Large Symptomatic Ventricular Fibromas: A Surgical Challenge

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INTRODUCTION

Primary cardiac neoplasms, including fibromas, and rhabdomyomas, are often detected early, with presentation in adulthood exceptionally uncommon.[1] They are extremely rare clinical entities, with a reported prevalence of 0.0017% to 0.019%.[2] Historically, noninvasive imaging such as transthoracic echocardiography (TTE) has had a pivotal role in the presumed diagnosis and detection of fibromas, with many cases incidentally discovered. Fibromas are commonly observed in children, being the second most common cardiac neoplasm following rhabdomyomas.[3] Management remains case by case, with resection typically reserved for symptomatic patients or those with life-threatening complications. We present two cases in which we used TTE and cardiovascular magnetic resonance imaging (CMR) to detect a cardiac fibroma and to guide management. Diagnosis was confirmed by histopathologic review.

CASE PRESENTATION 1

A 45-year-old woman with a medical history notable for gastroesophageal reflux disease presented with palpitations, atypical chest pains, and shortness of breath. The patient described palpitations as occurring several times per week, lasting 10 to 15 sec.

Physical examination revealed a well-appearing woman in no acute distress. Blood pressure was 124/82 mm Hg, heart rate was 93 beats/min, and oxygen saturation was 97% on room air. The patient had no evidence of jugular venous distension. Cardiac examination was normal.

Complete blood count and basal metabolic profile were within normal limits. Electrocardiography demonstrated normal sinus rhythm with no ST-segment changes concerning for ischemia (Figure 1A). Chest radiography revealed fullness of the left cardiomediastinal contour (Figure 1B). Given concern about a possible tumor, TTE was requested. TTE revealed a large, echodense mass involving the basal to midanterior and anterolateral walls, with mass effect on the left atrium and left ventricle (Figure 2A and B, Videos 1 and 2). CMR was requested for further tissue characterization of the mass and to provide additional diagnostic certainty; it provided further structural confirmation of the mass, which measured 6.0 × 5.0 × 5.0 cm (Videos 3 and 4). On further characterization, the mass was isointense on T1- and T2-weighted sequences and hyperintense homogenously on late gadolinium enhancement sequences, suggestive of a cardiac fibroma (Figure 2C-E). Cardiac computed tomography (CCT) was requested to assist in preoperative planning and for potential concomitant bypass surgery. It showed normal coronary arteries and the close proximity and anterior location of the left anterior descending coronary artery in relation to the mass (Figure 2F).

Because of significant symptom burden, a multidisciplinary heart team opted to excise the mass surgically. During the operation, it was observed that the mass was abutting the left anterior descending and diagonal arteries. The fibroma measured 3.0 × 5.0 × 7.0 cm (Figure 2H). Given the concern of risk for injury to the coronary arteries, a subtotal resection (27 g, roughly 80%) was performed (Figure 2G and H). The patient tolerated the procedure well, and histology revealed fibroblasts interspersed with collagen fibers, with no mitotic figures or cellular atypia observed, consistent with the diagnosis of cardiac fibroma (Figure 2I). The symptoms resolved, and imaging revealed no further growth of the remaining tumor. The patient has remained symptom free through 1 year of follow-up.

CASE PRESENTATION 2

A 53-year-old woman with a medical history notable for chronic hepatitis C, hypertension, transient ischemic attack, and diabetes mellitus presented with chest pain, described as a left-sided pressure, intermittent in nature, and typically occurring with exertion but also at rest. The patient had no alleviating factors and reported occasionally waking at night from the same pain.

Physical examination revealed a thin (39 kg) woman. Blood pressure was 159/95 mm Hg, heart rate was 87 beats/min, and oxygen saturation was 99% on room air. The cardiac examination revealed normal S1 and S2 heart sounds, without heave, murmur, or gallop. There was no evidence of jugular venous distention. Lungs were clear to auscultation.

The basic metabolic profile and complete blood count were within normal limits. Initial electrocardiography demonstrated normal sinus rhythm (Figure 3A). Initial chest radiography revealed a contour abnormality of the left heart near the apex but otherwise normal results (Figure 3B). TTE was requested to evaluate a possible cardiac mass. TTE demonstrated a large mass involving the mid to apical lateral and anterior wall and normal left ventricular systolic function and mass effect of the left ventricle (Figure 4A, Video 5). Computed tomography of the chest confirmed the same (Figure 4B). For further

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Keywords: Cardiac mass, Coronary artery, Fibroma, Thoracic surgery

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VIDEO HIGHLIGHTS

Video 1: Subcostal four-chamber view (anatomic format) demonstrating a large ventricular fibroma along the anterolateral wall in patient 1.

