CASE REPORT

Atrial myxoma presenting as acute ischaemic stroke and chronic right lower leg claudication

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SUMMARY

A 48-year-old man presented with acute onset of left facial numbness, ataxic gait and double vision. He also complained of chronic right lower leg pain with acute onset a year prior to presentation. His vital signs were within normal limits. Physical exam was notable for right-sided intranuclear opthalmoplegia, decreased sensation to light touch on the left side of his body, left-sided dysmetria and ataxic gait. Neuroimaging showed evidence of acute stroke in the cerebellum and brainstem, for which he was treated with thrombolytics. An echocardiogram revealed a 5×3 cm left atrial myxoma, which was surgically resected. Subsequent imaging of his lower extremity revealed a chronic common iliac artery occlusion for which he underwent angioplasty. His claudication symptoms resolved, and he was without any neurological deficits at a 2-year follow-up visit.

BACKGROUND

Cardiac myxoma is an endocardial-based neoplasm of uncertain histological origin that represents the most common primary cardiac tumour. It has an estimated annual incidence of approximately 0.5 cases per million with a female predominance and presentation typically in the fourth to sixth decade.[1] Patients typically present with at least one from the classic triad of signs consisting of heart failure, tumour emboli and constitutional symptoms.[2]

Here we present a case of acute stroke from myxoma embolisation in a patient with symptoms of lower extremity claudication for a year prior to presentation that were attributed to chronic lumbar radiculopathy.

CASE PRESENTATION

A 48-year-old man with a medical history of lumbar disc herniation and chronic right lower extremity pain presents to the emergency department with acute onset of left facial numbness, ataxic gait and double vision. His vital signs were within normal limits; however, his physical examination revealed right-sided internuclear opthalmoplegia, decreased sensation to light touch on the left side of his body, ataxia on finger-to-nose on the left and ataxic gait. On further questioning, he complained of acute right lower leg pain after running 1 year prior, which had been attributed to lumbar radiculopathy. He has no relevant family or social history.

A CT scan of his brain revealed no acute findings. After careful discussion with the patient, he was treated with intravenous tissue plasminogen activator. The following day an MRI of his brain showed multiple areas of left cerebellar and right paramedian mid-brain defects consistent with acute stroke (figure 1). As part of his stroke work-up, the patient underwent a transthoracic echocardiogram (TTE) that revealed a 5×3 cm mobile mass in the left atrium that prolapsed through the mitral valve, which was most consistent with left atrial myxoma (figure 2). The patient had urgent cardiac surgery consultation and underwent a coronary angiogram the same day that revealed patent coronary arteries. The next day the patient was taken to the operating room for resection of a 6.1×4.4×3.5 cm maroon to tan-white, rubbery to gelatinous mass. Surgical pathology confirmed atrial myxoma. His operation was successful and he was taken off cardiopulmonary bypass easily. However, on arrival to the intensive care unit, the nursing staff was unable to obtain Doppler pulses in his right foot. On physical

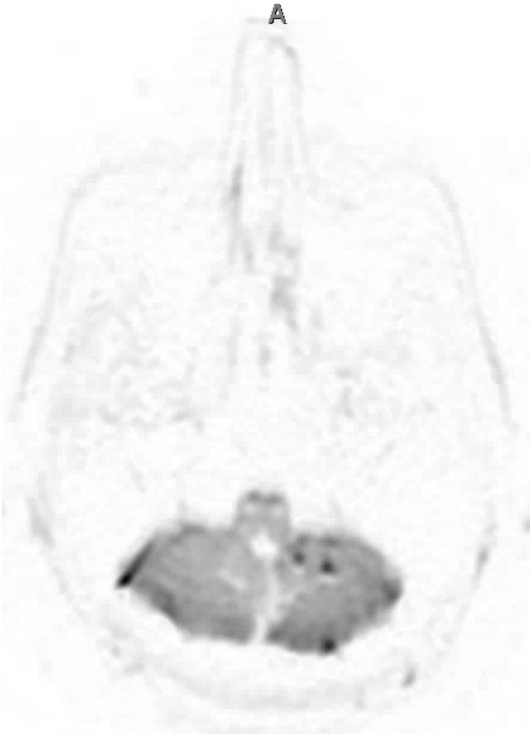

Figure 1 MRI of the brain revealing multiple areas of left cerebellar and right paramedian mid-brain defects consistent with acute stroke. A, anterior; V, view of ultrasound of probe.

