Atrial Myxoma: A case of acute aortic occlusion and discussion of radiological findings

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Learning Objectives

We report a case of left atrial myxoma presenting as acute aorto-iliac saddle embolus. This educational exhibit aims to outline the pathophysiology of cardiac myxoma, clinical presentation / management and findings on chest radiographs (CXR), echocardiography, computer tomography (CT) and magnetic resonance imaging (MRI).
CASE HISTORY

A 62-year-old man presented to the emergency department complaining of sudden onset bilateral lower limb pain. The exact time course of symptoms was unclear as the patient was confused on arrival. The patient's past medical history included ischemic heart disease with coronary artery bypass 6 years prior, hypertension, hyperlipidemia and type II diabetes mellitus. Medications included ACE inhibitor, statin and oral hypoglycemic agent.

On assessment, the patient was confused with a GCS of 14, pulse was regular at a rate of 85BPM and blood pressure was elevated at 200/100 mmHg. The patient's feet were white and cold and no lower limb pulses were palpable from the groin to feet. Physical examination was otherwise normal with no focal neurological deficits.

ECG was performed and confirmed sinus rhythm. CXR was normal, cardio-mediastinal contours were within normal limits and no features of heart failure were identified. Bedside abdominal ultrasound demonstrated no abdominal aortic aneurysm or intra-abdominal free fluid. Lactate was elevated at 5.5 mmol/L, results of FBC and ELFTs were within normal ranges.

The patient was referred for CT brain and CT angiogram (CTA) of the abdominal aorta and lower limbs. CTA demonstrated saddle thrombus at the aortic bifurcation and distal thrombus in the lower limbs bilaterally, no aneurysms were identified (Image 1). CT brain was normal.

Urgent vascular surgical review was undertaken and the patient proceeded to bilateral femoral embolectomy, popliteal embolectomies and fasciotomies. Clot was retrieved with Fogarty embolectomy catheter and distal flow was re-established. Bilateral fasciotomies were performed and the decompressed muscle was found to be swollen but viable. On close examination, the material retrieved at embolectomy was atypical for bland thrombus and appeared pale with a course, particulate consistency.

Transthoracic and transoesophageal echocardiogram were performed on the basis of intra-operative findings and demonstrated left atrial myxoma (Images 2-4). The patient underwent urgent cardiac surgery for resection of the left atrial myxoma. Histological examination confirmed the diagnosis of atrial myxoma and the patient made a full post-operative recovery.
CARDIAC MYXOMA

Cardiac myxoma is a rare disease with a variety of non-specific clinical manifestations [1]. Systemic embolism caused by tumour fragmentation is a well known entity [2-5]. Less commonly, aortic saddle embolus may occur and this presents as a vascular emergency requiring urgent intervention [6].

Primary cardiac neoplasms are rare, affecting patients of all ages, and have a reported prevalence in autopsy series of 0.001%-0.03% [7]. It is estimated that primary cardiac neoplasms are 100-1,000 times less prevalent than secondary neoplasms of the heart. The most common primary cardiac neoplasm is myxoma, which accounts for approximately half of all cases. Other benign primary tumours include papillary fibroelastoma, rhabdomyoma, fibroma, hemangioma, and lipoma [7]. Although sarcomas constitute less than 25% of primary cardiac tumours, they represent the second most common primary cardiac neoplasm [8]. Rarely, lymphoma may manifest as a primary cardiac tumour.

Cardiac myxoma is an intracavitary, round or ovoid neoplasm that is attached to the endocardium [10]. The majority (75%) occur in the left atrium, with a smaller proportion in the right atrium [1,7,8]. The histologic features of cardiac myxomas are distinctive but heterogeneous, the cellular origin is unknown, but it is likely a primitive mesenchymal cell capable of diverse forms of differentiation [10]. The majority of cardiac myxomas occur sporadically, with a small proportion demonstrating a familial predisposition or an association with a clinical complex (Carney complex) in which patients exhibit myxomas, spotty skin pigmentation and endocrine overactivity [11].

