

## IMAGING VIGNETTE

INTERMEDIATE

## CLINICAL VIGNETTE

# High-Grade Spindle Cell Sarcoma of the Heart



## A Rare Cause of Mitral Valve Disease

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## ABSTRACT

A young female with pulmonary congestion suspected to be secondary to mitral valve disease with left atrial appendage thrombus was given therapy for heart failure and anticoagulation. Subsequent multimodality imaging with echocardiography and magnetic resonance imaging established an accurate but rare diagnosis of spindle cell sarcoma of the heart. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:675-7) © 2019 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/>).

A 37-year-old female with known history of osteogenesis imperfecta (OI) had recurrent admissions for cough, fever, and pulmonary congestion over a 3-month period. Upon examination, she was euvolemic with no audible murmur. Results of her blood counts, serology, cardiac markers, brain natriuretic peptide level, and blood culture analyses were unremarkable. Initially, computerized tomography suggested pulmonary ground glass opacities, left atrial (LA) wall thickening, obliteration of the LA appendage and the right pulmonary veins (PV) (**Figure 1A**). Transthoracic (**Video 1**) and subsequent transesophageal echocardiography (**Video 2**) showed a diffuse echogenic thickening of the LA surface (**Figure 1B**), obliterating the LA appendage and extending into the PV and onto the mitral valve leaflets (**Figures 1B and 1C**). Doppler imaging showed severe mitral stenosis (mean diastolic gradient of 15 mm Hg at a heart rate of 100 beats/min) and moderate mitral regurgitation and tricuspid regurgitation (**Figure 1C**, Doppler), with an estimated pulmonary artery systolic pressure of 70 mm Hg. This led to a severely dilated right heart from pulmonary venous hypertension leading to a small, under-filled left ventricle (**Figure 1B**, **Videos 1 and 2**). Cardiac magnetic resonance imaging showed extensive hyperintense soft tissue thickening of the LA, mitral valve leaflets, and the appendage on T-1-weighted images (**Figure 1D**). Post-gadolinium contrast immediate T-1-weighted and delayed images showed extensive enhancement of the LA soft tissue mass, right PV region, and mediastinal extension (**Figures 1E and 1F**). These findings were most consistent with a probable primary cardiac sarcoma.

The patient was transferred to a tertiary sarcoma center, and efforts to obtain a tissue diagnosis using endobronchial ultrasonographic bronchoscopy and transeptal biopsy were nondiagnostic. Open surgical excision was considered a prohibitive risk given the extent of the disease. Instead, an empiric therapy for sarcoma using ifosfamide and doxorubicin chemotherapy was initiated. Unfortunately, the patient developed neutropenic fever and sepsis prior to her gradual demise 5 months after her initial presentation. An

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Informed consent was obtained for this case.

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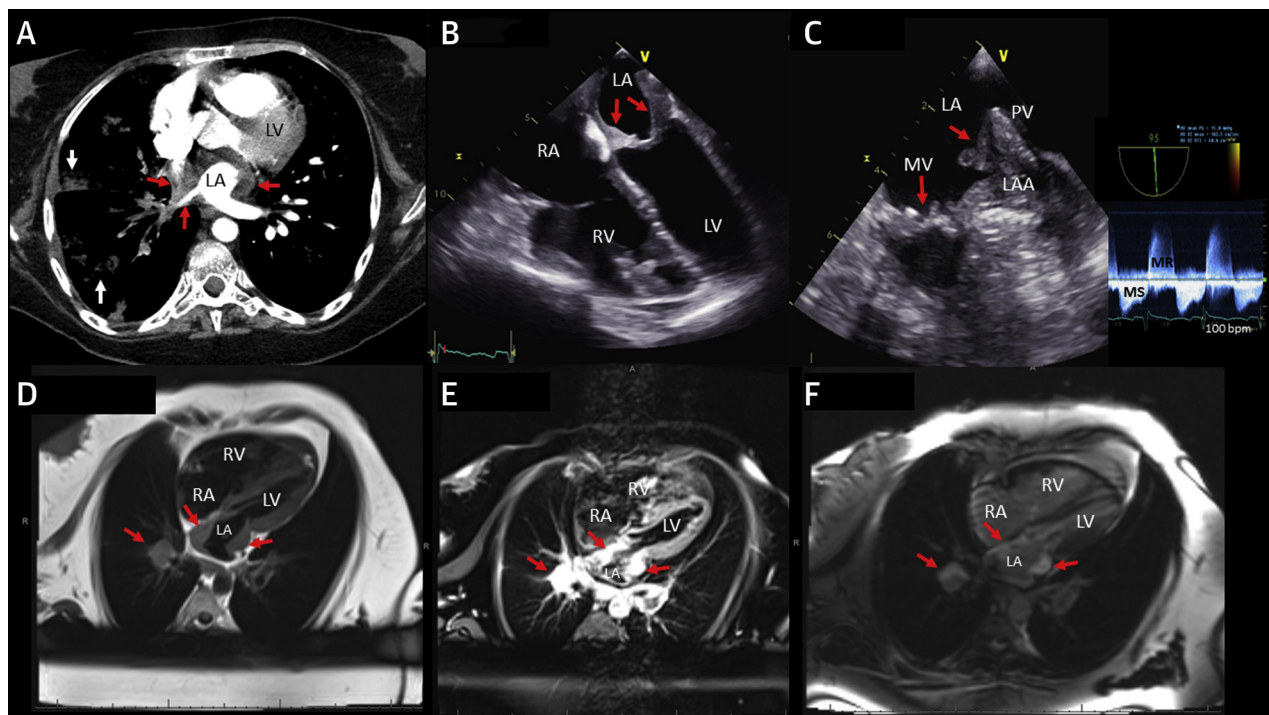
**ABBREVIATIONS  
AND ACRONYMS**

