Idiopathic fibrosing mediastinitis with unusual initial presentation of myelopathy

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Case Description: A 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. 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 T-spine showed a T2 hypointense enhancing soft tissue within epidural space from T3-T10 with severe compression of the thoracic spinal cord and mild spinal cord edema (Figure1). Contrast CT of the chest/abdomen revealed calcified mediastinal lymph nodes; soft tissue mediastinal density surrounding the aortic arch and descending aorta extending to paravertebral regions of the lower T-spine; mild narrowing of mediastinal pulmonary arteries and of the abdominal aorta (Figure2, A-D), compatible with fibrosing mediastinitis. 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.

Fibrosing mediastinitis is a rare disorder of fibrous tissue proliferation within the mediastinum with varying epidemiology[1]. Typically associated with infections, malignancies, and inflammatory disorders[2], it is termed idiopathic in the absence of an identifiable cause[1]. Symptoms arise when fibrosis causes compression of airways, vascular structures, or esophagus. Manifestations include cough, dyspnea, wheezing, chest pain, dysphagia, and hemoptysis[3]. Compression of great vessels results in pulmonary vessel obstruction, pulmonary hypertension, or SVC syndrome[4]. 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.

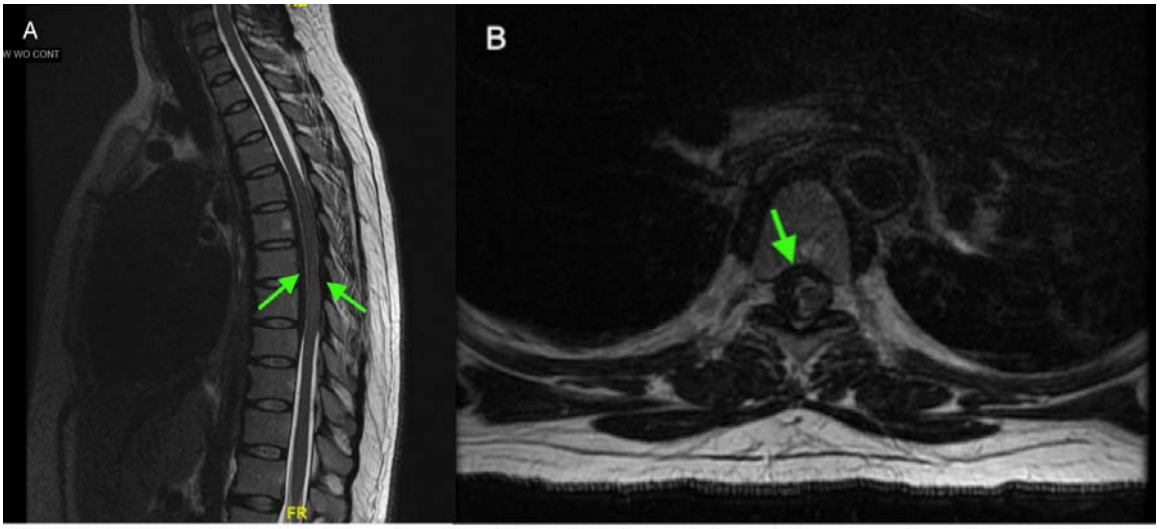

Figure 1, panels A-B. Thoracic spine MRI showing T2 hypointense enhancing soft tissue within the ventral, right lateral, and dorsal epidural space with severe compression and displacement of the thoracic spinal cord to the left with associated mild spinal cord edema (green arrows).

