An Unusual Cardiac Metastasis: Right Atrial Chondrosarcoma Diagnosed With Multimodality Cardiac Imaging

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INTRODUCTION

Arising from cartilage-producing chondrocytes, chondrosarcoma (CS) is the second most common bone tumor, with an estimated incidence of one in 200,000 people. CS typically presents as a local tumor in the pelvis, proximal long bones, ribs, scapulae, and vertebrae between ages 40 and 75 years, with a slight male predominance.[1] CS often follows an indolent course, with a favorable prognosis following surgical resection. However, hematogenous spread, most commonly to the lungs, portends a poor prognosis.[2] Metastatic CS to the myocardium has only rarely been described in case reports.[3] This article highlights the features of cardiac CS on echocardiography, cardiac magnetic resonance imaging (CMR), gross visualization, and pathology.

CASE PRESENTATION

A 56-year-old Hispanic woman presented to the hospital with acute onset of dyspnea. Her medical history included locally recurrent pelvic CS despite multiple resections and radiotherapy. In addition, she had a previous pulmonary embolus that was treated with 1 year of oral anticoagulation. Transthoracic echocardiography revealed a 58 × 29 mm heterogenous, multilobulated mass in the right atrium extending into the right ventricular inflow. Pericardial effusion was absent (Figure 1). The patient was started on unfractionated heparin for presumed thrombus in transit.

CMR was ordered to facilitate tissue characterization. A large, lobulated, mobile lesion was identified attached to the inferior wall of the right atrium, extending through the tricuspid valve into the right ventricle. The mass appeared avidly T2 hyperintense, had minimal uptake on first-pass perfusion images, was hypointense on early gadolinium enhancement images (inversion time 600 msec), and had mild patchy hyperenhancement on late gadolinium images (inversion time 230 msec). On the basis of these findings, tumor with surrounding thrombus was considered likely (Figure 2).

Transesophageal echocardiography was performed for preoperative planning (Videos 1 and 2). The mass had a highly irregular surface with a cystic/hypoechoic appearance. It had a broad-based attachment to the atrial wall with possible satellite lesions. The mass did not extend into the inferior vena cava or to the pulmonary artery. Importantly, a patent foramen ovale with a small right-to-left shunt at baseline was demonstrated on color Doppler (Figure 3). Given the patient's oncologic history, coupled with the irregular nature, tumor invasion suddenly seemed more likely than thrombus.

After right atriotomy, a large, gelatinous mass was identified in right atrium attached to a 1-cm stalk near the Eustachian valve (Figure 4). In addition, multiple large, cystic, firm tumor nodules were present in the right atrial wall. Frozen section was consistent with metastatic CS. The mobile mass was completely resected, but the satellite lesions were too extensive to safely remove and reconstruct the right atrium. A small, <1-cm patent foramen ovale was successfully closed. The final surgical pathology was consistent with metastatic, well-differentiated CS (Figure 5).

The patient remained hemodynamically stable throughout the case. After sedation weaning, the patient had new left-sided hemiparesis and hemineglect. Computed tomography revealed new hypodensities in the right posterior parietal and left frontal lobes concerning for multiple cardioembolic strokes. The patient did well postsurgically and was discharged to acute rehabilitation with oncologic follow-up.

DISCUSSION

Cardiac masses include thrombi, tumors, vegetations, and calcified lesions. Distinguishing among these entities can often be accomplished on the basis of clinical history, mass location, and echocardiographic appearance. When the diagnosis remains uncertain, CMR is a useful adjunct to ultrasound because of its superior tissue characterization. In this case, infective vegetation or calcified lesion did not fit the clinical situation. The patient's history of pulmonary embolus and malignancy made thrombus and tumor the most likely etiology of her mass. Thus, differentiating between thrombus and tumor became paramount.

Thrombus is the most common cardiac mass.[4] Thrombi can occur in any chamber of the heart and vary considerably in size, morphology, and mobility.[5] Given their avascularity, thrombi fail to show enhancement with ultrasound contrast after a high–mechanical index impulse destroys the contrast agent.[6] On CMR, thrombi appear hypointense on T1 imaging and hypointense on T2 imaging, and they do not enhance with contrast. Underlying conditions, such as atrial fibrillation, venous lines, pacemakers, mechanical valves, or clotting disorders, can predispose a patient to develop cardiac thrombus.

