Case report

An unusual presentation of atrial myxoma: thromboembolic phenomenon with a recent normal echocardiogram

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SUMMARY

Atrial myxomas are the most common primary neoplasms of the heart. Clinical presentation varies from asymptomatic incidental masses to serious life-threatening cardiovascular complications. We describe the case of a 37-year-old man who presented with both coronary and cerebral embolisation secondary to an atrial myxoma in the context of a recent normal transthoracic echocardiogram.

BACKGROUND

We discuss the importance of transthoracic echocardiography in the diagnosis of this rare condition and the unique challenges that their characteristic rapid growth poses to diagnosis and monitoring for recurrence.

CASE PRESENTATION

A 37-year-old man presented to our emergency department in May 2015 with palpitations and dizziness and was found to have new atrial fibrillation. His medical history was unremarkable, and he was a lifelong non-smoker. Physical examination was unremarkable, and a 12-lead ECG confirmed atrial fibrillation (AF) with a rapid ventricular rate of 140 beats/min. Pathology—full blood count, biochemistry and thyroid function test—were unremarkable. He was admitted to the coronary care unit where he received beta-blocker therapy and reverted spontaneously. The patient was discharged home without anticoagulation as his CHADSVASC score was 0. An outpatient transthoracic echocardiogram (TTE) revealed normal left ventricular systolic function with no significant valvular

pathology and 24 hours of Holter monitoring confirmed persistence of sinus rhythm.

Ten months later, in February 2016, he presented with severe retrosternal chest pain. An ECG showed ST elevation in inferior leads. He was admitted for emergency invasive coronary angiography, which revealed an occluded sub-branch of the posterior descending artery branch of a dominant right coronary artery. The remainder of his coronary arteries featured no disease. Left ventriculography confirmed minor inferoapical wall hypokinesis with overall preserved systolic function. The decision was made to manage his acute coronary syndrome conservatively with medical therapy. He was in sinus rhythm during his inpatient stay. On account of his recent normal TTE 10 months prior, he did not have a repeat study. He was commenced on aspirin, ticagrelor, atorvastatin and metoprolol, and was discharged home after 3 days. The aetiology of his acute coronary syndrome was likely secondary to an embolic source. He had a thrombophilia screen and no clinical or biochemical evidence to suggest infective endocarditis, and it was postulated that episodes of subclinical AF with thromboembolism could be a possibility.

In May 2016, he presented with transient left arm and leg weakness. His cardiac and neurological examination were unremarkable. CT of the brain did not reveal any intracranial abnormalities while brain MRI demonstrated evidence of embolic foci in multiple vascular territories ([figure 1](#)). Further investigations were performed to find a cause for his ischaemic stroke presentation. Carotid artery Doppler was unremarkable, and telemetry continued to show sinus rhythm. Transesophageal echocardiography revealed a 6 cm mass adjacent to

Figure 1 MRI showing pattern consistent with diffuse thromboembolic events arousing suspicion of central cause.

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Figure 2 A four-chamber view on transoesophageal echocardiography demonstrating a large mass on the mitral valve.

the anterior mitral valve leaflet prolapsing into the left atrium (figure 2). Cardiac magnetic resonance (CMR) imaging was performed to further characterise the mass and to assist with surgical planning. The CMR suggested intimate association with the anterior leaflet of the mitral valve, possible contact with interatrial septum and morphology and enhancement characteristics consistent with papillary fibroelastoma. It was concluded that the patient's presentations with acute coronary syndrome and then with ischaemic stroke were both secondary to embolic phenomena from the mass. This conclusion, coupled with the suggestion of malignancy on CMR, prompted a referral for surgical excision.

OUTCOME AND FOLLOW-UP

A large tumour attached to the fossa ovalis with a preserved mitral valve was observed intraoperatively, which is consistent with an atrial myxoma. The lesion was resected and was 6 cm in diameter (figure 3). Histology confirmed the mass as an atrial myxoma with characteristic large areas of haemorrhage. The patient recovered well with no recurrence of the mass on repeat transthoracic echocardiography 12 months later.

DISCUSSION

Atrial myxomas are the most common primary cardiac tumour. They typically occur in the left atrium and can occur at any age.^{1,2} Most patients present with symptoms of systemic embolisation

Figure 3 Resected atrial myxoma with ruler for scale.

