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Coil embolization of an enlarging fusiform myxomatous cerebral aneurysm

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ABSTRACT

Myxomatous cerebral aneurysms are rare sequelae of cardiac atrial myxoma. These aneurysms are generally fusiform, multiple, and distal. Pathogenesis and evolution of these aneurysms is still debated. There are currently no guidelines on the management of aneurysms secondary to atrial myxoma. We present a case of a 52-year-old man with multiple fusiform aneurysms 3 years after resection of a left atrial myxoma. One of these aneurysms was followed with cerebral angiography and showed substantial interval enlargement. This aneurysm was subsequently embolized. All aneurysms were stable 6 months post-embolization.

Case presentation

A 52-year-old man presented to the emergency department complaining of chronic cough. On review of systems, the patient reported several pre-syncopal episodes and substantial weight loss. On physical examination, he was found to have poor finger-nose-finger coordination bilaterally and up-going toes bilaterally. Clinical signs suggested congestive heart failure and the subsequent 2-dimensional echocardiogram revealed a mobile left atrial mass. Further evaluation with cardiac magnetic resonance imaging (MRI) confirmed a 4.5 × 2.9-cm left atrial mass, which prolapsed into the left ventricle during diastole. Computed tomography (CT) head demonstrated multiple scattered white matter lucencies, and subsequent MRI head showed multiple acute and subacute infarcts, consistent with embolic shower. Given patient's symptomatology, the decision was made to resect the left atrial mass, pathologically determined to be a myxoma.

Three years later, the patient presented with acute right lower extremity weakness and seizures. CT head demonstrated multiple small areas of intraparenchymal hemorrhage. MRI head showed multiple scattered nodular foci along the cortex/subarachnoid compartment, particularly prominent in the frontal and parietal lobes, suspicious for leptomeningeal metastases vs subacute cortical infarcts vs a granulomatous process, such as neurosarcoidosis. A search for a primary malignancy was undertaken, including a CT of the chest, abdomen, and pelvis, which was negative. Echocardiogram and cardiac MRI were also performed to evaluate for recurrence of the atrial

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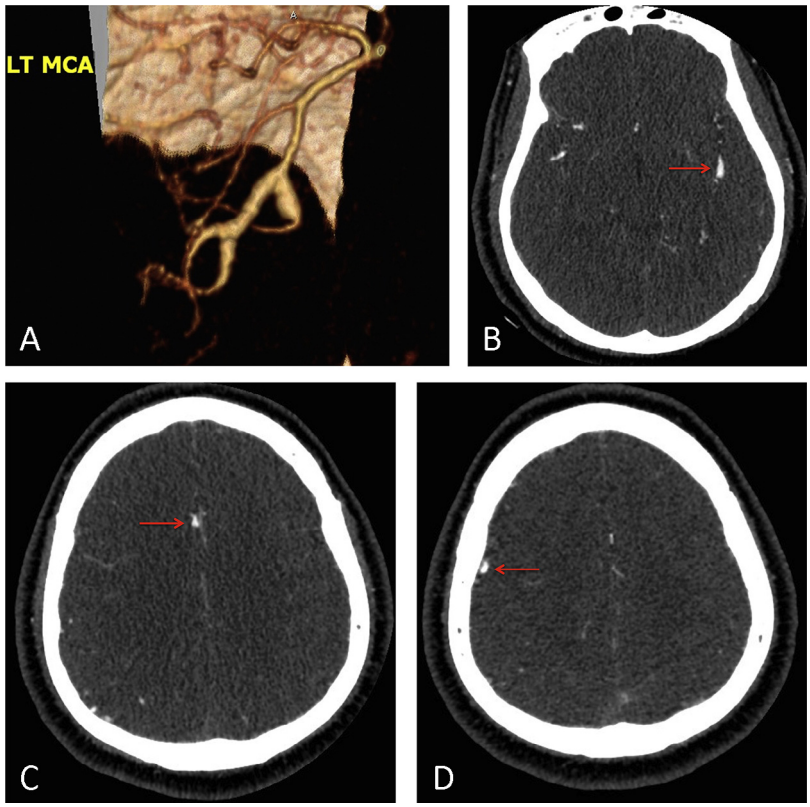