Patients with cardiac myxomas often present with one or more features of the classic triad: intracardiac obstruction (67%); embolic events (30%); and constitutional or systemic symptoms (34%) [12]. Cardiac obstruction commonly occurs at the mitral valve and affected patients present with dyspnea and orthopnea from pulmonary venous hypertension [1,2,9]. The pedunculated and prolapsing nature of these tumours allows for positional and intermittent mitral valve obstruction [9]. Right atrial myxomas may obstruct the tricuspid valve and cause symptoms of right-sided heart failure, peripheral oedema, passive hepatic congestion, and syncope [9]. The likelihood of obstruction by myxoma is dependent on tumour location, size and mobility. On physical exam, auscultation reveals systolic and diastolic murmurs in over 50% of patients [1].
Embolism occurs in 30-40% of patients with cardiac myxoma[1,8]. As most tumours reside within the left atrium, embolic events are normally systemic, involving the cerebral vessels most commonly. Peripheral vessels, abdominal viscera, renal arteries and coronary arteries may also be occluded. Right atrial tumours can cause pulmonary embolus, but these are rare[8]. Emboli may consist of portions of the myxoma itself or of thrombus, which often is seen in association with myxomas [12]. As well as causing symptoms due to vascular occlusion, emboli may also cause multiple aneurysms both in the CNS and elsewhere, this is thought to be due to invasion of the arterial wall by myxomatous material with subsequent weakening and dilation of the arterial wall [5].

Constitutional symptoms of fever, malaise, and weight loss have been reported and may be related to an autoimmune reaction initiated by the tumour [1,8,10]. These symptoms occur irrespective of tumour size or location [8]. Laboratory abnormalities are anemia, increase in erythrocyte sedimentation rate, C-reactive protein and globulin levels [8].

Patients with diagnosis of an atrial myxoma require early resection due to the high risk of valvular obstruction and embolization [13]. The general principles of surgical management are careful manipulation with excision of the stalk and endocardial origin of the tumour in an effort to prevent recurrence [13]. Cardiac valves in contact with the tumour require inspection to assess traumatic damage [14]. After resection, the recurrence rate is 1 to 3% for sporadic myxoma, but second myxomas can occur in 12% of patients with familial disease and 22% of patients with Carney complex [14].
Fig. 1: CT Angiogram of abdominal aorta (sagittal reconstruction), demonstrating saddle embolus at aortic bifurcation.

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Fig. 2: Transoesophageal echocardiography demonstrating left atrial myxoma

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Fig. 3: Transoesophageal echocardiography demonstrating left atrial myxoma

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Fig. 4: 4-Dimensional transoesophageal echocardiography demonstrating left atrial myxoma

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Imaging Findings OR Procedure Details

CHEST RADIOGRAPH

Classic radiographic findings of cardiac myxoma are related to the sequelae of mitral valve obstruction, causing cardiomegaly and elevated left atrial pressures. Specific findings related to elevated left atrial pressure include left atrial enlargement, vascular redistribution, prominent pulmonary trunk, pulmonary oedema and enlarged left atrial appendage [9]. These findings are not specific to left atrial myxoma and suggest more common diseases related to pathologic conditions of the mitral valve [9].

ECHOCARDIOGRAPHY

Two-dimensional echocardiography classically reveals a mobile tumour connected to the interatrial septum by a narrow stalk [15]. Myxomas usually have heterogeneous echogenecity and occasional calcifications [15]. The most important clue to the diagnosis is their location in the left atrium and origin from the mid-portion of the atrial septum [15]. Given a typical presentation, echocardiography is virtually diagnostic of myxoma. Transthoracic echocardiography is usually sufficient to make the diagnosis, but if the results are suboptimal, transoesophageal echocardiography should be employed [15].

