LEFT ATRIAL MYXOMA PRESENTING AS ACUTE LIMB ISCHEMIA OF 3 LIMBS WITH SADDLE EMBOLUS OF ABDOMINAL AORTA BIFURCATION AND INTRA CRANIAL BLEED – A RARE PHENOMENONE

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Abstract

Left atrial myxoma is the most common variant of cardiac tumour, rarely presenting with systemic embolization involving 3 limbs and intra cranial bleed simultaneously. Our case aims to discuss appropriate management strategy in such cases. This case presents a 34 year old lady with acute limb ischemia both lower limb and right upper limb with intracranial bleed with no signs of congestive heart failure diagnosed with LA Myxoma on preoperative routine 2d ECHO cardiography. She underwent embolectomy of right brachial artery and bilateral femoral artery, common iliac artery and abdominal aorta with excision of LA Myxoma planned for 4 weeks later. In patients presenting with acute limb ischemia and intracranial bleed, the definitive surgery of LA myxoma excision should be postponed for 4 weeks to prevent risk of systemic heparinisation causing worsening of neurological functions while acute limb ischemia should be addressed immediately.

INTRODUCTION

Cardiac tumours are rare disorders of heart, amongst which atrial myxoma are the most common variants. (1) median age at diagnosis is 50 years , and it usually presents at 3^{rd} to 6^{th} decade of life, with female to male ratio 2:1. (2) Clinical features are usually dependent on location of tumour, propensity of systemic embolization, or non-specific constitutional symptoms; usually independent of its pathological subtype. Patients can have cardiac manifestation of Congestive Heart failure including dyspnoea on exertion, orthopnoea, peripheral edema, pulmonary edema.(3) It can also present with valvular insufficiency , valvular stenosis or arrhythmia. Systemic embolization is other common variant of cardiac myxoma presentation. The emboli can consist of fragmented tumour and/or thrombus formed on the surface of the mass. Left sided emboli can present with systemic embolization and right side emboli can present with pulmonary embolization.(4) Constitutional symptoms like fever, anorexia, malaise are due to excessive production of IL-6 .(5) Treatment is complete excision of the myxoma with cuff of normal endocardium. The surgery requires Cardiopulmonary bypass, requiring full systemic heparinisation increasing the risk of aggravation of intracranial bleed.

CASE REPORT

This is a case of year old female presenting to PGIMER emergency with complaints of acute severe limb pain in both lower limbs and right upper limb for past 6 hours. Patient was evaluated clinically and acute limb ischemia was suspected for which patient underwent emergency CT Angiography of abdominal aorta till bilateral lower limbs and bilateral upper limbs. CT Angiography revealed hypo dense filling defect in the infra renal abdominal aorta extending till bilateral common iliac arteries and extending till right brachial artery till its bifurcation (Figure 1 Figure 2)

Patient was started on heparinisation as per Acute Limb Ischemia protocol with serial aPTT monitoring and aPTT was kept in the range of 45-70. Following initial stabilisation, patient underwent 2D-ECHO, which revealed 41* mm mass in Left Atrium attached to intra-artrial septum, prolapsing in LV cavity causing obstruction to flow of mitral valve resulting in Moderate Mitral regurgitation.(Figure 3 Figure 4)

Patient was planned for Emergency LA Myxoma excision under Cardio pulmonary bypass (CPB) with embolectomy of Right Brachial Artery and abdominal aorta, bilateral Common Iliac artery and bilateral femoral artery (superficial and deep branches). Although patient did not complain of headache, vomiting, loss of consciousness or ENT bleed, Preoperative NCCT head was done for risk analysis of heparinisation of acute limb ischemia and subsequent heparinisation required during CPB, which revealed acute bleed in right side fronto-temporal region. Considering the risks of systemic heparinisation during CPB with increase in intra cranial bleed causing life threatening complications, the plan was changed. Patient was then taken up for surgery and fogartisation was done via bilateral femoral approach and a large myxomatous tissue was removed. Fogartisation was also done from right brachial artery and similar myxomatous tissue was extracted (Figure 5 Figure 6)Post-operative patient had pulses in both lower limb Popliteal artery, femoral artery, ATA,PTA,DPA and in Right brachial artery and right radial and ulnar artery. Patient was kept on low dose of heparin infusion for 24 hours post-surgery. She has been planned for definitive LA Myxoma excision 4 weeks later.