Fig. 1 – (A) Three-dimensional reconstruction from CTA head and neck demonstrates second- and third-order left MCA branch vessel ectasia. (B) Axial CT image from the same CTA study shows the 4.6-mm aneurysmal left MCA M2 segment. (C) An axial CT image from the same study also demonstrates 3-mm ectasia of a distal branch of the right ACA. (D) Shows another ectatic vessel in the posterolateral right frontal lobe. ACA, anterior cerebral artery; CT, computed tomography; CTA, computed tomography angiogram; MCA, middle cerebral artery.

myxoma, which was also negative. Cytology obtained from lumbar puncture also failed to demonstrate malignant cells or any definitive evidence for infectious or inflammatory process. Computed tomography angiogram (CTA) head and neck demonstrated multifocal short segments of ectasia in the right anterior cerebral artery (ACA) and bilateral middle cerebral artery (MCA) branch vessels, with the largest aneurysmal dilatation seen in the Sylvian M2 segment of the left MCA, measuring 4.6 mm (Fig. 1A-D). Cerebral angiogram confirmed these findings, demonstrating diffuse distal cerebral arterial vasculopathy (Fig. 2). Given the patient's history of resected left atrial myxoma, and otherwise negative workup, this was favored to represent myxomatous vasculopathy.

Two years later, the patient presented with new headaches for several weeks. MRI brain showed significant enlargement of a fusiform aneurysm of the M2 segment of the left MCA over this time period, from 4.6 mm to 9.3 mm (Fig. 3). Patient was taken for pancerebral arteriogram for further evaluation, which confirmed dramatic progression of aneurysmal dilatation of a left inferior division MCA branch, measuring up to 9 × 14 mm (Fig. 4A-B). Interestingly, 2 smaller aneurysms (watershed distal right anterior cerebral artery/MCA at the posterior frontal lobe, and a small branch of the anterior inferior frontal distribution) showed interval resolution. However, all other fusiform aneurysms showed

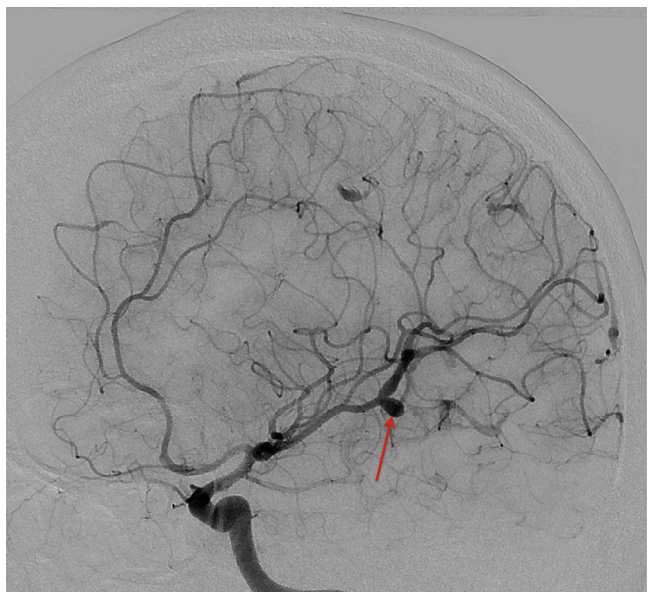

Fig. 2 – Cerebral angiogram of the left internal carotid artery (LICA) confirms diffuse cerebral arterial aneurysms. Arrow points to the left MCA M2 segment fusiform aneurysm measuring approximately 5 mm. MCA, middle cerebral artery.

