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Atrial Myxoma – An unusual cause of ischemic stroke in young

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ABSTRACT

Atrial myxomas are rare primary cardiac tumours with neurological manifestations being reported in 30% of cases. Though a rare cause of ischemic stroke in young patients, considering it as a possibility in absence of any obvious risk factors can help avoid misdiagnosis at early stages. We present a case of left atrial myxoma in a 36-year-old male with no known co-morbidities, showing an unusual clinical presentation of isolated bilateral painless vision loss. With multiple infarcts on Non Contrast Computerised Tomography (NCCT) and a suspicion of Atrial Myxoma on Transesophageal Echocardiography (TEE), patient was successfully managed surgically with confirmation of diagnosis on histopathology.

Keywords - Myxoma; Stroke; Thrombosis

INTRODUCTION

Primary cardiac tumors are found in 0.1% of total cases on autopsy,[1] represented mostly by the atrial myxoma. Myxoma is considered a "benign tumor" that arises in any of the cardiac chambers and can be uni or bilateral, though 75% occurs in the left atrium.[2] They have a spectrum of clinical presentations, with 30% cases presenting neurological signs, ischemic stroke being the most common.[3] Atrial myxoma accounts for only <1% cases of ischemic stroke in the young. However, it should be considered a possibility in the absence of any known risk factors to avoid misdiagnosis at early stages.[4] Transesophageal echocardiography (TEE) is considered the best diagnostic modality, and surgical excision remains the mainstay of treatment.[5]

We present an unusual clinical presentation of isolated bilateral loss of vision in a young male with left atrial myxoma, where a diagnosis of left ventricular thrombus was favored over atrial myxoma on cardiac magnetic resonance imaging) (MRI).

CASE REPORT

A 36-year-old previously healthy male presented to our hospital with sudden onset bilateral, painless loss of vision of 12 hours duration along with left-sided continuous headache of moderate to severe intensity and one episode of vomiting. 10 hours prior to the vision loss, he also had one episode of exertional pre-syncope after running a 5-km circuit. There were no other relevant past or family history.

On initial examination, the patient was well oriented, had stable vitals with a regular pulse of 60/min, blood pressure of 128/86 mmHg, and respiratory rate 14/min with SpO2 of 98%. His neurological examination was essentially normal. However, his visual acuity was < 6/60 though bilateral pupils were equally reactive to light with a normal fundus on examination. Cardiovascular examination was normal. ECG revealed ST depression in inferior leads with bradycardia.

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Non-contrasted computed tomography (NCCT) of the brain revealed an acute ischemic infarct in left Middle Cerebral Artery (MCA) -Posterior cerebral artery (PCA) watershed areas and distal MCA region along with multiple old lacunar infarcts (Figure 1A). His biochemical parameters remained normal except for a rise in ESR (Erythrocyte Sedimentation Rate, Normal Value ≤ 15 mm/hr) to 30 mm/hr within 3 days. The patient was managed as a case of Stroke in Young. As multiple infarcts on NCCT indicated a thromboembolic phenomenon, a TEE was done to rule out any cardiac cause. On TEE, a left atrial (LA) mass measuring 2.93 cm^2, mobile, attached to the interatrial septum (IAS) near the Inferior Vena Cava (IVC) junction was noted with suspicion of atrial myxoma (Figure 1B).

The ejection fraction was 65%. However, a cardiac MRI done subsequently favored a diagnosis of LA thrombus over myxoma as no stalk was demonstrated. After that, the patient was started on IV Heparin and was taken up for surgery 10 days later. Intraoperatively, a 3cm x 2cm pedunculated LA mass with rough villous surface and a calcified stalk arising from IAS, was noted. He underwent excision of the left atrial mass with autologous pericardial patch closure of the septal defect through the bi-atrial approach. The diagnosis of atrial myxoma was confirmed on histopathologic examination with characteristic features of stellate shaped myxoma cells embedded in a myxoid background (Figure 2). The post-operative period was uneventful.

