Recurrent Stroke in a Case of Left Atrial Myxoma Masquerading Vasculitis


Abstract
We report a case of left atrial myxoma presenting as recurrent neurological deficits with absent peripheral pulses mimicking systemic vasculitis. Due to absence of cardiac signs and symptoms, there was one year delay in diagnosis from initial symptom. Left atrial myxomas are rare but treatable cause of recurrent stroke.

Introduction
Atrial myxomas are rare cause of ischaemic stroke and seen most commonly in 3rd to 6th decade.1 The embolisation of particles or thrombotic material covered with tumour cells occurs in about 30 to 45 per cent in atrial myxoma patients. In nearly half of the cases cerebral arteries are affected leading to embolic stroke.2 Other rare neurologic complications include parenchymal brain metastases and intracerebral haemorrhage due to ruptured aneurysms.3 We report a case of a young male presenting with recurrent strokes and multiple peripheral arterial occlusions mimicking vasculitis.

Case History
A 30 year old male presented with history of sudden onset staggering gait and slurring of speech four days prior to admission. It was not associated with complaints of vertigo, double vision, weakness or sensory symptoms on either side of the body. He had maximum deficit at the onset and subsequently had good recovery and over next few days he was able to walk unaided.

He had past history of two episodes of neurological illness. First episode occurred one year back, when he had sudden onset right lower limb weakness which recovered over next two weeks. Second episode occurred four months back when he had acute right sided weakness with slurring of speech followed by fast recovery over next two days. For both the above episodes he didn’t seek any medical advice. There was no history of headache, recurrent vomiting or seizure. No associated complaints of fever, weight loss, joint pains, rash or photosensitivity were present. He did not have any history of palpitation, syncope, dyspnoea, hypertension, valvular heart disease or diabetes. He smoked 10 cigarettes daily for last 10 years and had no other addiction.

On examination his left radial artery, both dorsalis pedis and left posterior tibial arteries were not palpable; left brachial pulsations were weak. There was bluish discolouration and reduced temperature distally in left upper extremity. Both carotids and femoral arteries were normal. No significant difference of blood pressure was found in upper and lower limbs. Cardiac examination did not reveal any abnormality. He had mild cerebellar dysarthria and gaze evoked horizontal nystagmus. There was minimal right sided weakness, extensor plantar response, bilateral brisk deep tendon reflexes, mild in-coordination on left side and impaired tandem gait.
His haemoglobin was 10.2 gm%, ESR 35 mm and CRP was positive. Rheumatoid Factor, ANA and dsDNA, HIV, and VDRL were negative. ECG, chest X-ray and urine examination were normal. MRI brain showed multiple bilateral acute as well as chronic cortical and cerebellar infarcts (Figure 1).

In view of absence of multiple arterial pulsations, normal cardiac examination, chest X-ray and ECG, diagnosis of Takayasu arteritis, polyarteritis nodosa, prothrombotic state and systemic lupus erythematosus were considered and whole body CT angiography, 2-D echo and prothrombotic profile were planned.

CT angiography revealed thrombotic occlusion of left distal brachial, radial and ulnar artery (5 cm long). Distal radial and ulnar arteries were refilling through collaterals. There was near complete occlusion of bilateral tibio-peroneal trunk and proximal anterior tibial and peroneal arteries (4.8 cm long segment). Distal anterior tibial and peroneal arteries were refilling through collaterals bilaterally. The aortic arch and its major branch vessels, abdominal aorta, renal vessels, bilateral subclavian and axillary arteries were normal (Figure 2). It also revealed presence of filling defect in the left atrium, possibly a large saddle shaped thrombus (Figure 3). Trans-thoracic 2D-Echo (TTE) showed a mobile, homogeneous mass 39 x 25 mm in size attached to inter-atrial septum by thick pedicle (17 mm) at left aspect of inter-atrial septum prolapsing through mitral valve in diastole, suggestive of left atrial myxoma (Figure 4). Patient was put on anticoagulants and advised immediate surgical resection, the patient underwent surgery after 1 month of diagnosis, the histopathology report showed presence of stellate cells in a myxoid
background with scattered dilated congested blood vessels and chronic inflammatory cells, confirming the diagnosis of myxoma (Figure 5).

**Discussion**

This young patient of atrial myxoma presented with recurrent neurological events, absent peripheral pulses, raised ESR and positive CRP, simulating vasculitis without obstructive cardiac symptoms and signs.