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Keywords: Chondrosarcoma, Right atrial mass, Transthoracic echocardiogram, Transesophageal echocardiogram, Cardiac magnetic resonance imaging

Conflicts of interest: The authors report no actual or potential conflicts of interest relative to this document.

2468-6441

https://doi.org/10.1016/j.case.2019.03.005

VIDEO HIGHLIGHTS

Video 1: Four-chamber transesophageal echocardiogram demonstrating the extent of the mass.

Video 2: Bicaval transesophageal echocardiogram showing the mass's attachment to the right atrial wall without extension into the inferior or superior vena cava.

View the video content online at www.cvcasejournal.com.

The World Health Organization divides cardiac tumors into benign or tumorlike lesions, malignant tumors, and pericardial tumors.[7] Nearly 90% of primary cardiac tumors are benign. Although primary cardiac tumors are relatively rare (prevalence ~1:2,000), metastatic tumors are nearly 20 times more common.[8] The most common tumors to present in the right atrium include metastases, myxoma, angiosarcoma, and lymphoma. Perfusion imaging using contrast echocardiography can aid in the characterization of cardiac masses, as the degree of enhancement relates to the vascularity of the tumor. Typically, malignant or highly vascular tumors demonstrate greater enhancement than the adjacent myocardium, as opposed to other

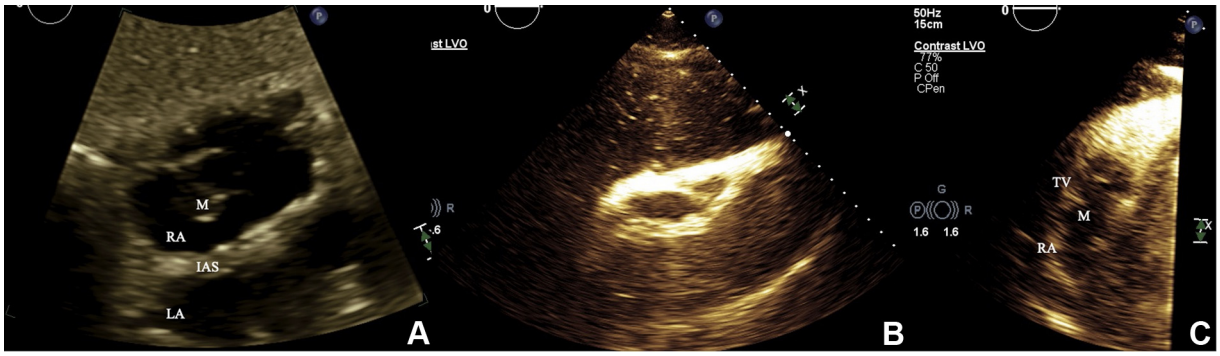

Figure 1 Transthoracic echocardiogram demonstrating the right atrial mass. **(A)** Subcostal view showing heterogenous mass in the right atrium (RA) with possible attachment to the right atrial wall. **(B)** Contrast-enhanced ultrasound provides border enhancement, allowing accurate measurement. **(C)** Right ventricular–focused apical four-chamber view with the mass protruding through the tricuspid valve (TV). *IAS*, Intra-atrial septum; *LA*, left atrium; *M*, mass; *RV*, right ventricle.