- LA** = left atrium
- LV** = left ventricle
- OI** = osteogenesis imperfecta
- PV** = pulmonary vein
- RA** = right atrium
- RV** = right ventricle
- TEE** = transesophageal echocardiography
- TTE** = transthoracic echocardiography

autopsy confirmed a diagnosis of primary cardiac high-grade spindle cell sarcoma with direct thoracic extension.

Primary cardiac sarcoma is rare (<0.01%), but spindle cell sarcoma (intimal sarcoma of the heart) is thought to be even rarer, as suggested by limited case reports (1,2). The diagnosis remains a challenge because of extended latency periods, atypical presentation, and aggressive nature (1,2). Presenting symptoms usually stem from obstruction, local invasion, metastasis, and embolic events and rarely result in sudden death (1). Although often encountered in larger arteries, LA infiltration with this tumor of connective tissue is known to occur (1,2). Tumor-inducing mitral valve pathology with LA appendage and lung infiltration can be misleading and result in potential erroneous use of anti-coagulation. Appropriate use of multimodal imaging techniques, as illustrated in the present case, can establish a prompt diagnosis of a rare invasive cardiac tumor. Roles of chemotherapy, radiotherapy, and surgical excision remain largely anecdotal from case experiences, as the overall prognosis is poor, with a mean survival of 3 to 12 months (1,2). OI is a rare, inherited autosomal dominant disease of type I collagen gene mutation with biologically plausible increased incidences of valvular regurgitation (aortic and mitral), ventricular dysfunction, vascular aneurysms, and dissection (3). Rarely, there are reported cases of osteogenic sarcoma, but to the authors' knowledge, there is no reported incidence of cardiac sarcoma in OI patients.

**FIGURE 1** Multimodality Imaging of the Case



Computerized tomography of the chest shows LA wall thickening, obliteration of the LAA, and the right-sided PV (A, red arrows), and spotty ground glass opacities of the right lung field (A, white arrows). Transesophageal echocardiography images of the LA wall thickening (B, arrows), RA, and RV dilation (B). Soft tissue thickening into the LAA and along MV leaflets are seen (C). Doppler echocardiography demonstrates severe MS and significant MR (C, Doppler inset). Cardiac magnetic resonance imaging of the 4-chamber view with hyperintense soft tissue thickening of the LA, LAA, and right PV regions on T1W image (D, arrows). Post-contrast immediate T1W and delayed images with contrast enhancement of the LA wall, right PV region with mediastinal extension (E, F, arrows). See Videos 1 and 2. LA = left atrium; LAA = left atrial appendage; MR = mitral regurgitation; MS = mitral stenosis; MV = mitral valve; PV = pulmonary veins; RA = right atrium; RV = right ventricular; T1W = T1-weighted.

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**KEY WORDS** cancer, cardiac magnetic resonance, echocardiography, mitral valve, thrombus, treatment

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 **APPENDIX** For supplemental videos, please see the online version of this paper.