Correlation between transillumination echocardiography and pathologic anatomy in a left atrial myxoma: a case report

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September 7, 2022

Abstract

Left atrium myxomas are the most frequent heart tumors. Assessment of this phenomenon is largely echocardiographic, since both TTE and TEE provide information on size, shape, and hemodynamic consequences of the tumor. In this publication, we bring forward the similarities between morphological assessment through transillumination echocardiography and pathologic anatomy, highlighting a possible study field for the future.

Introduction

The lack of ongoing cellular proliferation in the adult human heart makes myocardial tissue notably resistant to tumor formation, which is why cardiac tumors are very rare (1), with an overall incidence of about 0.5/million (2). A myxoma is a benign growth in the heart that may develop in the right or left atria, or, more rarely, in the ventricles (1)(3)(4). This rare condition's etiology remains unclear, yet its origin has been associated with a proliferation of primitive stroma/connective tissue cells (1) and undifferentiated mesenchymal cells (3).

Usually, clinical manifestations are non-specific and are defined by the location, size, and mobility of the tumor. Myxomas may present in several ways, the most frequent manifestations being hemodynamic consequences like dyspnea, arrhythmia, palpitations, syncope, congestive heart failure and sudden death; systemic embolism such as transient ischemic attack (TIA), cerebrovascular accidents (CVA); and constitutional or systemic manifestations such as fever, weight loss, arthralgia, and fatigue (2). Here, we present a typical and very illustrative case and literature review of a patient with a left atrium (LA) myxoma, in order to orient clinicians in the assessment of this rare condition.

Case description

A 67-year-old female with a history of diabetes and hypertension was admitted to the coronary care unit due to dyspnea associated with dizziness, intermittent holocranial headache (Verbal Pain Intensity Scale 3/10), cough, and stabbing chest pain that had begun three years prior. Dyspnea had evolved from grade I to IV on the New York Heart Association (NYHA) Scale in a one month period, after which she consulted her physician, who referred her to the cardiologist after ordering an anteroposterior chest-x ray that showed left cardiomegaly. An apex beat was found on the 5th left intercostal space with a split second sound secondary to a tumor plop. Follow-up transthoracic echocardiogram (TTE) was performed, showing a mobile, regular

edged 59 x 49 mm LA mass with a thin peduncle attachment to the atrial septum, that protruded towards the left ventricle during diastole. An electrocardiogram was performed, showing sinus rhythm with a slight left axis deviation. (**Figure 1)** Subsequent Transesophageal Echocardiography (TEE) was performed, observing a mass in the LA.**(Figure 2)** Surgical removal of the mass was performed successfully, and the patient recovered fully after a few days. Histopathologic analysis showed characteristic mucoid degeneration with stellate cells **(Figure 3)** , thus confirming the diagnosis of a left atrial myxoma that correlated in shape and size to the mass observed through transillumination echocardiography **(Figure 4)** . Follow-up TTE showed the absence of the mass with an adequately functioning mitral valve and left ventricle.

Discussion

Myxomas are benign growths in the heart. When in the LA, they are usually attached to the fossa ovalis region (83%). They may also be found in the right atrium (12.7%), both atriums (1.3%) or more rarely in the left or right ventricles. (1.7% and 0.6%, respectively) (1)(3)(4).

Cardiac myxomas are observed in approximately 0.5-1 cases per 1 million people per year. (5) Atrial myxomas occur predominantly in females between the ages of 40 and 60, with a women-to-men ratio of 2.05:1. (6)

There are two types of cardiac myxomas: typical and atypical. Typical cardiac myxomas are the most common. They usually grow in the LA and attach to the interatrial septum, typically forming sessile or pedunculated masses with smooth or papillary surfaces. (7). Myxomas may also be inherited in the Carney complex11The Carney Complex is a rare, autosomal dominant, multiple endocrine neoplasia and lentiginosis syndrome, caused in most patients by defects in the*PRKAR1A* gene, which encodes the regulatory subunit type 1α of protein kinase A. Characterized by abnormal cutaneous and mucosal pigmentation, myxomas predominantly of the heart, skin, and breast, endocrine neoplasms, psammomatous melanotic schwannomas (PMS), breast ductal adenomas, osteochondromas, and other non-endocrine tumors. (8) due to mutations of the PRKAR1A gene. (5)

Myxomas present with a classic triad: obstructive symptoms, embolic signs and symptoms and constitutional or systemic manifestations (4)(8). A study from the National Heart Center of Singapore (NHCS) observed no significant differences in gender, body size, tumor size, hemoglobin counts, blood cell or platelet counts between symptomatic and asymptomatic groups. They also found that the most common symptoms are dyspnea, ischemic stroke and palpitations. (8)

