenitis is an uncommon disease. It can be diagnosed by its characteristic manifestations on contrast enhanced CT scan. Due to its infiltrative pattern in mediastinum it can lead to complications ranging from benign course to narrowing of major airways, vessels and esophagus. For this reason it is crucial to be familiar with its different imaging features for accurate diagnosis as well as for planning noninvasive and surgical procedures. Computed tomography can play a vital role in its diagnosis, work up and follow up.

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