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Computed tomography imaging characteristics of primary atrial intimal sarcoma

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ABSTRACT

We present a 36-year-old man who presented to our emergency department with acute onset shortness of breath and syncope. He was found to have a large left atrial mass on initial computed tomography (CT) which was confirmed by echocardiography. Tumor biopsy and attempted excision were performed, showing a primary cardiac spindle cell sarcoma that was unable to be resected, to which the patient eventually succumbed to. Spindle cell sarcomas of the heart are very rare primary cardiac tumors, with a variable, non-specific presentation. The most effective treatment is surgical resection, with chemotherapy and radiotherapy showing some benefit. Despite these treatments, the prognosis is poor. Given the uncommon nature of this tumor, the objective of this report is to demonstrate the clinical presentation and CT imaging characteristics of a case of primary cardiac spindle cell sarcoma, to raise awareness of this entity, and to increase the index of suspicion as a potential differential diagnosis to cardiac tumors seen on imaging.

1. Introduction

Primary cardiac tumors are extremely rare, with an incidence at autopsy ranging from 0.001% to 0.030% [1]. Of the primary cardiac tumors, three-quarters are benign, and approximately half of those are myxomas [2]. Of the malignant primary cardiac tumors, 95% are reported as sarcomas, the most common being angiosarcomas (34%). The least reported primary cardiac tumors are spindle cell sarcomas [2].

To our knowledge, as of 2015, there have only been six reported cases of primary cardiac spindle cell sarcoma originating from the left atrium [3]. Our case describes the seventh reported case. Due to the rarity of these tumors and the similar imaging appearance to benign cardiac masses or thrombi, accurate diagnosis is challenging. Presenting signs and symptoms are variable, ranging from asymptomatic to signs of heart failure, palpitations, and chest pain, and to neurological symptoms such as headaches, or confusion [3,4]. Therefore, prompt multimodal imaging and expert pathological diagnosis are necessary to guide treatment decisions. Although recurrence is common and complete cure is difficult, surgical resection is the mainstay of treatment, with radiotherapy or chemotherapy showing benefit in patients without metastases [2,5].

2. Case report

The patient is a thirty-six-year-old man with no past medical history who presented to the emergency department with progressive dyspnea on exertion for seven months, worsening over the two weeks prior to presentation, during which he also experienced a syncopal episode while resting in a car. The evening prior to presentation, he had a second syncopal event during an episode of coughing. He described associated night sweats and occasional palpitations. Physical exam showed an irregularly irregular rhythm with normal breath sounds bilaterally. The patient was febrile to 100.5 °F, normotensive but tachycardic at one hundred and thirty beats per minute on initial evaluation. Oxygen saturation was within normal limits. EKG showed atrial fibrillation with rapid ventricular response, right axis deviation and right ventricular hypertrophy. Chest radiograph showed cardiomegaly with clear lungs. The patient underwent a CT chest pulmonary embolism protocol, which demonstrated a low-attenuation mass with lobular contours, centered in the left atrium and measuring approximately 7.0 × 10.1 × 6.2 cm, with extension beyond the interatrial septum into the right atrium and prolapse into the mitral valve. Findings were suspicious for a left atrial myxoma. Transesophageal echocardiography showed a large eight to nine centimeter left atrial mass attached to the interatrial septum causing significant mitral stenosis and obstruction of the right pulmonary veins, consistent with an atrial myxoma or other

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Vent. rate	178	BPM	ATRIAL FIBRILLATION WITH RAPID VENTRICULAR RESPONSE WITH PREMATURE VENTRICULAR
PR interval	*	ms	OR ABERRANTLY CONDUCTED COMPLEXES
QRS duration	78	ms	RIGHT AXIS DEVIATION
QT/QTc	256/440	ms	RIGHT VENTRICULAR HYPERTROPHY
P-R-T axes	* 142	-14	NONSPECIFIC ST ABNORMALITY , PROBABLY DIGITALIS EFFECT
			NO PREVIOUS ECGS AVAILABLE