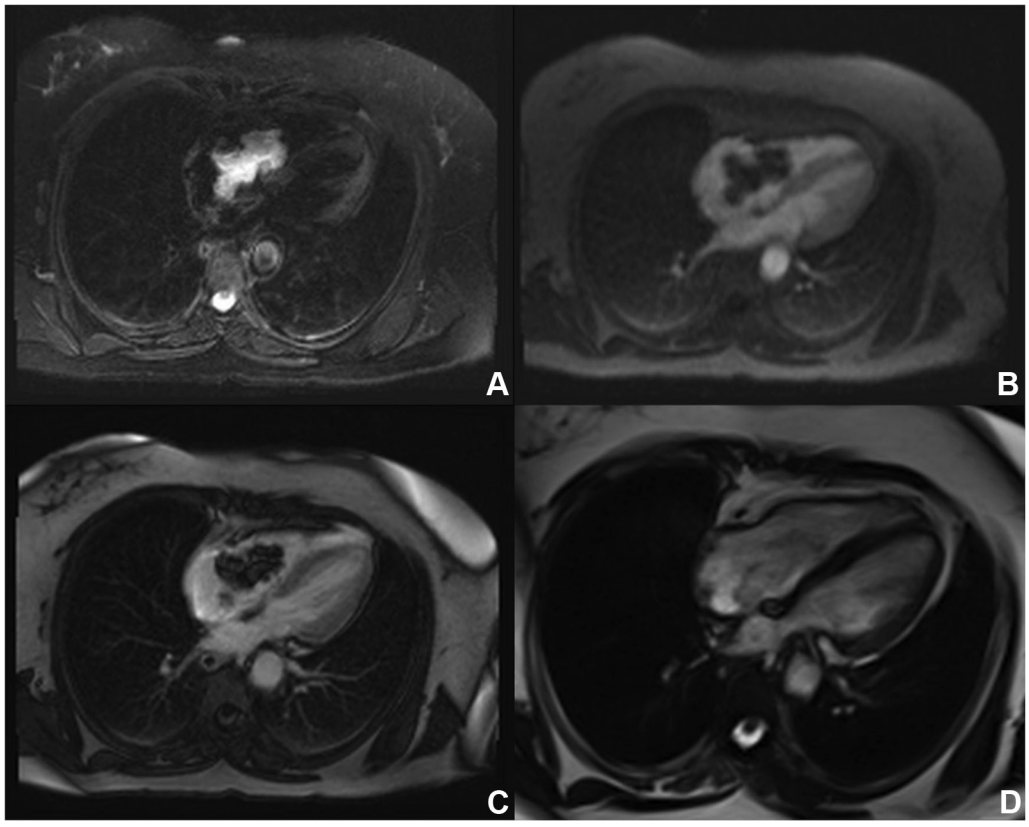

Figure 2 Four-chamber CMR sequences. **(A)** T2 short-tau inversion recovery sequence demonstrating avid T2 hyperintensity. **(B)** First-pass perfusion imaging with minimal uptake. **(C)** Hypointense early gadolinium phase-sensitive inversion recovery imaging (inversion time 600 msec). **(D)** Mild patchy enhancement on late gadolinium images (inversion time 230 msec).

