DIAGNOSIS OF LEFT ATRIAL MYXOMA FOLLOWING SYSTEMIC EMBOLISM

H C Tan, J Sebastian, M H Choo, G C Raju, C N Lee, J K Yeoh, M Tay

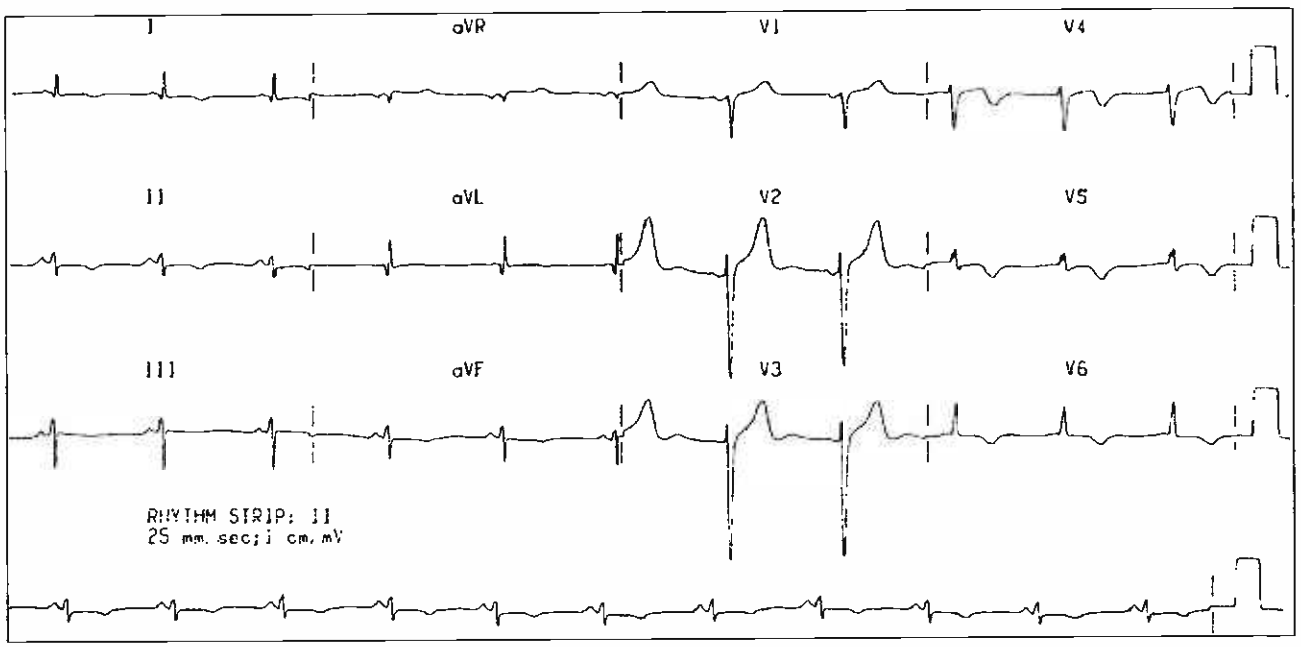
ABSTRACT

Cardiac myxomas are rare and noted for their varied clinical manifestations. Consequently, the diagnosis is often unsuspected until the symptoms and signs become advanced and obvious, or when the diagnosis is made fortuitously during echocardiography. This report illustrates a case in point in which the diagnosis was made on transthoracic echocardiography and amplified using transesophageal echocardiography.

Keywords: atrial myxoma, coronary and systemic embolism, transesophageal echocardiography.

SINGAPORE MED J 1994; Vol 35: 420-422

Fig 1 - Electrocardiogram showing anteroseptal myocardial infarction.



Cardiac Department
National University Hospital
Lower Kent Ridge Road
Singapore 0511.

H C Tan, M Med (Int Med), MRCP
Registrar

M H Choo, MD, M Med (Int Med), FRCP
Professor and Chief

C N Lee, FRCS, FRACS
Cardiothoracic Surgeon

J K Yeoh, M Med (Int Med), MRCP
Senior Lecturer and Consultant Cardiologist

M Tay
Chief Cardiac Technologist

Department of Pathology
National University Hospital

Raju GC, FRCPath, FCAP
Pathologist

RIPAS Hospital
Bandar Seri Begawan
Brunei Darussalam

J Sebastian, FRCP
Specialist Physician

Correspondence to: Prof M H Choo

INTRODUCTION

About 50% of cardiac tumours are cardiac myxomas. Cardiac myxoma classically manifests a triad of constitutional, embolic and obstructive symptoms[1,2]. Unless the presentation is typical, the diagnosis is rarely made on clinical grounds. Most cases are unsuspected, and often diagnosed during echocardiography. We report a case in which the diagnosis was confirmed after coronary and systemic embolism.

CASE REPORT

A month after presenting with angina pectoris, a 50-year-old Malay man suffered an acute anterior myocardial infarction (Fig 1). Following discharge, he continued to have exertional dyspnea, but no abnormal physical signs were detected. However, an echocardiogram to assess left ventricular function revealed, serendipitously, a large left atrial myxoma projecting through the mitral valve during diastole.

