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Case Report

Aggressive and progressive fibrosing mediastinitis involving the thoracic spine mimicking malignancy: A case report

Hee Kang, MD, PhD[a,*], *Min Jung Jung, MD, PhD*[b]

[a] *Department of Radiology, Kosin University Gospel Hospital, Kosin University College of Medicine, Busan, South Korea*

[b] *Department of Pathology, Kosin University Gospel Hospital, Kosin University College of Medicine, Busan, South Korea*

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ABSTRACT

Fibrosing mediastinitis is an uncommon benign disorder in which a chronic inflammatory reaction results in diffuse fibrosis of the mediastinum, potentially compromising the airways, great vessels and other mediastinal structures. Herein we describe a progressive course of fibrosing mediastinitis in a 72-year-old man. Computed tomography images depicted a diffuse, infiltrative, soft tissue mass involving the esophagus and superior vena cava in the mediastinum. Magnetic resonance imaging revealed destruction of the adjacent thoracic spine. Positron emission tomography-computed tomography also revealed increased metabolism in the periphery of the mass.

Introduction

Fibrosing mediastinitis (FM), also known as sclerosing mediastinitis, is a rare benign disease characterized by proliferation of dense fibrous tissue within the mediastinum. It may lead to life-threatening hemodynamic compromise caused by progressive fibrosis encasing the airway, great vessels and other mediastinal structures. Although FM is often preceded by granulomatous diseases such as histoplasmosis or tuberculosis, the precise cause and pathogenesis in most cases is unknown, with substantial variability in histopathologic appearance. We herein report the multi-imaging findings of FM in a 72-year-old Asian man that exhibited an aggressive and progressive course mimicking malignancy.

Case report

A 72-year-old man presented with a 3-month history of right posterior chest pain. His electrocardiogram was normal. The initial chest radiography showed slight widening of the right paratracheal stripe (Fig. 1). Postcontrast computed tomography (CT) revealed an ill-defined infiltrative mass with subtle peripheral enhancement, which showed loss of fat planes

* Corresponding author.
E-mail addresses: kanghi81@gmail.com, soinvain@naver.com (H. Kang).
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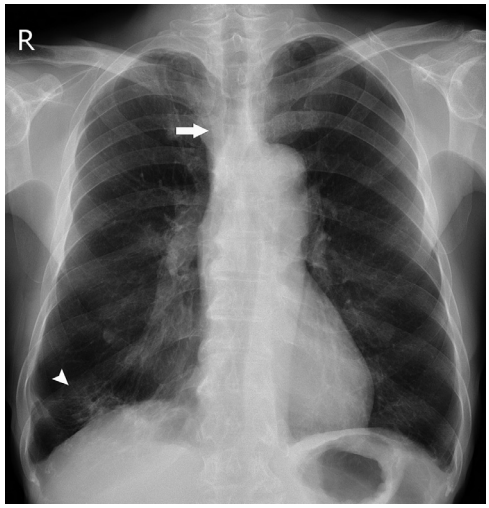

Fig. 1 – Initial posteroanterior chest radiograph showed slight widening of the right paratracheal stripe (arrow). Focal ill-defined patchy consolidation and linear opacities are noted in right lower lung zone (arrowhead), suggestive of pneumonia.

between the lesion and adjacent trachea, esophagus, and vertebral body (Fig. 2A). Endoscopy of the esophagus and bronchoscopy revealed no abnormal findings. The patient underwent endobronchial ultrasound biopsy, but there was no evidence of malignancy. Laboratory tests including antibodies for autoimmune disease, immunohistochemistry analysis for lymphoma, fungal serology, sputum smears, and cultures for acid-fast bacilli were all negative. Serum Immunoglobulin G4 (IgG4) level was also not elevated.

