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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Fibrosing Mediastinitis Causing High Degree AV Block in a Young Female Patient

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ABSTRACT

A 30-year-old woman presented with presyncopal episodes and was found to have high degree atrioventricular block. Computed tomography imaging demonstrated pericardial thickening extending from the main pulmonary artery to the aortic cusps. Here we present a rare case of fibrosing mediastinitis causing high-degree atrioventricular block. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;7:101717) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENT ILLNESS

A 30-year-old woman presented for multiple episodes of presyncope progressing to syncope. Her symptoms occurred intermittently over the 5 days preceding presentation and were not associated with any prodrome, chest pain, or palpitations. She also noted generalized fatigue and dyspnea on exertion over the previous 3 months. On the day of presentation, she syncopized after placing her child in its crib, prompting her to present to the emergency department.

On presentation, her vital signs were remarkable for bradycardia and examination for an early systolic murmur auscultated at the right upper sternal border. She was afebrile, with normal breath sounds, and no notable lymphadenopathy. Electrocardiogram was significant for a new 2:1 atrioventricular (AV) block with left bundle branch block (**Figure 1A**). She continued to experience episodes of presyncope that were correlated with periods of high-grade AV block on telemetry (**Figure 1B**). A temporary pacemaker was placed, which was later exchanged for a tempo-permanent pacing system.

LEARNING OBJECTIVES

- To understand the pathophysiology of FM and its place in the differential for a pericardial mass.
- To recognize AV block as a possible manifestation of FM.

PAST MEDICAL HISTORY

Medical history was significant for a systolic murmur noted 1 year before presentation during pregnancy. Echocardiography was unable to differentiate whether the murmur was secondary to a bicuspid aortic valve or aortic outflow tract compression. Cardiac magnetic resonance was performed 6 months before presentation, which uncovered a pericardial thickening extending from the main pulmonary artery to the level of the aortic cusps and mitral annulus. She otherwise had no previous medical

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ABBREVIATIONS AND ACRONYMS

AV = atrioventricular

FM = fibrosing mediastinitis

history or family history of congenital cardiac disease or malignancy. Additional workup was pending at the time of presentation.

DIFFERENTIAL DIAGNOSIS

Given her past medical history and new electrophysiologic disease at multiple levels of the conduction system, clinical concern was for possible infiltration versus mass effect from the extracardiac mass. Differential diagnosis included lymphoma, primary lung cancer, sarcoma, sarcoidosis, tuberculosis, and fibrosing mediastinitis (FM).

INVESTIGATIONS

Computed Tomography angiography of the thorax redemonstrated soft tissue thickening extending from the main pulmonary artery to the aortic cusps and possibly involving the pericardial recesses with extrinsic effect on the main pulmonary artery, anterior aspect of the left atrium, aortic root, and ascending aorta (Figures 2A to 2C). No lymphadenopathy was noted on computed tomography imaging of the thorax, abdomen, or pelvis.

Obtaining a tissue diagnosis was felt to be critical to formulate management decisions. The patient was transferred to a cardiac surgery-equipped facility and underwent successful aortic biopsy via median sternotomy with placement of epicardial pacing wires. Biopsy showed collagenous fibrosis with chronic inflammatory cell infiltrate extending into the adventitia and outer aortic media and scattered lymphoid follicles, consistent with idiopathic FM (Figures 3A to 3B). No granulomas were present. The performed immunostaining showed no evidence of lymphoma.