DISCUSSION

Cardiac tumours are amongst the rare diseases of heart with incidence of 0.02 %. Metastatic tumours are more common to heart as compared to primary tumours. Median age of presentation is 50 years (1). Atrial myxoma constitutes about more than 50% of all benign cardiac neoplasms. 75% of all Atrial Myxoma arises from left atrium, with majority originating at the edge of fossa ovalis, while 15-20% arises from right atrium; commonly at the edge of fossa ovalis. Bilateral myxoma represents extension from one atrium to another via foramen ovale. 3-4% of myxoma arises from ventricle with biventricular extension rarely seen. Vast majority are sporadic with male: female ratio 1:3. 5-10% are familial with slight male preponderance and are multicentric and more likely to recur.(2)

Clinical manifestations are due to location of the myxoma, propensity of embolization and nonspecific constitutional symptoms elaboration of cytokines by the tumour. Patient may present with symptoms of left sided or right sided cardiac failure, depending upon site of mass, point of obstruction caused by the mass. Valvular masses can cause insufficiency or relative stenosis of the mass. Left sided mass can embolise to systemic circulation causing stroke, acute limb ischemia.(3) Echocardiography is initial modality of choice, with advantage of temporal as well as spatial resolution the evaluating the motion of pedunculated tumour in cardiac chamber and its effect on the normal contraction , valvular function and blood flow during cardiac cycle(6). ECG gated CT and MRI can provide additional information in terms of tumour diagnosis, extent and involvement. Treatment is surgical excision of myxoma with cuff of normal endocardium to decrease chances of recurrence and it is usually curative. Resulting defect is closed primarily if possible or with autologous or bovine pericardium. Prognosis is excellent with expected survival equivalent to general population. Overall rate of embolization with cardiac tumour is around 25 %, with tumours in let atrium and aortic valve with maximum embolic potential (4) this is the reason the cases of LA Myxoma should be operated as soon as possible.

Resection of LA myxoma requires CPB support. Although there are no set guidelines for commencing cardiopulmonary bypass in cases of Myxoma, The society of Thoracic Surgeons Clinical Practice Guidelines published in 2011; recommend delaying surgery for Infective Endocarditis complicated by intra cranial bleed by up to 4 weeks from onset of cerebrovascular complications, due to the risk of extension of haemorrhage potentially causing lifelong morbidity or life threatening consequences from existing site of intracranial bleed.(7) Considering the risks of the extension of intracranial bleed, the surgery was delayed by 4 weeks in our patient.

CONCLUSIONS

Although, there are no set guidelines, the risk of postoperative neurological complications arising from CPB in patient with pre-existing atrial myxoma outweighs the propensity of risks of embolization and thus, although the emergent nature of requirement of surgery in cases of atrial myxoma, the surgery can be delayed by 4 weeks, while symptomatic treatment of Acute limb Ischemia can be done during those periods urgently. However, further evaluation is needed to establish the safe duration of surgery in cases of atrial myxoma complicated by the intracranial bleed.

CONFLICT OF INTEREST

The author declares no conflict of interest.

FUNDING

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ETHICAL CLEARANCE

Informed consent was received from the patient and was cleared by the departmental ethical committee.

