CASE REPORT

Atrial Myxoma Causing Stroke

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Abstract

Myxoma, though rare but most common among primary cardiac tumors, with the age of onset usually between 30 and 60 years, is more common in females.[1] On one hand atrial myxoma frequently comprises a diagnostic triad of obstructive symptoms, constitutional symptoms, and embolic phenomena. On the other hand, it can be asymptomatic or can present with nonspecific systemic symptoms that may be overlooked. Thereby, cardiac investigations may not be considered, and diagnosis of this rare condition is often missed so that it may present later with more significant embolic disease, such as stroke with neurological deficits, as in the case reported here. A healthy 50-year-old lady developed left hemiparesis with dysarthria. Imaging showed right middle cerebral artery (MCA) territory infarct. Cardiac evaluation revealed left atrial myxoma. The possible mechanism of stroke in this patient is discussed.

Keywords: Cardiac magnetic resonance imaging, Left atrial myxoma, Stroke.

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Case Vignette

A 50-year-old lady presented with sudden onset transient giddiness and fall followed by weakness of the left upper limb and left lower limb with facial droop on the left side and slurring of speech. The physical examination was restricted by the patient's confusional state, but it revealed weakness of the left arm and left leg with upper motor neuron type of facial weakness on the left side, dysarthria, and gaze preference to the right side. She had no history of hypertension, diabetes, or dyslipidemia. There was no history suggestive of recurrent syncopal attacks or transient ischemic attacks, joint pains or myalgia, weight loss, or concomitant cardiac disease in the past.

As the patient presented within 3.5 hours duration from the onset of symptoms, which was within the window period for stroke thrombolysis, and since she had no contraindications for thrombolysis with an NIHSS score of 18, she was initially treated with intravenous alteplase infusion.

Her lab investigations showed mild hypochromic microcytic anemia (Hb = 9.2 mg%) and ESR was 18 mm/hour with other blood reports being normal. Her initial computed tomography (CT) was normal but the post-thrombolysis CT showed an acute infarct in the right frontotemporal and capsuloganglionic region with mass effect over the ipsilateral lateral ventricle. Magnetic resonance imaging (MRI) brain showed acute infarct with hemorrhagic transformation in the right MCA territory (middle and inferior frontal gyrus, superior temporal gyrus, capsuloganglionic region, postcentral gyrus, corona radiate on the right side) with midline shift of 5 mm, and mass effect over the body of the right ventricle (Figs 1 and 2). Though her ECG was normal, transthoracic two-dimensional echocardiogram showed a hypermobile left atrial mass of 4 × 3.3 cm size, attached to the interatrial septum and anterior mitral leaflet, prolapsing into the left ventricle suggestive of myxoma versus thrombus. She was treated with heparin injections. Cardiac MRI showed 2.4 × 2.7 × 3.5 cm pedunculated T1 isointense, T2 isointense to the hyperintense lesion with lobular contour noted in the left atrium, possibly arising from the interatrial septum that prolapsed through the mitral valve

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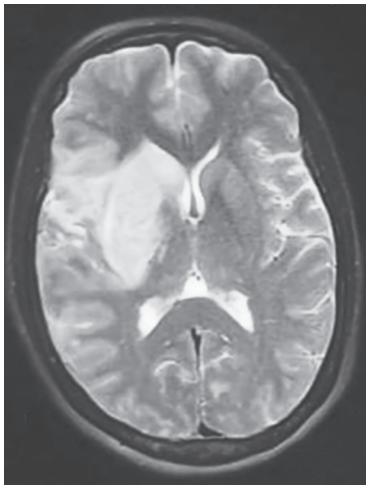


Fig. 1: T2 hyperintensities with foci of blooming in the right capsule-ganglionic region

