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Cardiac

Cardiac magnetic resonance imaging and a rare case of an atrial myxoma causing an atrial septal defect

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ABSTRACT

A 40 year-old athletic woman presented with worsening dyspnea on exertion over the preceding several months. Chest radiograph showed borderline cardiomegaly and subsequent echocardiography demonstrated a 5.0-cm left atrial mass as well as left-to-right interatrial shunting through a patent foramen ovale. Cardiac magnetic resonance imaging was performed, which demonstrated signal characteristics consistent with an atrial myxoma. The patient then underwent urgent surgical treatment with good technical and clinical outcome. Histologic examination confirmed an atrial myxoma. Cardiac magnetic resonance imaging was valuable in characterizing the nature of the atrial mass and patent foramen ovale, helping guide the surgical approach.

Introduction

Cardiac tumors, whether benign or malignant, require prompt diagnostic workup and therapeutic intervention. Transthoracic echocardiography (TTE) is widely available and remains essential in the initial workup of suspected cardiac tumors. However, it has significant limitations, to include limited fields of view of right heart and extracardiac structures, as well as operator and patient dependent factors. Additional imaging with transesophageal echocardiography (TEE) resolves some of these issues, but is invasive and requires sedation. Other imaging modalities, including computed tomography (CT) and cardiac magnetic resonance (CMR) imaging, are useful in evaluating cardiac tumors. Physicians should be aware that CMR has become increasingly valuable in both establishing a diagnosis and guiding appropriate therapeutic options for confirmed cardiac tumors [1].

The main differential of a left cardiac mass includes "pseudotumors" (eg, thrombus, anatomic variants), followed next by metastatic tumors and then primary cardiac tumors (benign and malignant) [1]. Metastatic tumors (eg, breast, lung, melanoma, lymphoma) are up to 40 times more common than primary cardiac tumors [2]. Primary cardiac tumors are rare,

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and greater than 75% are benign [1]. Of the benign primary tumors, myxomas make up the majority, followed by lipomas, papillary fibroelastomas (the most common valvular tumor), and rhabdomyomas (most common in children), to name a few [1]. Although making up less than 25% of all primary cardiac tumors, malignant sarcomas (angiosarcoma, fibrosarcoma, and rhabdomyosarcomas in children) are the second most common primary cardiac tumor [3]. Pericardial tumors can also mimic primary tumors, the 2 most common being teratomas and mesotheliomas [3]. The therapeutic options for some of these conditions vary widely. CMR helps differentiate benign from malignant tumors by the level of tissue invasion and extracardiac involvement, and thrombus from benign tumors by the presence of enhancement. Furthermore, the tissue type can be better characterized by the degree of sequence-dependent signal intensity.

Case report

A 40-year-old athletic woman presented to her primary care doctor complaining of dyspnea on exertion. The dyspnea had gradually worsened over the preceding months and significantly so over the few days before presentation, occurring after approximately 400 meters of running. She also admitted to audible wheezing, but denied other complaints on review of systems. Her only notable medical history was that her father suffered a myocardial infarction at age 43. Her vital signs and physical examinations, to include heart and lung examinations, were unremarkable. Given these nonspecific findings (wheezing and dyspnea on exertion), her primary care doctor elected to treat her for presumed exercise-induced asthma with albuterol before physical activity. Additionally, she was referred for pulmonary function tests and a chest radiograph.

Her pulmonary function tests revealed normal baseline spirometry. Her chest radiograph was notable for borderline heart enlargement without specific cardiac chamber enlargement. She continued to experience dyspnea on exertion, so an electrocardiogram (ECG) and TTE were ordered to evaluate her heart enlargement. Her ECG showed normal sinus rhythm, but with a P wave that was both 1.0 mm wide and deep in lead V1, consistent with left atrial abnormality. The TTE displayed normal left ventricular function and ejection fraction. A large left atrial mass, measuring 4.6 × 2.3 cm (Fig. 1A) and producing moderate functional mitral stenosis was present. Color Doppler imaging showed an atrial septal defect (ASD) with left-to-right interatrial shunting (Fig. 1B). A TEE, left heart catheterization, and CMR were subsequently performed to better characterize the mass and ASD. TEE revealed a normal-sized left atrium containing a multilobulated, heterogeneously echogenic mass (5.1 × 2.4 cm) attached anterior to the fossa ovalis, extending across the aorto-mitral continuity, and terminating 1.0 cm before the anterior mitral leaflet tip. Furthermore, the previously demonstrated ASD was shown to be a patent foramen ovale (PFO) that opened as the tumor descended into the left ventricle (Fig. 2A and 2B, and supplementary video). The left heart catheterization demonstrated faint calcifications and tumor blush of the mass, as well as atrioventricular and sinoatrial nodal arteries that were larger than usual, possibly supplying blood to the mass. The CMR displayed a well-circumscribed, noninfiltrative mass with a broad-based attachment to the interatrial septum as described above. This mass was mildly intense on T1-weighted imaging, highly intense on T2-weighted imaging, and showed no evidence of fat saturation. The mass showed very mild heterogeneous enhancement on both immediate and delayed post-gadolinium imaging. There was no evidence of malignancy such as a pericardial effusion, extracardiac masses, or pathologically enlarged hilar or mediastinal lymph nodes. Gradient echo CMR imaging redemonstrated the opening of the PFO and interatrial shunting as the attached mass descended into the left ventricle during diastole with excellent visual resolution (Fig. 3A-D).

A multidisciplinary team was convened to include members from the departments of cardiology, cardiothoracic surgery, and diagnostic radiology. Given the diagnostic information above, the mass was most likely benign and consistent with an atrial myxoma, for reasons detailed later. Regardless of the diagnosis,

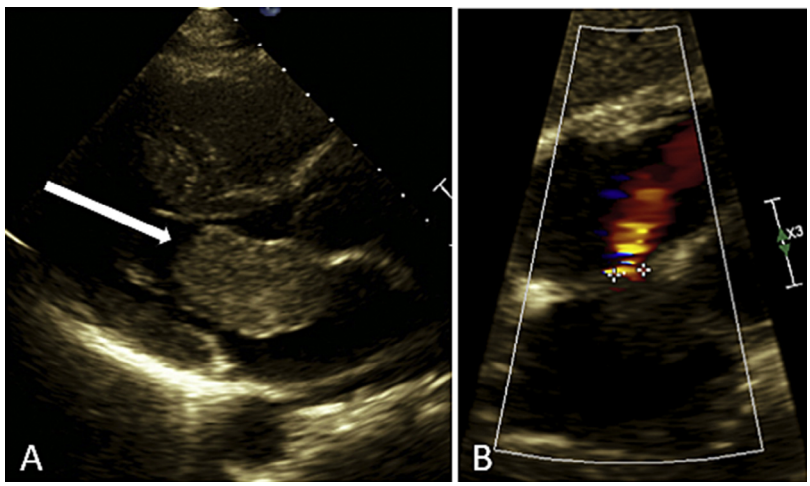

Fig. 1 – Transthoracic echocardiogram. (A) Gray-scale ultrasound image showing the left atrial mass with its distal tip (arrow) descending into the left ventricle during ventricular diastole. (B) Color Doppler image showing directional flow toward the ultrasound transducer and into the right atrium, signifying an atrial septal defect with left-to-right blood flow.