Video 2: Apical four-chamber view demonstrating a large ventricular fibroma along the anterolateral wall in patient 1.

Video 3: Two-chamber CMR, steady-state free precession sequence demonstrating a large ventricular fibroma and a small pericardial effusion in patient 1.

Video 4: Short-axis CMR, steady-state free precession sequence in patient 1 demonstrating a large ventricular fibroma with a small pericardial effusion noted anterolaterally and posteriorly. This subsequently resolved after surgery.

Video 5: Apical four-chamber view demonstrating a large ventricular fibroma involving the anterolateral wall in patient 2.

Video 6: Four-chamber CMR, steady-state free precession sequence demonstrating a large ventricular fibroma involving the anterolateral wall in patient 2.

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tissue characterization, CMR was obtained. A large mass measuring 7.5 × 4.6 × 4.3 cm was noted in the mid to apical lateral wall extending anteriorly, and borders were indistinguishable from the myocardium in the inferolateral wall, suggestive of possible infiltration (Figure 4C, Video 6). The mass was heterogenous on T1- and T2-weighted images and hyperintense with some nonhomogeneity on delayed gadolinium enhancement images (Figure 4D-F). On the basis of the location and other tissue characteristics of the mass, large fibroma, fibrosarcoma, and leiomyosarcoma were considered. Computed tomography of the abdomen and pelvis revealed no evidence of metastatic diseases. Holter monitoring revealed predominantly normal sinus rhythm with the occasional supraventricular ectopic beat. Given the patient's comorbidities of diabetes and transient ischemic attack, a vasodilator stress test was performed for preoperative risk stratification; it was negative for ischemia. After evaluation by the surgeon, it was deemed that full resection would be impractical, as it would likely lead to an incomplete reconstruction of the left ventricle because of inadequate remaining myocardium. The patient underwent anterior thoracotomy and incisional biopsy. Histology and electron biopsy confirmed that the mass was a fibroma (Figure 4G-I). The initial intraoperative frozen specimen demonstrated largely absent myocardial tissue and interspersed collagen fibers (Figure 4G), and additional frozen images (Figure 4G and H) showed collagen bundles with minimal cardiac myocytes remaining. There was a clear absence of inflammatory cells. The pattern of collagen and gross description of the specimen favored fibroma over fibrosis or malignancy.

Subsequent follow-up demonstrated continued tobacco abuse, preventing further evaluation of possible cardiac transplantation. Serial follow-up with CMR for 6 years failed to demonstrate further growth of the mass or change in the tissue characterization on T1, T2, and late gadolinium enhancement images. Additional serial monitoring with Holter monitors and stress testing was undertaken until the patient elected hospice care because of severe protein calorie malnutrition and overall failure to thrive; this serial monitoring was negative for both nonsustained and sustained ventricular arrhythmia.

DISCUSSION

Cardiac fibromas are considered benign tumors yet have been associated with arrhythmias, heart failure, and even sudden cardiac death.[1,4-6] Miyake *et al.*[5] found that fibromas were more likely than

