

Biatrial Myxoma and Atrial Septal Defect

Mumraiz Salik Naqshband¹, Abdul Rehman Abid², Raja Parvez Akhtar³, Muhammad Ayub⁴ and Jawad Sajid Khan³

ABSTRACT

Myxomas are rarely associated with congenital cardiac anomalies. We report a case of a young female presenting with symptoms of atrial myxoma but having co-existing ASD visualized on 64 slice multidetector computed tomography (MDCT) prior to cardiac surgery. Patient had a successful resection of biatrial myxoma and ASD repair. This case is unique as the myxoma originated from the inferior margin of the ASD straddling the inferior limbus. Over the years due to the left to right shunt at the ASD, the myxoma was initially prolapsing and oscillating between the two atria. As it grew larger it obliterated the ASD and got stuck in the right atrium (RA) and continued to grow giving false impression of a right atrial mass. Multi detector computed tomography is an alternative diagnostic modality to the gold standard transesophageal echocardiograph for detection of an ASD in the presence of atrial myxoma. Biatrial myxoma with associated atrial septal defect has optimal postoperative results.

Key words: *Biatrial myxoma. Atrial septal defect. Multi detector computed tomography.*

INTRODUCTION

Cardiac myxomas are benign intracavitary neoplasms and represent 5-10% of all neoplasms of the heart with an incidence of 0.002-0.3% in various autopsy studies.¹ Cardiac myxomas comprises 30-50% of the benign tumours in most series and over 70% in surgical series.¹ Left atrium is the most common site of origin of myxomas 75-80%, followed by right atrium 18% and biatrial in < 2.5%.¹ Since 1998, few cases of biatrial myxoma have been described in the literature.^{1,2} Furthermore, myxomas are rarely associated with congenital cardiac anomalies such as atrial septal defect.³ Because atrial myxomas frequently originate from the interatrial septum so atrial septal defects (ASD) are least expected to co-exist with myxomas.

We report a unique case of a young female presenting with symptoms of atrial myxoma but having co-existing ASD visualized on 64 slice multi detector computed tomography (MDCT) prior to cardiac surgery.

CASE REPORT

A 22-year-old unmarried female presented to the emergency department with a history of fever off and on for one year and vertigo and syncopal episodes for last 6 months. Fever was gradual in onset and intermittent for last one year for which she had taken wide range of antibiotics including anti-tuberculous treatment with no response. She also had vertigo and syncopal episodes

for 6 months which were attributed initially to long standing fever and chronic debilitated state. The frequency of syncopal episodes and palpitations had increased since one week so she was referred to our hospital.

At the time of presentation, she had pulse rate of 102/min, blood pressure of 100/70 mmHg and temperature of 100⁰ F. Apart from sinus tachycardia, her ECG was within normal limits. X-ray chest revealed normal cardiac shadow. On transthoracic echo-cardiography (TTE), a large mass almost filling RA and partially obstructing tricuspid valve flow was identified. Since the attachment of mass to interatrial septum could not be accurately visualized on TTE a transesophageal echo (TEE) was planned. After preparation TEE was started but patient had severe recurrent episodes of syncope so the test was abandoned. Patient was then taken for MDCT performed on 64 slice GE system. MDCT revealed biatrial myxoma with an atrial septal defect (Figure 1 a,b).

Once the diagnosis was established, the patient underwent an urgent operation. A median sternotomy was performed and the patient was placed on bypass after heparinization using double venous and aortic cannulation. Patient was cooled to 32⁰C, cross clamp applied blood cardioplegia delivered to the ascending aorta. The right atrium (RA) was opened obliquely and a huge encapsulated mass was seen to be occupying whole of the RA and protruding into the RV. The ASD was identified digitally and the mass was delivered and excised. It was originating from the inferior limbus of the ASD, straddling and extending onto the right and left side of the atrial septal defect margin. This part of the septum and margin was excised along with the myxoma. The ASD was closed using autologous pericardial patch with and 4-0 prolene (2-0 Ethicon Prolene Ethicon INC,

*Department of CME¹/Cardiology²/Cardiac Surgery³/
Cardiovascular Imaging⁴, Punjab Institute of Cardiology,
Lahore, Pakistan.*

Correspondence: *Dr. Raja Parvez Akhtar, 107 B, Street No. 12,
Cavalry Ground Ext., Lahore Cantt, Lahore.*

