Epicardial Cavernous Hemangioma: A Diagnostic Challenge

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INTRODUCTION

Primary cardiac tumors documented in the literature are exceedingly rare, with an incidence estimated at 0.001 to 0.3 per 100,000 patient-years, on the basis of a large autopsy study.[1-3] Although a large majority of these are benign tumors (75%), there is a broad differential diagnosis, including cardiac myxoma, papillary fibroelastoma, rhabdomyoma, lipoma, and hemangioma, all of which present their own unique clinical associations and management.[1] Secondary cardiac tumors are 40 to 100 times more common. Cardiac metastases originate most frequently from lung, renal cell, breast, and skin cancers.[4]

We present a case of a cardiac mass of unknown anatomic origin for which multiple imaging modalities were required for proper diagnosis and surgical planning.

CASE PRESENTATION

A 73-year-old woman with a medical history significant only for mild, diet-controlled hypertension and psoriasis presented to her primary care physician with several months of progressive fatigue and dyspnea. The patient denied symptoms of chest pain, orthopnea, lower extremity edema, and syncope and was an otherwise healthy nonsmoker, requiring no medications daily. Initial workup included a complete blood count, basic metabolic panel, electrocardiography, and stress myocardial perfusion scintigraphy; all results were within normal limits. Computed tomography was ordered and showed a cardiac mass, possibly pericardial, causing extrinsic compression of the left atrium. Follow-up transesophageal echocardiography (TEE) was ordered at an outside hospital and revealed a normal left ventricular ejection fraction of 60% to 65%, no wall motion abnormalities, and no hemodynamically significant valvular disease but reported a mass suggestive of left atrial myxoma on the left atrial free wall above the left atrial appendage (LAA), which appeared to be on a stalk. The patient was then referred to our center for surgical evaluation for mass excision. Given two contradicting reports of anatomic origin (computed tomography suggesting a pericardial location and TEE suggesting an intra-atrial location; Figure 1), cardiac magnetic resonance imaging (MRI) was ordered to further characterize the mass. MRI revealed a 6.3 × 6.2 × 3.9 cm, well-circumscribed, homogenous mass in the posterior pericardial space, compressing the left atrium (Figure 2A–F, Video 1). Rapid, central gadolinium enhancement was noted on T1-weighted gradient-echo sequences, suggesting a perfused mass (Video 2). Left heart cardiac catheterization revealed nonobstructive coronary disease, along with a vascular structure filling from the left circumflex coronary artery (Video 3).

She was scheduled for resection of the mass; however, surgical planning was complicated by the contradictory diagnoses made on preoperative imaging. Thoracic surgery was consulted by cardiac surgery to review the images in the event the mass appeared to be pericardial and amenable to minimally invasive thoracoscopic excision. On the basis of review of the outside hospital's TEE, cardiac MRI, and cardiac catheterization, it was believed that the mass was extracardiac, but not necessarily pericardial, and would require sternotomy with cardiopulmonary bypass (CPB).

Intraoperatively, general anesthesia was induced without hemodynamic compromise. Pre-CPB TEE showed a fixed mass occupying the exterior aspect of the LAA, with no visible tissue plane between the mass and the LAA wall. The mass was surrounded by pericardial fluid with a sharp pericardial inflexion, which from some angles made it appear intracardiac; however, it was completely immobile, suggesting an epicardial location (Figure 3A and B, Videos 4–7). Again, it was noted that the patient had normal biventricular function and no significant valve pathology. Surgical exploration found a 5.2 × 4.3 × 2.2 cm, red, spongy mass adherent to the LAA wall. This required resection together with the LAA and prolonged CPB duration to adequately repair the remaining left atrial tissue and to obtain surgical hemostasis. The surgical team noted one artery and one vein exsanguinating at the excision site; both were successfully clipped. Following a second return to CPB, platelets, fresh frozen plasma, and cryoprecipitate were given, but the patient continued to bleed and we again returned to CPB. BioGlue and Surgicel were placed over the atrial incision, and hemostasis was achieved. The patient remained hemodynamically stable after CPB on minimal pressor support. Postoperatively, histopathology was consistent with an epicardial cavernous hemangioma (Figure 4A and B).