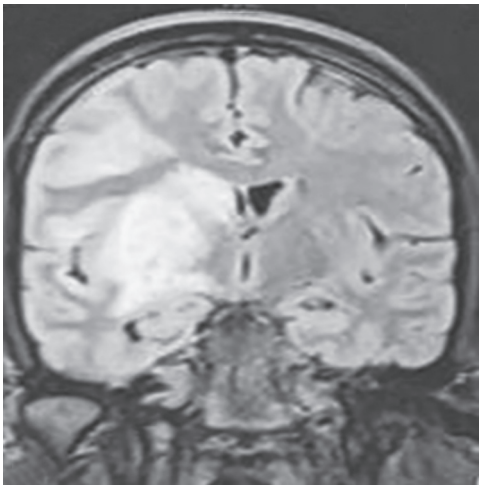


Fig. 2: FLAIR hyperintensities with foci of blooming in the right capsule-ganglionic region

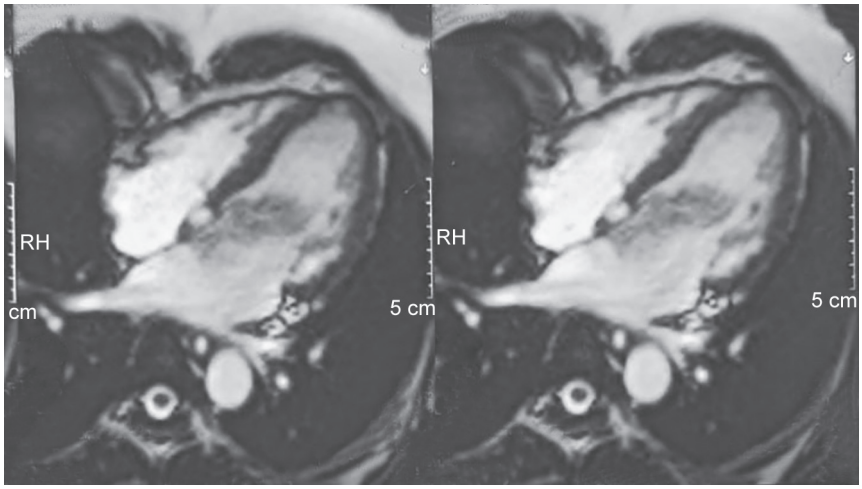


Fig. 3: Cardiac T2w MRI showing left atrial myxoma

during diastole. The lesion showed subtle enhancement suggestive of left atrial myxoma (Figs 3 and 4).

A cardiothoracic consultation was taken and excision of atrial myxoma was planned for a later date. At the time of discharge, the patient's neurological complaints improved fully.

Discussion

About 14–30% of ischemic strokes are of cardioembolic in origin.[2] The major sources of cerebral emboli are acute myocardial infarction, atrial fibrillation, cardiac valvular disease, infective endocarditis, and cardiac myxoma. Cardiac myxoma, though uncommon, is an important treatable etiology for stroke in the young, but the diagnosis of cardiac myxoma is often elusive in stroke patients.[3] Morphologically, cardiac myxoma may have an irregular surface with a soft consistency or with a smooth surface with a compact consistency. The embolic potential is determined by the mobility and lobulated morphology, but not the size, of cardiac myxoma.[4]

Although, atrial myxoma is mostly sporadic and located in the left atrium.[5] Atleast 7% of cases are familial and described as Carney complex, which is caused by an autosomal dominant mutation of the *PRKAR1*α gene located on chromosome 17q.

Clinically, cardiac myxoma can present with varied symptoms which comprises a diagnostic triad (Flowchart 1). Neurologic complications have been documented in 26–45% of atrial myxomas, the most frequent being cerebral infarcts (83%),[2] caused by detached thrombus from the myxoma. Constitutional symptoms are often associated with elevated erythrocyte sedimentation rate and C-reactive protein. The inflammation is thought to be mediated by IL-6 produced from the tumor. Although transthoracic echocardiography is usually done for the screening of a cardiac etiology in patients with stroke, transesophageal echocardiography provides better results with 100% sensitivity.[6] Attention must be given to differentiate between intracardiac tumors and thrombus, which can be done by using cardiac MRI which can further delineate tumor size, attachment, and mobility.