DISCUSSION

Atrial myxoma is the commonest of the otherwise rare primary cardiac tumors. It is usually seen between the third and sixth decades of life with 2: 1 female preponderance and left atrium being the commonest site accounting for 75% of the cases.[1,2,6] Though mostly sporadic, a familial association has been noted in 7% of the cases.[3,6]Recurrences are reported, and malignant transformation remains a controversial issue.[2]

Clinical presentation of atrial myxoma often comprises a diagnostic triad of symptoms due to the obstruction of the cardiac outflow or due to embolism or constitutional symptoms.[2-4,6]Due to a myriad of presentations, diagnosis can be missed in a great number of cases at early stages, increasing the risk of morbidity and mortality. Clinical signs or echocardiography abnormalities may be absent in up to 36% of the cases,[6] as was noted in our case. Thus, the presence of an embolic phenomenon, especially in young patients with neurological symptoms, should prompt early neuroimaging and echocardiography.[6]

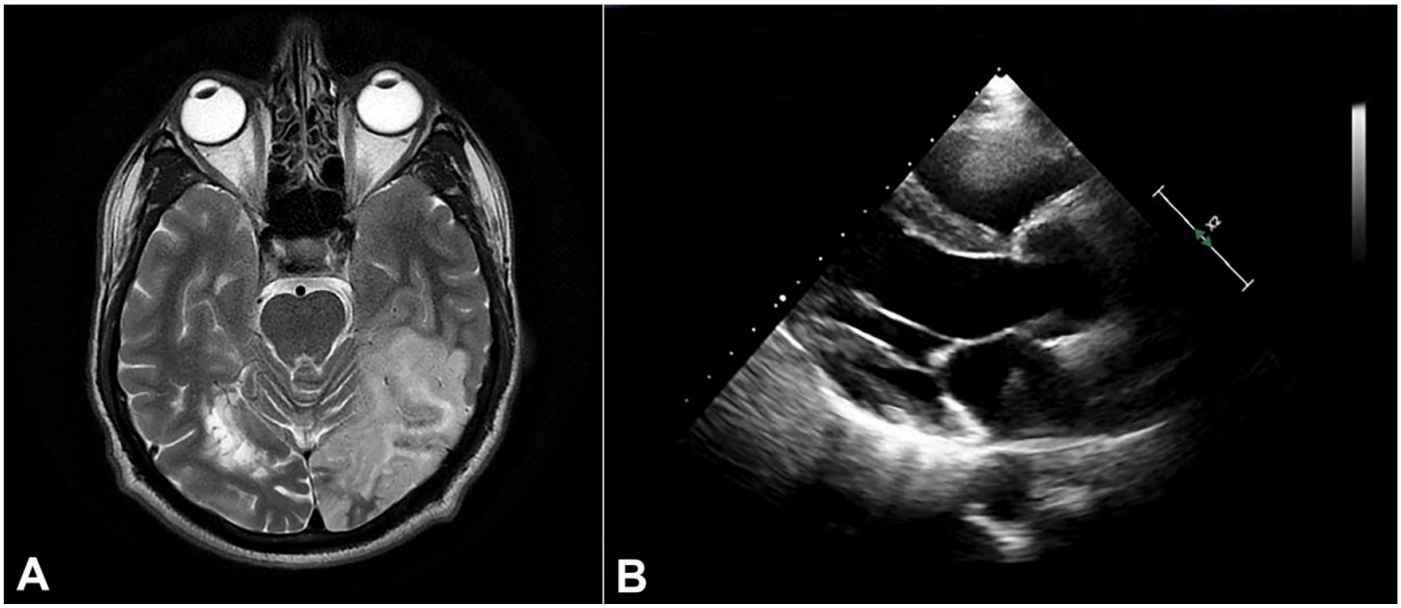

Figure 1. A – NCCT brain showing an acute wedge-shaped infarct in left hemisphere, **B –** Transesophageal echocardiography (TEE) showing a left atrial mass arising from Interatrial septum.

