Left atrial myxoma in a patient with nonspecific clinical presentation: A case report

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ABSTRACT

Introduction: Myxoma, the most common primary cardiac tumor, may cause a variety of clinical manifestations depending on its size and anatomical location. But also it may be presented with non-specific symptoms or remains asymptomatic. In this case report, the non-specific clinical manifestation is discussed.

Case Report: A case of cardiac myxoma in a 60-year-old female is presented. The patient was referred to our hospital with mild non-specific symptoms, with no prior history of syncope, shortness of breath, or chest pain. The elliptical mobile myxoma was diagnosed by transthoracic echocardiography. It was located in the left atrium, prolapsed through the mitral orifice during diastole, obstructing diastolic filling of the left ventricle. The patient was referred for surgical intervention.

Conclusion: The patient in this case report did not present with the common symptoms associated with an atrial myxoma, which was revealed by transthoracic echocardiography. Thus, echocardiography, as a screening method and a non-invasive, widely available and practical technique, plays a prominent role in the diagnosis of cardiac masses.
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Keywords: Echocardiography, Myxoma, Nonspecific symptoms

INTRODUCTION

Cardiac tumors are differentiated into primary and secondary. The prevalence of primary cardiac tumors is 0.001–0.03% in autopsy series [1]. Seventy-five percent of primary tumors are benign in origin. Myxoma is the most common, accounting for more than 50% of all primary cardiac tumors. Most cases are sporadic. The female-to-male ratio is 2–4:1.

Approximately, 7% of cardiac myxomas arise as components of a heritable disorder with spotty pigmentation of the skin and endocrinopathy, which is recently referred as Carney complex, which is a familial, autosomal dominant syndrome of recurrent cardiac myxomas and is previously described cardiac myxoma syndromes such as LAMB (lentigines, atrial myxoma, mucocutaneous myxomas and blue nevi) and NAME (nevi, atrial myxoma, mucinosis of the skin and endocrine overactivity) [2]. Mutations in the gene PRKARIA (which encodes enzyme cAMP-dependent protein kinase type I-alpha regulatory subunit) on chromosome 17 (17q24), which may function as a tumor-suppressor gene, plays an essential role in cardiac
development and myxomagenesis [3] and cause Carney complex. Multiple tumors occur in approximately 50% of familial cases and are more frequently located in the ventricle (13% versus 3% in sporadic cases).

About 75% are pedunculated and may prolapse through the mitral valve and obstruct ventricular filling during diastole. Tumors vary widely in size, ranging from 1 to 15 cm in diameter, and weighing between 15 g and 180 g [4]. They may be myxoid and gelatinous; smooth, firm, and lobular; round, polypoid, oval. As previously reported, the myxoma surface is friable or villous in 35% of the cases, and smooth in the other 65% [4]. Friable, villous, irregular tumors increase risk of systemic embolism. Myxomas usually are composed of areas with hemorrhage, necrosis, calcification, and cyst formation.

Myxomas are thought to originate in undifferentiated and totipotent mesenchymal stem cells [1]. They produce vascular endothelial growth factor (VEGF), which probably contributes to the induction of angiogenesis and the early stages of tumor growth [2].

Although cardiac myxomas commonly present as a benign neoplasm, there are many reports suggesting its malignancy, including recurrence of the tumor, locally invasive myxoma, extension from the heart, and distant metastasis or peripheral tumor mass [2, 5]. Cardiac myxomas rarely metastasize, but if they do so, their common sites are the brain, sternum, vertebrae and scapula, pelvis.

CASE REPORT

A 60-year-old woman was referred to our hospital after complaining of episodic palpitations and anxiety after stressful event in her life a month ago. No prior history of syncope, dizziness or fever. She denied chest pain and shortness of breath. No weight loss or appetite changes. No urinary or bowel problems. Past medical history was unremarkable. Family history: noncontributory.

Patient was in no acute distress. Vital signs: Blood pressure was 120/80 mmHg, heart rate was 67 beats/ min. ECG a week ago showed tachycardia (114 beats/ min). Physical examination: Neck: no jugular venous distention. Lungs were clear to auscultation bilaterally with no rales or wheezing. Cardiovascular exam was notable for a mild diastolic murmur at the apex, apical impulse was not displaced, regular rate and rhythm. No rubs or gallops. Abdomen was soft, non-tender, non-distended, no hepatosplenomegaly. Extremities: peripheral pulse 2+, symmetric in upper and lower extremities. No edema, cyanosis, or clubbing.