Cardiac myxoma can be treated successfully with surgical resection with 1–3% of recurrence rate and perioperative mortality rate found to be less than 1%. Incomplete removal, intraoperative

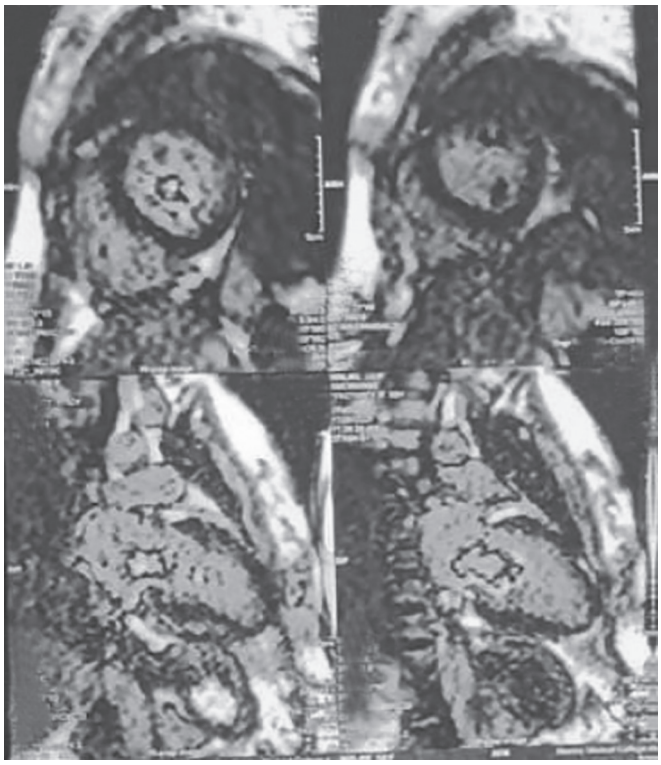
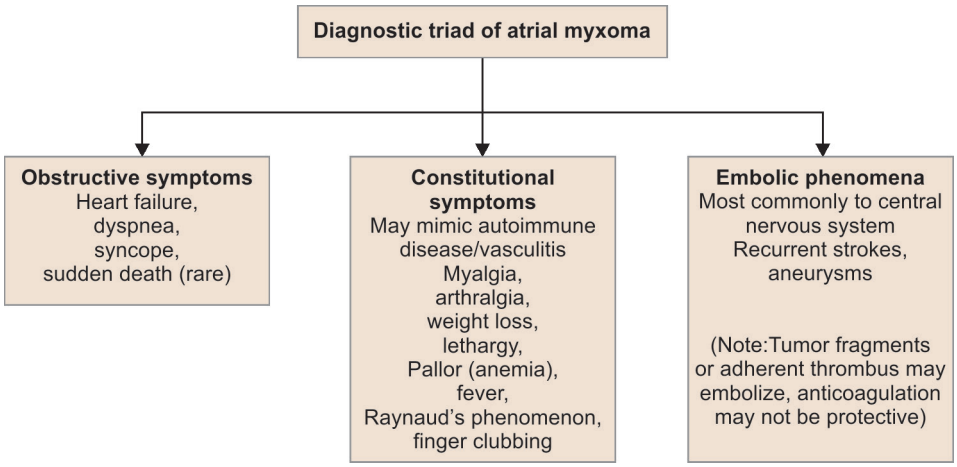


Fig. 4: T1 cardiac MRI showing hypointense lesion in left atrium

Flowchart 1: Diagnostic triad of atrial myxoma



embolization, and multifocal sites are thought to be causes of the recurrence.

Conclusion

Cardiac myxoma can be asymptomatic and may present with mild constitutional symptoms or cardiac obstruction and cerebral embolism, which can be severe and life-threatening. Neurological complications can be the presenting manifestations of cardiac myxoma, especially in young patients. Early diagnosis is paramount to prevent further episodes and other complications. Though echocardiography is the initial imaging modality of choice, an additional cardiac MRI is also helpful in differentiating it from other mimickers. Though excellent prognosis is assured by surgical removal, recurrence can still happen in a small number of cases. Therefore, it is advisable for long-term follow-up in these patients.

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