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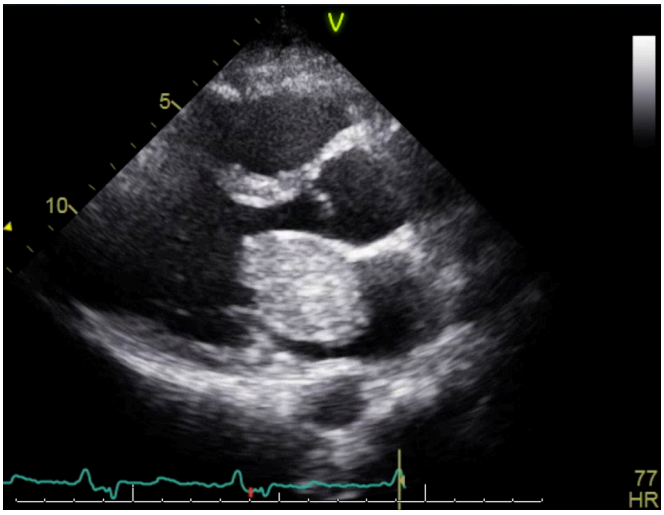

Figure 2 TTE revealing a 5×3 cm mobile mass in the left atrium consistent with atrial myxoma.

exam his right femoral artery pulse was diminished with absent right pedal pulse. An ultrasound Doppler study of the right leg showed monophasic waveforms throughout the right lower leg. His ankle brachial index (ABI) was 0.4 on the right leg and 1.0 on the left leg. A subsequent CT angiogram found a chronic right common iliac artery occlusion (figure 3). The patient recovered neurologically from his stroke and was discharged home with outpatient vascular surgery follow-up. His discharge medications included aspirin 81 mg and atorvastatin 40 mg daily.

Due to persistent right leg claudication, he underwent peripheral angiogram 2 months later with successful angioplasty of a chronically occluded right common iliac artery (figure 4A,B).

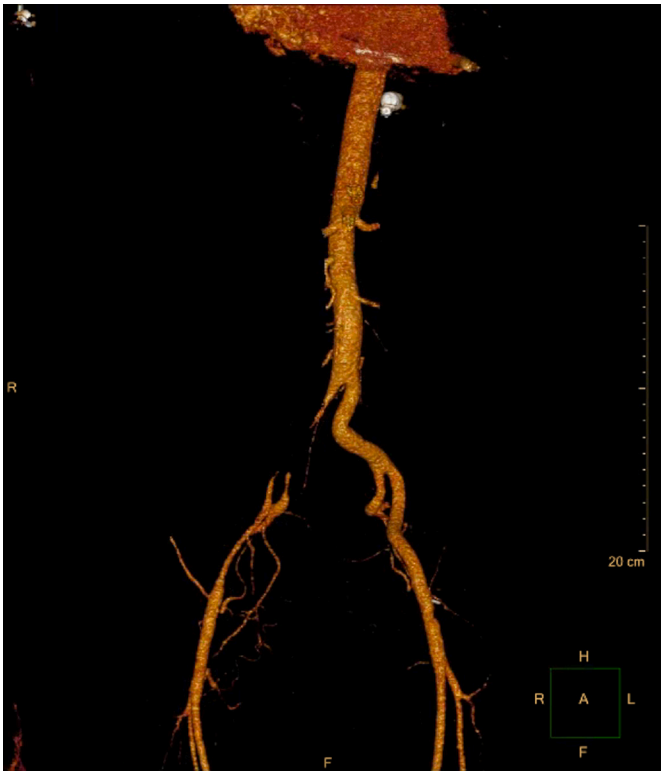

Figure 3 CT angiogram of the aorta revealing a chronically occluded right common iliac artery.

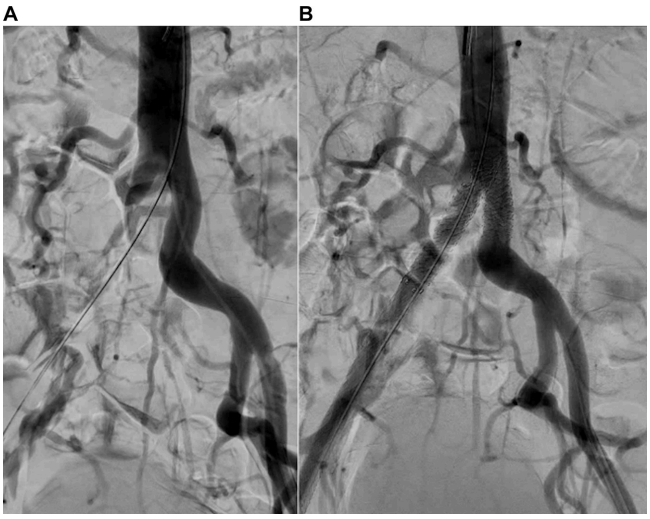

Figure 4 Intraoperative aortogram before (A) and after (B) successful angioplasty and stenting of the right common iliac artery.