While awaiting cardiac surgery, he developed acute bilateral lower limb ischaemia necessitating emergency operation. Myxomatous tumour tissue, confirmed on histopathology (Fig 2), was removed from the ilio-femoral arteries. Post-operative transthoracic and then transesophageal echocardiography showed a residual myxoma attached to the low atrial septum (Figs 3 and 4). Apart from an occluded left anterior descending coronary artery, coronary arteriography

was normal (Fig 5). There was severe antero-apical left ventricular hypokinesia and mild mitral regurgitation.

At surgery, the myxoma was resected together with its base, and the resultant septal defect was closed with a Dacron patch. The left anterior descending artery was deeply intramyocardial, and could not be safely biopsied. All visible and palpable epicardial coronary arteries were normal.

DISCUSSION

Cardiac myxomas are true intracavitary neoplasms that can occur anywhere in the heart. There is no sex preponderance, and it is commonest between 30 and 60 years of age. Occasional familial cases have been reported and the mode of transmission is believed to be autosomal dominant or recessive. About 75% are found in the left atrium and most have a pedunculated attachment to the atrial septum near the fossa ovalis. Pedunculated myxomas tend to be soft, friable, mobile and more likely to embolise than sessile tumours which often present with constitutional symptoms rather than mechanical complications.

The classical triad of constitutional, embolic and obstructive manifestations results in protean presentations. Constitutional symptoms, seen in up to 90% of cases, include myalgia, muscle weakness, arthralgia, fever, weight loss, fatigue, Raynaud's phenomenon, and clubbing. Myxomatous embolism can affect any part of the arterial bed, but involves the cerebral circulation in about 50% of cases[3]. Obstruction can mimic mitral stenosis, with two additional features – the patient improves when supine and may develop syncope when reclining, and a pathognomonic "tumour plop" may be

Fig 2 - Slide of femoral embolectomy showing myxomatous mass composed of polygonal or stellate cells in loose abundant mucoid matrix.

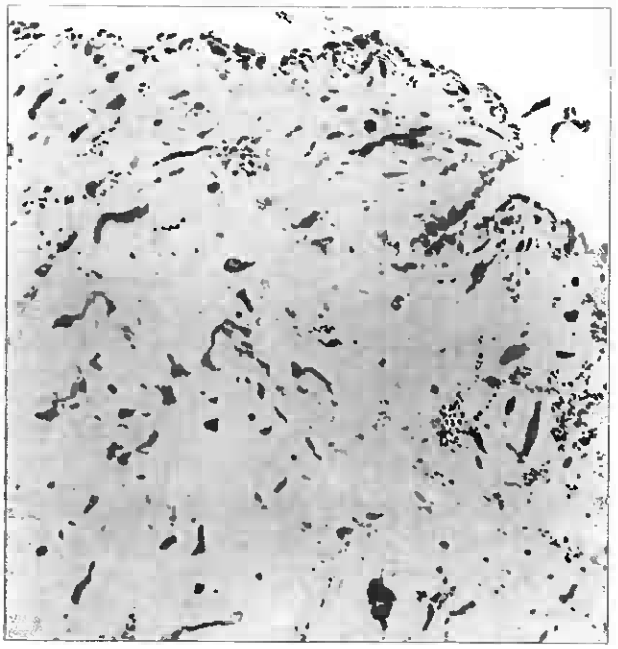


Fig 3 - Transthoracic echocardiographic print.

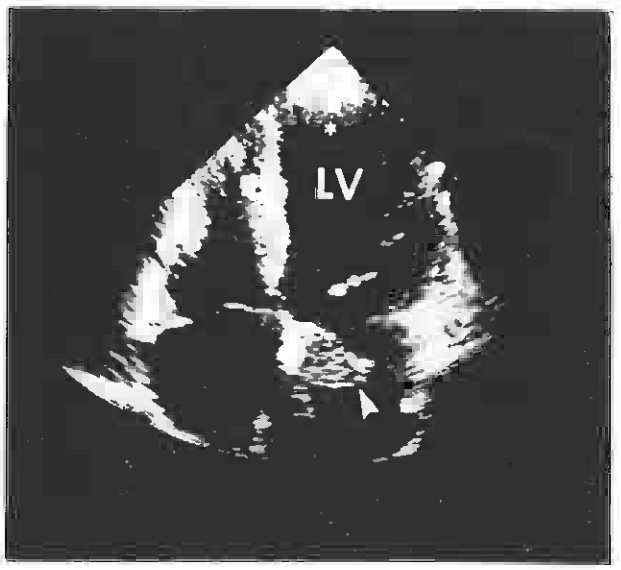


Fig 4 - Transesophageal echocardiographic print.