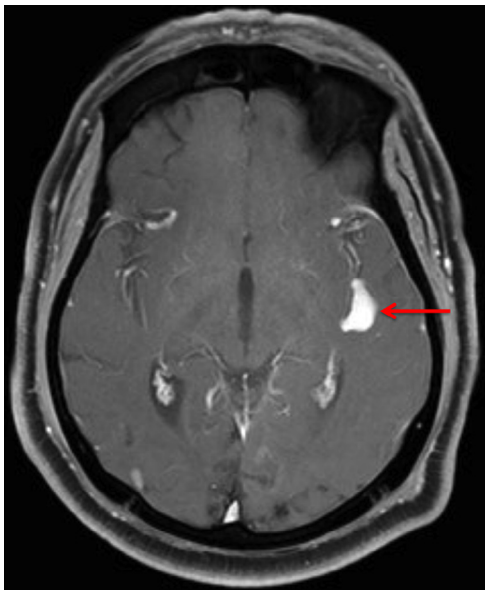

Fig. 3 – Enhanced T1 axial MRI demonstrates substantial interval enlargement of the left MCA aneurysm, now measuring up to 9.3 mm. MCA, middle cerebral artery; MRI, magnetic resonance imaging.

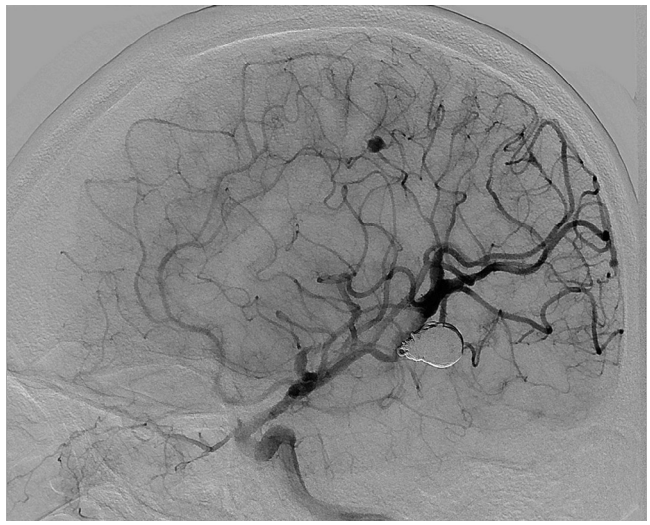

Fig. 5 – Lateral projection of a cerebral aneurysm of the LICA status post treatment of the enlarging left MCA fusiform aneurysm with a series of detachable platinum aneurysm coils. Additional smaller fusiform aneurysms are again noted more distally. LICA, left internal carotid artery; MCA, middle cerebral artery.

progression. Given this substantial interval enlargement and the patient's symptoms, the dominant ovoid fusiform aneurysm of the inferior division of the left MCA branch was subsequently embolized with a series of P-400 detachable platinum aneurysm coils (Fig. 5). The day after the procedure, magnetic resonance angiogram (MRA) head demonstrated a completely occluded aneurysm. Six months later, repeat MRI/MRA head showed stable branch occlusion (coil mass) without recurrence. Other smaller aneurysms were unchanged.

Discussion

Myxoma is the most common type of cardiac tumor, with neurologic symptoms occurring in 26%-45% of patients [1]. However, intracranial myxomatous aneurysms are a very rare complication of left atrial myxoma, with only about 40-50 cases reported in the literature [2–4]. The presentation of intracranial myxomatous aneurysms may be concurrent with, or, as in our case, several years after, the initial diagnosis of the cardiac tumor. This delayed presentation, in conjunction with the rarity of myxomatous aneurysms and the nonspecific symptoms such as headaches, weakness, paresthesias, and seizures, often leads to a prolonged and extensive workup.