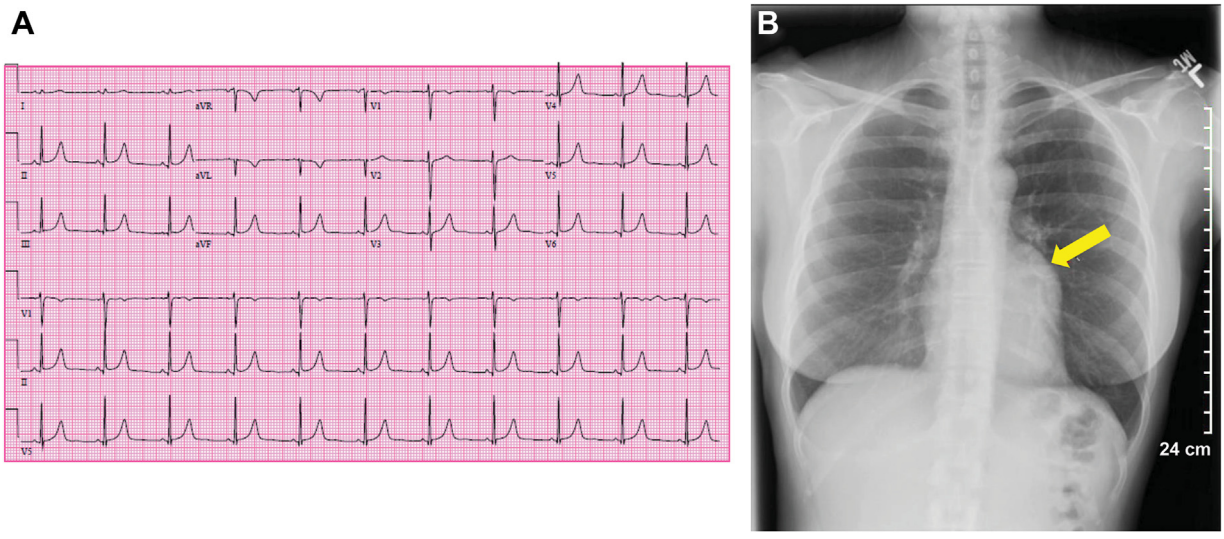

Figure 1 Initial electrocardiography and chest radiography of patient 1. **(A)** Initial presenting electrocardiography demonstrates normal sinus rhythm with no evidence of ST/T-wave changes suggestive of ischemia. **(B)** Initial chest radiography demonstrates a subtle smooth prominence (*arrow*) of the upper lateral left heart border.