Diagnosis is difficult in atypical presentations due to echocardiographic limitations such as lack of tissue characterization and a restricted field of view (3), therefore, definitive diagnosis of a myxoma should be made following surgical removal of the tumor and subsequent histopathological assessment, where characteristic stellate or globular cells may be observed. (8)

Cardiac myxomas are most commonly diagnosed through TTE or transesophageal echocardiography (TEE). Additional diagnostic tools are useful, like electrocardiography, chest computed tomography (CT) and chest or heart magnetic resonance imaging (MRI). There are no specific blood tests for this condition. (5)

TTE is useful for determining a myxoma's location, size, shape, attachment, mobility and hemodynamic impact. Characteristically, cardiac myxomas are found as a mobile pedunculated mass attached to the endocardial surface, usually in the interatrial septum, ranging in size from 3 to 4 cm in diameter. They are attached by a broad or narrow based stalk, frequently without involvement of adjacent structures. An homogenous or heterogenous aspect may be found depending on the presence of hemorrhage, calcification and surface thrombi. Distinctively, these heart tumors are mobile, which makes them prone to prolapsing into the left ventricle through the atrioventricular valves during diastole. TEE is also useful in these instances, improving spatial resolution and allowing for a better visualization of implantation and extension to adjacent veins (3)(5)(10). Our patient's case provides an excellent example of typical echocardiographic findings in patients with this clinical entity.

Typical LA myxomas may cause obstructive complications, such as mitral valve obstruction or regurgitation, left-sided heart failure and secondary pulmonary hypertension. The most common symptom referred by

patients is exertion dyspnea, followed closely by orthopnea. Furthermore, myxomas present with a high risk of systemic embolization, resulting in TIA, CVA, hemiplegia, loss of vision, chest pain and dyspnea. A wide number of patients (approximately 3.2% to 46.4%) with cardiac myxomas are asymptomatic, which toughens the clinical diagnosis, thus highlighting the need for imaging techniques like TTE, TEE, CT or MRI. These studies are also important for the characterization of the mass and subsequent surgical removal, which is the only definitive treatment for this condition. (5)

LA myxomas are the most common cardiac tumors, and as such, they should be the first clinical suspicion when a heart mass is observed through an imaging study. Moreover, the first step towards diagnosis should always be TTE. Our study suggests the usefulness of transillumination echocardiography for the assessment of tumor morphology, for it resembles macroscopic pathologic findings post-surgically and may be useful for surgical planning. However, more evidence is needed on the significance of this matter.

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Figure Legends

Figure 1. Electrocardiogram showing sinus rhythm with no significant anomalies.

Figure 2. A. TEE showing severe dilation of the LA (LAVI 58.7 mL/m^2) with a 59 x 49 mm mass **(*)** adhered to the interauricular septum. **B.** TEE at 139° showing the prolapse of the mass **(*)** towards the left ventricle during diastole.**C.** TEE at 0° showing protrusion of the mass **(*)**towards the LV during diastole. **D.** TEE at 0° showing a pedunculated mass **(*)** adhered to the interauricular septum in the LA, measuring 59 x 49 mm. Abbreviations: LA: left atrium; LV: left ventricle; AV: aortic valve; Ao: aorta; RA: right atrium; RV: right ventricle.

Figure 3. A. Macroscopic architecture of the resected left atrial myxoma. **B.** Mucoid degeneration with stellate cells (pseudo-neuronal tissue) **(*)** and septations **(black colored arrow)** lined by endothelial cells **(red colored arrow)** . A slight resin artifact is observed in the right side of the image.**C.** Mucoid degeneration **(*)** in the midst of fibrous tissue **(+)** forming pseudovascular canals **(black colored arrow)** . **D.** Slide showing the division between myxomal tissue**(*)** and myocardial tissue **(+)** .

Figure 4. A. TTE showing a pedunculated mass **(white arrow)** in the LA measuring approximately 60

x 50 mm. **B.** 3D reconstruction of the pedunculated mass **(white arrow)** .**C.** Transillumination echocardiography during systole showing a septated, regular, pedunculated mass **(*)** in the LA with its attachment **(white arrow)** to the interatrial septum. **D.**Macroscopic anatomy of the resected mass showing similar morphology to the one observed through transillumination echocardiography. Abbreviations: LA: left atrium; LV: left ventricle; AV: aortic valve; Ao: aorta.