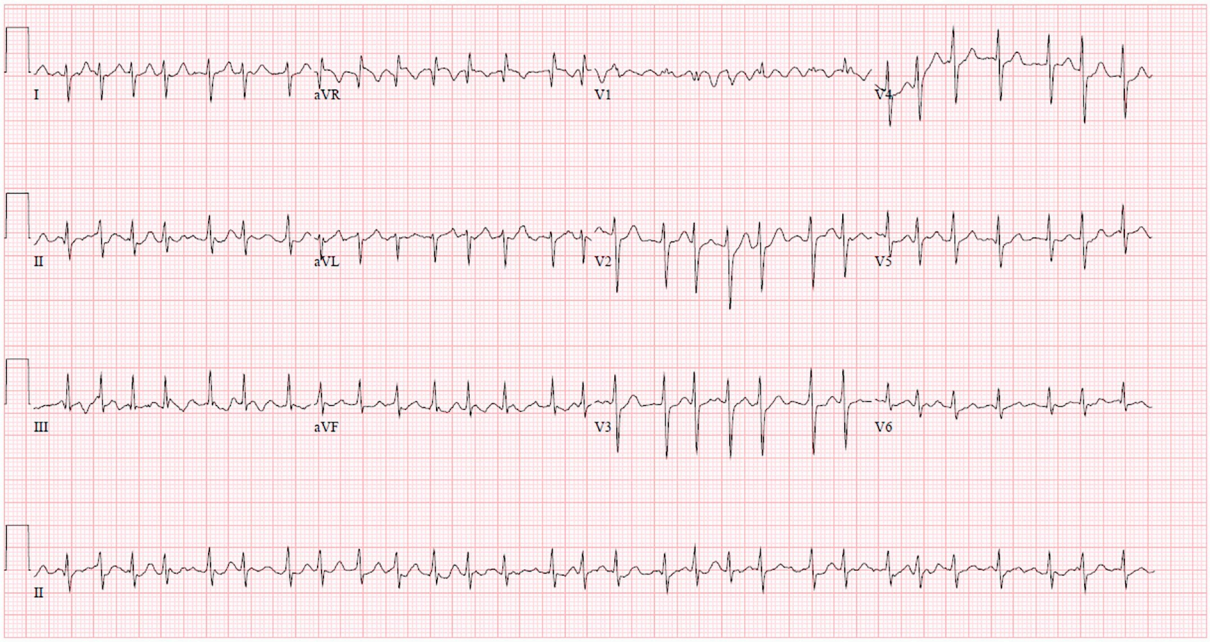

Fig. 1. Initial EKG demonstrating atrial fibrillation with rapid ventricular response, right axis deviation and right ventricular hypertrophy.

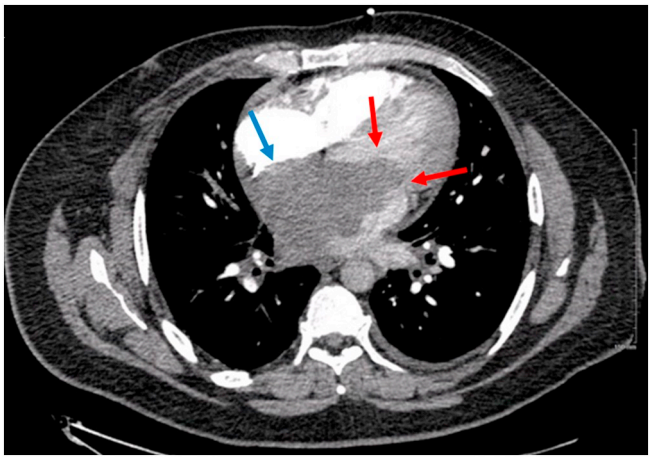

Fig. 2. Axial image from CT chest (pulmonary embolism protocol) showing a large, low-attenuation mass with lobular margins centered in the left atrium with involvement of the interatrial septum and extension into the right atrium (blue arrow) and mitral valve (red arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

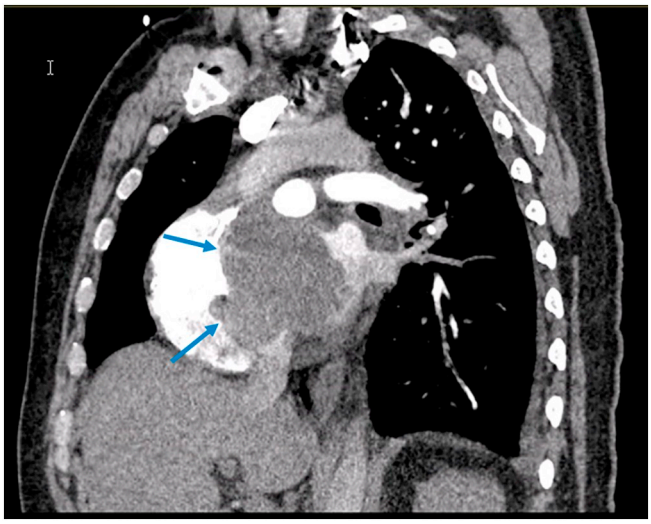

Fig. 3. Reconstructed short axis view through the atria showing extension of the mass into the right atrium (blue arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

atrial mass (Fig. 1–6).