DISCUSSION

Cardiac hemangiomas are exceedingly rare occurrences. They can derive from all three cardiac layers, endocardium, myocardium, and epicardium, as well as pericardium.[2] Furthermore, these tumors can be located in multiple anatomic locations, the majority of which present in the right ventricle (36%), followed by the left ventricle (34%), then the right atrium (23%), and last the left atrium (7%).[1,4,5] Furthermore, hemangiomas are classified into cavernous type, those with large, thin-walled vascular spaces; arteriovascular type, with an unorganized malformation of arteries and veins; and hypervascular capillary type.[4,5] Cavernous hemangiomas are notoriously slow growing and rarely invade surrounding structures; however, they commonly cause symptoms of external compression, including outflow tract obstruction, atrial compression, ventricular dysrhythmias, and embolization.[3-5] Improved imaging modalities have largely simplified diagnosis of these cardiac tumors. Typically,

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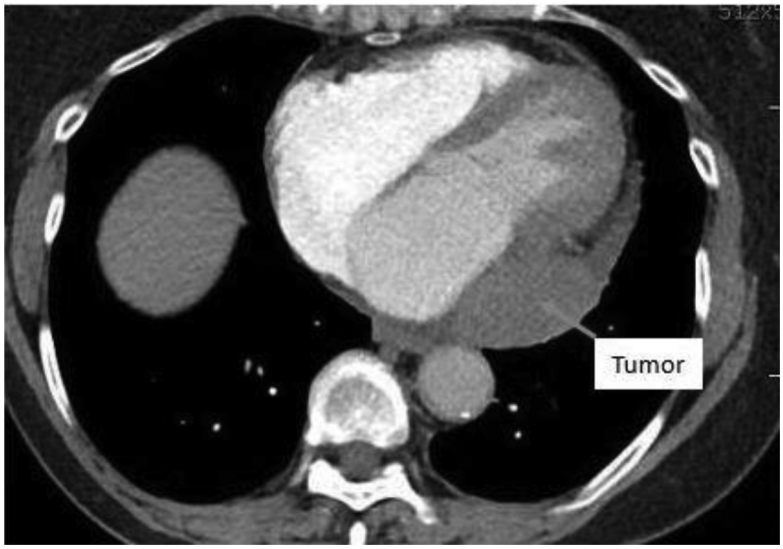

Figure 1 Computed tomography: modified four-chamber view showing well-circumscribed, homogenous mass abutting the left atrium within the confines of pericardium. There appears to be neither intra-atrial or intracardiac nor extracardiac extension.

echocardiography is the methodology of choice, and in the case of a hemangioma, contrast, if used, will be rapidly taken up by the highly vascular tumor. This can also be helpful when differentiating an intracardiac mass from a thrombus, which would be nonenhancing. In retrospect, we could have used this technique of contrast TEE when anatomic location appeared unclear; however, our transesophageal echocardiographic images were obtained intraoperatively, before CPB, and we were unable to use contrast, because of time constraints. Furthermore, hemangiomas have classical characteristic features seen on other imaging modalities discussed earlier; for one, computed tomography with contrast will also show rapid, intense tumor enhancement. Cardiac magnetic resonance images show an isointense or hyperintense signal compared with myocardium (T1-weighted and T2-weighted images, respectively), often because of high water content, and again, first-pass gadolinium images show rapid fill of the tumor with subsequent washout of contrast, suggestive of a vascular structure.[6,7] Cine magnetic resonance images can be useful to evaluate a mass's mobility relative to surrounding structures.[8] Late enhancement of gadolinium is not usually appreciated, as was the case in our patient's MRI study (see Figure 2A–F). Finally, cardiac catheterization can reveal a source of blood supply with rapid filling of the tumor with characteristic "tumor blush" and washout.[6,8] Despite these many reliable imaging techniques, it is often difficult to distinguish one type of benign cardiac tumor from another without histologic confirmation. Specifically, there were multiple clinical factors initially pointing toward a diagnosis of cardiac myxoma. Myxomas are far more common than cardiac hemangiomas, comparing an incidence of 50% and 2%, respectively. Myxomas typically present in elderly women, such as our patient, whereas hemangiomas present at all ages with a male predominance.[3,9] Furthermore, our patient presented with vague constitutional symptoms, commonly associated with intracavitary myxomas, thought to be secondary to excess cytokine release, which may have added support to a clinical diagnosis of myxoma. We could have additionally ordered preoperative erythrocyte sedimentation rate and serum C-reactive protein level (both typically elevated) and evaluated for the presence of hypergammaglobulinemia and anemia, also frequently associated with increased interleukin-6 levels in atrial myxomas.[9,10] Yet neither our transesophageal echocardiographic images nor complementary imaging were consistent with a classic myxoma. These tumors often arise from the interatrial septum with a stalk and are highly mobile, features not present in our TEE, and although cardiac catheterization, computed tomography with contrast, and gadolinium-enhanced MRI can show neovascularization and mass perfusion in the case of a myxoma, one would not necessarily expect the vascular fill-in and washout present with a hemangioma with these same modalities.[6,9,11]