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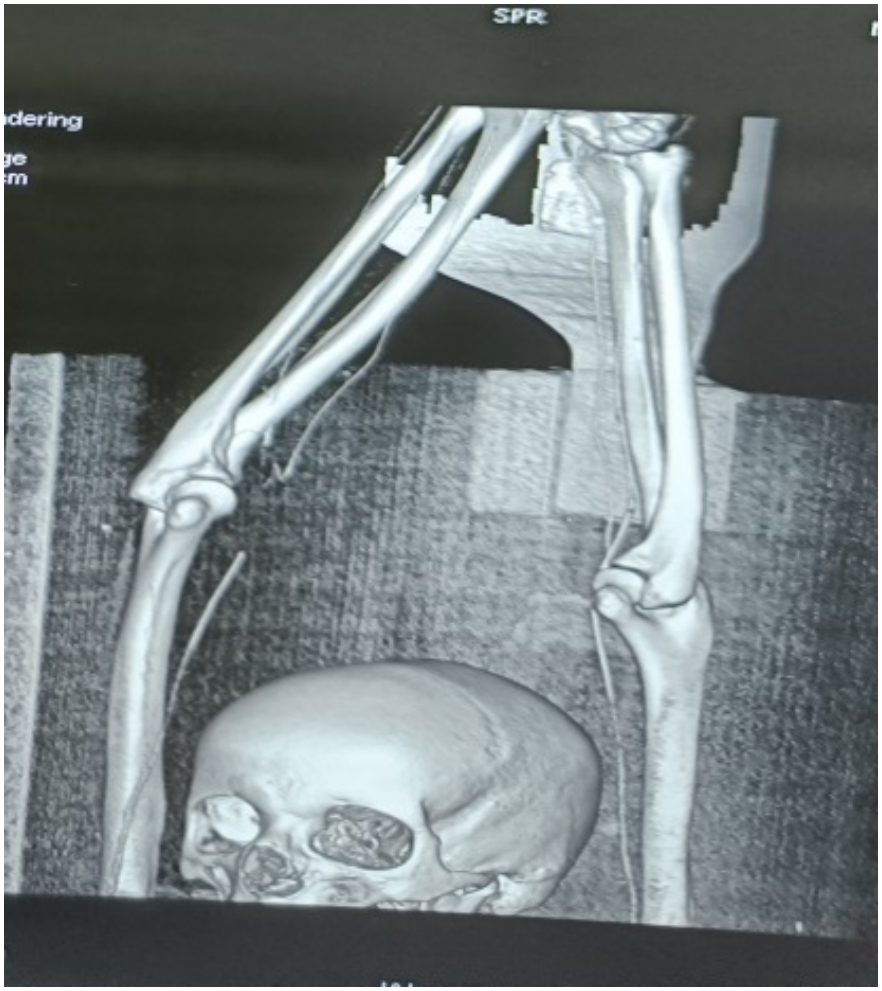

Figure 1 right brachial artery showing filling defects with distal opacification of radial and ulnar artery

