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Tuberculosis-associated Fibrosing Mediastinitis: Case Report and Literature Review

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ABSTRACT

Fibrosing mediastinitis is a rare condition defined by the presence of fibrotic mediastinal infiltrates that obliterate normal fat planes. It is a late complication of a previous granulomatous infection, such as histoplasmosis or tuberculosis (TB). Due to its rarity, fibrosing mediastinitis is often under-recognized, and the clinical presentation is variable and dependent on the extent of infiltration or encasement of structures within the mediastinum. We present a case of fibrosing mediastinitis in a man with a prior history of TB, who presented with progressive dyspnea and was found to have chronic mediastinal soft tissue opacities and pulmonary hypertension. His diagnosis was delayed due to the lack of recognition of this clinical/radiographic entity. Fibrosing mediastinitis is a rare entity usually caused by granulomatous disease. Most cases develop as a late complication of histoplasmosis or TB. The presence of calcified mediastinal soft tissue infiltrates on advanced chest imaging can be diagnostic of fibrosing mediastinitis in patients with a prior history of a granulomatous infection once active processes such as malignancy are excluded.

Key words: Fibrosing mediastinitis, histoplasmosis, mediastinal fibrosis, sclerosing mediastinitis, tuberculosis

INTRODUCTION

Fibrosing mediastinitis, also known as sclerosing mediastinitis or mediastinal fibrosis, is a rare late sequela of a thoracic inflammatory process. Most cases arise from prior histoplasmosis infection, and less commonly from tuberculosis (TB). Fibrosing mediastinitis is characterized by the development of dense mediastinal infiltrates that obliterate normal fat planes and is best viewed through advanced imaging modalities, such as computed tomography (CT) or magnetic resonance imaging (MRI).

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unrecognized and is often misdiagnosed. We present the
case of a patient with TB-associated fibrosing mediastinitis
whose diagnosis was delayed.

**CASE REPORT**

A 71-year-old man with a history of chronic obstructive
pulmonary disease (COPD), pulmonary hypertension,
and TB status posttreatment in 1985, presented for
evaluation of chronic, progressive dyspnea. The patient
had previously been seen at our institution for dyspnea and
had been diagnosed with and treated for numerous COPD
exacerbations. He had been treated with bronchodilators,
corticosteroids, and occasional antibiotics without
improvement. He had no history of tobacco use, significant
alcohol consumption, or any illicit drug use.

On examination, the patient was in no significant distress
and was able to speak in full sentences. His vital signs were
normal except for a respiratory rate of 20 breaths/min and
oxygen saturation of 90% on room air at rest. Cardiovascular
examination was notable for jugular venous distension
to the angle of the mandible. A parasternal heave, an
accentuated P2, and an S3 were appreciated. Lung
auscultation was notable for decreased breath sounds. The
remainder of the examination was unremarkable.

Transthoracic echocardiography revealed right ventricular
dilatation with flattening of the interventricular septum
and an estimated pulmonary artery systolic pressure of
85 mmHg. Pulmonary function testing (PFT) demonstrated
severe obstructive ventilatory defect with diffusion
impairment.

Chest radiograph demonstrated bilateral perihilar soft
tissue densities with right apical scarring, pleural thickening
and volume loss [Figure 1]. Contrast-enhanced chest CT
revealed ill-defined, infiltrative bilateral hilar soft tissue
densities with calcifications, enlargement of the main
and right pulmonary artery, and irregular narrowing of
the right mainstem and upper lobe bronchi [Figure 2].
Positron emission tomography (PET) showed mild
fluorodeoxyglucose uptake in the bilateral perihilar
regions, right more prominent than left. Of note, the
patient had undergone a non contrast-enhanced thoracic
CT three years prior, which documented relative stability all
the abnormal findings, including the bronchial narrowing
and pulmonary artery dilatation [Figure 3].

The patient had previously undergone a bronchoscopy
with bronchoalveolar lavage, brushings, and biopsies which
were negative for fungal organisms and acid-fast bacilli.

The patient’s known history of TB and findings of old
granulomatous disease on imaging along with negative bronchoscopy results established fibrosing
mediastinitis as the likely cause of his mediastinal infiltrates.
Although the PET scan showed mild uptake in the perihilar
regions consistent with an active inflammatory process,
the stability noted on imaging over a prolonged course
was reassuring that the mediastinal and hilar infiltrates
did not represent malignancy. The patient had compression
of his pulmonary artery due to fibrosing mediastinitis and
findings of pulmonary hypertension on the examination
and by echocardiography. He also had tracheobronchial
narrowing as a result of this entity as seen on chest
imaging which likely manifested as obstruction on
spirometry. The patient's respiratory symptoms stabilized
on supportive medications, and no further interventions
were required.

