A case of left atrial myxoma presenting with multiple embolic manifestations

JEGADEESWARI ARUMUGAM
Department of Cardiology,
MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract: Myxomas are the most common cardiac primary tumours. They account for 30 to 50 of all benign cardiac tumours. Seventy five percent of myxomas occur in the left atrium. The clinical presentations of cardiac myxoma varies from asymptomatic to symptoms of systemic manifestations, primary cardiac symptoms and those due to embolization. Tumor embolism is not an uncommon mode of clinical presentation and can affect the systemic or pulmonary circulation, depending upon the location of the tumor and the patency of the foramen ovale. Cardiac myxoma is the most common primary cardiac tumor to produce tumor emboli. It has been reported to embolize to any organ or tissue. Large saddle emboli in the abdominal aorta have been reported, causing acute lower limb ischemia. We are presenting a case of left atrial myxoma, presenting with aortoiliac and coronary arterial embolic manifestation. Our patient is a 42 year old female, presenting with one episode of syncope, followed by pain, numbness and weakness of right lower limb. Clinical examination showed weak lower limb pulses and a mid diastolic murmur in the mitral area. Electrocardiogram revealed symmetrical, deep T wave inversion in the precordial leads. Echocardiography showed left atrial myxoma and regional wall motion abnormality involving the left anterior descending coronary artery territory, with mild left ventricular dysfunction. Peripheral angiogram showed tumour embolus in the aortoiliac bifurcation. She underwent successful catheter embolectomy, followed by surgical removal of the left atrial tumour.

Keyword: left atrial myxoma, embolisation

INTRODUCTION:
Left atrial myxomas are an uncommon cause of acute lower limb ischemia and myocardial ischemia. These benign intracardiac tumors can rarely give rise to emboli large enough to cause macrovascular occlusion. Peripheral embolization results from tumor fragmentation, or, rarely, from complete tumor detachment, causing syncope, chest pain, dyspnea, neurologic symptoms, or ischemic limb pain. We present a rare case of left atrial myxoma, presenting with multiple tumour fragment embolization, causing acute lower limb ischemia and myocardial ischemia.
CASE REPORT:
42 year old female had presented to her primary care physician with complaints of one episode of sudden and transient loss of consciousness, a brief episode of retrosternal chest pain and pain, numbness and weakness of her right leg and foot, five days earlier. She was referred to our hospital for further management. She had been previously healthy. On admission to our hospital, her pulse rate was 80 beats/min and her blood pressure was 110/70 mm of Hg. Her right femoral and popliteal pulses were weak; right dorsalis pedis, left femoral, popliteal and dorsalis pedis pulses were absent. Tips of the right toes were discolored and cold. Cardiac auscultation revealed a mid diastolic murmur in the mitral area, which varied in intensity with the position of the patient. The erythrocyte sedimentation rate was 95mm/hour. Twelve lead surface electrocardiogram revealed deep, symmetrical T wave inversion in the precordial leads (Fig - 1). Chest x-ray was normal. Transthoracic echocardiogram showed a large mobile mass in the left atrium, attached to the interatrial septum and prolapsing into the left ventricle during diastole, consistent with a diagnosis of left atrial myxoma (Fig 2, 3). There was regional wall motion abnormality involving the left anterior descending coronary artery territory and the left ventricular ejection fraction was 40%. Doppler study of the lower limb arterial system showed loss of triphasic flow in bilateral femoral and right popliteal arteries and absent flow in the left popliteal, bilateral dorsalis pedis and posterior tibial arteries. Coronary angiogram showed normal epicardial coronary arteries, with slow flow in the left anterior descending artery. Peripheral angiogram (Fig 4) showed partial occlusion with saddle embolus in the aortic bifurcation and partial embolic occlusion of bilateral common iliac and external iliac arteries, with reduced flow in the bilateral femoral, popliteal and the pedal arteries.