The patient underwent evaluation by cardiothoracic surgery, including a tumor biopsy and attempted excision. The mass was deemed non-resectable intra-operatively. Pathology showed intimal sarcoma with focal infiltration into the myocardial tissue. The patient opted to wait for his parents to arrive from Ecuador before making a decision in regard to starting chemotherapy. Over the course of the hospital stay, the patient became increasingly short of breath with Rapid Responses for repeated episodes of supraventricular tachycardia and uncontrolled atrial fibrillation. Despite resuscitative efforts, the patient eventually succumbed to cardiopulmonary arrest prior to the arrival of his parents.

3. Discussion

Primary cardiac masses are diagnostically and therapeutically challenging. The common presenting signs and symptoms are variable and nonspecific, and the rarity of these diagnoses make initial suspicion very low. For these reasons, a higher index of suspicion and subsequent

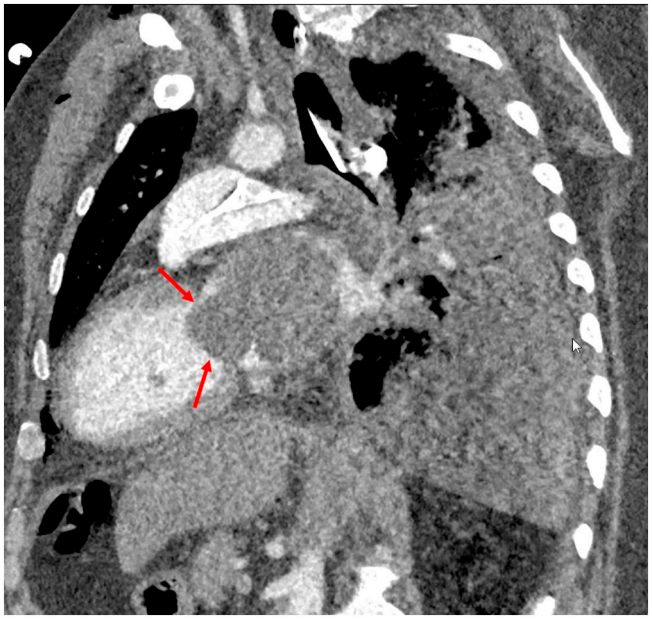

Fig. 4. Reconstructed vertical long axis view through the left atrium and left ventricle from CT chest, abdomen and pelvis following attempted excision again showing involvement of the mitral valve (red arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

multimodal approach to imaging can lead to earlier identification of a potential cardiac neoplasm. The benefits of early identification include diagnosis of the tumor while it is still considered operable which allows an aggressive surgical approach, prevention of dangerous or irreversible structural and electrophysiological damage, prevention of metastases, and improving overall long-term outcome. Even without early pathological diagnosis of an intimal sarcoma, an increased index of suspicion for aggressive cardiac neoplasms on imaging would prevent delay in delivery of the appropriate treatment.

According to the 2013 clinical recommendations from the American Society of Echocardiography, echocardiography is the recommended initial imaging test to assess cardiac masses [6]. Advantages to echocardiography include widespread availability, no radiation or contrast exposure, and dynamic visualization of cardiac activity. Echocardiography is not without disadvantages, though. The patient's body habitus can limit visualization, soft-tissue characteristics are poorly elucidated, there is an operator-dependent nature to the quality of image acquisition, and variable evaluation of extracardiac structures can result in incomplete understanding of the pathology [8]. Cardiac MR has been shown to be the preferred imaging modality for the diagnosis or cardiac masses, and is recommended for pediatric patients, where radiation is a concern. Cardiac CT is a viable alternative to cardiac MR in patients with contraindications to MR imaging and is considered superior for the evaluation of calcified masses and the assessment of concurrent coronary artery obstruction, which may impact the surgical timeline and decision-making.

Common imaging features associated with cardiac malignancy include: large (> 5 cm) lesions, right heart origin, broad-base, demonstration of enhancement, irregular/ill-defined margins, invasion, pericardial effusion, and calcification [8]. Clinical knowledge of a primary malignancy, preexisting hypercoagulable state, arrhythmia predisposing to intracardiac thrombus, or infection should also influence the differential diagnosis when encountering a cardiac mass on imaging. Although these features can favor malignancy over a benign tumor, definitive diagnosis is achieved with tissue specimen pathological examination. Due to the rarity of cardiac spindle cell sarcomas, specific imaging patterns have not yet been described. Our patient's tumor was larger than 5 cm, invaded into the interatrial septum and mitral valve, involved more than one chamber, had a broad-based attachment, an intra-cavitary component, and an associated pericardial effusion. These are all features favoring primary malignancy as described above, and are common features described in the other case reports of this rare condition. In comparison, cardiac myxomas often demonstrate a smooth border, often are pedunculated, can be mobile on cine, may be heterogenous if necrosis or hemorrhage is present, and are often not associated with a pericardial effusion. Calcification is present in only 10% [8]. They also commonly demonstrate hypointensity on T1 and hyperintensity on T2-weighted images, which help differentiate myxomas from thrombi. Thrombi are often associated with prior myocardial infarction, history of arrhythmia or hypercoagulable state, and signs of peripheral embolization. They often are homogenous, non-enhancing on CT and delayed-enhancing on MR, and do not enhance with perfusion studies due to their avascularity [7]. Metastases to the heart are most often located in the pericardium, followed by the epicardium and myocardium. Rarely do metastases involve the endocardium or intracavitary regions [8].