Myxomas may be asymptomatic in about 10 to 20 per cent of the cases and discovered as an incidental finding on echocardiography. About 30 to 40 per cent of myxomas may embolise. Neurologic symptoms have been reported in 26 to 45 per cent of patients. Atrial myxomas can cause up to 0.5 per cent of ischaemic strokes. Embolisation most frequently presents with cerebral infarcts which may be recurrent as in our case, followed by TIA and seizure. Our findings are in collaboration with another series of 74 patients with cardiac myxoma, in which 9 (12%) patients presented with neurologic manifestations, out of which ischaemic cerebral infarct was the most common presentation (89%). No patients had associated cardiac symptoms. Cerebral aneurysm and myxomatous metastasis were the other complications noted which can mimic vasculitis or endocarditis. In another analysis of 113 cases of atrial myxoma with neurologic presentation, 83% of patients presented with ischaemic stroke, most often at multiple sites (41%). The other presentations included syncope (28%), psychiatric symptoms (23%), headache (15%) and seizures (12%).

Systemic emboli are frequent in myxomas located in the left atrium. Embolisation may occlude peripheral arteries as well as visceral, renal and coronary artery. Rarely, complete occlusion of abdominal aorta by tumour emboli has also been reported. Occlusion of retinal arteries may lead to temporary or permanent visual loss. In our case his left radial, both dorsalis pedis and left posterior tibial pulsations were absent clinically. In addition CT Angiography demonstrated occlusion of several other arteries including left distal brachial, left radial, left ulnar and bilateral tibio-peroneal arteries. The differential diagnosis of peripheral arterial embolism should therefore include left atrial myxoma. Right atrial myxomas may embolise to pulmonary vessels leading to pulmonary hypertension and lethal outcome.

Obstructive cardiac symptoms are most commonly related to mitral valve obstruction and can manifest as dizziness, palpitations, dyspnoea, syncope and congestive heart failure. These may be sudden in onset and positional in nature, owing to the effects of gravity on position of tumour. A characteristic low-pitched sound, referred to as a “tumour plop,” may be appreciated on auscultation. But there was lack of these obstructive symptoms in our patient and he did not show any auscultatory abnormalities. In about one-third of these patients auscultation may be normal. A murmur suggestive of mitral stenosis has been reported in only about half of patients.

There was evidence of anaemia, raised ESR with positive CRP in our patient, however significant constitutional symptoms were lacking. Constitutional signs and symptoms like fever, weight loss, cachexia, malaise, arthralgias, rash, and Raynaud’s phenomenon are present in about half of these patients and they may also have increased globulins, anaemia, elevated
ESR, thrombocytopenia, or thrombocytosis. Because of these systemic constitutional features, patients with myxomas are frequently misdiagnosed as having vasculitis, endocarditis, collagen vascular disease, or paraneoplastic syndromes.1

The absences of obstructive and constitutional symptoms with lack of auscultatory findings along with normal chest X-Ray and ECG are the factors responsible for a delay in diagnosis in our patient. A median delay of 36 months in diagnosing myxoma has been reported in patients with neurological manifestations or peripheral embolisation.1

Two dimensional echocardiography for cardiac chambers and CT angiography for thoracic and peripheral vessels were diagnostic in the present case.

Transathoracic echocardiography (TTE) has a sensitivity of around 90 per cent,10 whereas Transoesophageal echocardiography has a sensitivity of nearly 100 per cent.9 Transathoracic echocardiography (TEE) is the method of choice for the detection and description of the size, shape, location, attachment to the cardiac wall and movement of the myxoma, while TEE should be considered when TTE fails to detect the pathology and clinical suspicion is high, as it can pick up even smaller (1-3 mm in diameter) vegetations or tumour.1 TEE may also improve the detection of other major cardio-embolic sources (e.g., intra-cardiac thrombus, vegetation or aortic arch plaque), as well as less common potential sources (e.g., patent foramen ovale, atrial septal aneurysm or left ventricular aneurysm).1

It is important to differentiate atrial myxoma from thrombi on 2D-Echo, which can be done on the basis of size, origin, mobility and prolapse.11,12 Presence of characteristic narrow stalk is the most important distinguishing feature followed by tumour mobility and distensibility.13,14 Other than this thrombus is usually situated in the posterior portion of the atrium and it has a layered appearance.15 Thrombi usually occur in patients with regional or global wall abnormalities (such as myocardial infarction and dilated cardiomyopathy) or atrial fibrillation, valvular heart disease especially enlarged left atrium with mitral valve disease.21 In the present case there was no cardiomegaly or atrial fibrillation which also favours our diagnosis of myxoma. Cardiac MRI may further delineate tumour size, attachment and mobility.1

Surgical excision of atrial myxoma gives excellent short term and long term results. Surgery should not be deferred even in asymptomatic cases. Resection may also lead to resolution of constitutional symptoms as well.1

Conclusion

Left atrial myxoma is a rare treatable cause of recurrent ischaemic stroke in young and clinical picture may mimic vasculitis. Early diagnosis needs high index of suspicion and can prevent significant morbidity and mortality.

References