After 4 months, follow-up CT revealed an increased extent of the mass along the pleura and mediastinum (Fig. 2B) and also showed circumferential enhanced wall thickening and luminal narrowing of the right main bronchus (not shown here). ^{18}F-fluorodeoxyglucose positron emission tomography-CT (^{18}F-FDG PET-CT) images showed avid uptake (maxSUV: 4.8) in the periphery of the mass invading the adjacent thoracic spine (Fig. 3). Postcontrast axial T1-weighted magnetic resonance image (MRI) revealed an ill-defined low-signal intensity mass with peripheral rim enhancement (Fig. 4A). The sagittal image showed increased signal intensity of the adjacent thoracic vertebral bodies which is suggestive for involvement of bone marrow (Fig. 4B). The mass had diffuse low-signal intensity in T1-weighted MRI (Fig. 4C) and mixed high- and low-signal intensity in T2-weighted MRI (Fig. 4D) suspected of FM. The patient underwent video-assisted thoracoscopic surgery for biopsy of the mediastinal mass. The specimens obtained from this hard and adhesive mass in the mediastinum were dense and fibrotic lesion with variable cellularity, including inflammatory cells such as lymphocytes and plasma cells (Fig. 5). There was no evidence of malignancy, other neurogenic tumor or granulomatous lesion.

Three months later, the patient visited the hospital again complaining of worsened pain, difficulty swallowing, and swelling of the face. The follow-up CT showed an increased extent of tumor infiltration, with invasion into adjacent

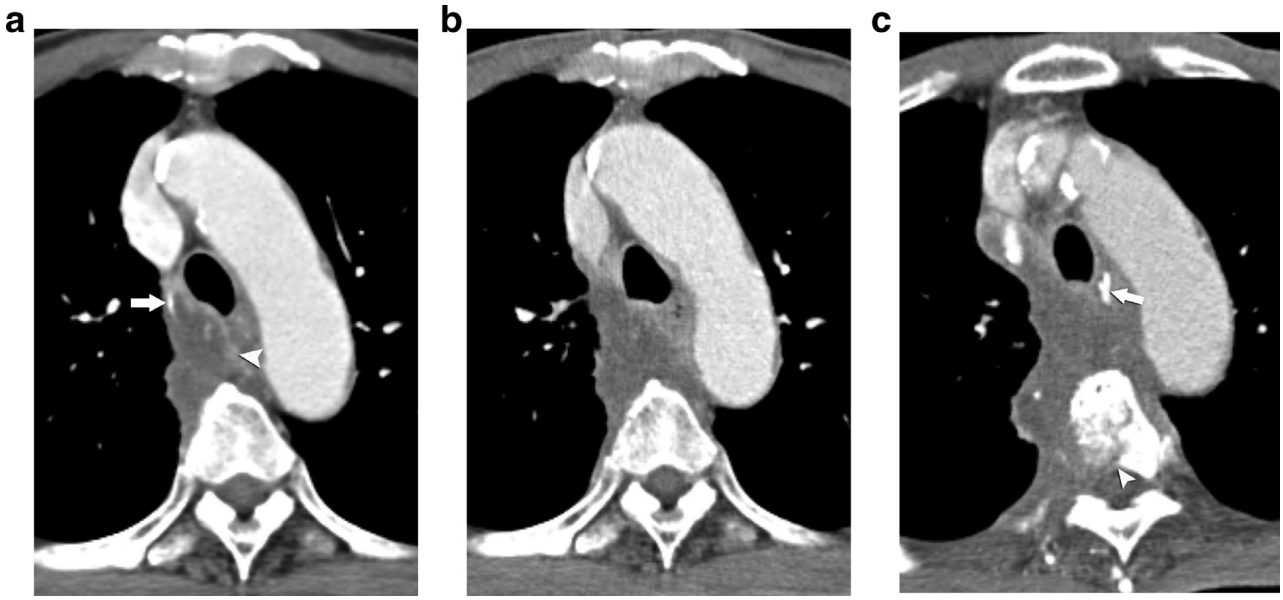

Fig. 2 – (A) Postcontrast CT scan at the level of the aortic arch shows about 38 x 25 x 57 mm sized ill-defined, infiltrative, low-attenuating mass engulfing the azygos vein (arrow) with subtle peripheral enhancement (arrowhead) in the right paratracheal area. There is loss of fat plane between the lesion and the adjacent trachea, esophagus, and vertebral body. (B) After 4 months, CT scan reveals increased extent of the mass along the pleura and mediastinum. (C) Follow-up CT scan in 3 months shows increased infiltration of the tumor with invasion into adjacent structures including the esophagus and vertebral body. The infiltration of the esophageal wall by the tumor is seen. There is subtle residual oral contrast material in the esophageal lumen (arrow). The bony destruction of the fourth thoracic vertebral body (arrowhead) is also noted.