The prognosis of primary cardiac sarcoma is very poor, with a median survival of 3 months, although one case report describes a patient who survived 11 years after diagnosis [2,3]. Surgical resection is the essential treatment for these tumors, but early recurrence and

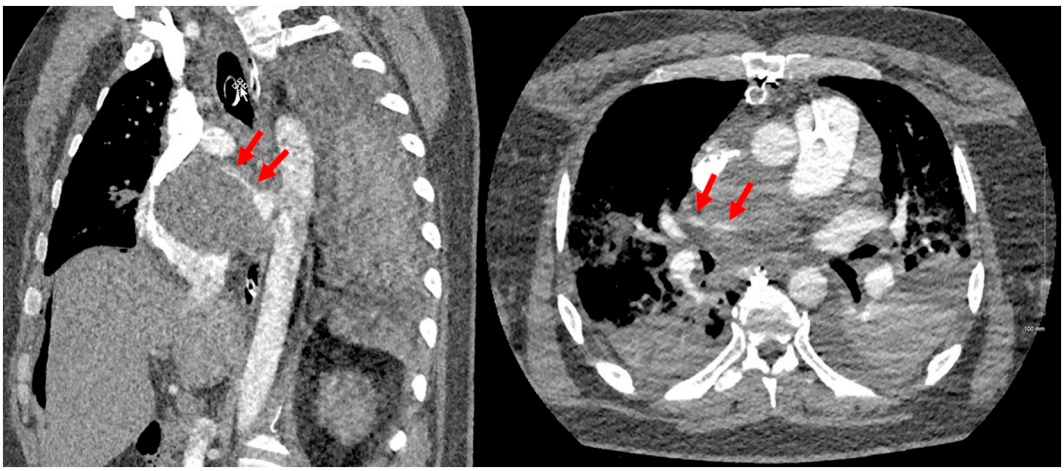

Fig. 5. Sagittal oblique reconstruction and axial image from CT chest, abdomen and pelvis following attempted excision showing severe narrowing of the superior right pulmonary vein (red arrows) secondary to extrinsic compression by the mass. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

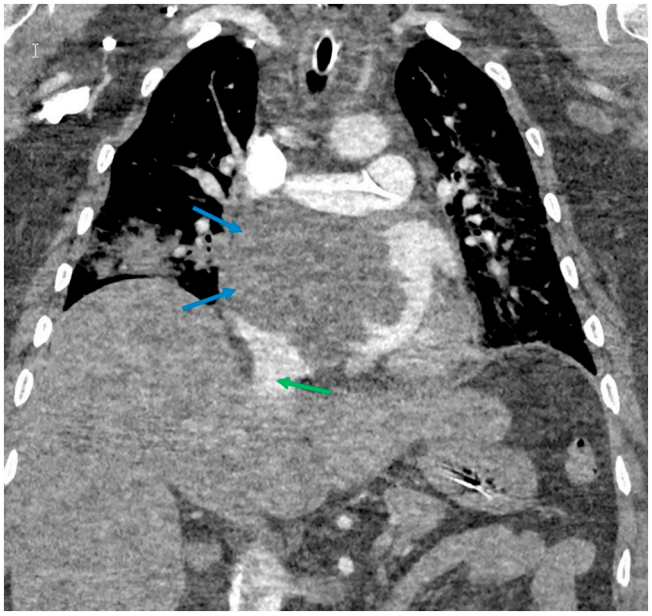

Fig. 6. Coronal reconstruction from CT chest, abdomen and pelvis following attempted excision again showing tumor invasion of the right atrium (blue arrows), as defined by the chamber receiving the IVC (green arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

metastases are common. As with our patient, arrhythmia is a common cause of death in affected patients; and even with successful resection, scar formation can lead to arrhythmia and death or can necessitate the need for lifelong anticoagulation. As more of these cases are reported, our hope is to increase the index of suspicion for this entity and other cardiac neoplasms, leading to earlier diagnosis, less delay in the appropriate therapy and therefore improved prognosis.

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