Azene Dessie, Sunita Maheshwari, Colin John, Sejal Shah, *Ethiop Med J, 2016, Vol. 54, No. 4*

CASE REPORT

RECURRENT MYXOMA ARISING FROM MULTIPLE CARDIAC CHAMBERS WITH SYSTEMIC EMBOLIZATION:A CASE REPORT

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ABSTRACT

Myxomas occupying multiple chambers of the heart are extremely rare, being more common in familial cases. Recurrence of these tumors after surgical excision is also a rare condition, observed in about 3% of patients in sporadic cases. Systemic embolization especially to the brain is one of the common presentations of cardiac myxomas apart from intracardiac obstruction and systemic symptoms.
We report an adolescent girl presenting with recurrent myxoma arising from multiple cardiac chambers with systemic embolization. As a diagnostic and preventive measure of recurrence and chronic systemic embolization, we recommend a regular follow up of such patients with echocardiographic evaluation following surgical resection.

Keywords: *cardiac myxoma,recurrence,systemic embolization*

INTRODUCTION

The exact incidence of cardiac tumors has been difficult to ascertain primarily due to the use of different denominators in various studies. However, echocardiography based new incidence studies suggest that one or two new primary cardiac tumors will be detected for every 1000 first-time pediatric echocardiograms. Myxomas do constitute about 50% of primary cardiac tumors in patients of all ages (1,2). Cardiac myxomas are usually solitary and develop in the atria 75% originating in the left atrium and about 25% in the right atrium. Multiple myxomas occupying multiple chambers of the heart are extremely rare, being more common in familial cases (2,3). More than 90% of the myxomas are sporadic and solitary and are more common in females (1,2-4).

The size, location and mobility of myxomas influence clinical presentation which typically includes constitutional symptoms and/or symptoms of intracardiac obstruction or embolism. Signs and symptoms are sometimes vague and unusual and this can contribute to misdiagnosis and diagnostic delay in the pediatric population (1,3).

Peripheral embolization occur in >70% of pediatric patients with myxoma, including newborns in whom embolization has been reported to have occurred in utero (2,5,6). Systemic embolization which occurs from left sided tumors can occludecerebral, coronary, pancreatic, thyroid, adrenal, renal, splenic and extremity arteries resulting in infarction of corresponding tissue (6,7).

The treatment of myxomas is surgical removal and should not be delayed even in asymptomatic patients because of the risk of embolization and cardiovascular complications including sudden death. The long term outcome following surgical excision is excellent (8-10).In this report, we describe an adolescent girl presenting with recurrent myxoma arising from multiple cardiac chambers with systemic embolization.

CASE REPORT

We report an 11 year old girl who was initially referred to the outpatient pediatric cardiology clinic of our institution in 2013. The patient presented with a one year history of headache, occasional vomiting and palpitations. Her history was negative for any family history of heart disease including tumors or other cardiovascular problems.

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Physical examination revealed heart rate of 94/min, regular, and amid-diastolicmurmur (MDM) was noted at the cardiac apex. The remaining physical examination findings were unremarkable. Laboratory tests were normal. Two-dimensional echocardiography showed a 52cm × 38cm nonhomogenous, pedunculated mass in the left atrium and a 37cm × 22cm cystic mass in the left ventricle. There was moderate mitral regurgitation. Brain MRI revealed chronic infarct related changes in the left posterior frontal lobe with no other significant neuroparenchymal abnormality. Surgical excision of both left atrial and ventricular masses was done and histopathological evaluation disclosed the tumor to be myxoma. The postoperative course was uneventful and the patient was discharged in a stable condition with follow-up arrangement.

One year after the girl had undergone the initial surgery, she presented with a one month history of nausea and headache. There was no associated history of skin color changes or weakness of extremities. The only relevant physical examination finding was an ejection systolic murmur of grade 2/6 at the lower left sternal border. Routine laboratory tests including hematologic evaluations were reported normal. Transthoracic echocardiography revealed a 39 cm × 27 cm, pedunculated and freely moving mass in the left ventricle (Figure 1).

Left ventricular mass excision was done by a redo sternotomy. Histopathology evaluation again confirmed the diagnosis of cardiac myxoma. Postoperative recovery was uneventful and the patient was discharged with follow-up appointment.

