CASE REPORT

Multiple cerebral aneurysms in a patient with cardiac myxoma: what to do?

Olafur Sveinsson,¹ Lars Herrman²¹Department of Neurology, Karolinska University Hospital, Stockholm, Sweden²Department of Neuroradiology, Karolinska University Hospital, Stockholm, Sweden

Correspondence to

Dr Olafur Sveinsson, olafur.sveinsson@karolinska.se

Accepted 21 January 2015

SUMMARY

Cardiac myxomas are a very uncommon cause of cerebral aneurysms. We present a case of a young woman with neurological symptoms attributed to small cortical infarctions and multiple cerebral aneurysms, caused by a cardiac myxoma.

BACKGROUND

1. An aneurysm caused by cardiac myxoma can be missed if an angiography is not performed.
2. These aneurysms can appear many years after the myxoma has been resected.
3. Our case presents a dilemma where there are no guidelines or large series to guide treatment in a patient with multiple aneurysms caused by a myxoma.

CASE PRESENTATION

A formerly healthy 19-year-old woman presented with a 3-year history of episodic loss of consciousness and right-sided weakness. The attacks were interpreted to be seizures with Todd's paresis, and medication with levetiracetam was initiated. Neurological examination was normal. MRI of the brain revealed multiple microbleeds and multiple aneurysms (figure 1). Conventional cerebral angiography and CT angiography showed a large number of distal well-demarcated fusiform aneurysms (figures 2 and 3). Most of them were small, but some were up to 11 mm in diameter. Transthoracic and transoesophageal echocardiography showed a 3.3×2 cm large mass protruding from the septum into the left atrium; it appeared to be a myxoma. The tumour was excised without any complications and pathologically confirmed to be a myxoma (figure 4). There were no signs of a connective tissue disorder such as Ehlers-Danlos syndrome presentation such as the Carney complex.

DIFFERENTIAL DIAGNOSIS

Multiple fusiform aneurysms caused by infection, connective tissue disease or choriocarcinoma.

TREATMENT

A conservative approach was chosen because of the following:

- The patient was doing well.
- The multiplicity, peripheral location, fusiform and wide necked shape of most of the aneurysms and their possibly friable nature.
- Little evidence is to be found in the literature regarding coiling/clip obstruction of multiple fusiform aneurysms caused by a heart myxoma.

OUTCOME AND FOLLOW-UP

A year later, the patient was well and had no symptoms, although some of the aneurysms had increased in size. Interleukin 6 levels were normal. Since none of the aneurysms were suitable for endovascular coiling or surgical clipping, a conservative approach with annual conventional angiography was chosen.

DISCUSSION

Even though cardiac myxomas have been recognised for over 150 years, relatively little is known about them.¹ In 1953, the first atrial myxoma was surgically resected. Before that they were exclusively diagnosed at autopsy. Echocardiography has since then revolutionised the diagnosis of cardiac myxomas. Myxomas occur first and foremost in the left atrium (90% of cases). In fewer than 10% they are found in the right atrium. The tumours usually arise from the interatrial septum and grow into the left atrium.

Symptoms

Besides constitutional and cardiac symptoms such as dyspnoea and chest discomfort, myxoma can cause neurological symptoms.

Neurological symptoms

TIA/stroke is the most common neurological symptom caused by myxomatous embolisation to brain arteries. The embolisation can very infrequently lead to aneurysm formation and the risk for subarachnoid haemorrhage. Unlike berry aneurysms, which are usually located around the circle of Willis, these myxomatous aneurysms are often multiple and located peripherally in the intracranial vascular tree. They also have a tendency to be fusiform instead of saccular. Because of this they are often thought of as being mycotic aneurysms.²⁻⁴ Aneurysms caused by cardiac myxoma can be found many years after a successful atrial myxoma resection.^{2,5}

It is not quite clear how the aneurysms develop but a few hypotheses have been put forward. First, they are not caused by blood-flow dynamics such as berry aneurysms. In all likelihood they are caused by myxomatous tumour invasion into the vessel wall causing its weakening.⁶ Pathologically proven myxomatous emboli have been demonstrated in cerebral arteries.⁷ The tumour emboli cells can infiltrate the arterial wall leading to its weakening and cause subsequent aneurysm formation.^{8,9} Another theory is that the tumour cells embolise into the vasa vasorum