Retrograde trans femoral tumour embolectomy was done using Fogarty catheters, which were passed through both common femoral arteries. Large numbers of embolic fragments were extracted from both the femoral arteriotomy sites. Embolic fragments were gelatinous and were mixed with thrombus. Microscopic examination of the embolectomy specimen showed myxoid stroma, with many stellate stromal cells, consistent with myxoma (Fig 5,6). Both the dorsalis pedis arterial pulsations and vascular doppler flow signals were restored immediately after arteriotomy closure. Sternotomy was then performed and the cardiac myxoma was removed from the left atrium. The tumour was attached to the fossa ovalis by a stalk and was removed by right atriotomy and pericardial patch closure of the atrial septal defect was done. Macroscopically, the tumour measured 30 x 42 mm, and had a nodular, glistening surface and interspersed areas of hemorrhage (Fig 7). Histopathological examination confirmed the diagnosis of myxoma. There were no post operative complications and the patient was discharged after ten days. At 3 months after surgery, follow up examination showed normal doppler flow in both the lower limb arteries. On transthoracic echocardiographic examination, there was no tumour recurrence or regional wall motion abnormality and the left ventricular ejection fraction had improved to 55%.

DISCUSSION:
Primary tumors of the heart are rare, with an incidence of 0.0013 and 0.03% in collected autopsy series \(^1\), \(^2\), \(^3\). In adults 50% of benign cardiac tumors are myxomas \(^4\). 75% of primary
cardiac tumors are benign. While most cases are located in the left atrium, these tumors are also found in the right atrium, right ventricle and left ventricle. Cardiac myxomas usually originate from the region of the fossa ovalis, but may arise from a variety of locations within the atria. Although histopathologically benign, cardiac myxomas can cause chronic systemic inflammation, embolism, or intracardiac obstructions, leading to increased morbidity. In a retrospective study of 32 patients by Muthubaskaran et al, obstructive symptoms was the commonest mode of presentation followed by embolic manifestation. In the embolic group, five patients (41.6%) had peripheral artery embolism and six (50%) had a cerebrovascular event and one presented with pulmonary embolism (8.33%). Cardiac myxoma is the most common primary cardiac tumor to produce tumor emboli, and it has been reported to embolize to virtually any organ or tissue. Embolic complications occur in 30-40% of patients with myxomas. As most myxomas are located in the left atrium, systemic embolism is particularly frequent especially to cerebral arteries and peripheries. Cases of cardiac myxomas embolizing to coronary arteries, kidney, liver, spleen, eye, and skin have also been reported. Coronary artery embolism associated with myxoma is very rare and has been documented by both angiography in living patients and histology at post mortem study. Myocardial infarction is sometimes the first manifestation of a myxoma. Total detachment of cardiac myxoma causing saddle embolization of the abdominal aorta has also been rarely reported. Acute lower extremity ischemia due to the complete embolization of a left atrial myxoma have also been reported. Echocardiography remains the diagnostic test of choice. In addition, coronary and peripheral angiogram are useful in providing additional information regarding the embolic complications. Surgical excision of the tumour under cardiopulmonary bypass is the typical treatment for cardiac myxoma.

Recurrence of the tumor has been reported in up to 17% of cases and can occur at the original site of implantation, at other foci, or as a result of direct embolization. Malignant transformation has been described. This patient was a previously healthy middle aged female, with no systemic manifestations or obstructive symptoms who presented with features of emboli to both the coronary artery and the lower limb simultaneously. This serves as a reminder that cardiac embolic source should be contemplated in a relatively young patient presenting with acute lower limb ischemia. The diagnosis in this case was established by echocardiography and confirmed by histopathological examination. Our patient’s presentation with a single acute event prompted immediate treatment and resulted in complete functional recovery with minimal complications.

Fig 1: ECG showing anterior and inferior wall ischemia  
Fig 2: PLAX view of LA myxoma
An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Sciences

Fig 3: A4C view of LA myxoma

Fig 4: Peripheral angiogram showing saddle embolus and partial occlusion of bilateral common iliac arteries

Fig 5: Microscopic appearance of the tumour specimen

Fig 6: Myxoid stroma of the tumour

Fig 7: Macroscopic appearance of the tumour
Reference:
4. Prichard RW. Tumors of the heart: review of the subject and report of 150 cases. AMA Arch Pathol. 1951;51:98–128.