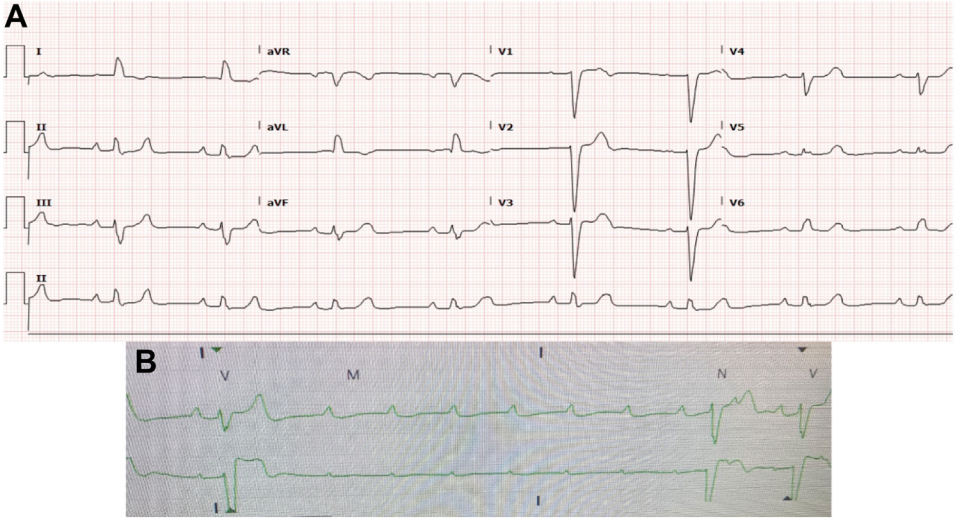
MANAGEMENT

Before the results of the biopsy, she was started on intravenous glucocorticoids under rheumatology guidance owing to suspicion for an inflammatory lesion. Given the lack of known efficacy in this disease process, she was tapered off steroids. Her hospital course was complicated by pericardial effusion causing hemodynamic compromise requiring emergent pericardiocentesis. An analysis of the pericardial fluid showed numerous neutrophils on the background of blood. Throughout the rest of her hospital stay, telemetry demonstrated improvement in conduction, and the patient was placed on a backup pacing rate of 30, which she tolerated well. Both AV block and left bundle branch block resolved before discharge, and a tempo-permanent pacer was removed.

DISCUSSION

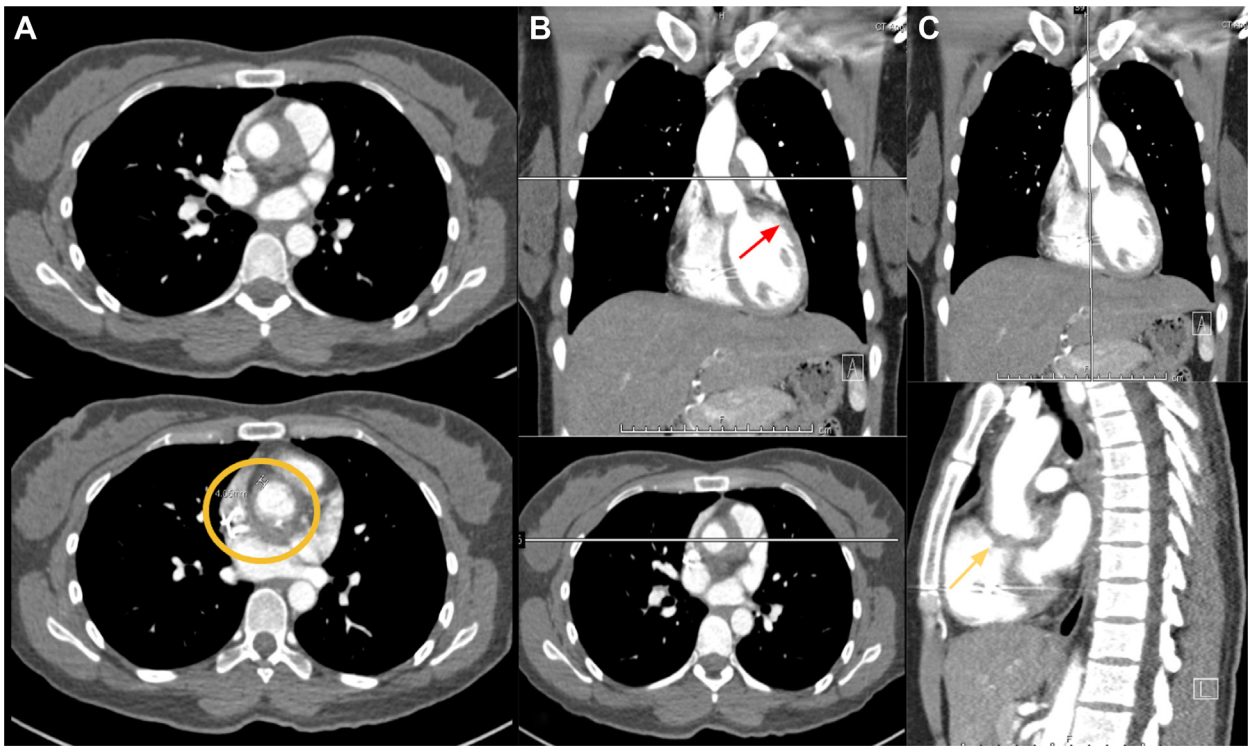
High-degree AV block is uncommon in this patient's age group and should prompt consideration for reversible causes including infections, metabolic derangements, drug effects, excessive vagal tone, and infiltrative disease.[1] This is crucial to determining if

