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A Butterfly Floating in Heart: Rare Presentation of Biatrial Myxoma

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ABSTRACT

Myxoma, a sporadic form of cardiac malignancy, constitutes only a minor section of the presentations in cardiology. Myxoma arising from single atria are relatively common, while cases of biatrial myxoma are extremely rare. We will present an unusual case of a 43-year-old female having two independently-growing atrial myxomas floating like a butterfly. We will highlight the key steps in the management strategy of biatrial myxoma that include timely detection of symptoms, echocardiographic confirmation of myxoma, and early removal by surgical procedure.

KEYWORDS

Myxoma; Cardiac malignancy; Surgical procedure

INTRODUCTION

Myxoma is a sporadic form of cardiac malignancy with approximately 0.0017% cases seen in the general population [1]. Cardiac myomas most commonly arise from the left atrium (LA) 75%-80%; the right atrium (RA) constitutes the second important site, at 10%-20%. Cardiac myxomas can be multiple (≈5%), out of which half of these (2.5%) are biatrial [1,2]. In the literature, only a few cases of biatrial myxoma have been found to date, and they mostly define a single tumor extending to both atria in these cases [3]. We will present a very rare case of a middle-aged lady having two independently-growing atrial myxomas floating like a butterfly.

CASE REPORT

A 43-year-old female came to our hospital with four months' history of recurrent palpitations and atypical chest pain of increasing frequency and duration; three months' history of progressive exertional dyspnea; and, at presentation, had NYHA class III dyspnea. She did not have previous co-morbidity or any cardiac illness. Cardiac auscultation revealed a loud pulmonary component of the second heart sound, a mid-diastolic murmur at apical area, and another diastolic sound (tumor sound). The patient's electrocardiogram had sinus rhythm, incomplete RBBB and right axis deviation. Transthoracic echocardiogram showed two cardiac masses in right and left atria that were attached to

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interatrial septum (IAS) around fossa ovalis; these cardiac masses were highly mobile and were prolapsing through RV and LV inlet, respectively. On color Doppler imaging, moderate eccentric MR was also noted; however, mitral valve apparatus was normal, and moderate pulmonary hypertension was also present. To further delineate these cardiac masses, a transoesophageal echocardiogram (TEE) planned, which showed a typical butterfly-shaped mass attached to interatrial septum (Figure 1), highly suggestive of biatrial myxoma. On echocardiogram, it was not confirmed whether it was a single mass attached to interatrial septum and protruding across it, or two separate masses. Although on TEE, it seems to be separate masses. The decision was made to perform immediate surgery on the patient; resection of these myxomas was done through right atrium with cardiopulmonary bypass. Both atrial masses were removed successfully (Figure 2). The interatrial septum was replaced with a pericardial patch. Preoperatively, it was confirmed there were two separate cardiac masses attached to interatrial septum. Histopathology of the atrial masses revealed myxoma containing spindle-shaped cells and myxomatous material confirming the diagnosis of myxoma (Figure 3). The left-sided mass measured 6.5 cm × 5 cm × 3 cm, while the right-sided mass measured 5 cm × 3 cm × 2 cm (Figure 4).

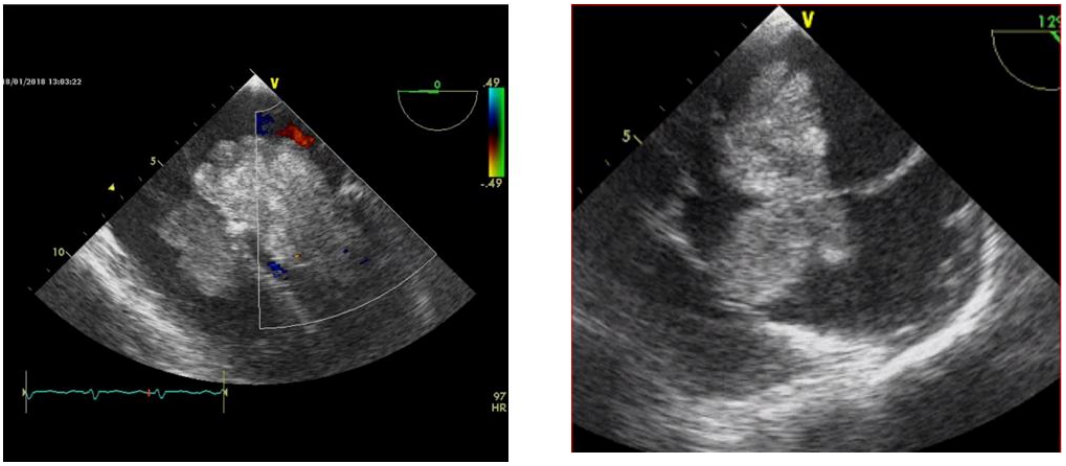

Figure 1: Transoesophageal echocardiogram showing typical butterfly-shaped biatrial myxoma.