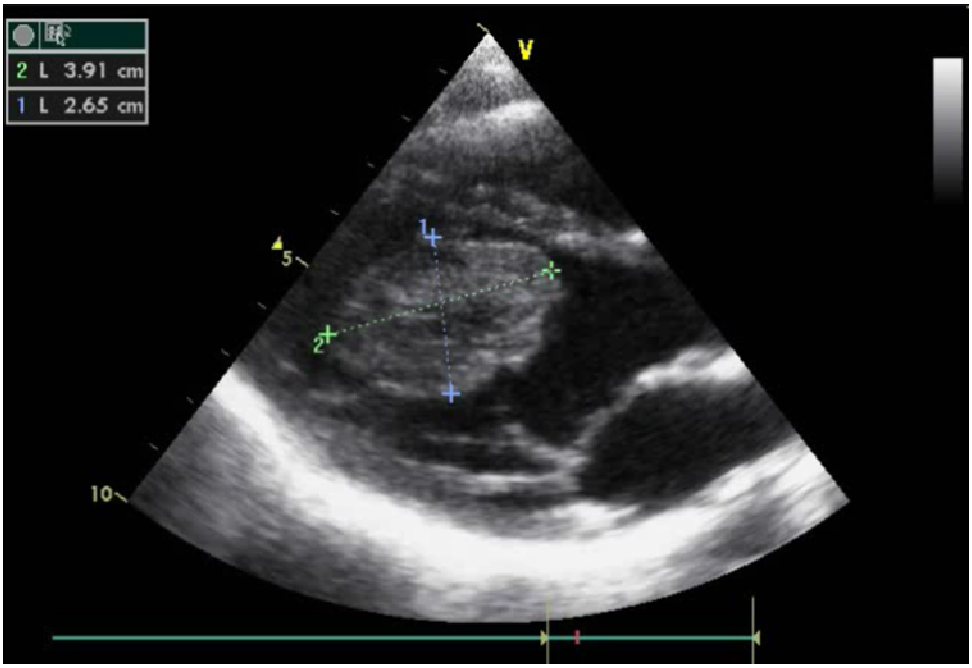

Figure 1. Parasternal long-axis view showing left ventricular mass during the recurrent episode of myxoma

DISCUSSION

Almost 90% of primary cardiac tumors excised surgically are benign, with nearly 80% of these tumors representing myxomas. Cardiac myxomas are more common in females and in patients aged 30-60. However, as in our case, they do occur in neonates and children, sometimes mimicking congenital heart disease. Although myxomas usually occur sporadically, about 7% may be familial, inherited as autosomal dominant disorder associated with skin lentigines and endocrine neoplasm, an association termed as Carney Complex (1,12). Despite the fact that the age, sex, and occurrence of tumor in multiple cardiac chambers with recurrence in our case favor a familial nature of the tumor, there is no other clinical evidence for a “syndromic myxoma”.

The clinical presentation of myxomas is often enigmatic because of the vague constitutional symptoms. The clinical features are determined by location, size and mobility of the tumor. The primary diagnostic procedure for the evaluation of cardiac tumors in pediatric patients is two-diamentional Doppler echocardiography which is expedient, noninvasive and accurate. Different studies showed that transthoracic echocardiography (TTE) has a sensitivity of around 90% in the diagnosis of left atrial myxoma. The sensitivity of transoesophageal echocardiography (TEE) is higher (13,14).

Systemic embolization is one of the commonest presentation of cardiac myxomas apart from intracardiac obstruction and systemic symptoms. Emboli are related to fragmentation of tumor substance or embolization of thrombi adherent to the tumor external surface. Studies have shown that the prevalence of embolization to the brain in atrial myxoma occurs in 20-45% of cases, and cerebral arteries are affected most (7,15-18). In our case, MRI of the brain that was performed prior to the initial surgery revealed chronic infarct-related changes in the left posterior lobe. Such chronic infarction is probably due to recurrent embolic events that commonly occur in myxomas (7,8). The occurrence of recurrent embolic events prior to intervention is one of the reasons that justify the urgent surgical resection of myxomas once the diagnosis has been made.

Recurrence of cardiac myxomas after surgical intervention is rare and is estimated at 2-3% in solitary tumors and 12-22% in familial forms. Carvalho et al. (4) suggested four different possible mechanisms to explain recurrence: inadequate resection, totipotent multicentricity, inheritance (familial type) and metastatic recurrence. However, it is difficult to confirm the exact mechanism of recurrence in individual scenarios as in our case. Studies suggest that familial and multicentric tumors have an increased risk of recurrence (12,16).

In our case, at the time of the initial diagnostic and surgical intervention the patient had both left atrial and ventricular tumors. One year after the surgical resection and follow-up visit, the recurrent myxoma was observed only in the left ventricle. It is quite difficult to explain the exact mechanism of the recurrence of the left ventricular myxoma as opposed to the left atrial tumor. There is also little evidence in the literature to support the concept of intracardiac seeding of myxoma through deposition of detached fragments carried by a high- velocity blood stream (15). Manfroi and colleagues (17) reported that the time for the recurrences varies from 3 months to 14 years after the initial resection, and 87% occur in five years. Based on this observation several investigators suggest echocardiographic follow-up to be done every six months following surgical resection, particularly in the first four years. Early detection of new recurrences facilitates early surgical management (3,11,16,17).

In conclusion, this rare case illustrates the recurrence of a myxoma that initially arose with in multiple cardiac chambers with possible recurrent systemic embolization to the brain. As a diagnostic and preventive measure of recurrence and systemic embolization of cardiac myxomas, we emphasize the importance of the periodic follow-up of such patients with echocardiographic assessment.

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