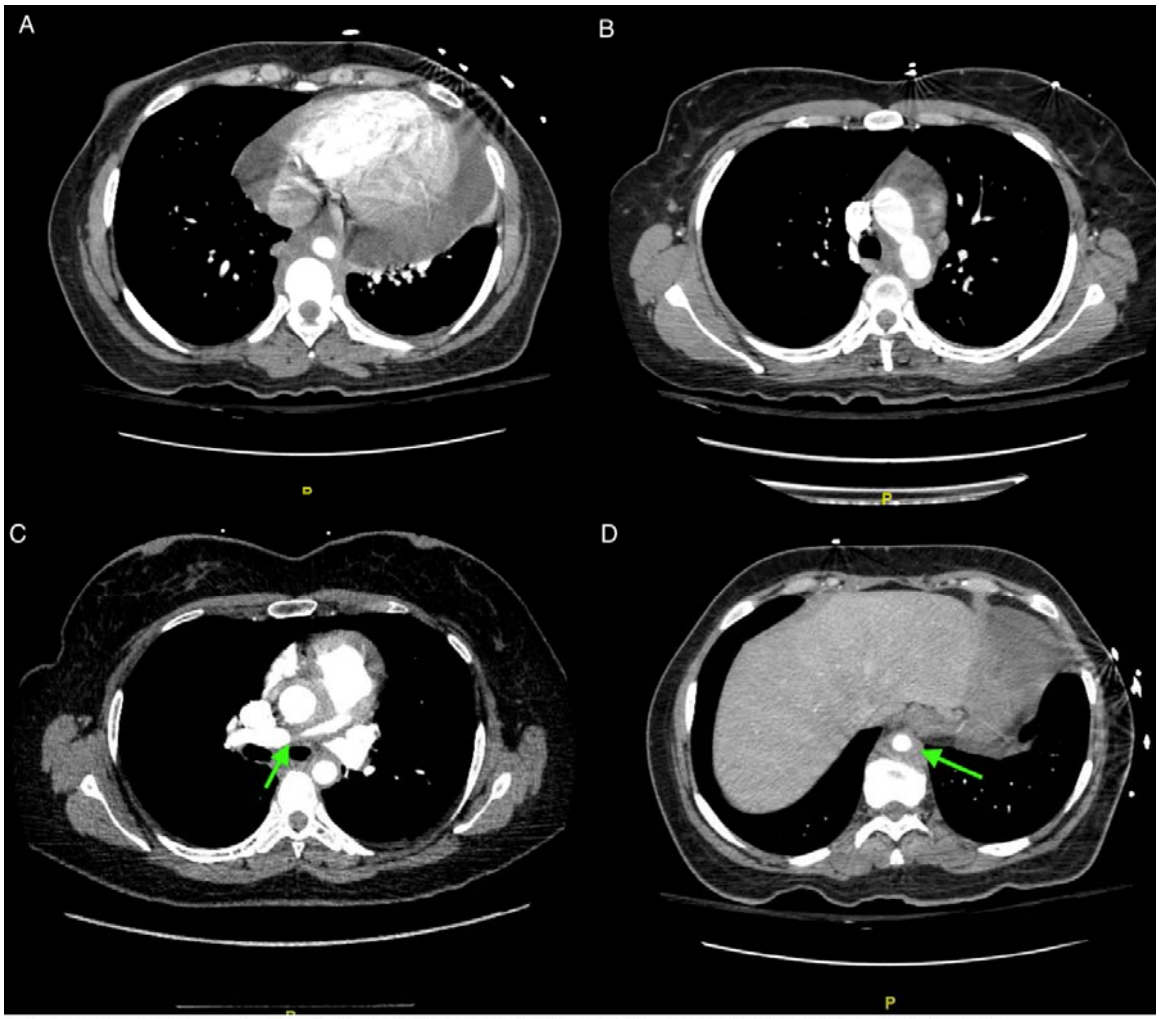

Figure 2, panels A-D. A chest CT scan with contrast showing (A) a soft tissue mediastinal density, (B) also surrounding the aortic arch and descending thoracic aorta extending to the prevertebral and paravertebral regions of the lower thoracic spine, with (C) mild narrowing of both pulmonary arteries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

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[1] Peikert, T., Colby, T. V., Midthun, D. E., Pairolero, P. C., Edell, E. S., Schroeder, D. R., & Specks, U. (2011). Fibrosing mediastinitis: Clinical presentation, therapeutic outcomes, and adaptive immune response. *Medicine*, *90*(6), 412–423. https://doi.org/10.1097/MD.0b013e318237c8e6

[2] Jain, D., Fishman, E. K., Argani, P., Shah, A. S., & Halushka, M. K. (2011). Unexpected sclerosing mediastinitis involving the ascending aorta in the setting of a multifocal fibrosclerotic disorder. *Pathology - Research and Practice*, *207*(1), 60–62. https://doi.org/10.1016/j.prp.2010.05.010

[3] Athanassiadi, K. A. (2009). Infections of the Mediastinum. *Thoracic Surgery Clinics*, *19*(1), 37–45. https://doi.org/10.1016/j.thorsurg.2008.09.012

[4] Lin, J., & Jimenez, C. A. (2022). Acute mediastinitis, mediastinal granuloma, and chronic fibrosing mediastinitis: A review. *Seminars in Diagnostic Pathology*, *39*(2), 113–119. https://doi.org/10.1053/j.semdp.2021.06.008