CASE REPORT

Arrhythmias are not to blame for all cardiac syncope patients: left atrial myxoma causing syncope in a middle-aged man

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SUMMARY

A 47-year-old man presented with a history of syncope that lasted for 3 min and was not accompanied by jerky movement of limbs or incontinence. After regaining consciousness, he felt generalised weakness. There was no history of chest pain or palpitation. ECG showed normal sinus rhythm. All blood investigations were normal. Transthoracic echocardiography showed a large multilobulated echo dense mass in the left atrium. The mass was prolapsing through the mitral valve during diastole. Transoesophageal echocardiography verified these findings and also showed the stalk of the mass attached to the interatrial septum near the fossa ovalis. The mass was highly suggestive of myxoma. The patient underwent surgical resection of the mass and histopathology confirmed the diagnosis of left atrial myxoma.

BACKGROUND

Primary tumour of the heart is a very rare entity. Cardiac myxoma is the most common primary tumour in adults and usually arise from the left atrium. Their presentations vary from asymptomatic to obstructive symptoms causing syncope, peripheral embolisation presenting with ischaemic stroke, cardiac failure mimicking mitral stenosis or constitutional symptoms. Syncope is the principal symptom of obstruction caused by myxoma. Therefore, echocardiography plays a key role in the search for cardiac aetiology in the workup of syncope.

CASE PRESENTATION

A previously healthy 47-year-old labourer presented to the emergency department with a history of sudden loss of consciousness lasting for 3 min. It was not associated with any jerking movement of the limbs or incontinence of urine. After regaining of consciousness, the patient reported transient numbness and weakness in the left side of the body. The patient did not have a history of palpitations, dizziness, chest pain or shortness of breath prior to the episode of syncope.

The patient's medical history was unremarkable.

On physical examination he was conscious and alert. The blood pressure was 131/70 mm Hg, heart rate was 65 bpm and regular, respiratory rate was 18 breaths/min and he was afebrile. In cardiovascular examination there was a third heart sound, plop and soft systolic murmur were audible. The respiratory and neurological examinations were unremarkable.

INVESTIGATIONS

ECG showed normal sinus rhythm. There was no evidence of long or short QT syndrome, brugada syndrome, pre-excitation or epsilon wave. The patient did not develop any arrhythmia during the course of his stay in hospital. His blood investigation showed haemoglobin of 13.6 g/dL, white cell count $7.9 \times 10^3/\mu\text{L}$, platelet count $270 \times 10^3/\mu\text{L}$ and glycosylated haemoglobin 5.4%. His prothrombin time 14.1 s, partial thromboplastin time 38.4 s and international normalised ratio 1.13. Sodium was 140 mEq/L and potassium 4.4 mEq/L. Cardiac enzymes and troponin were normal. C reactive protein level was 30 mg/L and erythrocyte sedimentation rate was 37 mm in the first hour. Chest X-ray was normal. MRI of the brain was normal.

Transthoracic echocardiography showed normal-sized cardiac chambers. There was a large, fragile, multilobulated and pedunculated mass floating in the left atrium originating from the interatrial septum and measuring 6.9×2.4 cm. The anterior mitral leaflet was free of the mass. It prolapsed through the mitral valve and into the left ventricle during diastole (figure 1). There was a mild mitral regurgitation and the mitral valve pressure half time was 122 ms, giving a mitral valve area of 1.8 cm^2 ; mean gradient was less than 3 mm Hg. There was mild tricuspid regurgitation with right ventricular systolic pressure of 30 mm Hg. The left ventricular systolic function was fair and the ejection fraction was 55%. The transoesophageal echocardiography verified the initial findings and additionally showed the stalk of the mass attached to the interatrial septum near the fossa ovalis (figure 2). These findings were highly suggestive of left atrial myxoma.

DIFFERENTIAL DIAGNOSIS

- ▶ Transient ischaemic attack
- ▶ Seizure
- ▶ Hypertrophic cardiomyopathy
- ▶ Mitral stenosis
- ▶ Aortic stenosis

TREATMENT

The patient was admitted and monitored. Brain imaging was carried out and was normal. After completing cardiac imaging the patient was referred for cardiac surgery. He underwent surgical resection of the left atrial mass.

Macroscopically, the mass was irregular and nodular. It measured $6.0 \times 3.5 \times 1.5$ cm and

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Figure 1 (A) Transthoracic echocardiography in parasternal long-axis view and (B) apical long-axis view showing prolapsing of the left atrial myxoma.

weighed 20.3 g. The external surface was haemorrhagic (figure 3A) and its slicing revealed greyish white jelly-like soft tissue. Microscopic examination revealed nodular tumour tissue composed of a few spindle and stellate cells embedded in abundant pale eosinophilic myxoid stroma (figure 3B). There were areas of haemorrhage and hemosiderin pigments were seen in between. Histopathology confirmed the diagnosis of left atrial myxoma.

OUTCOME AND FOLLOW-UP

After the operation, the patient was clinically and haemodynamically stable. He was discharged and came for follow-up after 1 month. Transthoracic echo was repeated, showing the left atrium free of mass; there was mild mitral regurgitation and normal left ventricular function. The patient was advised to return for echocardiography yearly in order to monitor for recurrence of the myxoma.