CONCLUSION

Primary cardiac tumors continue to be rare; however, each histopathologic type has individual clinical consequences. Despite advances in medical imaging and echocardiography, this case highlights preoperative diagnostic limitations and resultant clinical implications.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.07.004.

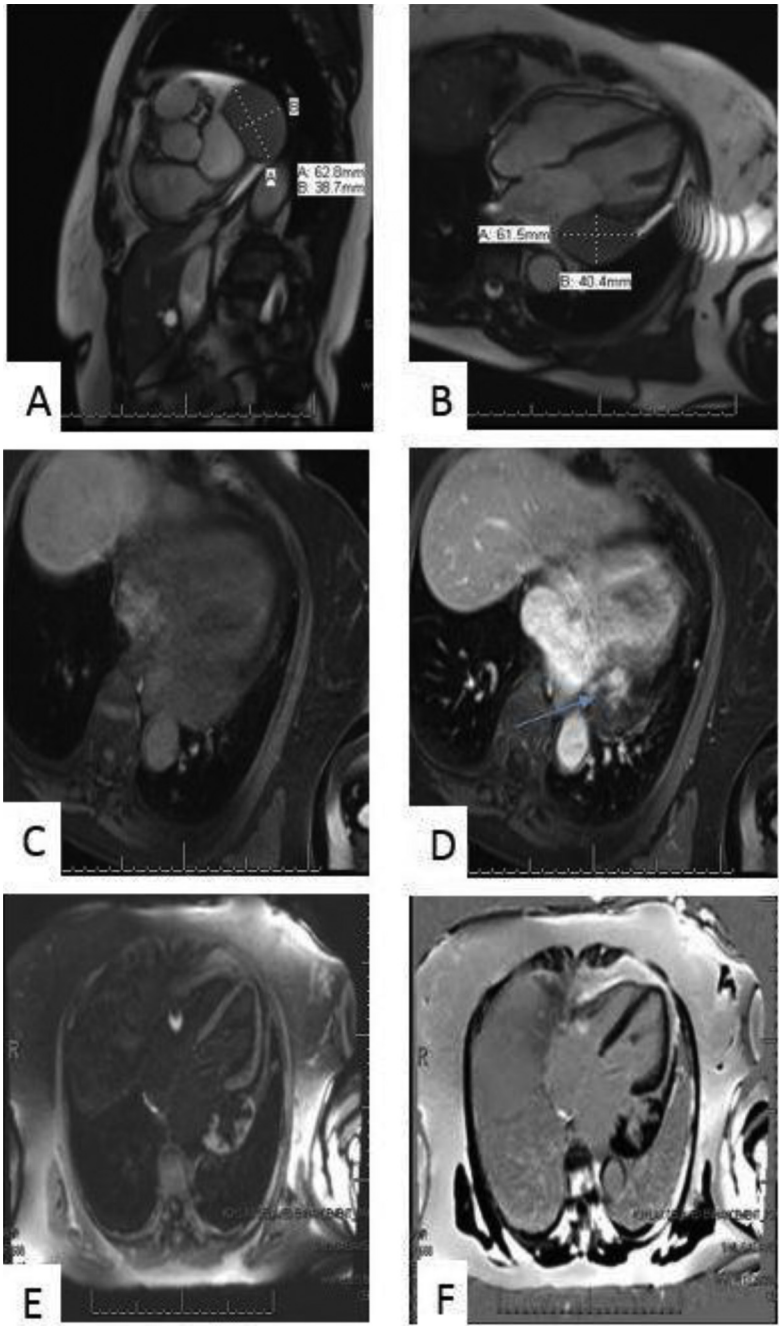

Figure 2 Cardiac MRI: **(A)** T1-weighted short-axis, sagittal view of cardiac mass compressing the left atrium. **(B)** T2-weighted four-chamber axial view of cardiac mass that appears within pericardium and does not extend into the left atrium. **(C,D)** T1-weighted four-chamber gradient-echo sequence with fat saturation before and after contrast showing rapid central, heterogeneous enhancement of the mass (*blue arrow*) indicative of significant blood supply. Delayed enhancement MRI: **(E)** T1-weighted and **(F)** T2-weighted four-chamber axial views showing mass that is isointense or slightly hyperintense to surrounding myocardium.