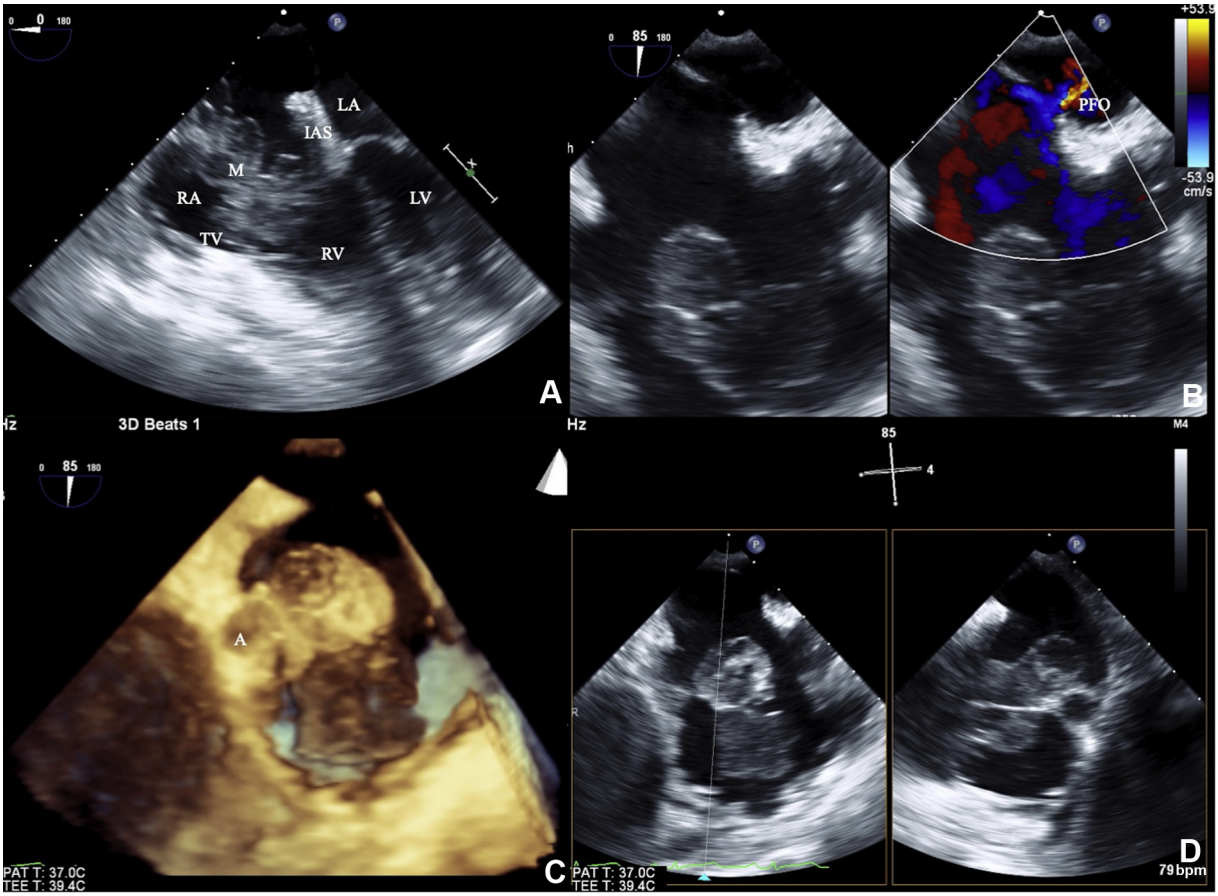

Figure 3 Transesophageal echocardiogram demonstrating right atrial mass. **(A)** Four-chamber view demonstrating large mass in the right atrium (RA) with extension into the right ventricle (RV). **(B)** Color-compare image highlighting a small patent foramen ovale (PFO) with right-to-left shunt by color Doppler. **(C)** Three-dimensional rendering of the right atrial mass and attachment site. **(D)** xPlane through the attachment site in the right atrial wall. *IAS*, intra-atrial septum; *LA*, left atrium; *LV*, left ventricle; *M*, mass; *TV*, tricuspid valve.

benign lesions in which the uptake of contrast is partial. CMR can distinguish between benign and malignant pathologies on the basis of tissue characterization. Metastases generally demonstrate hypointensity on T1 imaging, hyperintensity on T2 imaging, and heterogeneous enhancement with contrast.[9]

Because of their rarity, the cardiac manifestations of CS are limited to case reports. In a 1988 review, Leung *et al.*[3] documented 18 cases of metastatic CS to the heart. The most common site of metastasis was the right atrium (66%), followed by the right ventricle (33%), left atrium (28%), and left ventricle (17%). Patients most often reported dyspnea or pleuritic chest pain on presentation. Median survival following the development of cardiac symptoms was 18 months in patients treated with surgical resection ($n = 2$) and 2 months for those treated nonsurgically ($n = 16$). Since this case study, fewer than a half dozen new cases have been reported. One patient experienced an acute myocardial infarction due to tumor or thrombus embolization to the left anterior descending coronary artery.[10] The most recent example documents a left atrial CS leading to recurrent transient ischemic attacks.[11]

This case adds to the limited data available regarding the diagnosis and treatment of cardiac CS. The patient's presentation and history made tumor and thrombus the most likely diagnosis. However, the CMR tissue characterization strongly supported intracardiac thrombus. The unusual surface structure, cystic appearance, and broad-based attachment noted on transesophageal echocardiography made thrombus highly unlikely. Contrast uptake in perfusion images with echocardiography was equivocal, likely because of the concomitant presence of tumor and thrombus. Although CMR is generally viewed as the gold standard in cardiac imaging, in this case the findings on transesophageal echocardiography provided better tissue characterization that led to the diagnosis.

CONCLUSION

Cardiac masses can be differentiated by clinical context, anatomic location, and appearance on echocardiography. CMR can provide incremental information regarding tissue characterization. Cardiac CS is extremely rare, but the diagnosis can be achieved using a multimodality approach. The prognosis is extremely poor, but surgical correction may improve survival. This case adds to the limited experience with cardiac CS.