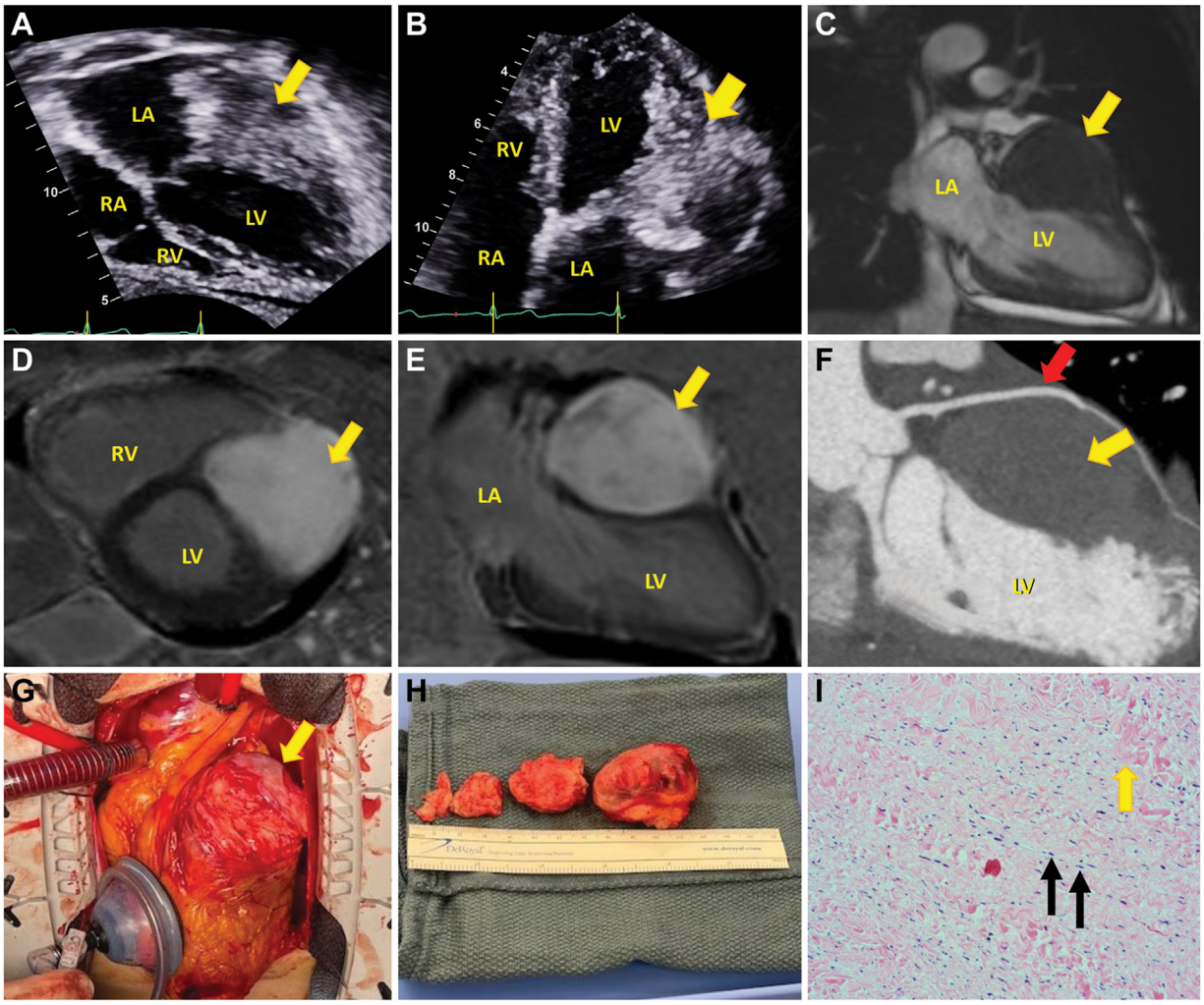

Figure 2 Multimodality imaging and histopathology of patient 1. **(A)** Two-dimensional TTE obtained in the subcostal view in systole (in congenital format) demonstrates a large mass consistent with a fibroma (*arrow*), with mass effect on both the left ventricle (LV) and left atrium (LA). **(B)** Two-dimensional TTE obtained in the apical four-chamber view in mid-diastole demonstrates a large mass consistent with a fibroma (*arrow*). **(C)** A two-chamber steady-state free precession cine CMR T1 image demonstrates the fibroma (*arrow*) involving the left ventricular myocardium. **(D)** Short-axis late gadolinium enhancement (LGE) sequence demonstrates a large homogenous, hyperintense mass consistent with a fibroma (*arrow*). **(E)** Two-chamber LGE sequence demonstrates a large homogenous, hyperintense mass consistent with a fibroma (*yellow arrow*). LGE shows the hyperintensity of the mass. **(F)** CCT shows the left anterior descending coronary artery (*red arrow*) in close proximity to the fibroma (*yellow arrow*). **(G)** An intraoperative photograph demonstrates the gross tumor (*arrow*) before surgical removal. **(H)** The gross specimen is seen upon removal. **(I)** Histopathology of the fibroma. The *yellow arrow* highlights the abundance of collagen; the *black arrows* indicate the spindle-shaped fibroblasts with no pleomorphism and no mitotic activity. *RA*, Right atrium; *RV*, right ventricle.

other primary cardiac tumors to cause arrhythmia in children. Reports of cardiac fibromas in adults are rare, and the true incidence remains unclear. Much of the data regarding cardiac fibromas have been extrapolated from studies in children to adults.

The most common location of a cardiac fibroma is generally the left ventricle, followed by the right ventricle and the interventricular septum.[7] Tumor location appears to be related to the outcome, with the ventricular septum being the most lethal location of a cardiac fibroma predisposing to conduction system disease and subsequent lethal arrhythmias.[7] Fibromas are derived from fibroblasts and thus can interdigitate with ventricular muscle. This results in both contractile dysfunction of the left ventricle and predisposition to arrhythmia by interruption of the normal conduction system.

Echocardiography is generally the imaging modality initially used, given its timely availability and feasibility.[8] The use of an ultrasound enhancing agent can be helpful for obtaining clearer delineation of the endocardial border when noncontrast images are suboptimal. In addition, ultrasound enhancing agents can determine vascularity of masses and better define tissue planes and their invasion by malignant

