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Stroke due to a cardiac myxoma

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Abstract

Cardioembolic stroke is an important cause of stroke in young people. Cardioembolism classically causes infarcts in multiple vascular territories. In the case described here a young woman developed cardioembolic stroke secondary to atrial myxoma. The presentation was atypical in that the woman had no systemic manifestations of her myxoma.

BACKGROUND

Although jobbing neurologists are mindful of the rarer causes of cardioembolic stroke in young adults, rare diseases may bear none of their “textbook” hallmarks.

CASE PRESENTATION

A woman in her late twenties suddenly became unsteady on her feet with a tendency to stagger to the left. She had a moderate frontal headache at the onset. Over the next hour she developed numbness of the whole of her lower lip, left upper lip and the left side of her face. These symptoms were accompanied by very mild weakness of the right arm, nausea, slight photophobia, oscillopsia, a quiet voice, and a need to gulp twice in order to swallow. Her only past history was of migraine equivalents without headache. There was a family history of migraine affecting her mother, and of stroke affecting her great aunt in her seventies. She had been on the progestogen-only contraceptive pill (norethisterone 350 μg daily) for three years, and was a non-smoker.

On examination, she had fine nystagmus on left lateral gaze, diminished cutaneous sensation over the left cheek, and mild left hemi-ataxia. On standing, she had pronounced truncal ataxia which instantly made her vomit. The rest of the neurological examination was normal. General examination including the skin was normal, heart sounds were pure with no added sounds, and she had a regular pulse of 60 beats/minute with a blood pressure of 120/70 mmHg.
INVESTIGATIONS

The following blood tests were normal: urea and electrolytes, liver function, full blood count, fasting cholesterol and glucose, erythrocyte sedimentation rate, coagulation screen, complement, rheumatoid factor, extractable nuclear antigens, anti-nuclear antibody, anticardiolipin IgG, anti-neutrophil cytoplasmic antibodies, thrombophilia screen, total plasma homocysteine, and C-reactive protein. A 12-lead electrocardiogram revealed normal sinus rhythm, with a rate of 60 beats/minute and a corrected QT interval of 473 ms (reference range 360–430 ms). A chest x-ray was normal. Unenhanced brain computed tomogram (CT) was normal but magnetic resonance imaging (MRI) (fig 1) revealed multiple high signal intensities on T2-weighted and FLAIR in the cerebellar vermis, left middle cerebellar peduncle, medulla, and the right occipital, left frontal and left parietal regions. Some of these defects showed restricted diffusion, and on gradient echo sequences some had evidence of haemosiderin deposition. A transthoracic echocardiogram revealed a dilated left ventricular cavity with mild left ventricular systolic impairment—and a large left atrial myxoma (fig 2; see also http://www.practical-neurology.com for online video).

TREATMENT

She was treated with aspirin (loading dose 300 mg and 75 mg daily thereafter) and her symptoms started to improve within 24 hours. She stopped the contraceptive pill. Twenty five days after her initial presentation a median sternotomy was performed via a horizontal submammary incision. The left atrium was opened on full cardiopulmonary bypass at 34°C with cardioplegic arrest of the heart. The left atrial myxoma was confirmed to be arising from the inter-atrial septum and radical excision including the base was performed (fig 3). The defect in the septum was closed primarily.

OUTCOME AND FOLLOW-UP

She recovered over the following nine months and returned to work part-time, with only a little residual unsteadiness attributable to her initial stroke.

DISCUSSION

Despite the passage of more than one-and-a-half centuries since the first description of cardiac myxomas in 1845, 3 relatively little is known about them. A search of Ovid Medline from 1966 to November 2005 yielded only 136 references (combining the terms “atrial myxoma.tw” and “exp Cerebrovascular accident/OR exp Cerebral Infarction/or exp Cerebral Hemorrhage/or exp Cerebrovascular Disorders/”). Unfortunately, the limitations of case reports, case series and literature reviews preclude reliable statements about, for example, the frequency of constitutional symptoms due to myxomas (which our patient definitely did not have, even after being asked on many occasions).

