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MINI-FOCUS ISSUE: IMAGING

INTERMEDIATE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Cardiac Myeloid Sarcoma

A 1-kg Heart

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ABSTRACT

A 53-year-old man with a background of acute myelomonocytic leukemia in remission presented with pleurisy. Repeat transthoracic echocardiography over several weeks revealed thickening left ventricular walls and decreasing systolic function. He died of decompensated heart failure due to cardiac myeloid sarcoma, with autopsy revealing an enlarged heart weighing >1 kg. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:963–5) © 2021 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 53-year-old man presented with pleuritic chest pain. His medical history was unremarkable except for a background of acute myelomonocytic leukemia (AML) that had been in remission for 3 years after treatment with an allogenic bone marrow transplant and fludarabine, cytarabine, and filgrastim chemotherapy. Clinical examination revealed atrial fibrillation with a heart rate of 120 beats/min, blood pressure of 108/60 mm Hg, oxygen saturations 91% on room air, temperature of 36.1°C, and a respiratory rate of 19 breaths/min. Jugular venous pressure was raised at 6 cm, and there were bibasal inspiratory crackles. Heart sounds were dual, muffled with no murmurs audible. There was mild pitting edema in his ankles. Transthoracic echocardiogram revealed severe left ventricular hypertrophy and a small pericardial effusion (Video 1).

Despite aggressive medical therapy with furosemide 120 mg BD intravenously, spironolactone 50 mg daily orally, and metoprolol 50 mg twice daily, the patient continued to deteriorate with increased work of breathing. His blood pressure dropped, and inotropes (including high-dose dobutamine and norepinephrine infusions) were initiated. Extracorporeal membranous oxygenation was considered; however, the patient died of acute heart failure before commencing therapy. On autopsy, his heart weighed >1 kg with a left ventricular thickness of up to 3 cm. There was diffuse fibrinous pericardium and multiple firm nodular masses on the epicardium. The myocardium was largely replaced by a firm homogeneous pale tissue comprising dense infiltrates of myeloid cells in keeping with extramedullary myeloid leukemia (myeloid sarcoma) (**Figure 1**).

Myeloid sarcoma, also known as granulocytic sarcoma, is a rare cancer most commonly associated with AML. It can present as isolated extramedullary infiltrations of immature myeloid cells. Such infiltrates may be seen concurrently or with relapse of previously diagnosed AML. Sites of occurrence include bone, periosteum, skin, and soft tissue and very rarely the heart. Patients can present with chest pain, arrhythmias, or heart failure. There is no consensus on treatment, with prognosis remaining poor despite chemotherapy, radiotherapy, and surgical intervention. To our knowledge, only one heart at autopsy has been reported as weighing >1 kg that similarly was caused by cardiac myeloid sarcoma (1,2).

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

AML = acute myelomonocytic leukemia

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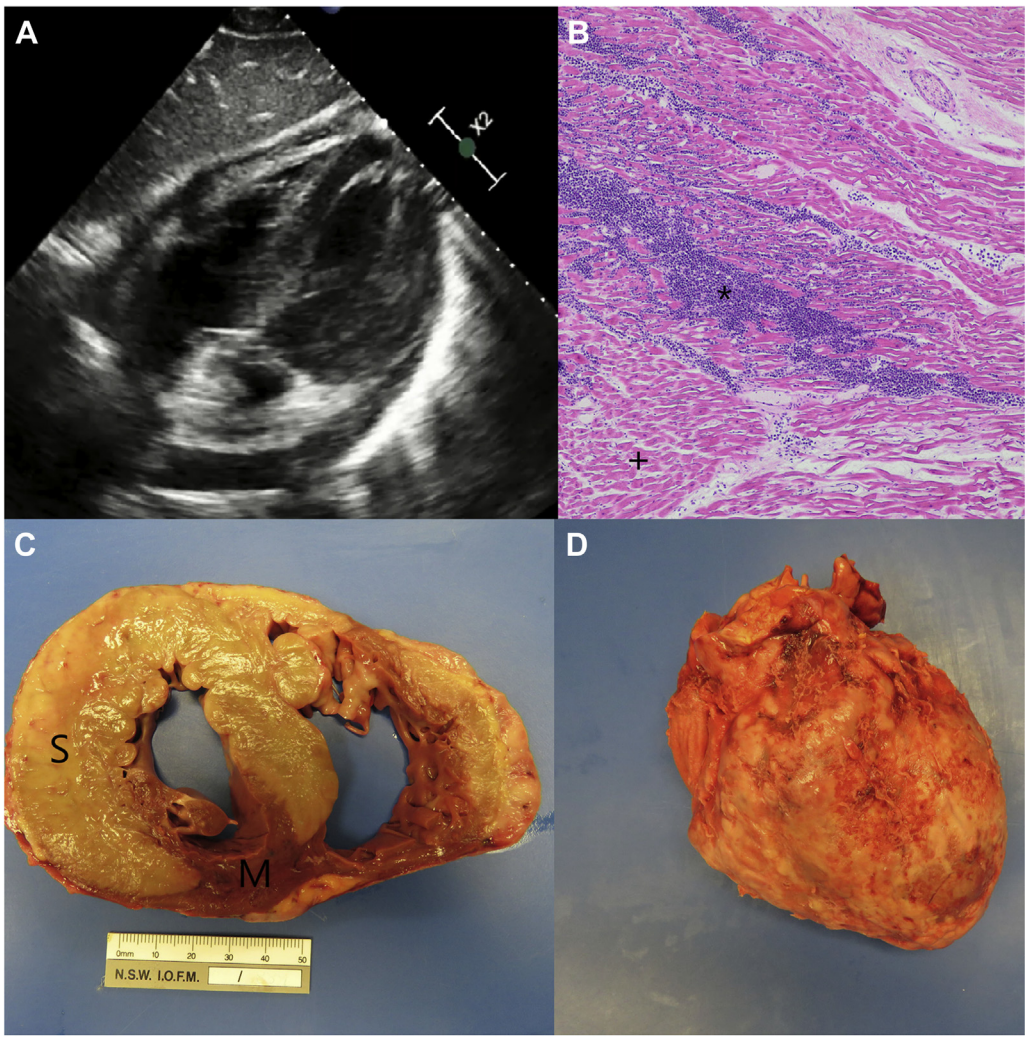
FIGURE 1 Imaging and Pathological Features of Cardiac Myeloid Sarcoma



(A) Transthoracic echocardiogram revealed severe left ventricular hypertrophy and a small pericardial effusion. **(B)** Histopathology sections of right and left ventricle show widespread infiltration and replacement of the myocardium (marked with +) and epicardial adipose tissue by a dense population of homogeneous small round blue cells (marked with *). Special stains (CD34, CD43, myeloperoxidase) confirmed myeloid origin. **(C)** Autopsy revealed that firm nodular masses (marked with "S") had replaced normal myocardium (marked with "M"). Left ventricle diameter of >3 cm and **(D)** the heart weighing 1 kilogram.

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KEY WORDS cancer, cardiomyopathy, left ventricle

APPENDIX For a supplemental video, please see the online version of this paper.