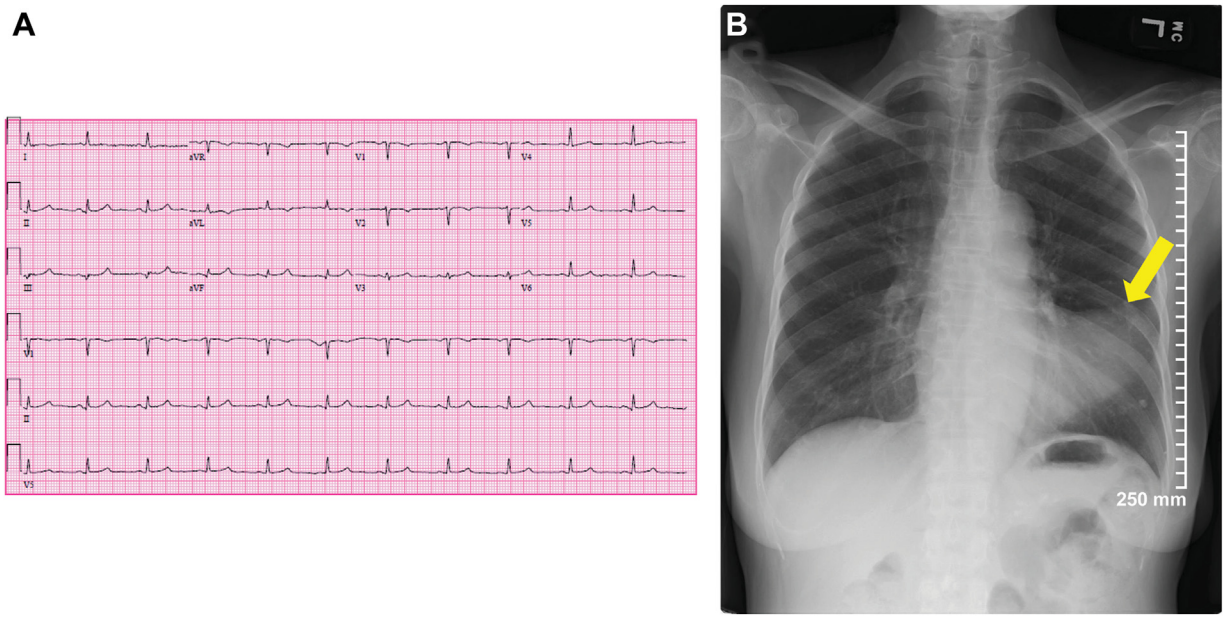

Figure 3 Electrocardiography and chest radiography of patient 2. **(A)** Initial electrocardiography demonstrates normal sinus rhythm. **(B)** Initial chest radiography demonstrates an abnormality near the apex (*arrow*).

masses. In our cases, the noncontrast image windows were optimal, and ultrasound enhancing agent was not used. However, in clinical practice, additional imaging with other modalities such as CCT and CMR is often required to determine further tissue characterization, anatomic extension, and invasion of adjoining structures for surgical planning.[8]

The left ventricle is the most common site for the tumor, as seen in both of our cases. Because of the infrequency, there are no true consensus guidelines for managing fibromas in adults. In the first case, we felt that the patient's clinical symptoms were due to mass effect, thus the requisite for surgical excision. Resection was challenging, however, with evidence of the tumor abutting the left anterior descending coronary artery, which was most clearly noted on CCT. After multidisciplinary discussion within our team of cardiologists and cardiac surgeons, we opted for partial resection to preserve the integrity of the epicardial coronary artery. Reports of partial resection remain scarce, but one study did not find evidence of recurrence in either partial or complete resection of cardiac fibromas, with follow-up extending to 30 years.[9] Rarely, there can be recurrence of a fibroma. One case noted a recurrence resulting in ventricular tachycardia, and with removal, cessation of the ventricular tachycardia.[10] Our second case demonstrated a fibroma that remained unresectable because of its size and anatomic location. Serial cardiac imaging revealed that the fibroma remained stable in size for 6 years.

