CASE REPORT

A large left atrial myxoma causing multiple cerebral infarcts

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SUMMARY

A 52-year-old man presented with a history of sudden onset diplopia. On neurological examination, the only abnormality was a right-sided oculomotor (third nerve) palsy. A brain CT was performed and reported as showing no abnormality. He was discharged to be investigated as an outpatient. He presented 1 month later with a new expressive dysphasia and confusional state. MRI was performed which revealed multiple cerebral infarcts. He was discharged on secondary stroke prevention medication. Six months elapsed, before a transthoracic echocardiogram was performed. This showed a large left atrial myxoma. The patient underwent an emergency resection and made a good postoperative recovery. This case report showed the importance of considering a cardiogenic source of emboli in patients who present with cerebral infarcts. Performing echocardiography early will help to detect treatable conditions such as atrial myxoma, and prevent further complications.

BACKGROUND

Cardiac tumours are rare and the majority are benign. Myxomas account for more than half of cardiac tumours. Myxomas are a recognised source of emboli, and can lead to stroke and systemic embolisation. It is potentially treatable, and therefore early detection can prevent complications.

CASE PRESENTATION

A 52-year-old man presented with sudden onset of diplopia. He had no medical history of note.

Neurological examination revealed ptosis of the right eye and impaired adduction with a non-dilated pupil. On auscultation the heart sounds were normal. In particular no diastolic murmur or 'tumour plop' was heard. The rest of the physical examination was unremarkable, with no other neurological deficit.

His blood pressure (BP) was 130/70 and his pulse regular 70 bpm. His ECG showed sinus rhythm.

A diagnosis of partial oculomotor nerve palsy was made. The brain CT was normal. As the patient was not diabetic, autoimmune pathology was considered as a possible cause. The patient was discharged with the plan of reviewing him with the result of his autoimmune profile. A month later he was readmitted with acute confusional state and a new expressive dysphasia. MRI of the brain was performed which revealed multiple ischaemic areas.

The patient was started on secondary stroke prevention medication and discharged. The echocardiogram and carotid Doppler were to be performed

as an outpatient. Unfortunately, there was a significant delay in requesting the echocardiogram.

It was performed 6 months following his first presentation.

This revealed a large left atrial myxoma. The result was immediately brought to the attention of the cardiologists who arranged his urgent transfer to a tertiary centre, where he underwent an emergency resection.

INVESTIGATIONS

On his first presentation, the erythrocyte sedimentation rate was 50 and white cell count and C reactive protein slightly raised. The rest of his routine blood results including blood glucose were normal. His autoimmune profile was normal. ECG, chest X-ray, CT of the head and carotid Doppler were normal.

On his readmission MRI of the brain (figure 1) was performed with and without contrast and this revealed multiple ischaemic areas. There were multiple predominantly cortical lesions in hemispheres as well as the cerebellum which were bright on T2-weighted sequences and dark on T1-weighted sequences without intravenous contrast enhancement. All the lesions showed evidence of restricted diffusion.

The echocardiogram (figure 2A,B), which was performed 6 months following his readmission revealed a large left atrial myxoma which was intermittently obstructing the mitral valve orifice and prolapsing into the left ventricle.

DIFFERENTIAL DIAGNOSIS

- ▶ Thrombus
- ▶ Benign tumours: papillary fibroelastoma, fibroma
- ▶ Malignant primary cardiac tumours: angiosarcoma and rhabdomyosarcoma
- ▶ Normal structures and variants
- ▶ The eustachian valve, chiari network, crista terminalis, pectinate muscles, moderator band, trabeculations, interatrial septal aneurysm and lipomatous hypertrophy of the interatrial septum.

TREATMENT

Following the result of the echocardiogram, he was immediately transferred to a tertiary centre, for an emergency excision of the myxoma. He also required atrial septal defect repair and coronary artery bypass grafting.

OUTCOME AND FOLLOW-UP

The patient made a good postoperative recovery. A year following surgery his speech had returned to

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Figure 1 (A–F) MRI of the brain showing left frontal and parietal infarct (A), bilateral parietal infarct (B), bilateral cerebellar and left temporal infarct (C), infarct in the cerebellar vermis and left temporal lobe (D), occipital infarct (E) and left parietal infarct (F).

normal. The right eye ptosis had resolved, but adduction was still impaired causing persisting diplopia.