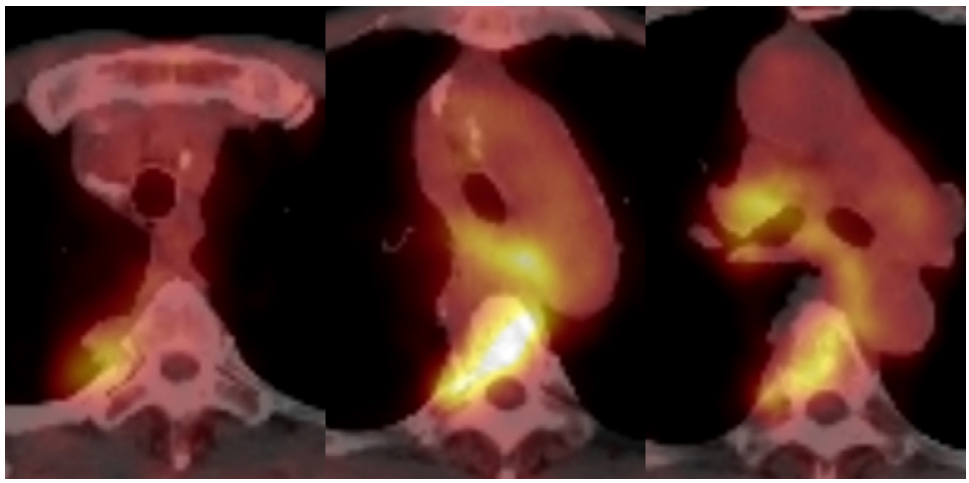

Fig. 3 – ^{18}F-FDG PET-CT shows avid uptake in the peripheral rim of the mass. Hypermetabolism of the adjacent thoracic vertebral body was also seen (maxSUV: 4.8).

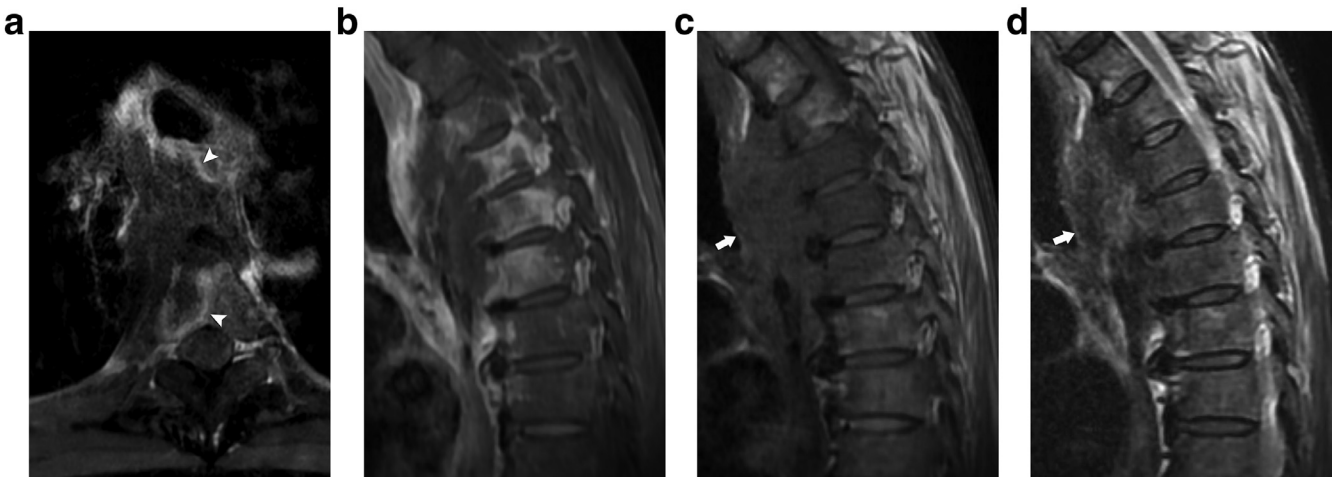

Fig. 4 – (A) Postcontrast axial T1-weighted MRI with fat suppression reveals an ill-defined low-signal intensity mass with peripheral rim enhancement (arrowheads) including the thoracic spine. (B) Postcontrast sagittal T1-weighted MRI with fat suppression shows increased signal intensity of the adjacent third to sixth thoracic vertebral bodies suggestive for bone marrow involvement. The mass (arrow) has diffuse low-signal intensity in T1-weighted MRI (C) and mixed high- and low-signal intensity in T2-weighted MRI (D).