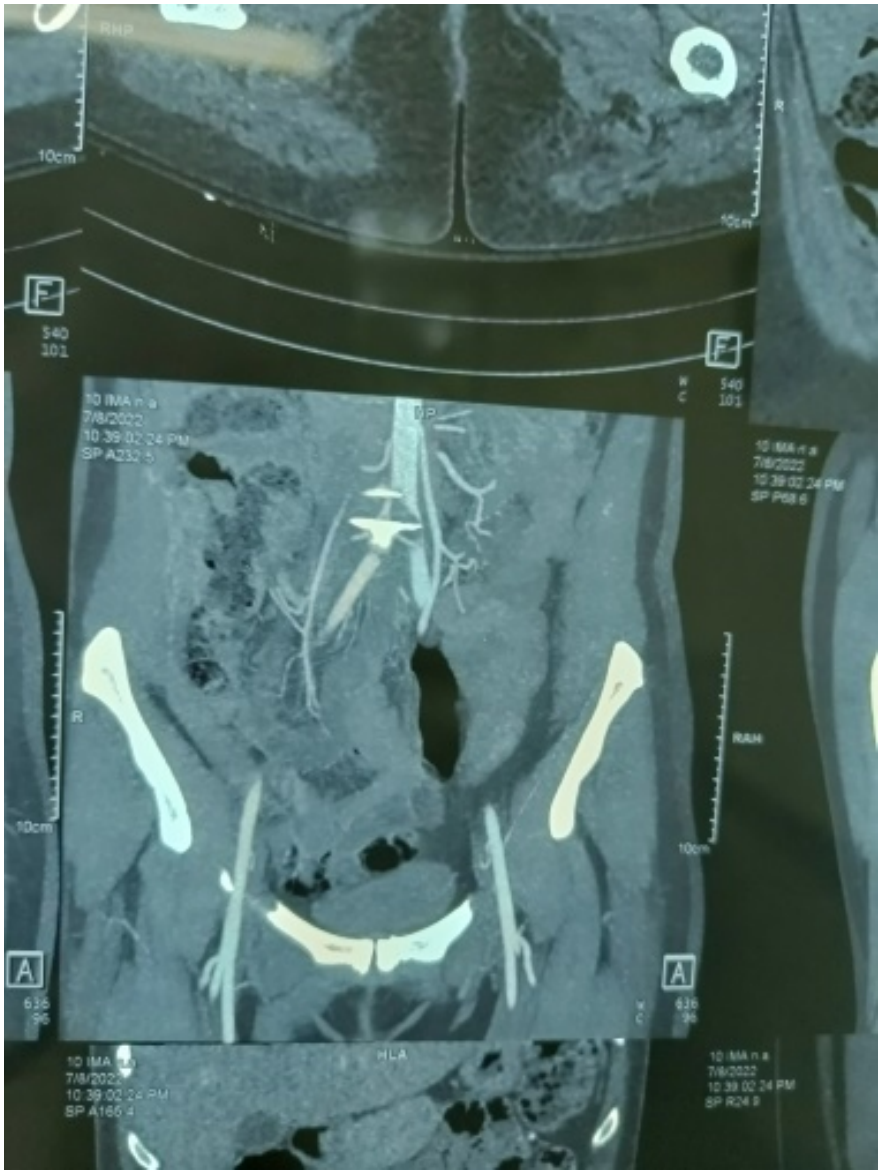

Figure 2 filling defect in bilateral common femoral artery and infra renal abdominal aorta

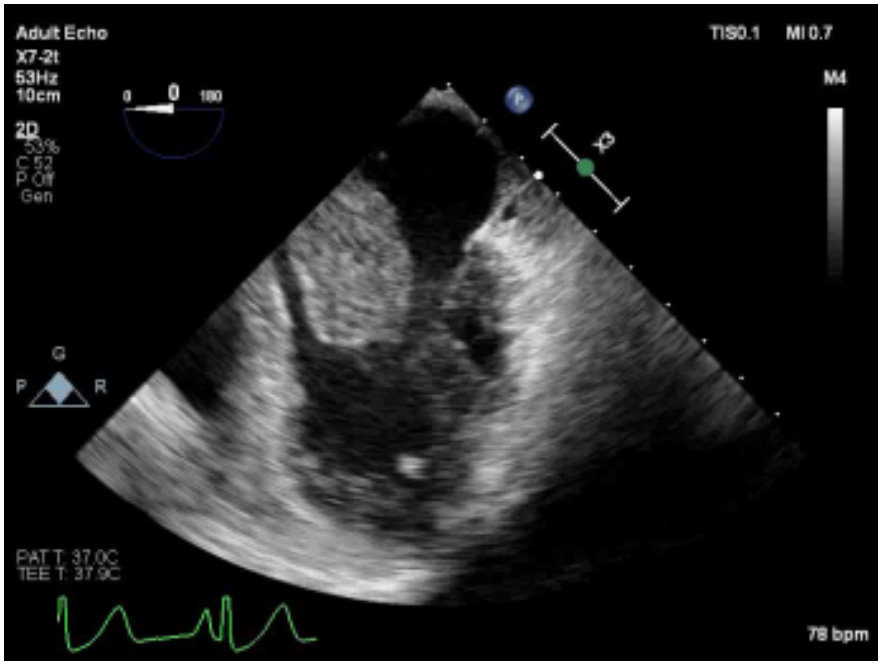

Figure 3 echo image showing large mass in LA cavity attached to intraartrial septum and Mitral valve prolapsing in LV