FIGURE 1 Electrocardiogram and Telemetry Data on Initial Presentation



(A) Electrocardiogram demonstrating 2:1 AV block and LBBB. **(B)** Telemetry showing high degree AV block correlating with patient's symptoms of presyncope. LBBB = left bundle branch block.

FIGURE 2 **Axial, Coronal, and Sagittal CT Imaging of Periaortic Thickening**

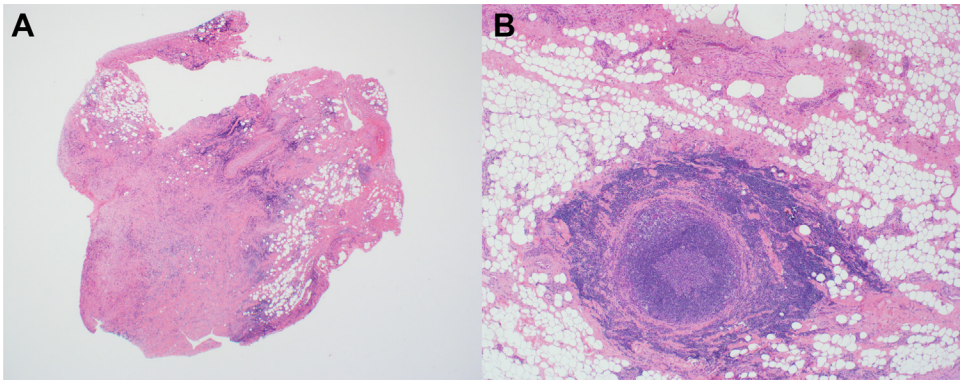


(A) Axial CT images demonstrating periaortic thickening **(yellow circle)**. **(B)** Coronal **(top)** and corresponding axial CT images **(bottom)** depicting extension of periaortic thickening to aortic cusps **(red arrow)**. **(C)** Coronal **(top)** and corresponding sagittal CT images **(bottom)** periaortic thickening extending to aortic cusps and infiltrating or compressing AV node and left bundle branch **(yellow arrow)**. CT = computed tomography.