DISCUSSION

Primary cardiac tumours are rare, and are found in less than 0.2% of patients at postmortem. The majority of primary cardiac tumours are benign, of which approximately 50% are myxomas. They are more common in women, and occur most frequently between the third and sixth decade of life.¹

As in our case, the most common location of cardiac myxomas is the left atrium. A right atrial location is very rare.^{2–4}

Large myxomas may cause obstruction of the atrioventricular valves and lead to dyspnoea (secondary to impairment of ventricular filling), presyncopal or syncopal episodes and sudden death.^{3 4}

Cardiac manifestation is the most frequent mode of presentation, followed by embolic manifestation. Embolic events occur in 30–40% of patients. In the majority of cases the cerebral arteries are affected leading to ischaemic stroke.^{5–8}

In one review, patients who presented with neurological complications of cardiac myxoma were young and stroke was the most common presentation (83%).⁹

Stroke, associated with myxomas is usually caused by embolisation of tumour or thrombus, and classically involves several vascular territories. However, aneurysm formation and intracerebral or subarachnoid haemorrhage have been reported.^{10 11}

Cerebral infarction and systemic embolism are usually caused by a left-sided myxoma. However, in the presence of an intracardiac shunt they can also be caused by a right-sided myxoma.

Pulmonary embolism may originate from a right-sided myxoma.^{12 13}

Non-specific constitutional symptoms such as weight loss and fever are common.

In a minority of cases (2–10%) patients may be asymptomatic and the myxoma may be discovered as an incidental finding.^{5 14}

On auscultation a diastolic murmur suggestive of significant mitral stenosis may be heard. The classic ‘tumour plop’, an extra heart sound found in patients with prolapsing myxoma is uncommon. In a review of 112 cases, 64% of the patients had a murmur suggestive of mitral valve disease and 15% the tumour plop.⁵

In our case the patient initially presented with an isolated right-sided oculomotor (third nerve) palsy with no cardiac auscultatory findings. Initially a head CT was performed, which did not reveal any abnormalities. Following his readmission with an unexplained confusional state and expressive dysphasia, MRI was performed, which revealed multiple ischaemic areas in both hemispheres.

In suspected posterior fossa lesions, CT has a limited role and MRI is the procedure of choice. However, a head CT was performed in our case, as MRI was not readily available.

Unfortunately there was a significant delay in requesting an echocardiogram. Once the large left atrial myxoma was detected, the patient was transferred to a tertiary centre, where he underwent emergency surgery.

In their 2008 guidelines for Management of Ischaemic Stroke, the European Stroke Organisation (ESO) recommend echocardiography when cardiac source of embolism (eg,

Figure 2 (A) Parasternal long axis view of the transthoracic echocardiogram showing a left atrial myxoma obstructing the mitral valve orifice (marked by arrows). (B) Apical view of the transthoracic echocardiogram showing a left atrial myxoma prolapsing into the left ventricle (marked by arrows). LA, left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle; AV, aortic valve; IVS, interventricular septum.

infarctions in multiple cerebral or systemic arterial territories) is suspected.¹⁵

Two-dimensional echocardiography is an excellent initial diagnostic technique to detect a cardiac mass. It is non-invasive and widely available. It has sensitivity and specificity of 90% and 95%, respectively. In comparison to transthoracic echocardiography, transesophageal echocardiography provides superior image resolution and better visualisation of anatomic details of the tumour, its location, attachments and relation to other anatomical structures. Although TEE appears to be superior to TTE, in many patients it is not performed on a routine basis and should be considered when the transthoracic study is sub-optimal. Doppler echocardiography can show the haemodynamic consequences of atrial myxoma.^{16–18}

Cardiac MRI may provide additional information to that obtained by echocardiography. It can distinguish between tumour and non-tumour masses with sensitivity and specificity of 94% and 91%, respectively, and between benign (tumour or non-tumour) and malignant masses with a sensitivity and specificity of 89% and 100%, respectively.^{19–21}

MRI has a better tissue characterisation than CT and does not have the risk of radiation.

Finally, the possibility of recurrence after surgical resection makes regular follow-up with echocardiography essential. The risk of recurrence ranges between 1% and 3% for sporadic cases, and 12%–22% for familial and complex myxomas, respectively.^{1–5, 22}

Learning points

- ▶ It is important to consider a cardiogenic source in patients who present with stroke.
- ▶ CT of the brain may be normal and MRI may be required to confirm the diagnosis.
- ▶ Multiple infarcts in different vascular territories are a clue to a cardiogenic source.
- ▶ Performing echocardiography early will help detecting treatable conditions such as atrial myxoma, and prevent further complications.

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