Transthoracic echocardiographic (TTE) study showed elliptical, mobile hyperechoic mass, 47x39x32 mm (Figures 1, 2), with smooth surface, which contained anechoic area 8 mm, inside the left atrium that prolapsed through the mitral orifice during diastole, obstructing diastolic filling of the left ventricle (Videos 1-3). The peduncle was not visualized. All chambers dimensions were normal (left atrium size was on the upper limit 3.9 cm; left atrium area 20 cm²). Minimal pericardial layers separation (3 mm during diastole) was noticed. Ejection fraction of the left ventricle was 60%. Color Doppler revealed increased velocity of blood flow across the mitral valve. Mitral valve area was 1.1 cm², using pressure half-time method. No mitral regurgitation. No signs of pulmonary hypertension. Ultrasonographic longitudinal views of the inferior chest regions showed moderate pleural effusion bilaterally. A diagnosis of left atrial myxoma was made and the patient was referred to another hospital for surgical intervention.

Figure 1: Myxoma in the left atrium. Transthoracic echocardiographic (TTE) study, parasternal long axis view. Left atrium elliptical and mobile mass (44x32 mm), containing anechogenic area (8 mm), protruding through the mitral valve orifice. Normal dimensions of the chambers.

Figure 2: TTE, apical 4 chamber view. Dimensions of the mass are 47x39 mm.
VIEWS

Video 1: TTE, parasternal long axis view. Left atrium myxoma, protruding through the mitral valve orifice, minimal pericardial effusion, normal left ventricle function.

Video 2: TTE, parasternal short axis view at aortic valve level. Mobile mass in the left atrium.

Video 3: TTE, apical four-chamber view. Left atrium myxoma, protruding through the mitral valve during diastole.


DISCUSSION

In the reported case, myxoma was located in the left atrium, which is typical situation. About 75% of myxomas occur in the left atrium (the usual site of attachment is the area of the fossa ovalis), and 15-20% occur in the right atrium. Atypical locations were reported and included: posterior or anterior left atrium wall, atrial appendage ridge, ventricles (only 3-4%), chordae of the mitral valve [6], the aortic valve [7]. Mitral valve myxomas may be localized to the anterior or posterior mitral leaflets or mitral annulus.

The patient in this report had nonspecific clinical presentation. Commonly myxoma, located in the left atrium, causes mitral valve obstruction, mimicking rheumatic heart disease (mitral stenosis). Symptoms may occur at any time, but often depend on a change in body position. Clinical manifestations can be classified into cardiac (67%), embolic (29%), and systemic (34%) symptoms [4]. Cardiac symptoms include valvular obstruction (dyspnea, orthopnea, pulmonary edema, if left sided; symptoms of right heart failure, if right sided), the direct invasion of the myocardium, which leads to decreased contractility, arrhythmias and heart blocks. Patients also can experience dizziness and episodes of syncope. Embolic events - most are left sided, and, therefore, most are systemic. Constitutional symptoms (fever, weight loss, fatigue, arthralgia, myalgia and Raynaud’s phenomenon) are present in 30% of patients. But also cases of asymptomatic tumors have been reported [8].

In developing countries certain clinical manifestations depend not only on site and size of the tumor but age of the patient. Cardiac myxomas present at a younger age in developing countries [9].

Echocardiography is a simple and noninvasive screening method of diagnosing heart masses, detecting 95% of myxomas. But 2D echocardiography is limited to planar imaging which provides representative measures for symmetric structures, but not for asymmetric masses, it relies heavily on geometrical assumptions to provide quantitative parameters of left ventricular function [8]. Real-time three-dimensional echocardiography is a technique capable to acquire a pyramidal volume of information, which can be rotated and cropped to focus on any region of interest [10].

Magnetic resonance imaging and computed tomography scans provide additional important information (tissue characteristics, precise location and thus resectability). Treatment is surgical removal. Long-term outcomes following complete resection are excellent, with a post-operative mortality rate of 0–3%.

Myxoma recurrence may be secondary to incomplete resection of the tumor, implantation from the original tumor, unrecognized multicentric origin, or the new growth of pretumor or reserve cells [2, 5]. Recurrence of cardiac myxoma has been observed in about 3% of patients in sporadic cases, and 20% in Carney complex. These recurrences may grow faster and be more infiltrative than the original tumor [2]. Serum interleukin-6 levels may be raised and can be used as a marker of recurrence [2].

CONCLUSION

Taking into consideration that myxomas may remain asymptomatic and may be occasionally found on routine examinations, symptoms may develop rather late in the course of tumor development and clinical presentation may be unclear or nonspecific, it is importantly to emphasize that transthoracic echocardiography, being a non-invasive, not expensive and practical technique, plays a crucial role in the diagnosis and assessment of cardiac tumors, as well as it is a useful diagnostic tool for a follow-up of these patients.

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Acknowledgment

I am grateful to Dr. Taras Stetsiv for technical support, and to IJCRI editors for their helpful advice and constructive suggestions that improved the manuscript.

Author Contributions

Tetyana Okan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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