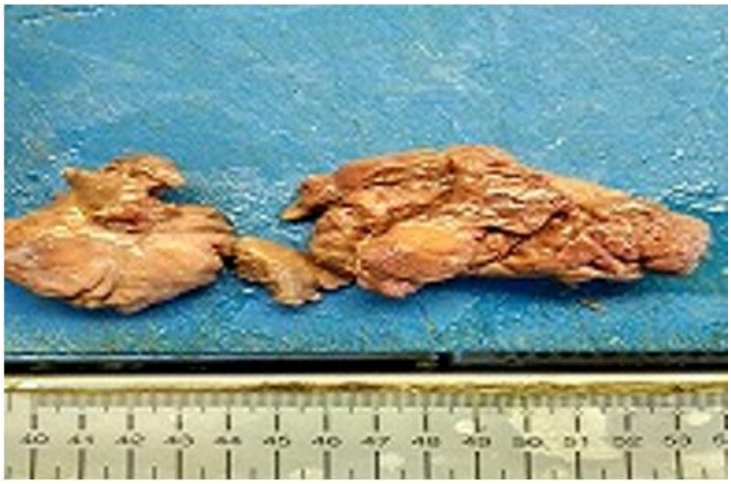

Figure 2: Gross appearance of biatrial myxoma; dark yellowish brown pedunculated gelatinous texture.

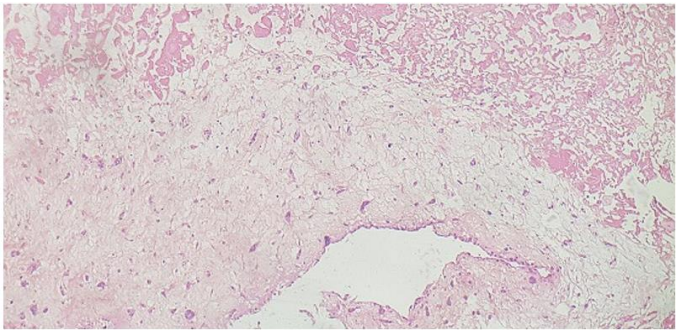

Figure 3: Histopathological appearance of myxoma containing spindle-shaped cells and myxomatous material.

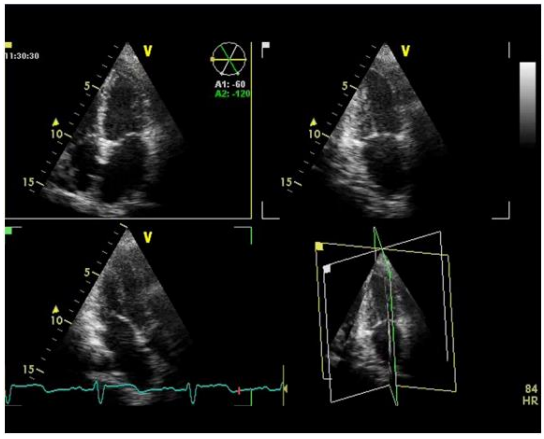

Figure 4: Post-operative echocardiogram did not show any residual tumor.

DISCUSSION

Atrial myxomas are the primary intracardiac tumor which commonly arise from the LA. Atrial myxomas arising from the right atrium (RA) are uncommon, around three to four times less than those arising from left side [4]. Around 5% of all cardiac myxomas are multiple, while less than 2.5% are biatrial in origin, a very low percentage [4].

The symptoms of atrial myxoma are relatively nonspecific, which is why early diagnosis of this condition remains a difficult task [5]. For those patients who are symptomatic, they are usually due to blood flow obstruction or distant embolization of the tumor. Symptoms in patients with myxoma can be intermittent because these symptoms will appear only when this pedunculated mass prolapse from the LA into the ventricle obstructs the forward blood flow through the atrioventricular valve [6]. Due to valvular obstruction by myxoma, a wide range of symptoms like dyspnea, chest discomfort, cardiac arrhythmias, precordial uneasiness, vertigo syncope, and acute pulmonary edema can occur in these patients. Some patients of myxoma may have a variety of presentations such as Raynaud's phenomenon, fever anemia, cachexia, and raised erythrocyte sedimentation rate, based on those facts that tumor cells release interleukin factor-6 [7].

On auscultation these patients can have similar findings to mitral stenosis due to obstruction of the valve and can be associated with the sound produced by the tumor. Echocardiogram remains the confirmatory test in patients with high sensitivity, which provides information regarding location and extension of this cardiac tumor. Chest X-rays or CT scans are relatively non-diagnostic tests; however, these tests could yield some information regarding pulmonary metastasis in cases with metastasis [8].

Atrial myxoma is a benign tumor on histopathology, but its recurrence and transformation into malignancy have been also defined in literature. The

recurrence of myxoma is around 5% at five years after surgical removal of the tumor. The recurrence rate is usually higher in younger patients, especially those with family history and in patients with myxomas having multilocular structure [9]. A few other important aspects of tumor recurrence include incomplete resection of myxoma, implantation of tumor cells during surgical excision, and re-formation in another site. To avert these factors, surgeons formulate various strategies such as detailed examination of the heart chambers at the end of the procedure and minimal operative manipulations to reduce risk of intraoperative embolization. During surgical resection of the tumor, extensive resection involving adjacent cardiac tissue and septum may sometimes be needed. The surgical excision of myxoma and small portion of surrounding IAS in our case was done through the right atrial approach. The rent in the IAS was closed with a pericardial patch. This was important to prevent myxoma recurrence.

The biatrial myxoma is an extremely rare entity, and, in most cases, it is the left sided tumor that extends through the interatrial septum into the right side. Ours is among few reported cases where both tumors have separate origins and have typical butterfly shape and prolapsing through RV and LV inlet. After establishing the diagnosis of biatrial myxoma, surgical resection is required in order to prevent these cardiac or neurologic complications. The recurrence rate is usually less than 5% after complete resection, and we have a follow-up of our patient: six months with no symptoms and no evidence of recurrence found.

CONFLICT OF INTERESTS

All authors declare no conflict of interest.

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