Idiopathic Fibrosing Mediastinitis with Unusual Initial Presentation of Myelopathy

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Introduction

- Fibrosing mediastinitis is a very rare disorder characterized by proliferation of fibrous tissue within the mediastinum
- Affected patients are typically young
- The precise pathogenesis of fibrosing mediastinitis remains unknown
- There is no curative treatment, and management is targeted at symptom alleviation

Case Report

- 44-year-old Hispanic female presented with acute bilateral lower extremity weakness and numbness following a year of progressively worsening thoracic back pain and exertional dyspnea
- Physical examination showed a sensory deficit to light touch from T7 down bilaterally, with motor strength of 2/5 hip flexion, knee flexion and extension bilaterally, 2/5 dorsiflexion bilaterally, absent plantar flexion, and decreased rectal tone
- Hemoglobin was 9.3 gm/dL, MCV 60.6 fl, ESR 123 mm/hr, CRP 91 mg/L, alkaline phosphatase 189 U/L, with rest of chemistries unremarkable
- Chest X-ray was unremarkable
- MRI of the thoracic and lumbar spin with and without contrast was obtained (Fig 1)

Imaging

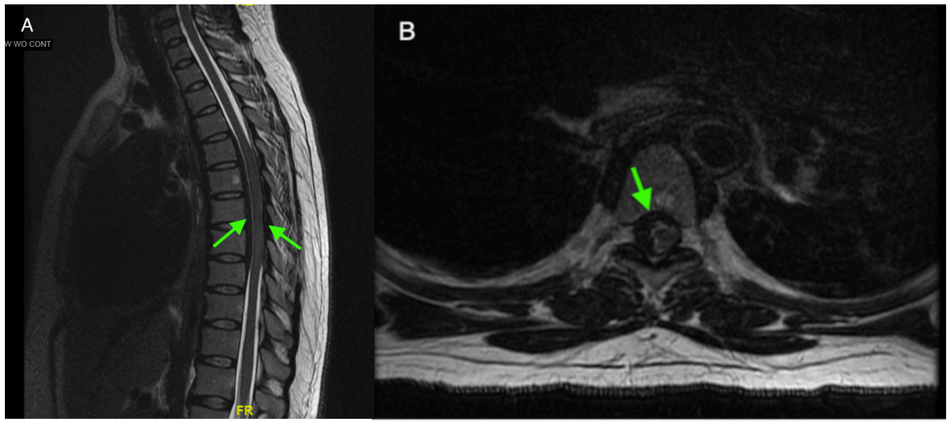

Figure 1, panels A-B. Thoracic spine MRI showing T2 hypointense enhancing soft tissue within the ventral, right lateral, and dorsal epidural space with severe compression and displacement of the thoracic spinal cord to the left with associated mild spinal cord edema (green arrows).

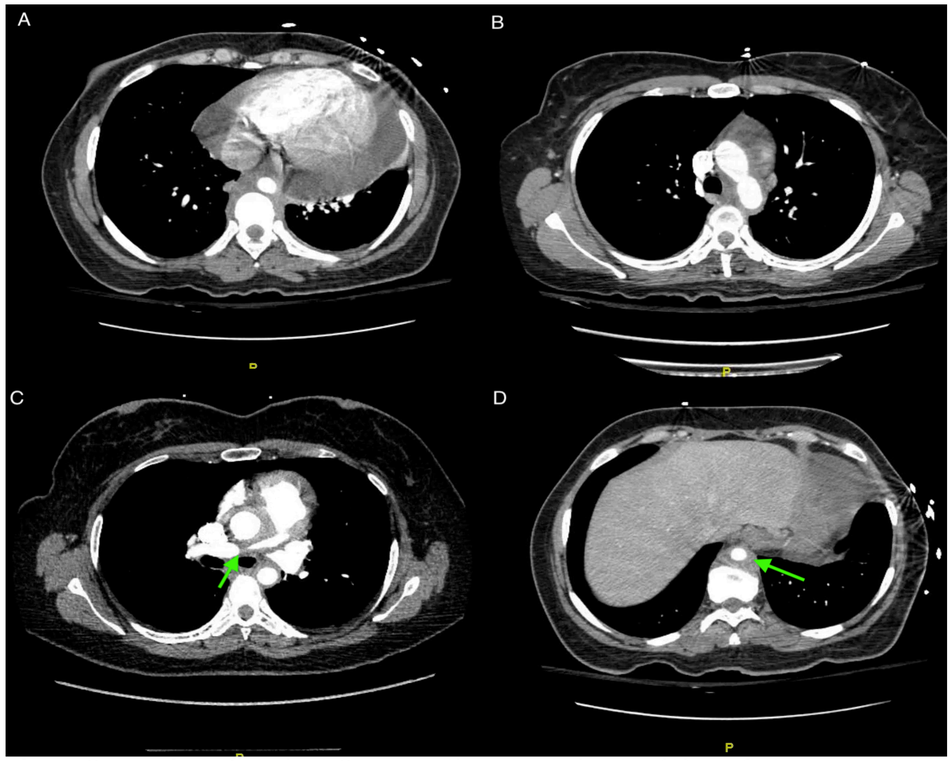

Figure 2, panels A-D. A chest CT scan with contrast showing (A) a soft tissue mediastinal density, (B) also surrounding the aortic arch and descending thoracic aorta extending to the prevertebral and paravertebral regions of the lower thoracic spine, with (C) mild narrowing of both pulmonary arteries within the mediastinum (green arrow on the right pulmonary artery), as well as of the (D) abdominal aorta (green arrow).

Hospital Course

- Echocardiography revealed pulmonary arterial systolic pressure of 155 mmHg with severely dilated right atrium and right ventricle
- Biopsy through partial excision of mediastinal mass via video assisted thoracoscopic surgery showed fibroadipose tissue without evidence of malignancy
- Bacterial and fungal cultures, histoplasma antigen, quantiferon TB gold, ACE, ANA, ANCA, PR3, and MPO were negative with normal serum IgG and IgG4
- Her weakness and numbness improved with dexamethasone. Rituximab was planned but she was lost to follow up

Discussion

- Fibrosing mediastinitis is typically associated with infections, malignancies, and inflammatory disorders, and it is termed idiopathic in the absence of an identifiable cause
- Symptoms arise when fibrosis causes compression of airways, vascular structures, or esophagus
- Manifestations include cough, dyspnea, wheezing, chest pain, dysphagia, and hemoptysis
- Compression of great vessels results in pulmonary vessel obstruction, pulmonary hypertension, or SVC syndrome
- Myelopathy from spinal compression has never been reported as a presentation of idiopathic fibrosing mediastinitis and should be recognized as a complication of this exceedingly rare disorder

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