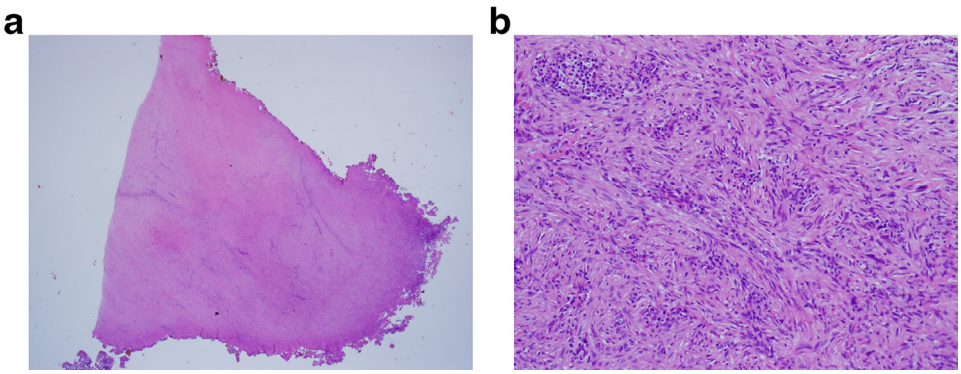

Fig. 5 – (A) Low magnification photography of the biopsy specimen with hematoxylin-eosin (H-E) staining reveals a dense fibrotic lesion with variable hypo- to hypercellularity. (B) High-magnification photography (H-E staining, ×200) shows some inflammatory cells including lymphocytes and plasma cells in the hypercellular portion.

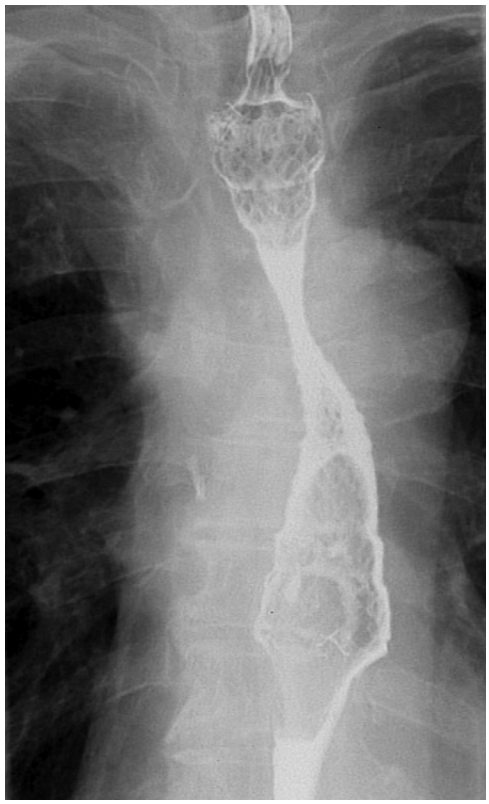

Fig. 6 – Esophagography after a stent placement shows an incomplete expansion of the stent graft but the passage of the contrast material is observed.

mediastinal structures including the superior vena cava (SVC), esophagus and vertebral bodies (Fig. 2C). An esophageal stent graft was placed to facilitate swallowing. On esophagography after stent placement, the stent was not fully expanded, but the passage of contrast material was observed (Fig. 6). A corticosteroid and itraconazole were administered, but his symptoms were refractory to the medication, and he was transferred to a long-term care facility.

