Dumbell Shaped Giant Left Atrial Myxoma Extending into Left Ventricle Across Mitral Valve in an Elderly Male Presenting with Cardiac Failure

GT Sudarshan, KN Nagle, N Makarand, Aironi Balaji, Kamlesh Jain, V Narendra

Abstract
Although rare, atrial myxomas constitute the most common benign cardiac tumour. Left atrial myxoma presents a fascinating problem because of its rarity, protean manifestations and curability. Symptoms may be related to the haemodynamic effects of obstruction, to embolic phenomena or to systemic manifestations which may mimic collagen disease. They have an excellent prognosis following surgical excision. We report a 65 year old male who presented with features of both right and left heart failure. Ausculation of the heart revealed mid-diastolic murmur. Two-dimensional colour flow Doppler echocardiography revealed a large pedunculated left atrial myxoma that extended into left ventricle across the mitral valve causing obstruction to flow across mitral valve. The patient underwent a successful surgical excision of the tumour.

Introduction
Atrial myxomas are the most common benign primary tumour of the heart and occur in as many as 3 in 1000 patients. These tumours are a major cause of patient morbidity and mortality. Clinical manifestation varies from no symptoms and very poor or no clinical signs to various manifestations of chronic or acute congestive heart failure, syncope and arrhythmias with or without systemic findings such as high erythrocyte sedimentation rate, anaemia, leucocytosis, elevated gamma globulin, thrombocytopenia or low grade fever, as well as cerebrovascular accidents due to tumour embolization, hence the suspicion of the diagnosis on the basis of clinical symptom is very rarely achieved.

Operation for left atrial myxoma can be undertaken solely on the basis of echocardiographic findings, but coronary angiography should be performed in older patients who are at risk for coronary artery disease. Surgical excision of left atrial myxomas must be performed as soon as possible after diagnosis is established because of the high risk of valvular obstruction or systemic embolization. Thus, surgical intervention can be curative for patients with left atrial myxomas and most of these can expect an excellent outcome. Here we present an unusually large atrial myxoma presenting in an elderly male with signs and symptoms of congestive cardiac failure.

Case History
The patient was a 65 year old male who presented with dyspnoea on exertion with both lower limb swelling associated with dry cough, palpitations and orthopnoea since 2 months. He had these complaints on and off for the past 4 years. On examination there was pitting type of pedal oedema with facial puffiness and pallor along with mild icterus. The JVP was raised and he had tachypnoea with usage of accessory

Department of Cardiovascular and Thoracic Surgery, BYL Nair Hospital and TN Medical College, Mumbai Central, Mumbai 400 008.
muscles of respiration. Vitals were normal. Cardiac examination revealed a Grade II/III pansystolic murmur at the apex and a plopping sound heard at the same area. Bilateral basal crepitations were present. There was a moderate liver enlargement. The electrocardiogram was within normal limits. Chest radiogram showed an enlarged heart with basal hilar pulmonary congestion. Two dimensional echocardiogram revealed a large left atrial homogenous mass measuring 8 by 4 cm occupying the whole of the atrium and abutting the mitral valve causing a mild to moderate mitral regurgitation. The peak to mean gradient across the mitral valve was 11/6 mm Hg. Other valves were normal. There was moderate right ventricular systolic dysfunction with the ejection fraction of 55%. The pulmonary pressure was high (PASP by TR jet of 70 mm Hg). Patient was treated for the cardiac failure and later subjected to coronary angiography which was found to be normal.

Patient was operated with midline sternotomy and was put on cardiopulmonary bypass. Right atrium was opened. The base of the tumour peduncle on the inter-atrial septum was identified and a rim of the septum around it was cut to enter the left atrium. A large gelatinous dumbbell shaped mass measuring around 10 x 6 cm was found occupying the entire left atrial cavity and extending into left ventricle across mitral valve. The tumour was delivered intact from left atrium and left ventricle. The defect in the inter-atrial septum was closed with Dacron patch and the right atrium was closed.

The post operative period was uneventful and the patient was discharged on the 7th post operative day. Follow up 2D Echo was found to be normal and the patient is doing well at 2 months follow up.

Discussion

Primary cardiac tumours are rare. They have been found in 0.05% of autopsied material. The antemortem diagnosis of primary cardiac tumours is rarer still but with the advent of angiocardiography and the
adequate techniques of their surgical removal, the consideration of this diagnosis has become critical. The most common primary tumour is the myxoma which accounts for 50% of all primary tumours.

Cardiac myxoma have been reported in the age group of newborn to 70 years, but most frequently manifests in the third to sixth decade and are often reported to be more common in females. Approximately 50% of patients with myxomas may experience symptoms due to central or peripheral embolism or intracardiac obstruction, but 10% of patients may be completely asymptomatic. Symptoms of mitral valve obstruction, the first arm of the classic triad of myxoma presentation, is present in 75 patients (67%), with mostly cardiac failure or malaise. Symptoms of embolism, the second frequent presentation in the classic triad, were observed in 33 cases (29%) with 1 or several locations, essentially cerebral emboli with stroke. Males are statistically at greater risk than females of developing embolic complications. The third arm of the classic triad consists of constitutional symptoms (34%) with fever, weight loss, or symptoms resembling connective tissue disease, due to cytokine (interleukin-6) secretion. Younger and male patients have more neurologic symptoms, and female patients have more systemic symptoms. Screening for myxomas should involve a thorough history and physical examination and a transthoracic and/or transoesophageal echocardiogram. Transthoracic echocardiography is approximately 95% sensitive for the detection of cardiac myxomas, and transoesophageal echocardiography approaches 100% sensitivity.

A high index of suspicion seems important for early diagnosis. Immunologic findings may play an additional role in confirming the diagnosis and the recurrence of a myxoma. Immediate surgical treatment is indicated because of the high risk of embolization or of sudden cardiac death. Biatrial approach allows for the inspection of the four cardiac chambers, limits manipulation of the mass, and facilitates the complete excision of the tumour. Complete surgical resection of primary cardiac tumours prolongs symptom-free survival and is associated with a low recurrence rate. Since late recurrence, although rare, has been reported, especially in familial myxomas, long-term clinical and echocardiographic follow-up is recommended.

References