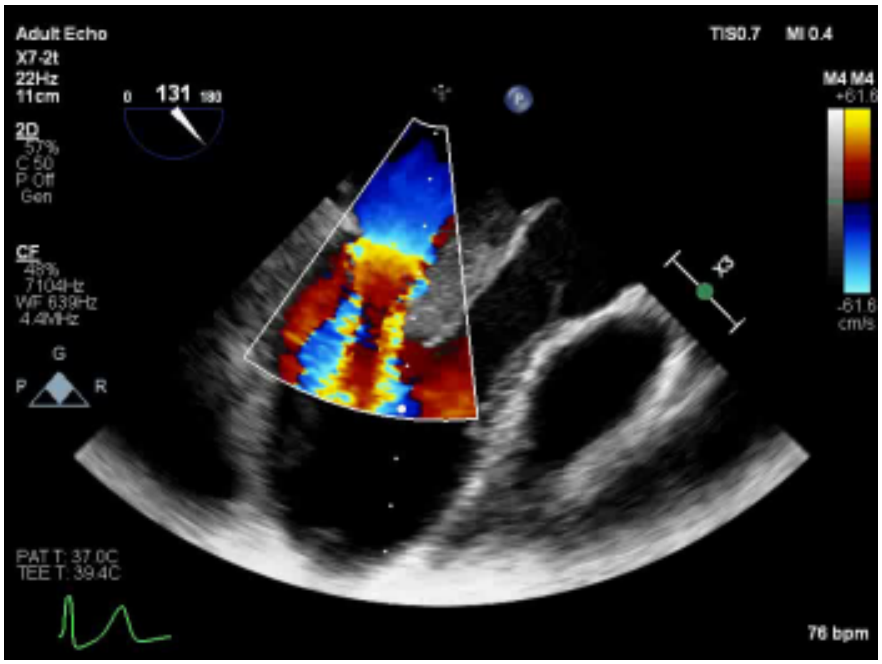

Figure 4 LA mass showing moderate mitral valve regurgitation

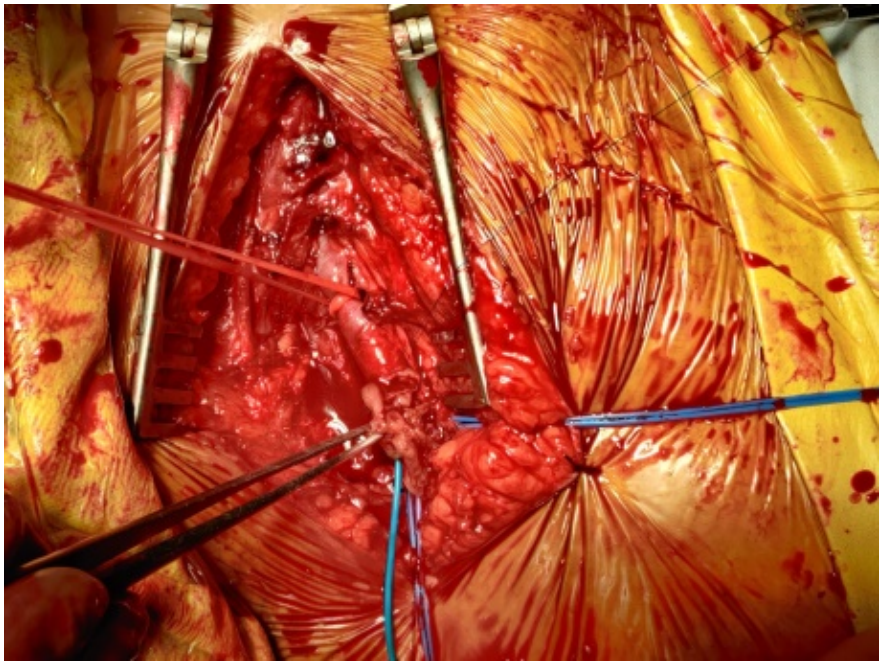

Figure 5 access via right superficial femoral artery post embolectomy showing myxomatous tissue retrieed from Right CIA

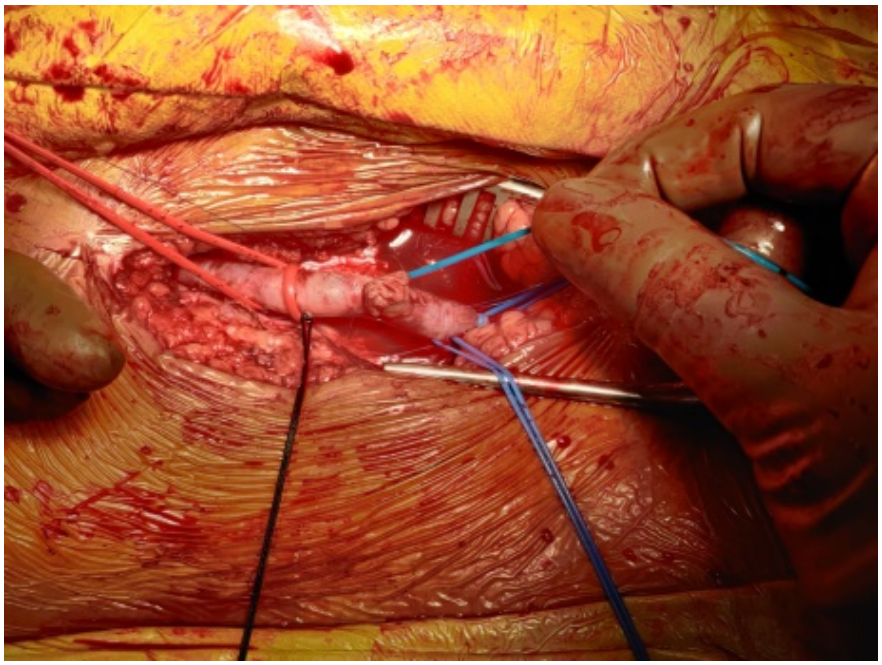

Figure 6 left SFA access shoeing retrieved myxomatous tissue from Abdominal aorta and CIA