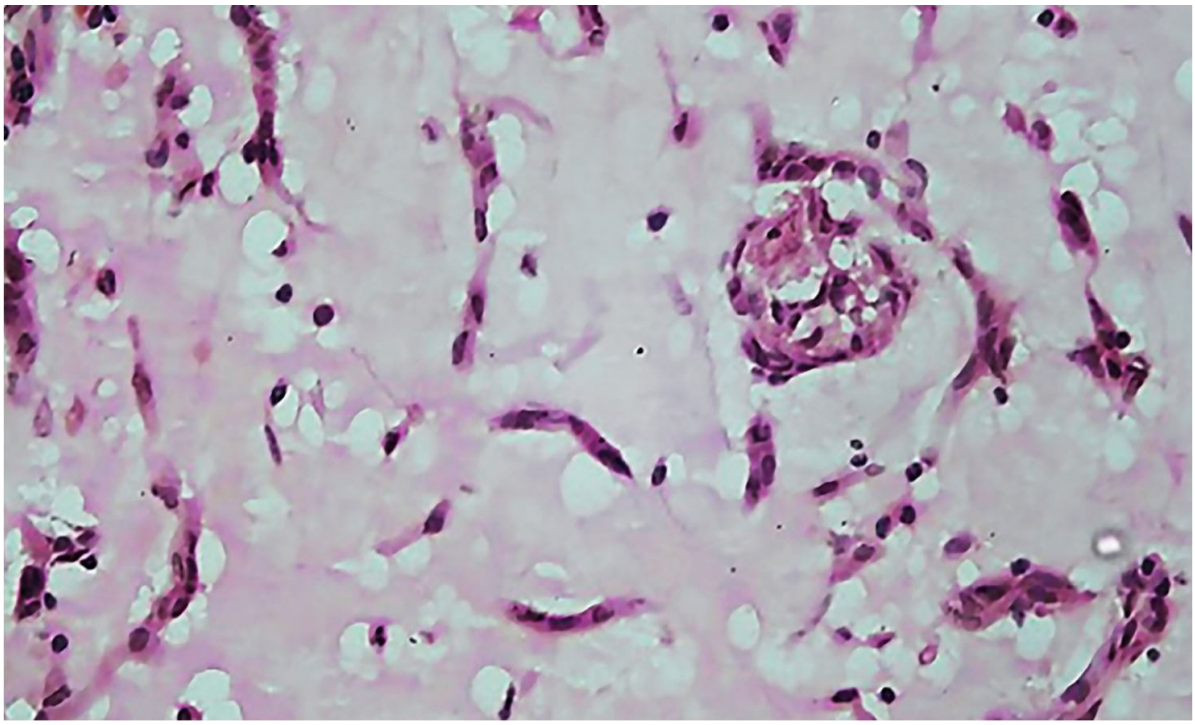

Figure 2. Photomicrograph of the surgical specimen showing the microscopic features of atrial myoma: Stellate shaped myxoma cells embedded in a myxoid stroma.

The ultrastructural analysis and immunohistochemical investigation suggest that atrial myxoma is more likely derived from a pluripotent mesenchymal stem cell or sub-endothelial cell.[7] Angiocardiography was the first diagnostic modality introduced for cardiac tumors, which led to the antemortem diagnosis of atrial myxomas, as prior to which, cases were diagnosed only on autopsy.[2] At present, TEE is the best diagnostic modality for diagnosis of atrial myxomas.[5] Cardiac MRI is useful in determining the tumor size, attachment, and mobility, which helps the surgical management and planning.[6] In cases of ischemic stroke caused by a left atrial myxoma, thrombolytic treatment is considered a safe option of initial management. However, the extent of anticoagulation therapy should be carefully monitored, as hemorrhagic transformation might alter the timing of definitive surgery. In this setting, a delay can increase the overall morbidity and mortality.[4]

Histopathology is confirmatory for benign atrial myxoma. Few studies,[8,9] which attempted classifying myxomas based on myxoma cell population and differentiation, reported that all recurrences were observed in patients with active myxoma with poor differentiation. Recurrences are reported in 1-3% of cases, primarily due to inadequate surgical resection.[10] Annual follow up with echocardiography is suggested for a period of 3 to 4 years, especially in sporadic cases.[11]

CONCLUSION

Albeit rare, considering a differential diagnosis of atrial myxoma in cases of stroke in young, evaluation by early imaging and prompt treatment can help decrease morbidity and mortality resulting from the associated thromboembolic phenomenon.

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