CrossMark

To cite: Sveinsson O, Herrman L. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-200767

Figure 1 (A) Susceptibility-weighted MRI showing multiple microhaemorrhages in both hemispheres. (B) Axial T2 (left) and T1 contrast-enhanced (right) images showing multiple aneurysms in both hemispheres. The aneurysms are best seen on the T1 contrast-enhanced image as the intravenous contrast fills the dilated vessel segments.

of peripheral arteries and from there proliferate into the vessel wall. This in turn can lead to a weakening of the internal elastic lamina and induce aneurysm formation. In either mechanism it is possible that an inflammatory reaction ensues because of interleukin 6 production by the myxoma cells; this plays an important role.

Treatment and follow-up

The myxoma should be excised as quickly as possible to prevent further embolic complications or valve obstruction.¹⁰ Not much is known about the natural history of these aneurysms to guide treatment. The many distal fusiform aneurysms are not well suited for coiling. There are case reports of

Figure 3 Three-dimensional angiography with contrast injection into the right internal carotid artery showing multiple fusiform aneurysms in peripheral branches of the middle and anterior cerebral artery (arrows).

clipping the largest aneurysms. Fortunately, bleeding is not frequent. In the largest series of 34 patients with aneurysms caused by myxoma, only 4 (13%) patients bled.¹¹ Therefore a conservative approach is most often recommended. There are a few case reports using chemotherapy and radiation, with some effect.¹²

Figure 2 (A and B) Digital subtraction angiographies of the left internal carotid artery, 1 year apart, showing increase of an aneurysm of the middle cerebral artery, M2 branch (thick arrow). The fusiform aneurysm (thin arrow) disappears on the latter investigation most likely due to an occlusion of a distal branch supplying flow to the aneurysm. The figure also shows multiple peripherally located aneurysms (small arrows).

Figure 4 Morphology of the myxoma. The lighter area is the base that was attached to the septum of the left atrium.

Learning points

- ▶ Aneurysms caused by cardiac myxoma can be missed if angiography is not performed.
- ▶ The aneurysms can appear many years after the myxoma has been resected.
- ▶ Currently there are no guidelines or large clinical series to guide treatment, but a conservative approach is usually recommended.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 King TW. On simple vascular growths in the left auricle of the heart. *Lancet* 1845;2:428–9.
- 2 Jean WC, Walski-Easton SM, Nussbaum ES. Multiple intracranial aneurysms as delayed complications of an atrial myxoma: case report. *Neurosurgery* 2001;49:200–3.
- 3 Burton C, Johnston J. Multiple cerebral aneurysms and cardiac myxoma. *N Engl J Med* 1970;282:35–6.
- 4 Nucifora PG, Dillon WP. MRI diagnosis of myxomatous aneurysms: report of two cases. *AJNR Am J Neuroradiol* 2001;22:1349–52.
- 5 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.
- 6 Attum AA, Johnson GS, Masri Z, *et al.* Malignant clinical behavior of cardiac myxomas and 'myxoid imitators'. *Ann Thorac Surg* 1987;44:217–22.
- 7 Matsuoka S, Ito M, Shinonome T, *et al.* An autopsy case of cerebral embolism caused by atrial myxoma. *No Shinkei Geka* 1992;20:255–9.
- 8 Furuya K, Sasaki T, Yoshimoto Y, *et al.* Histologically verified cerebral aneurysm formation secondary to embolism from cardiac myxoma: case report. *J Neurosurg* 1995;83:170–3.
- 9 New PFJ, Price DL, Carter B. Cerebral angiography in cardiac myxoma: correlation of angiographic and histopathological findings. *Radiology* 1970;96:335–45.
- 10 Reynen K. Cardiac myxomas. *N Engl J Med* 1995;333:1610–17.
- 11 Sabolek M, Bachus-Banaschak K, Bachus R, *et al.* Multiple cerebral aneurysms as delayed complication of left cardiac myxoma: a case report and review. *Acta Neurol Scand* 2005;111:345–50.
- 12 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.

Copyright 2015 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow