Case Report

Cerebral aneurysms in atrial myxoma: a delayed, rare manifestation


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Atrial myxomas are the most common primary tumors of the heart. Neurologic involvement usually occurs as a stroke with ischemic episodes. Following excision of cardiac myxomas, delayed neurologic events owing to aneurysms are rare and have not been reported from India. We report an operated case of left atrial myxoma. The patient initially presented with a stroke and 6 months after the surgery, developed multiple intracerebral hemorrhages due to the rupture of fusiform cerebral aneurysms, without recurrence of the cardiac tumor.

Key Words: Cardiac myxoma, cerebral aneurysm, intracerebral hemorrhage

Atrial myxomas are the most common primary tumors of the heart. At least one-fourth of the patients with left atrial myxoma develop a neurologic event suggestive of ischemia secondary to embolism.[1] Associated with cardiac myxomas, intracerebral hemorrhages can also occur due to rupture of cerebral aneurysms (termed ‘myxomatous aneurysms’ by some),[11] although this is a rarity and its pathogenesis is still speculative.

We report the case of a patient with left atrial myxoma who developed multiple aneurysms with intracerebral hemorrhages 6 months after successful resection of the cardiac tumor and the probable pathogenic mechanisms are discussed.

A month later, he underwent a trans-septal left atrial tumor resection with pericardial patch closure of interatrial septum. The excised hemispherical oval sessile mass measured 5 x 3 x 2.5 cm and had numerous blunt finger-like projections on its surface. Translucent gelatinous myxomatous tissue and areas of hemorrhage were seen throughout. Histological sections showed a zonation characteristic of a ‘myxoma’ [Figure 1].

The postoperative period was uneventful. At 2 months follow up, he had good recovery in his motor power and aphasia and remained functionally independent, with only mild residual spasticity of the right upper and lower limbs.

Six months later, he developed left focal motor seizures with secondary generalization lasting an hour. Magnetic resonance imaging (MRI) of the brain showed multiple hemorrhages of different ages in the right frontal region posteriorly, both parietal regions and right occipital region of average 1- to 2-cm size. The lesions showed moderate contrast enhancement [Figure 2]. Cerebral 4-vessel angiography revealed multiple, small, distal, fusiform aneurysms along both middle and anterior cerebral arteries [Figure 3A and B]. A repeat transthoracic and transesophageal echocardiogram showed no evi-

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A 54-year-old man with no known cardiovascular risk factors presented with acute onset of right hemiplegia and motor aphasia. A computerized tomography (CT) scan of the head showed a left perisylvian infarct in the middle cerebral artery territory. Echocardiogram revealed a 50 x 35 mm echogenic left atrial mass protruding into the left ventricle. A coronary angiogram showed normal anatomy. The patient did not manifest any constitutional symptoms and the other laboratory investigations were noncontributory.

Figure 1: Blood vessels in atrial myocardium show mucoid degenerative changes in their wall and contain plugs of myxomatous tissue [hematoxylin and eosin (H&E) x240].
The patient was treated with antiedema measures and Phenytoin. He improved symptomatically, his seizures did not recur, and he became functionally independent again.

Discussion

Myxomas are the most common tumors of the heart accounting for more than 50% of all primary cardiac neoplasms. Patients often manifest cardiac symptoms, nonspecific constitutional symptoms, and sequelae of cerebral or systemic embolization. Neurologic events occur in more than 25% of patients. Delayed cerebrovascular events may occur months after excision of the atrial myxoma. The pathogenesis of these aneurysms remains to be elucidated. The different postulates include a ‘vascular damage theory’ proposed by Stoane et al. in which tumor emboli cause perivascular damage with scarring and pseudoaneurysm formation; the other is a ‘neoplastic theory’ in which hematogenous metastases of myxoma cells penetrate and damage the vessels with subsequent fibroblastic proliferation. This view was supported by histopathological findings in a few reports. Myxomas may occur rarely as part of familial and inherited disorders such as the Carney’s complex or Marfan’s syndrome; however, such associations were absent in our patient. Furuya et al. refer to neovascularization by prominent vasa vasora in the hyperplastic arterial wall as a mechanism of aneurysm growth. However, similar changes noted in cardiac myxomas and in organizing vascular thrombi by Salyer et al. suggested that myxomas were organizing thrombi with abnormal reparative processes and not neoplasms. We suggest the use of the term ‘myxoma-related’ aneurysm to be more appropriate than ‘myxomatous’ or ‘oncological’ aneurysm as designated by some.

To our knowledge, this is the first report of atrial myxoma with cerebral aneurysm from India. The true incidence of myxoma-related aneurysms is unknown. Because of the rar-
ity of the disease, little is known about the natural history and nature of such aneurysms. Hence, the management of these aneurysms is controversial. Spontaneous regression of aneurysms has been well documented in patients in whom the primary atrial tumor has been excised.\cite{2} Considering the occurrence of ‘multiple’ hemorrhages in many of the reports including ours, it appears that the risk of bleeding may be more than the usual nonmyxoma-related cerebral aneurysms.\cite{1,4,6} Although fusiform aneurysms cannot be clipped as they lack a stem, successful aneurysmectomy and aneurysm wrapping for moderate-sized aneurysms have been reported.\cite{6} Chemotherapy with Doxorubicin was tried for 6 months in another patient with no apparent success.\cite{2} As our patient had multiple aneurysms in the distal branches of middle and anterior cerebral arteries, no surgical intervention was feasible; no other specific therapy was given as they are of no proven benefit.

Therefore, patients with myxoma presenting with delayed neurological events after surgery should be evaluated for the presence of ‘myxoma-related aneurysms.’

References


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