Discussion

FM is a rare benign disease also known as sclerosing mediastinitis or mediastinal fibrosis. It was first reported by Oulmont in 1855, who classified it as idiopathic fibrous mediastinitis; it was thought to be secondary to tuberculosis or syphilis until 1925 when Knox reported an association with fungal infections [1]. Histopathologically, it is characterized by abundant, paucicellular fibrous tissue infiltrating adipose tissue in mediastinum. This fibrous tissue can contain some mononuclear cells [2]. A definite cause of FM has not been established. Many if not most cases in the United States have been linked to *Histoplasma capsulatum* infection. However, a definite organism of infection could not be identified in many cases. This disease is thought to result not from direct infection, but rather from an abnormal immunologic reaction to *H. capsulatum* antigens [3]. Other infections such as tuberculosis, aspergillosis, blastomycosis, and cryptococcosis can also cause granulomatous fibrous mediastinitis [4]. The focal granulomatous type is well encapsulated and does not affect adjacent mediastinal fat or organs. However, it can enlarge and rupture, causing diffuse infiltrative FM in a minority of patients [5]. The diffuse nongranulomatous type is considered an idiopathic disease related to autoimmune syndromes, radiation therapy, or drug reaction [4]. Recently, immunological changes consistent with IgG4-related disease (IgG4-RD) were demonstrated in some cases of idiopathic FM. IgG4-RD is a multiorgan immune-mediated fibroinflammatory disease similar to retroperitoneal fibrosis, sclerosing cholangitis or Riedel thyroiditis [6]. In this patient, serum IgG4 was not elevated and there was no other evidence of autoimmune disease or infection.

Diffuse infiltrative FM can cause obstruction or compression of mediastinal structures including airways, great vessels, and the esophagus. SVC syndrome and obstruction of airways or the esophagus are rare complications of FM [7–10]. FM involving the thoracic spine has not been reported previously, to the best of our knowledge. Cortical destruction and bone marrow invasion appeared to be caused by locally invasive proliferation of active inflammatory and fibrous tissue.

CT depicts the extent and severity of visceral encroachment, as indicated by the obliteration of fat planes of the mediastinum and the presence of a soft tissue mass with circumferential encasement of the mediastinal structures. It depicts foci of calcification better than radiography or MRI. On contrast-enhanced imaging, fibrotic tissue shows variable enhancement. Differential diagnosis of this disease upon CT includes other infiltrative lesions of the mediastinum, such as lung cancer, metastatic carcinoma, lymphoma, and mediastinal desmoid tumors [8,11]. Fibrotic tissue has intermediate signal intensity on T1-weighted MRI and variable intensity on T2-weighted MRI [12]. ^{18}F-FDG PET-CT is seldom used for the evaluation of diffuse infiltrative-type FM. A few case reports have shown variable FDG avidity [13–16]. In this case, the uptake of the lesion was thought to be correlated with the aggressiveness of the disease, similar to a previous case [13].

Extensive surgical biopsy sampling of the mediastinum is necessary to confirm the diagnosis and exclude malignancy. Biopsy samples obtained via the percutaneous needle technique may be insufficient to rule out malignancy [1,17].

The prognosis of idiopathic FM is uncertain, with both spontaneous remission and exacerbation of symptoms being reported. Patients with extensive bilateral or subcarinal disease usually have worse outcomes than those with more localized disease. There is no proven effective medical treatment for idiopathic FM. Corticosteroid and rituximab or tamoxifen have been shown to be effective in selected cases [4,18,19]. However, no prospective randomized controlled trials have been conducted so far. Symptomatic airway constriction and SVC syndrome can be treated with balloon dilatation and/or stent placement, although restenosis of the stent is frequent and retreatment is often needed; some patients may still require surgical repair [20,21]. In this case, we administered a

corticosteroid and antifungal agent, but the symptoms were refractory. To address dysphagia, a metallic stent was placed in the obstructed portion of the esophagus. Difficulty swallowing was somewhat improved, although the stent was not fully expanded.

In conclusion, idiopathic FM can be aggressive and progressive despite being benign. Postcontrast CT and MRI effectively demonstrate the extent of fibrosis and complications such as SVC syndrome and airway or esophageal constriction. It would be accompanied by bone involvement causing cortical and medullar destruction like this extremely rare case. The MRI and ^{18}F-FDG PET-CT are more sensitive for detection of the early inflammatory proliferation of FM.

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