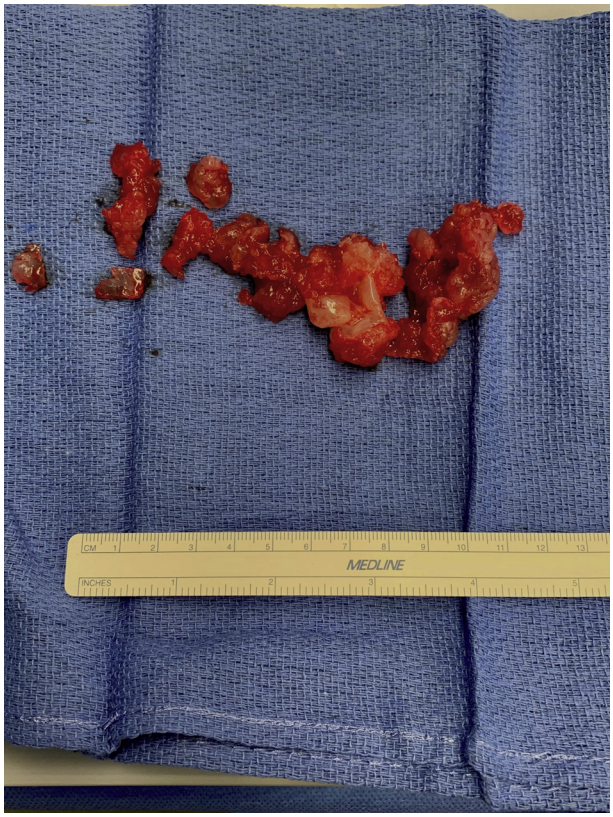

Figure 4 Gross pathology of the right atrial mass. The mass was large, highly mobile, and gelatinous in nature, consistent with cartilage. It attached to the right atrial wall near the Eustachian tube. There were multiple satellite lesions within the right atrial wall that could not be safely resected.

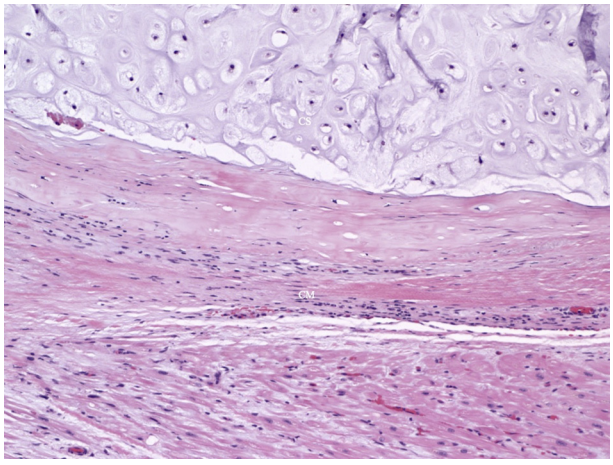

Figure 5 Surgical pathology of the right atrial mass. The top third shows a well-differentiated CS, with single chondrocytes and doublets within lacunae in a myxohyaline matrix. The cells have small round nuclei without atypia. Mitotic activity was not seen. The bottom third shows the adjacent cardiac muscle (CM), with a layer of intervening dense fibroconnective tissue.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.03.005.

REFERENCES

1. Ferguson JL, Turner SP. Bone cancer: diagnosis and treatment principles. Am Fam Physician 2018;98:205-13.
2. Chow WA. Chondrosarcoma: biology, genetics, and epigenetics. F1000Res 2018;7:F1000. Faculty Rev-1826.
3. Leung CY, Cummings RG, Reimer KA, Lowe J. Chondrosarcoma metastatic to the heart. Ann Thorac Surg 1998;45:291-5.
4. Basso C, Rizzon S, Valente M, Thiene G. Cardiac masses and tumours. Heart 2016;102:1230-45.
5. Waller BF, Grider L, Rohr TM, McLaughlin T, Taliercio CP, Fetters J. Intracardiac thrombi: frequency, location, etiology, and complications: a morphologic review. Clin Cardiol 1995;18:477-9.
6. Kirkpatrick JN, Wong T, Bednarz JE, Spencer KT, Sugeng L, Ward RP, et al. Differential diagnosis of cardiac masses using contrast echocardiographic perfusion imaging. J Am Coll Cardiol 2004;43:1412-9.
7. Travis WD, Brambilla E, Burke AP, Marx A, Nicholoson AG. WHO classification of tumours of the lung, pleura, thymus and heart. Lyon, France: IARC Press; 2015.
8. Camm JA, Luscher TF, Serruys PV. ESC textbook of cardiovascular medicine. New York: Oxford University Press; 2009:735-61.
9. Desai MY, Jellis CL. Differentiation of cardiac masses by CMR: judging a character by the company it keeps. JACC Cardiovasc Imaging 2014;7:906-8.
10. Di Michele S, Mirabelli F, Galzerano D, Mankad S. An unusual myocardial infarction. Echo Res Pract 2014;1:K9-12.
11. Sundboll J, Hansson NHS, Baerentzen S, Pareek M. A fatal case of primary cardiac chondrosarcoma presenting with amaurosis fugax. BMJ Case Reports 2015. bcr2015212178.