DISCUSSION

Primary tumour of the heart is a very rare disease and its incidence is very low. In a large review of more than 12 000 autopsies by Lam *et al*,¹ the incidence of primary cardiac tumour was 0.056%. Most primary cardiac tumours are benign and cardiac myxoma is the most common benign tumour in adults.² Myxomas are mostly sporadic but occasionally they can be familial, as in Carney complex tumour syndrome due to mutation in the PRKAR1 gene.³

Myxomas typically originate from the endocardium of any cardiac chamber and, rarely, from heart valves. They commonly present in the age group between 30 and 60 years, with

50 years being the mean age at time of presentation. Two-thirds of patients are female.⁴ A review article by Reynan mentioned that 75–80% of myxomas arise from the left atrium, 15–20% from the right atrium and, rarely, they can be biatrial.⁵ In our case report, the patient was a male and his age was 47 years, which is near the mean age of other studies.

The clinical presentation of cardiac myxoma varies from asymptomatic to that with serious cardiovascular complication. The classic triad of clinical presentation includes valvular obstruction, embolic phenomenon and constitutional symptoms. However, in a case series of 112 patients of left atrial myxomas analysed by Pinede *et al*,⁶ the three most common symptoms were cardiac failure, embolism and constitutional symptoms. Manduz *et al*,⁷ in another small study of 12 patients, also found the most common presentations were cardiac failure and embolisation. Lee *et al*⁸ reviewed case records of 74 patients of cardiac myxoma; 12% of them presented with neurological symptoms. Ischaemic cerebral infarction was the most common manifestation.

In a case report by Mehmet, a 65-year-old woman was diagnosed initially as cardiac failure, 3 months after diagnosis she developed syncope. Her ECG showed atrial fibrillation and echocardiography revealed large right atrial myxoma.⁹ Nogueira *et al*¹⁰ cited a case report of a 14-year-old girl with a history of recurrent syncope. Her investigation exhibited left atrial myxoma. Similarly, Azevedo *et al*¹¹ cited a case of a 74-year-old woman with massive right atrial myxoma causing tricuspid obstruction, and presenting as syncope and exertional dyspnoea.

Sunnar *et al* reported an interesting case of a 58-year-old woman presenting with features of cardiac failure with signs of pulmonary hypertension. On physical examination, she had a systolic murmur and mid diastolic rumbling murmur. She was diagnosed as mitral stenosis. The transthoracic echocardiography showed dilation of both atria and right ventricle, with a large left atrial myxoma prolapsing into the left ventricle during diastole. There was moderate mitral regurgitation and severe tricuspid regurgitation with severe pulmonary hypertension. That case was an example of left atrial myxoma resembling mitral stenosis.¹²

Cardiac conditions account for a small proportion in the aetiology of syncope. Brignole and his colleagues carried out a prospective multicentre study of 941 syncope patients. They found that 6% of the patients had a cardiac cause of syncope.¹³ Echocardiography is an important tool for the assessment and diagnosis of cardiac causes of syncope.

Transthoracic echocardiography is mandatory in high-risk groups of patients to rule out structural heart disease, for

Figure 2 Transoesophageal echocardiography showing stalk of the mass attached to interatrial septum above fossa ovalis.

Figure 3 (A) Haemorrhagic external surface of myxoma and (B) histopathology of mass showing myxoid cell and stellate cell.

example, aortic stenosis, hypertrophic cardiomyopathy, myxoma or arrhythmogenic right ventricular dysplasia.¹⁴

Echocardiography can delineate the mass and it produces better images of attachment of the stalk of the myxoma. CT scan and MRI provide a higher degree of soft tissue discrimination but mobility and attachment site of myxoma cannot be assessed accurately.¹⁵

Cardiac myxomas are well treated by complete surgical resection. Mishra published his experience of surgery for myxoma over a period of 4 years with 50 patients. He found the mortality rate was 8% and the risk of recurrence was low. Recurrence was more common in the familial form.¹⁶ Surgical results by Centofanti *et al*¹⁷ of 83 patients with cardiac myxoma found that in-hospital mortality was 3.6% and there was no recurrence in long-term follow-up.

In our case of left atrial myxoma, the patient presented the first time as syncope. The mitral inflow velocities were mimicking mild pseudo-mitral stenosis. The syncope in our patient was most likely due to the myxoma causing obstruction of the mitral valve.

Learning points

- ▶ History, physical examination and assessment of ECG play vital roles in the diagnosis of syncope.
- ▶ Do not forget cardiac masses in the differential diagnosis of syncope.
- ▶ Echocardiography is an important tool in the work up of cardiac syncope.
- ▶ Primary cardiac tumour is a very rare disease and myxoma is the most common tumour in adults.
- ▶ The long-term outcome after surgical resection of cardiac myxoma is favourable.

Contributors RNM and MHM were involved in the case study and discussion preparation. PRG helped in the acquisition of echo images.

Competing interests None.

Patient consent Obtained.

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