COMPUTER TOMOGRAPHY

Cardiac myxomas generally appear as filling defects within a contrast material filled cardiac chamber, and are either hypoattenuated or isoattenuated relative to the myocardium [9]. Previously described CT characteristics of cardiac myxoma include a lobular contour and heterogeneous attenuation [13]. Scheffel et al evaluated CT findings in patients with myxoma and atrial thrombus and concluded that origin, mobility and prolapse were helpful features in differentiation myxoma from thrombus. Size and shape were sometimes useful, while CT attenuation, location and calcification were not differentiating features (Table 1) [16]. Myxomas typically originate from the fossa ovalis, while thrombus is most often seen in the atrial appendage. Myxomas often demonstrate stalk like attachment to the myocardium and are found to be more mobile than thrombus, while prolapse through the mitral valve is highly specific for myxoma [16].
ECG-gated CT coronary angiogram demonstrating left atrial myxoma, which is attached to the intra-atrial septum via thin stalk and prolapses through the mitral valve. Small calcifications are also identified. (Image 5)

MAGNETIC RESONANCE IMAGING

Cardiac MRI plays a significant role in the evaluation of cardiac masses and is of greatest value when echocardiographic findings are equivocal, suboptimal, or when the lesion has an atypical location or appearance [8]. Heterogeneous signal intensity in myxomas likely correlates with the high frequency of haemorrhage, calcification, fibrosis, and cystic change, as well as the variable amounts of myxomatous components that characterize these tumours [8].

Studies that have directly correlated signal intensity of myxomas with the underlying pathologic entity reveal that calcified areas are low in signal intensity with both T1- and T2-weighted sequences and that chronic haemorrhage exhibits high signal intensity with both sequences [17]. Myxomatous elements with a polysaccharide-rich ground substance have low signal intensity on T1-weighted images and high signal intensity on T2-weighted images [18].

Case 3

Cardiac MRI of a left atrial myxoma. Myxoma was identified on echocardiography and due to atypical location, further evaluation with MRI was performed. MRI demonstrates a mobile lesion attached to the posterior wall of the left atrium, with heterogeneous signal intensity on double inversion recovery and fat suppression sequences and progressive enhancement on delayed myocardial enhancement phases. (Images 6 & 7)
### Table 1: CT Features For Differentiating Cardiac Myxoma And Thrombus

<table>
<thead>
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<th>Feature</th>
<th>Significance</th>
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<tr>
<td>Origin</td>
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<td>Mobility</td>
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<td>Shape</td>
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<td>Location</td>
<td>-</td>
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<tr>
<td>Calcifications</td>
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(++) = p < 0.01  (+) = p < 0.05  (--) = p not significant

*Scheffel* et al. Atrial Myxoma and Thrombi: Comparison of Imaging features on CT. AJR 2009

**Fig. 5:** ECG-gated CT coronary angiogram: Axial images at upper and mid section of lesion, 4 Chamber and long axis reconstruction (Clockwise from top left). (Images from The Prince Charles Hospital, Department of Medical Imaging.)

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Fig. 6: Double inversion recovery T1 (DIR), DIR + fat suppression (FS), Myocardial delayed enhancement (12 minutes), Myocardial delayed enhancement (5 minutes) (Clockwise from top left). (Images from The Prince Charles Hospital, Department of Medical Imaging.)

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**Fig. 7:** Fast imaging employing steady state acquisition (FIESTA) of left ventricle in long axis

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Conclusion

Cardiac myxoma is a rare disease with a variety of non-specific clinical manifestations [1]. Systemic embolism caused by tumour fragmentation is a well known entity and most frequently involves the cerebral vessels [2-5]. We present a less common case of aortic saddle embolus caused by left atrial myxoma and highlight the radiological findings to aid in prompt diagnosis and facilitate urgent management.

Chest radiographs of affected patients demonstrate findings related to mitral valve obstruction and elevated left heart pressures, however, these findings are non-specific and normally relate to more common conditions involving the mitral valve [8]. Echocardiography is a useful first-line investigation for cardiac myxoma and transoesophageal imaging can be performed to further evaluate suboptimally visualized or atypical lesions [15]. CT can accurately differentiate atrial thrombus from cardiac myxoma [16] and can be included when CT angiogram is performed to investigate embolic events. Cardiac MRI is of greatest value when first line investigations are equivocal or for further evaluation of atypical lesions [8].

The cases discussed in this article highlight some of the unique characteristics and clinical presentations of cardiac myxoma. An awareness and understanding of these features is important in all aspects of clinical practice, so that potential cases of cardiac myxoma can be correctly diagnosed and managed.
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References