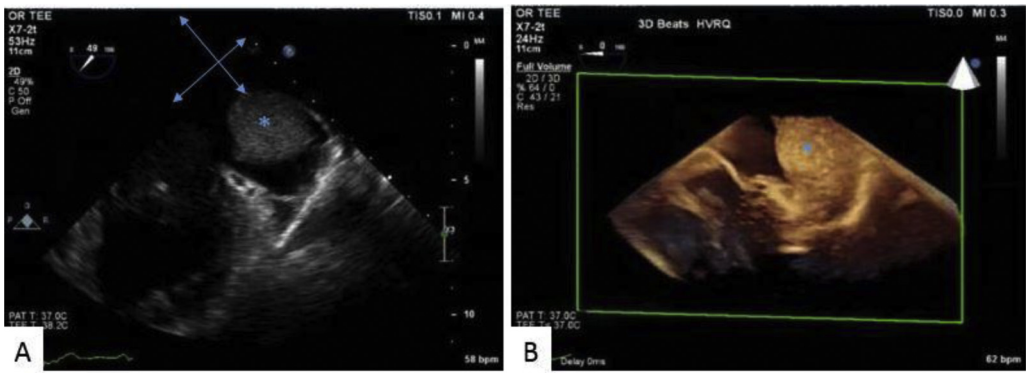

Figure 3 **(A)** Two-dimensional TEE: midesophageal mitral commissural view showing fixed mass (*blue asterisk*) in what appears to be the LAA. **(B)** Three-dimensional TEE: midesophageal view of LAA with noted mass (*blue asterisk*) with pericardial inflexions and small surrounding pericardial effusion.

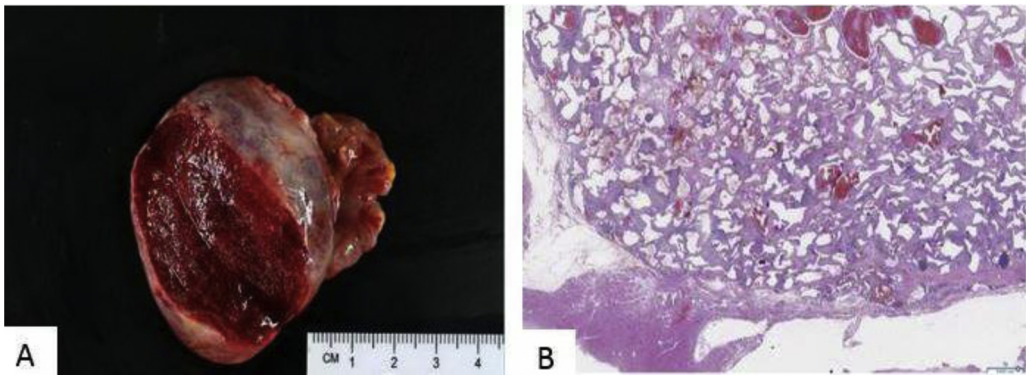

Figure 4 **(A)** External cardiac tumor, red-brown in color with semitranslucent capsule with attached portion of the LAA. **(B)** Histopathologic specimen with Hematoxylin and Eosin stain at 1× magnification showing tumor juxtaposed to myocardium in left lower corner. Tumor compared with myocardium has extensive endothelial-lined, thin-walled capillaries, consistent with a cavernous hemangioma.

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