OUTCOME AND FOLLOW-UP

The patient had complete resolution of his claudication symptoms. He has had normal ABI in both legs after 2-year follow-up. He also remains asymptomatic from a neurological standpoint.

DISCUSSION

Cardiac myxoma is an endocardial-based neoplasm of uncertain histological origin that represents the most common primary cardiac tumour. It has an estimated annual incidence of approximately 0.5 cases per million with a female predominance and presentation typically in the fourth to sixth decade.[1] Approximately 60%–80% of myxomas arise within the left atrium near the fosae ovalis border but may be diagnosed in the other chambers as well.[2] Macroscopically, they appear as polyploid myxomas, which are compact, solid tumours and less frequently as the softer, gelatinous papillary myxomas prone to embolisation. A classic triad of presentation consists of heart failure from valvular obstruction, tumour emboli and constitutional symptoms.[3] Diagnosis can be made confidently by echocardiography and confirmed by pathological examination after surgical resection.

Distant embolism is only seen in 30%–40% of patients with the majority being to the central nervous system with the lower extremities being the next common anatomical site.[2] In our patient, stroke work-up revealed no evidence of carotid artery stenosis and no atrial fibrillation. Although no pathology specimen was obtained, his neurological deficits resolved following thrombolytic therapy. To the best of our knowledge, the role of thrombolytic therapy in acute stroke from myxoma embolism is unclear; the predisposition of embolised cells to invade cerebral vasculature forming 'pseudo-aneurysms' carries an increased risk for cerebral haemorrhage.[4] Moreover, a myxoma may embolise associated with thrombus or as just tumour tissue itself, less amenable to thrombolytic therapy. Although only a few case reports are available, the rate of intracranial haemorrhage was 1/7 for all cases of thrombolysis in patients with embolic stroke from a myxoma.[5] Since myxomas are rarely diagnosed at the time of presentation from a stroke, our case further adds to current literature in suggesting that thrombolytic therapy may be a safe and effective option even before a definitive diagnosis is made.

Notably, our patient also experienced distant embolism with a CT angiogram showing a right common iliac artery occlusion. Although there are several case reports of acute lower extremity ischaemia from myxoma embolism, this is the first case of claudication from chronic occlusion in a patient without other risk factors for peripheral vascular disease. Whereas prior case reports have discussed surgical embolectomy with relief of acute ischaemia, our patient recovered to baseline with endovascular recanalisation.[6]

Prompt surgical removal is warranted with no data to support recurrence following surgery with negative margins.[7] However, the optimal time for myxoma resection following an embolic stroke is still unclear with the competing risks of anticoagulation and cardiopulmonary bypass during surgery balanced with the incidence of further embolic events. A review of the literature has shown good outcomes with both emergent surgery and delayed surgery weeks after the stroke.[8] In our patient, the small foci of infarction informed the decision to proceed with surgery using full perioperative anticoagulation and reduce the risk of further tumour embolisation. In patients at higher risk of haemorrhagic transformation based on their tumour burden, preoperative imaging may be beneficial to allow further risk-stratification by identifying early reperfusion haemorrhages.[9]

In this case, the patient's worsening lower extremity pain over an extended time period had been attributed to his known lumbar radiculopathy, leading the clinician away from a wider differential including an embolic event. A higher index of suspicion and earlier diagnostic work-up may have prevented subsequent cerebral embolism.

Learning points

- The classic triad of cardiac myxoma includes symptoms of acute heart failure, central nervous system or peripheral emboli, and constitutional symptoms although all three may not be apparent at time of presentation.
- Diagnosis is via echocardiography and surgical resection is curative.
- The optimal timing of surgical resection after cerebral embolism is not well established; the risk of haemorrhagic transformation of an ischaemic stroke must be weighed against the risk of further embolic events on an individual basis by a multidisciplinary team.
- The use of thrombolytic in an acute stroke secondary to myxoma embolisation has not resulted in increasing intracranial haemorrhage despite the propensity of myxomas to form cerebral pseudo-aneurysms prone to bleeding.

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