Primary tumours of the heart are 20–40 times less common than tumours that have metastasised to the heart. The prevalence of primary cardiac tumours in a pooled analysis of autopsy studies was 0.021%. 4 Three quarters of these tumours are benign, and almost half of the benign intracardiac tumours are myxomas. Although classified as benign tumours, myxomas seem to grow and spread, but whether they metastasise or whether fragments simply embolise is debated. 5 Myxomas usually arise from the inter-atrial septum on a short pedicle and grow into the left atrium (fig 2), but they also occur in the right atrium, and occasionally they are bi-atrial.

Myxomas tend to affect women and present between the third and sixth decades of life with at least one of the typical triad of: cardiac failure due to outflow obstruction (mimicking mitral or tricuspid stenosis), cerebral and/or systemic embolism (usually with left atrial myxomas), and constitutional...
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Therefore, cardiac myxomas can mimic infective, malignant and connective tissue diseases by causing symptoms such as malaise, fatigue, weight loss, fever, rash, arthralgia and myalgia.

Stroke appears to be responsible for 80% of the neurological presentations of myxomas, and only 40% of these have the typical pattern of cardio-embolism involving several vascular territories. Myxomas usually cause ischaemic stroke by embolism of tumour or thrombus, but aneurysmal dilatations at sites of earlier embolic vascular occlusion can cause intracerebral or subarachnoid haemorrhage.

Clinical neurologists who use their stethoscopes may suspect a myxoma when they hear a diastolic murmur, split first heart sound or “tumour plop”. Further suspicion may be aroused by blood tests revealing anaemia, polyclonal immunoglobulin proliferation and a raised erythrocyte sedimentation rate and C-reactive protein (but not in this case). The electrocardiogram is usually unhelpful (revealing, if anything, signs of atrial overload or cardiac failure) and the chest x ray may reveal signs of left or right heart failure, but echocardiography is usually diagnostic. This case was easily diagnosed with transthoracic echocardiography but if there is diagnostic uncertainty transoesophageal echocardiography provides superior views of both the inter-atrial septum and the atria.

The anatomy of myxomas makes them relatively easy to excise. First reported in 1955, surgery is usually performed swiftly in an effort to prevent embolic complications, is generally curative, and appears to have a low risk of complications. Patients can be reassured that the risk of recurrent myxoma is 1–3% for sporadic cases. Any recurrence is usually attributable to multifocal myxomas, myxoma embolisation, or incomplete resection. Some advocate regular follow up echocardiography.

LEARNING POINTS

- Brain CT is often normal in posterior circulation ischaemia, necessitating MRI of the brain for confirmation in many cases.
- Infarcts in different vascular territories on brain imaging are a clue to a cardioembolic cause.
- The routine use of bubble/contrast echocardiography in the investigation of ischaemic stroke in the young does occasionally identify abnormalities other than patent foramen ovale.
- Not all atrial myxomas are accompanied by constitutional symptoms, or even a raised erythrocyte sedimentation rate and C-reactive protein.

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Footnotes

Competing interests: None.

REFERENCES


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Figures and Tables

Figure 1

Magnetic resonance imaging of the brain, revealing: a vermian infarct on (A) FLAIR (B) T₂-weighted, and (C) diffusion-weighted imaging; a right occipital infarct on (D) FLAIR and (E) diffusion-weighted imaging; and a left frontal infarct on (F) FLAIR imaging with (G) haemosiderin evident on gradient echo.
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**Figure 2**

Parasternal view of the transthoracic echocardiogram demonstrating a left atrial myxoma (solid arrow), measuring $4.6 \times 2.3$ cm, protruding through the mitral valve, with its stalk attached just behind the anterior leaflet of the mitral valve and below the aortic valve (dashed arrow).

**Figure 3**
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Macroscopic view of the left atrial myxoma.