

SHORT REPORT

Multiple cerebral aneurysms secondary to cardiac myxoma

Kuriakose Joshi George¹, Adam Rennie² & Amar Saxena¹

¹Neurosurgery, University Hospitals of Coventry and Warwickshire, Coventry CV2 2DX, UK, and ²Radiology, University Hospitals of Coventry and Warwickshire, Coventry CV2 2DX, UK

Abstract

A 45-year-old lady presented with transient ischemic attack symptoms. Subsequent CT and DSA revealed multiple fusiform dilatations of cerebral vessels. A search for sources of emboli led to an atrial myxoma being found on echocardiogram. She made a good recovery after resection of the atrial myxoma. The aneurysms were managed conservatively. We discuss the association of multiple cerebral aneurysms with atrial myxoma.

Keywords: aneurysms; atrial; cardiac; cerebral; multiple; myxoma

Case history

A 45-year-old lady presented with a gradual onset of headache and transient left arm weakness which lasted only for a few minutes.

A CT scan of the head revealed a 1-cm hyperdensity in the right sylvian fissure, suspicious of a middle cerebral artery aneurysm but with no evidence of subarachnoid haemorrhage. A subsequent CTA followed by a digital subtraction angiogram (DSA) (Fig. 1 and 2) revealed that in addition to a 1-cm-wide neck, right middle cerebral artery aneurysm, there were numerous fusiform dilatations of the distal intracranial arteries numbering at least 12, most of which were less than 4 mm in size.

None of the aneurysms were thought to be suitable for either endovascular embolisation or surgical clipping as they looked either very friable, fusiform or wide necked. MRI of the head showed multiple small vessel infarcts and enhancement of the wall of the fusiform dilated vessels, suggesting an acute inflammatory process. She was investigated for possible vasculitis and endocarditis. A consequent echocardiogram showed that there was a 1 inch diameter mass in the left atrium attached to the septal wall. She was put on aspirin initially and two weeks later underwent open heart surgery for operative removal of the mass.

Histology revealed this to be an atrial myxoma. She made a full recovery and was discharged from hospital 2 weeks later. Repeat DSA showed stable appearances of the fusiform aneurysms 6 months later. She remains on Aspirin in an effort to prevent further stroke.

Discussion

Multiple fusiform-cerebral aneurysms are rarely found in choriocarcinoma, infective endocarditis, in collagen disease or cardiac myxoma.

Cardiac myxoma (CM) is a potentially treatable cause of cerebral embolism. They are slowly growing intra-cavitary benign neoplasms of the heart. It is the most common primary tumour of the heart with an incidence of 0.05% of all necropsies. Patients with cardiac myxoma usually present with one or more of the triad of embolism, both cerebral and peripheral, intracardiac obstruction, and constitutional symptoms like fever, weight loss, anemia and raised erythrocyte sedimentation rate. Systemic embolism has been reported in 30–50% of patients with left atrial myxoma, half of them with cerebral involvement.

A recent study¹ of 74 patients with CM reported that only one case had concurrent cerebral aneurysm indicating that development of cerebral aneurysms in CM a rare.

Cerebral aneurysm formation in association with left cardiac myxoma was first described by Marchand in 1894. Pinede et al.² found a median age of 53 years for women and 58 years for men in total of 112 cases. The male/female ratio was 71% female. Preoperative symptoms were non-specific. Only about one-fourth of the patients showed cardiac signs. Recent findings suggest that cardiac myxoma both produce and release the cytokine interleukin-6 (IL-6) and that this cytokine may be responsible for the reported constitutional symptoms such as weight loss and fever. Nearly all patients suffered from embolism into cerebral (74%) or peripheral (23%) vessels. Surgical resection of the cardiac myxoma dramatically reduced the number of deaths from cerebral infarction.

Sabolek et al.³ were able to analyze a total of 101 aneurysms in 33 patients with atrial myxoma with a median number of 3.0 aneurysms per patient (range: 1–8). Most aneurysms (91%) were of the fusiform type, the remaining ones were saccular dilatations (ranges from 3 to 10 mm in diameter).

On MRI, myxomatous aneurysms appeared as fusiform tubular dilatations of cerebral arteries within the sulci on T1- and T2-weighted images; in some cases a hypointense

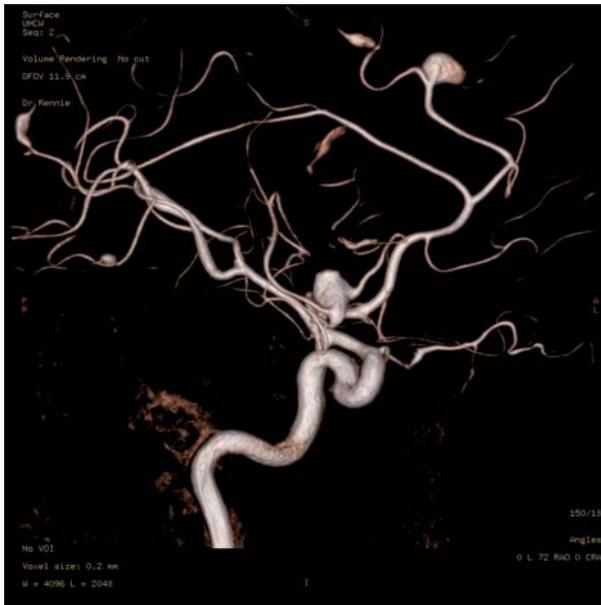


Fig. 1. Digital subtraction angiogram: Right carotid injection volume rendered image- shows multiple fusiform and wide necked aneurysms 270 × 270 mm (96 × 96 DPI).