Learning points

- ▶ The importance of considering central embolic phenomenon in patients who present with acute coronary syndrome, or stroke without obvious risk factors or with atypical pathology.
- ▶ The importance of considering rare unifying diagnoses in the context of a constellation of symptoms or signs that are not typical or a patient who is not demographically typical for the condition.
- ▶ Practitioners should not be reassured by a normal echocardiogram performed over 6 months prior when considering either new or recurrent atrial myxoma in their differential diagnosis.

or left atrial obstruction, but up to 10% of patients may be completely asymptomatic.^{1,2}

This case features the unusual heterogeneity of atrial myxoma growth and a rare clinical presentation characterised by multiple thromboembolic events.²⁻⁷

In the case described above, the patient initially presented with atrial fibrillation and his transthoracic echocardiography did not reveal any left atrial masses. Ten months later, he presented with an acute coronary syndrome and finally a further 4 months later with an ischaemic stroke. This illustrates the rapid progression of atrial myxomas and the importance of repeat echocardiography in young patients with no significant cardiovascular risk factors. In this case, repeat echocardiogram was omitted on account of recent normal TTE (10 months prior) and an opportunity to make an earlier diagnosis was missed.

Cases describing the rate of growth of atrial myxomas are rare as patients usually do not have an echocardiogram documenting the absence of the lesion in the months prior to their diagnosis. In this case, the atrial myxoma grew at a presumed rate of 0.43 cm/month. This rate relies on several assumptions, it is calculated from the time of last normal echocardiogram to the time of excision and so may reflect time to diagnosis rather than rate of growth, especially considering that the patient was symptomatic of the lesion some time prior to excision. However, a rate of 0.43 cm/month is consistent with a case series by Walpot *et al*⁸ that reported rates from 0 to 1.36 cm/month with the majority growing at a rate between 0.1 and 0.9 cm/month. This gross variability in growth rate makes it difficult to establish appropriate interval period for follow-up echocardiography given a recurrence rate of 5%–14% and a reported time to recurrence of 0.5–6.5 years.^{1,2,7,8}

Finally, the combination of coronary and neurological manifestations of embolic phenomenon in the same patient has rarely been reported in the literature.^{1,3-5} Typically, patients present with abnormalities in a single system. If they present with evidence of embolic phenomenon this is usually only limited to a single event.^{1,2} A case series by Pinede *et al*¹ described 2 or more embolic events in only 7 out of 112 cases and coronary emboli in only 4 of 112 cases. The most common presenting complaint reported in this case series were signs and symptoms of cardiac failure which were notably absent in the case described in this report.

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REFERENCES

- 1 Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. *Medicine* 2001;80:159–72.
- 2 Yu K, Liu Y, Wang H, *et al.* Epidemiological and pathological characteristics of cardiac tumors: a clinical study of 242 cases. *Interact Cardiovasc Thorac Surg* 2007;6:636–9.
- 3 Sha D, Fan G, Zhang J. Multiple cerebral infarction as the initial manifestation of left atrial myxoma: a case report and literature review. *Acta Cardiol* 2014;69:189–92.
- 4 Rahman SMM, Kibria MG, Rahim AMA, *et al.* Clinical presentation of left atrial myxoma in relation to anatomic and pathologic type. *Cardiovascular Journal* 2015;8:19–22.
- 5 Zheng Z, Guo G, Xu L, *et al.* Left atrial myxoma with versus without cerebral embolism: length of symptoms, morphologic characteristics, and outcomes. *Tex Heart Inst J* 2014;41:592–5.
- 6 Arcenas RF, Ali MI. Left atrial myxoma: a rare nonatherosclerotic cause of acute myocardial infarction. *Case Rep Cardiol* 2013;2013:1–4.
- 7 Buksa M, Gerc V, Dilic M, *et al.* Clinical, echocardiographic and echophonocardiographic characteristics of the atrial myxomas in 22 years period. *Med Arh* 2009;63:320–2.
- 8 Walpot J, Shivalkar B, Rodrigues I, *et al.* Atrial myxomas grow faster than we think. *Echocardiography* 2010;27:E128–31.

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