E-mail: rajapakhtar@gmail.com

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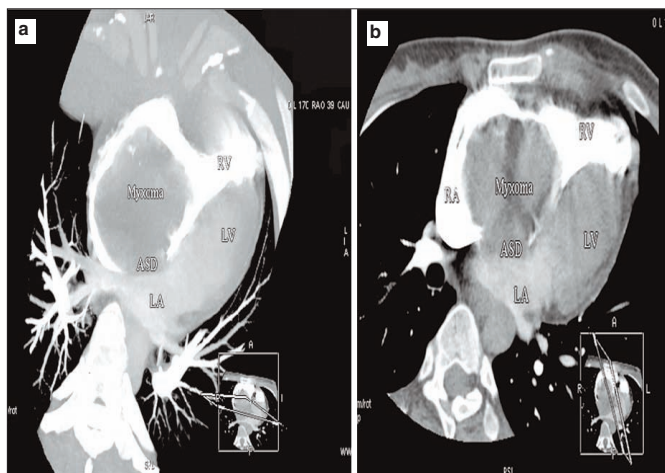


Figure 1(a,b): MDCT images showing mass in RA and atrial septal defect.

USA). Patient was rewarmed, RA was closed, heart was de-aired and cross clamp was removed. Patient regained sinus rhythm and got weaned off from cardiopulmonary bypass without any problem. The specimen was sent for histopathology, which showed it to be a cardiac myxoma.

Her postoperative course was uneventful and she was discharged from the hospital on the 5th postoperative day. Prior to discharge a whole body scan was done to look for tumours on any other site but there was no evidence of any extracardiac tumour. On 3 months postoperative follow-up, she was asymptomatic.

DISCUSSION

Cardiac myxomas are intracavitary, round or ovoid neoplasm attached to the endocardium.⁴ A vast majority of these are attached at the fossa ovalis of the interatrial septum, they can arise from any endocardial surface.⁴ Biatrial myxomas are extremely rare. Imperio *et al.* in 1980 described only 3 cases of successful removal of biatrial myxomas in the literature.⁵ Since then very few cases of biatrial myxoma have been described. Association of congenital cardiac anomaly like atrial septal defect is also rarely reported in the literature.³

This patient was a female and most series of myxomas have shown a female preponderance.⁶ However, the degree of female preponderance is unclear, varying from 3-1 ratio in some series to a 1-1 ratio in others.¹

In this patient the diagnosis was not clearly established on transthoracic echocardiographic examination and ASD was missed. The diagnosis of ASD was confirmed on 64 slice MDCT prior to the operation. Transthoracic and transesophageal echocardiography are currently the primary imaging modalities used. TTE is used to determine size, mobility, shape and location of the tumour.⁷ TEE is considered more effective at detecting atrial tumours and may be particularly effective at identifying the site of tumour attachment and characteristics of the mass.⁷

Kosar *et al.* reported a case of left atrial (LA) myxoma associated with an atrial septal defect. Their patient was a 72-year-old woman presenting with loss of consciousness and prior history of dyspnea on exertion and episodes of palpitations. Her CT brain revealed an ischemic necrotic zone. TEE revealed pedunculated and lobulated mobile non-homogenous mass and an ostium secundum ASD. This patient luckily did not have an embolic event although she had frequent episodes of syncope. Due to unstable condition of the patient, TEE could not be performed and instead MDCT scan was performed. In another report of LA myxoma complicated by ASD, Takashi *et al.* reported a 55 years old man with a myxoma that originated in the left atrium and grew through a secundum ASD into the right atrium.⁸ The tumour was attached by a pedicle to the lateral wall of the left atrium near the right pulmonary vein, was resected under cardiopulmonary bypass. TEE was important in the successful outcome of surgical treatment. In this patient, the myxoma was originating from the inferior margin of the ASD and was straddling the inferior limbus. Over the years, due to the left to right shunt at the ASD the myxoma initially was prolapsing and oscillating between the left and right atrium. As it reached a critical size i.e. larger than the ASD, it got stuck in the RA and continued to grow into RA towards the tricuspid valve and right ventricle.

Yoon *et al.* reported a case of multicentric biatrial cardiac myxoma in a 29-year-old female.⁹ By using TEE, a biatrial myxoma attached to the interatrial septum was identified. Pre-operatively it was observed that there was a large right atrial myxoma extending through the fossa ovalis into the left atrial free wall. An ASD was created by resection of interatrial septum and free wall which was safely repaired by bovine pericardial patch.

Tsukamoto *et al.* reported a case of LA myxoma complicated by a secundum ASD diagnosed intra-operatively.¹⁰ The tumour was attached by a pedicle to the posterior wall of the left atrium near the posterior commissure of the mitral valve and was removed together with a small portion of the left atrial wall. Since the defect was functionally cancelled by the myxoma, the diagnosis of an atrial septal defect was not confirmed pre-operatively even by color Doppler echocardiography and cardiac catheterization. Similarly, in this patient the ASD was functionally cancelled due to the myxoma which was obstructing it. Hence, it was missed on echo-cardiography.

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