rim was present around the aneurysm. These lesions were often characterized by vascular dilatation and might be surrounded by edema and hemorrhage. By far the greatest part of the aneurysms (74%) was found in the area of middle cerebral arteries on both sides with slight laterality toward the right side. In some patients, there were no cerebral aneurysms at the time of cardiac surgery as confirmed by angiography demonstrating that aneurysm formation can occur after resection of the cardiac myxoma. Sequential angiography in some patients revealed that the time course of aneurysm formation and growth is very irregular.

The pathogenesis of aneurysm formation in myxoma patients

Current hypothesis favours that tumour material from cardiac myxoma embolizes into the vasa vasorum of peripheral arteries and subsequently proliferate in the vessel wall leading to a weakening of subintimal tissue such as the internal elastic lamina with subsequent aneurysm formation. Due to the very slow growing behaviour of the myxomatous tissue, there is a variable delay between embolism of the tumour material and the destruction of the vessel wall with aneurysm formation.

Management strategies

Patients found to have multiple fusiform aneurysms should have echocardiography to rule out atrial myxoma or endocarditis.

Cardiac surgery to remove the primary cardiac tumour is the first choice procedure as this minimizes the risk of embolization of tumour material, but cannot completely abolish the risk of delayed cerebral aneurysm formation, presumably as a result of metastatic seeding prior to surgery.



Fig. 2. Digital subtraction angiogram: Right carotid injection 64 × 264 mm (96 × 96 DPI).

Patients with current or prior cardiac myxoma and new neurological symptoms should undergo neurovascular imaging. Since the majority of myxomatous aneurysms are located distal to the circle of Willis, MRA is likely insufficient to rule out a myxomatous aneurysm. A CTA in a high resolution CT scanner or a DSA is necessary. Follow-up can be with either CTA or MRA depending on the size of the aneurysm of interest. MR of the brain may disclose an inflammatory reaction in parenchyma underlying a myxomatous aneurysm, and can guide angiography. By analogy with un-ruptured non-myxomatous aneurysms, aneurysm enlargement may warrant consideration for treatment.

Clip reconstruction of myxomatous aneurysms has been reported but clipping of very friable aneurysms can be associated with very high rates of mortality.⁴ Clip occlusion or trapping may serve as a definitive surgical strategy. Some fusiform aneurysms can be wrapped with good surgical results.⁵ For ruptured myxomatous aneurysms, surgical hematoma evacuation with clip ligation/trapping of the offending aneurysm is reasonable.

The finding that dividing tumour cells are responsible for aneurysm formation suggests the possibility of using chemotherapy to prevent aneurysmal growth, but the results of doxorubicin alone are equivocal. Low-dose radiation therapy in conjunction with chemotherapy revealed more encouraging results.⁶

Conclusion

Our case is unusual that the myxoma was discovered after the cerebral aneurysms were found. Subsequent CTA and DSA revealed multiple fusiform dilatations of cerebral vessels. A search for sources of emboli led to the myxoma being found on echocardiogram. Our patient is also unusual in that she had the largest reported number of aneurysms.

It is emphasised that patients with multiple aneurysms (particularly if the aneurysms are peripheral or distal) must be investigated with echocardiography to find a source of infective or tumour emboli as the cause of multiple aneurysms.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References

1. Lee VH, Connolly HM, Brown RD Jr. Central nervous system manifestations of cardiac myxoma. *Arch Neurol* 2007;64:1115-20.
2. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. *Medicine (Baltimore)* 2001;80:159-72.
3. Sabolek M, Bachus-Banaschak K, Bachus R, Arnold G, Storch A. Multiple cerebral aneurysms as delayed complication of left cardiac myxoma: A case report and review *Acta Neurol Scand.* 2005;111:345-50.
4. Pepin M, Schwarze U, Superti-Furga A, Byers PH. Clinical and genetic features of Ehlers-Danlos syndrome type IV, the vascular type. *N Engl J Med* 2000;342:673-80.
5. Park SH, Yim MB, Lee CY, Kim E, Son EI. Intracranial Fusiform Aneurysms: It's Pathogenesis, Clinical Characteristics and Management. *J Korean Neurosurg Soc* 2008;44:116-23.
6. Bernet F, Stulz PM, Carrel TP. Long-term remission after resection, chemotherapy, and irradiation of a metastatic myxoma. *Ann Thorac Surg* 1998;66:1791-2.