CONCLUSION

These cases highlight the importance of echocardiography in the initial diagnosis of a cardiac mass and the utility of CCT and CMR in delineating the etiology of the cardiac tumor and perioperative planning. Although complete resection is ideal, partial resection may prove sufficient in certain selected cases given that the recurrence growth rate is extremely slow.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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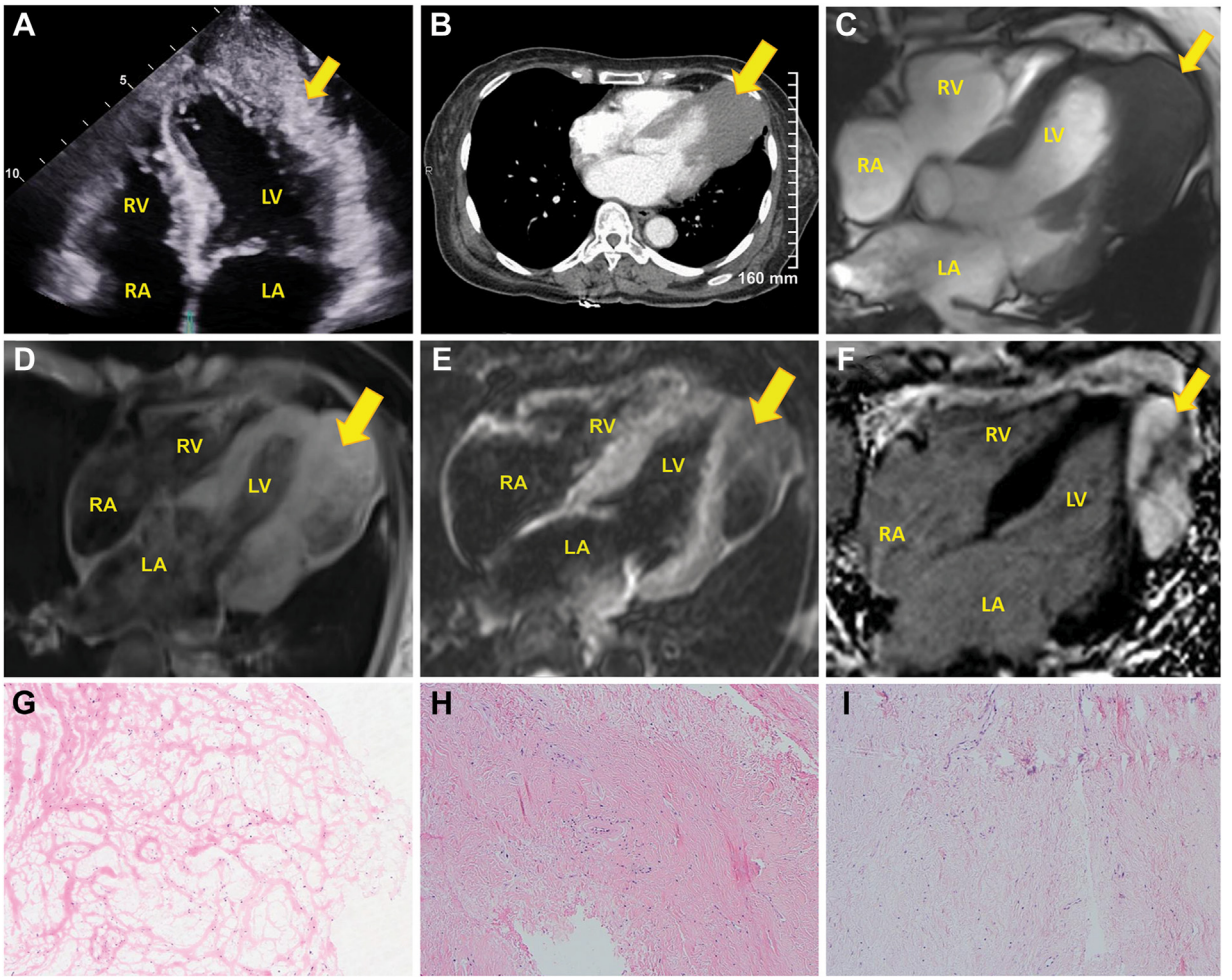

Figure 4 Multimodality imaging and histopathology of patient 2. **(A)** The two-dimensional transthoracic echocardiographic apical view in systole demonstrates the fibroma (*arrow*), with mass effect on the left ventricle (LV). **(B)** Chest computed tomography demonstrates the fibroma (*arrow*) extending from the lateral to the anterior wall in the short-axis view. **(C)** Four-chamber steady-state free precession cine CMR imaging demonstrates the fibroma (*arrow*) involving the left ventricular myocardium. **(D)** Four-chamber, T1-weighted double inversion sequence image demonstrates that the mass (*arrow*) is heterogenous and isointense. **(E)** Four-chamber, T2-weighted triple inversion sequence image demonstrates that the mass (*arrow*) is heterogenous and hypo- to isointense. **(F)** Four-chamber, LGE sequence image demonstrates the hyperintensity with some nonhomogeneity of the fibroma (*arrow*). **(G)** An initial intraoperative frozen specimen demonstrates largely absent myocardial tissue and interspersed collagen fibers. **(H)** A pathologic specimen again demonstrates collagen bundles with minimal remaining cardiac myocytes. There is a clear absence of inflammatory cells. **(I)** A pathologic specimen with large amounts of collagen and minimal to no remaining cardiac myocytes is seen. *LA*, Left atrium; *RA*, right atrium; *RV*, right ventricle.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.04.007.

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