**DISCUSSION**

The pathophysiology of fibrosing mediastinitis is
postulated to stem from the chronic inflammation and
associated profound fibrotic changes of mediastinal
structures from the adjacent primary granulomatous
disease, either via direct infiltration from granuloma
rupture or indirectly via local inflammatory processes
within regional mediastinal lymph nodes. The
etiology of this inflammation includes granulomatous
diseases (such as histoplasmosis, TB, sarcoidosis, and
Behcet’s disease), malignancy (such as bronchogenic
cancer and lymphoma), trauma, and medication-induced
(methylsergide); however, a large number of cases remain
idiopathic. A subset of idiopathic fibrosing mediastinitis

![Figure 1](http://www.clinicalimagingscience.org)
now encompasses an emerging disease entity mediated by IgG-4, a presumed anti-inflammatory immunoglobulin whose exact role in inflammation modulation is yet to be elucidated but can manifest as retroperitoneal fibrosis with or without thyroiditis.

The development of fibrotic infiltrates/masses has the potential to encase and compromise mediastinal structures including the airway, esophagus, and/or major vessels. Airway involvement can vary widely ranging from hoarseness, if the laryngeal nerve is affected, to obstructive pneumonitis and/or atelectasis, and hemoptysis when erosion of the vascular bundle due to local inflammation into the pulmonary parenchyma occurs. Gastrointestinal involvement can include dysphagia and odynophagia through a direct mass effect. Vascular involvement can compromise any of the following: superior vena cava (SVC), aorta, pulmonary arteries, and veins through compression secondary to mass effect. Direct invasion can result in fistula formation. Clinically significant SVC syndrome and pulmonary hypertension can occur. Of note, the insidious onset of vascular compromise often allows for the development of significant collaterals and the potential to remain asymptomatic.

The diagnosis of fibrosing mediastinitis is established via clinical impression and imaging, and must exclude other active infiltrative processes involving the mediastinum, particularly bronchogenic carcinoma and lymphoproliferative disorders. Chest radiographs are usually nonspecific and often underestimate the extent of mediastinal disease. Therefore, advanced imaging modalities such as CT and MRI are preferred. Characteristic findings on CT or MRI include soft tissue obliteration of normal mediastinal fat planes with or without encasement and invasion of adjacent structures.

The management of fibrosing mediastinitis is largely directed toward symptom palliation, if present. There are currently, no medical therapies that have shown to prevent this sequela of granulomatous disease or alter it’s natural progression. Although controlled trials have not been performed, glucocorticoids do not appear to be beneficial. Corticosteroids, however, may be effective in IgG-4 related fibrosing mediastinitis. Treatment of any underlying infectious process is still warranted for patients’ well-being and public health purposes. Other management modalities can include percutaneous stenting of vascular structures such as the SVC to relieve obstruction, or placement of endoscopic stents in cases of airway obstruction and for the management of dysphagia. Surgery has a limited benefit and generally leads to poor outcomes given the extent of the fibrosis, calcifications, and the presence of collaterals.

Terminal outcomes with fibrosing mediastinitis include the development of cor pulmonale and respiratory compromise from recurrent infections, bronchial obstruction, or hemoptysis.

CONCLUSIONS

Although rare, fibrosing mediastinitis should be considered in the differential diagnosis of an ill-defined, calcified soft tissue mass in the mediastinum. The clinical presentation
and symptomatic involvement is directly proportional to the extent of infiltration of mediastinal structures. Fibrosing mediastinitis is a diagnosis of exclusion and is best established via advanced imaging modalities. It is most often a late sequela of granulomatous inflammation, however, active processes such as infection or malignancy must first be excluded. The management is primarily directed toward palliation of symptoms, if present, particularly as no medical therapy or surgical intervention has shown any benefit.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Rossi SE, McAdams HP, Rosado-de-Christenson ML, Franks TJ, Galvin JR. Fibrosing mediastinitis. Radiographics 2001;21:737-57.
2. Fijolek J, Wiatr E, Blasinska-Przerwa K, Roszkowski-Sliz K. Fibrosing mediastinitis as an atypical complication of tuberculosis. Case report. Pol Arch Med Wewn 2009;119:752-5.
3. Goodwin RA, Nickell JA, Des Prez RM. Mediastinal fibrosis complicating healed primary histoplasmosis and tuberculosis. Medicine (Baltimore) 1972;51:227-46.
4. Zider, A, Kamangar N. An 80-year-old female with progressive shortness of breath and a mediastinal mass. Chest 2016;150:19-22.
5. McNeeley MF, Chung JH, Bhalla S, Godwin JD. Imaging of granulomatous fibrosing mediastinitis. AJR Am J Roentgenol 2012;199:319-27.
6. Koksal D, Bayiz H, Mutluay N, Koyuncu A, Demirag F, Dagli G, et al. Fibrosing mediastinitis mimicking bronchogenic carcinoma. J Thorac Dis 2013;5:E5-7.
7. Peikert T, Colby TV, Midthun DE, Pairolero PC, Edell ES, Schroeder DR, et al. Fibrosing mediastinitis: Clinical presentation, therapeutic outcomes, and adaptive immune response. Medicine (Baltimore) 2011;90:412-23.
8. Kant S, Walsh GL. Fibrosingmediastinitis and consequent superior vena cava syndrome – A case report. J Thorac Dis 2012;4:428-30.
9. Mathisen DJ, Grillo HC. Clinical manifestation of mediastinal fibrosis and histoplasmosis. Ann Thorac Surg 1992;54:1053-7.
10. Sherrick AD, Brown LR, Harms GE, Myers JL. The radiographic findings of fibrosing mediastinitis. Chest 1994;106:484-9.