the patient will require permanent pacemaker placement. FM is not commonly considered among reversible causes, but this case highlights the need to consider FM in the right clinical context. FM is believed to be the result of an adverse host response in the mediastinal lymph nodes leading to excessive fibrotic proliferation that may invade into adjacent structures.[2] Common provoking insults vary regionally, oftentimes coming in the form of fungal infection.[2] Noninfectious causes include sarcoidosis, IgG4-related diseases, or prior mediastinal irradiation. When no prior insult is identifiable, FM is deemed idiopathic.[2] A rare disease, FM is often discovered as a result of complications from infiltration into adjacent mediastinal structures. FM has been shown to manifest as pulmonary vascular obstruction, superior vena cava syndrome, or postobstructive pneumonia.[3,4] This case highlights a rarely described cardiac manifestation of FM, the development of conduction abnormalities including high-grade AV block resulting in syncope. The mechanism of these conduction abnormalities may be secondary to direct infiltration of the conduction system or extrinsic compression by the periaortic mass. In a previously described case of complete heart block owing to FM, fluorodeoxyglucose positron emission tomography showed both extracardiac and myocardial uptake that were similarly metabolically active, suggesting direct infiltration as the cause.[5] FM can be difficult to differentiate from pulmonary malignancy or lymphoma on imaging. In the correct clinical context, classical findings on contrast enhanced computed tomography imaging may be sufficient to make the diagnosis of FM.[6] These findings include a calcified mediastinal mass or surrounding lymph nodes.[5] A definitive diagnosis can only be made by histological analysis.[6] Given the rare manifestations and severe symptomatology in this case, a tissue diagnosis was crucial to rule out other causes of disease.

This case highlights the need to expand the literature around cardiac involvement of FM. In the future, physicians may have the tools to confidently identify FM from clinical history and imaging alone. In patients without risk factors for malignancy, or with

FIGURE 3 Histologic Sections From Aortic Root Biopsy



(A) Histologic sections show a fibroinflammatory lesion composed of mature adipose and dense fibrous tissue with scattered lymphoplasmacytic infiltrates (stain: hematoxylin and eosin; original magnification ×20). **(B)** Occasional lymphoid follicles with germinal centers are present (stain: hematoxylin and eosin; original magnification ×40).

evidence of common insults that lead to FM, this may allow physicians to forgo an invasive biopsy and monitor the disease over time. As seen in this case of a 30-year-old nonsmoking woman without a family history of malignancy, if a noninvasive diagnosis were possible, this could prevent the need for median sternotomy and its associated morbidity. Unfortunately, FM is thus far an incurable disease and treatment is centered around interventions to manage its complications.[7] Further research is needed into understanding its pathophysiology for both prevention, early recognition, and treatment.

FOLLOW-UP

At a follow-up appointment 2 weeks after discharge, she had no further episodes of presyncope or syncope. She then switched insurance providers and established care within a different medical system, but is reportedly doing well with no need for a pacemaker.

CONCLUSIONS

We describe a rare case of cardiac involvement of FM. Although usually recognized after complications from involvement of pulmonary or other mediastinal structures, manifestations of her underlying fibrosis presented as high grade AV block leading to syncope. Recognition of FM as a cause of heart block will assist physicians in its diagnosis and in making the decision to pursue invasive biopsy.

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REFERENCES

1. Kusamoto FM, Schoenfeld MH, Barrett C. 2018 ACC/AHA/HRS guideline on the evaluation and management of patients with bradycardia and cardiac conduction delay: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol*. 2019;74(7):e51-e156.

2. Rossi SE, McAdams HP, Rosado-de-Christenson ML, Franks TJ, Galvin JR. Fibrosing mediastinitis. *Radiographics*. 2001;21(3):737-757.

3. Jain A. Mediastinal fibrosis, pulmonary vein stenosis, and the role of transesophageal echocardiography. *J Cardiothorac Vasc Anesth*. 2020;34(3):835-836.

4. Loyd JE, Tillman BF, Atkinson JB, Des Prez RM. Mediastinal fibrosis complicating histoplasmosis. *Medicine (Baltimore)*. 1988;67(5):295-310.

5. Bharadwaj R, Madakshira MG, Bharadwaj P, Sidhu HS. Sclerosing mediastinitis presenting as complete heart block. *J Clin Diagn Res*. 2017;11(5):ED12-ED14.

6. McNeeley MF, Chung JH, Bhalla S, Godwin JD. Imaging of granulomatous fibrosing mediastinitis. *Am J Roentgenol*. 2012;199(2):319-327.

7. Kern R, Peikert T, Edell E, et al. Bronchoscopic management of airway compression due to fibrosing mediastinitis. *Annals ATS*. 2017;14(8):1353-1355.

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