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A Roller Coaster Ride of Idiopathic Fibrosing Mediastinitis

Qian Zhang
Abington Jefferson Health

Khine Shan
University of Maryland Medical Center

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SESSION TITLE: Medical Student/Resident Disorders of the Mediastinum Posters

SESSION TYPE: Med Student/Res Case Rep Postr

PRESENTED ON: October 18-21, 2020

INTRODUCTION: Fibrosing mediastinitis (FM) is a rare benign pulmonary condition characterized by excessive fibrosis in the mediastinum due to an abnormal immune response to infections or inflammatory conditions [1, 2].

CASE PRESENTATION: A 39-year-old man presented to the clinic with a few weeks of left-arm edema with negative ultrasound findings of deep venous thrombosis but found to have axillary lymphadenopathies which warranted further investigation. A chest CT scan revealed a 2.5x2.4 cm round soft tissue mass in the left upper mediastinum that occluded the left brachiocephalic and subclavian vein. Thoracoscopic excisional biopsy of the mass showed hypocellular fibrosis, collagen bands with intervening mesenchymal cells without the presence of Reed-Sternberg cells, granulomas, or morphology suggestive of malignancy. The diagnosis of FM was confirmed by three different Pathologists. Serum antinuclear antibody, histoplasmosis titer, Quantiferon gold tuberculosis assay, and angiotensin-converting enzyme levels were unremarkable. He denied living in or had a travel history to endemic fungal or mycobacterial disease regions. Interval CT scans throughout the next three years showed a stable lung mass without progression. He remained to be clinically stable until he developed right-sided hearing loss due to a transverse sinus thrombosis seen on the cerebral angiogram. A repeat chest CT discovered a new 1.9x1.5 cm spiculated mass in the left upper lung lobe with small satellite nodules adjacent to the previous FM mass. An extensive hypercoagulable workup was unremarkable. The new mass was deemed to be not amenable to navigational bronchoscopy or interventional radiology guided biopsy. He was discharged with anticoagulation and will have repeated imaging studies in the outpatient setting as there were low clinical indications of an urgent biopsy.

DISCUSSION: It is extremely rare to have FM with a negative histoplasmosis test result in addition to an extensive unremarkable workup for other potentially related pathologies. Furthermore, there was a low clinical suspicion of a new onset of malignancy in this low-risk patient despite that this new thrombosis could be a prothrombotic effect of malignancy from the new mass or simply just an incidental finding that led to the discovery of the new mass. A few case reports detailed a new lymphoma diagnosed in the setting of an original biopsy-proven FM [3]. However, the new mass could also just be a progression of the original FM given the proximity in location.

CONCLUSIONS: Patients with FM are often at a crossroads when there are new suspicious incidental findings found on repeated imaging studies. Close monitoring with interval imaging studies is important and a repeat of invasive biopsy may be required to rule out a possible new malignancy.

Reference #1: Rossi SE, McAdams HP, Rosado-de-Christenson ML, et al. Radiographics. 2001;21(3):737-57. PMID: 11353121 https://doi.org/10.1148/radiographics.21.3.g01ma17737.

Reference #2: Harman M, Sayarlioglu M, Arslan H, et al. Fibrosing Mediastinitis and Thrombosis of Superior Vena Cava Associated With Behçet's Disease. Eur J Radiol 2003; 48(2):209-12. PMID: 14680916 https://doi.org/10.1016/S0720-048X(0300006-8).

Reference #3: Kakuta N, Sumitani M, Sugitani A, et al. Mediastinal Peripheral T-cell Lymphoma Diagnosed by Repeated Biopsies After an Initial Diagnosis of Fibrosing Mediastinitis. Respirol Case Rep 2017;5(6):e00272. PMID: 28932400 https://doi.org/10.1002/rcr2.272