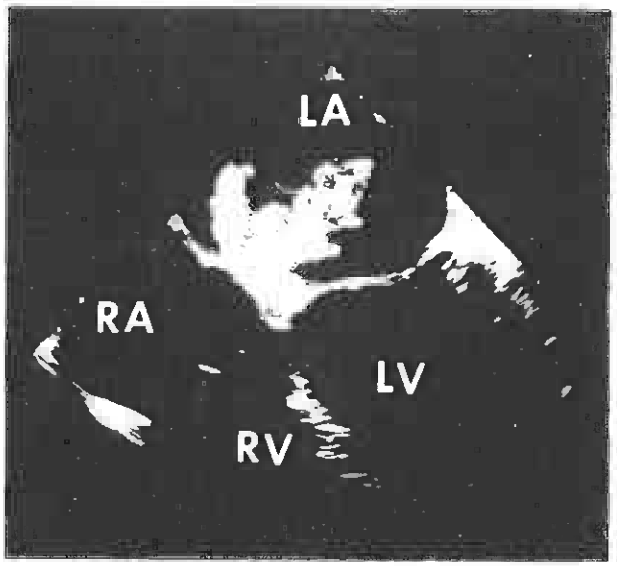
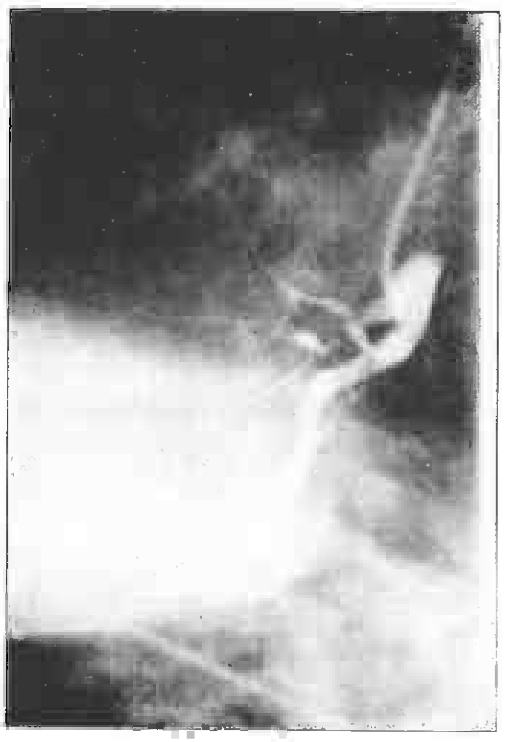


Fig 5 - Coronary angiogram showing blocked left anterior descending artery.



auscultated.

Despite these classical features, the clinical presentation is often initially subtle, and recognition is frequently delayed. Transthoracic and, more recently, transesophageal echocardiography are often the means by which the diagnosis is revealed or confirmed.

In systemic embolism, atrial myxoma should be suspected if the embolism is recurrent or neurologic, especially if the patient is young. Embolic myocardial infarction is uncommon[1]. Careful clinical assessment may uncover symptoms and signs suggesting mitral obstruction. The index of suspicion is even higher if the patient also has a disorder resembling collagen disease or vasculitis, accompanied by an elevated sedimentation rate, hypocomplementemia and positive anti-double stranded DNA antibodies[5]. An echocardiogram should then be performed as soon as possible. If cerebral embolism is suspected, magnetic resonance imaging may reveal clinically inapparent but therapeutically important disseminated lesions in the brain[6].

The treatment of choice is prompt resection for symptomatic improvement, avoidance of complications and early normalisation of intracardiac pressures. Pre-operative transesophageal echocardiography, as in this report, is invaluable and superior to transthoracic echocardiography and even cine-angiography for morphologic characterisation of the tumour to facilitate precise surgery[7].

In the clinical context, and especially since the remainder of the coronary circulation was normal, our patient most likely had recurrent embolisation to the left anterior descending coronary artery manifesting initially as chest pain and subsequently infarction. Further embolisation of the myxoma affected his iliofemoral arteries. As in many such patients, diagnosis was made during echocardiography.

References

1. Difilo T, Cantelmo NL, Haudenschild CC, Watkins MT. Atrial myxoma with remote metastasis: case report and review of the literature. Surgery 1992; 111 : 352-5
2. Markel ML, Waller BF, Armstrong WF. Cardiac myxoma: a review. Medicine 1987; 66 : 114-25
3. Obeid A, Marvasti M, Parker F, Rosenberg J. Comparison of transthoracic and transesophageal echocardiography in diagnosis of left atrial myxoma. Am J Cardiol 1989; 63 : 1006-8.
4. Reichmann H, Romberg-Hahnloser R, Hofmann E, Becker T, Mertens HG. Neurological long-term follow-up in left atrial myxoma: Are late complications frequent or rare? J Neurol 1992; 239 : 170-4.
5. Romisher SC, Cannon LA, Davakis N. Atrial myxoma associated with inferior myocardial infarction. Ann Emerg Med 1991; 20 : 1236-8.
6. Marazuela M, Garcia-Merino A, Yebra M, Brasa JM, Diego J, Durantez A. Magnetic resonance imaging and angiography of the brain in embolic left atrial myxoma. Neuroradiology 1989; 31 : 137-9.
7. Coughlin WF, Knott PE. Right atrial myxoma: A cause of septic pulmonary emboli in an adolescent female. J Adolesc Health Care 1990; 11 : 351-4.