The pathogenesis of myxomatous aneurysms is unclear. One hypothesis posits that the aneurysms are the result of myxomatous emboli that lodge in the cerebral vessels, penetrate the vessel wall, and infiltrate via subintimal growth, leading to weakening of the artery wall and aneurysm formation [1]. A similar hypothesis is that the embolized myxoma tissue blocks the vasovasorum of the intracranial arteries, predisposing to ischemic weakening of the vessel wall and subsequent dilatation [5–7]. A third explanation is that tumor emboli occlude the vessel lumen, causing scarring and pseudoaneurysm

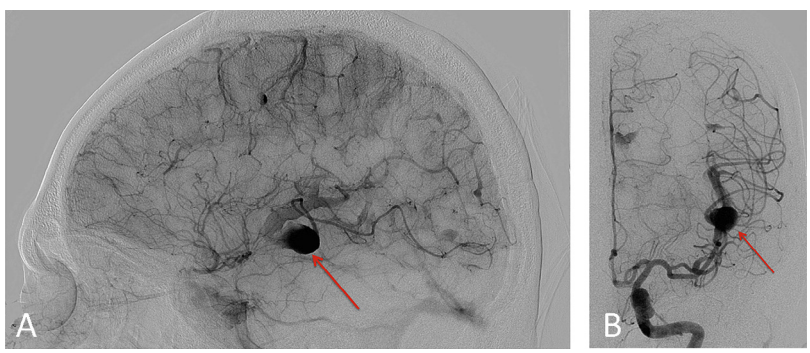

Fig. 4 – Cerebral angiogram of the LICA in frontal oblique (A) and lateral (B) projections confirms substantial interval enlargement of the fusiform aneurysm of the left MCA M2 segment (arrows). Additional smaller fusiform aneurysms can be seen more distally. LICA, left internal carotid artery; MCA, middle cerebral artery.

formation [5–7]. Other authors have proposed aneurysm formation is secondary to a reaction to secretion of a proinflammatory cytokine, interleukin-6 (IL-6), by myxoma cells [4,8]. Studies have shown atrial myxoma cells are capable of producing IL-6 [9,10], and elevated IL-6 levels have been detected in patients with intracranial myxomatous aneurysms, both before and after myxoma resection [4,8,9,11]. Although the exact mechanism of aneurysm induction by IL-6 has not been elucidated, it has been proposed that IL-6 induces overexpression of multiple proteolytic enzymes, which can degrade the extracellular matrix of myxoma cells and encourage embolization to the cerebral vessels [9,12]. It has also been suggested that these embolized myxomatous cells continue to produce IL-6, directing weakening cerebral vessel walls and leading to aneurysm formation [9,11].

There is no consensus on the management of patients with myxomatous aneurysms. Patients are often followed with serial imaging to assess for aneurysm growth and progression. The natural history of these aneurysms is also not clear, as some cases have shown stability, others improvement (self-occlusion/spontaneous thrombosis), and others an increased number and size of aneurysms [5,13–16]. IL-6 has been proposed as a marker for myxomatous aneurysms, but this is not used regularly in clinical practice [4,7,9,11,17]. In rare instances, chemotherapeutic agents have been tried, with limited equivocal results [7,18,19].

Myxomatous aneurysms are often multiple, fusiform, and distal in location, which limits invasive treatment options such as clipping or coil embolization. In only a minority of cases has an invasive intervention been undertaken, most often due to aneurysm rupture [8,20,21]. Single saccular myxomatous aneurysms have also been managed surgically [3,8]. Only very rarely has intervention been undertaken in patients with multiple aneurysms, and in these cases, patients continued to require close follow-up to evaluate for growth of the untreated aneurysms [15,22].

In our case, the fusiform aneurysm at the inferior division of the left MCA was embolized with coils secondary to its significant enlargement and the patient's new headaches; however, multiple other aneurysms persist and the patient is at continued risk for hemorrhage.

Myxomatous aneurysms are very rare, and given their multiplicity and fusiform morphology, intervention is infrequently undertaken. Our case is unique in that the dominant enlarging fusiform aneurysm was successfully coiled, and stable at 6-month follow-up. Additionally, our case also demonstrated the complicated evolution of myxomatous aneurysms, with auto-sacrifice of a few small aneurysms, but substantial progression of others in a 2-year time frame. Further investigation into the long-term outcomes of such intervention and results of imaging follow-up are necessary to better establish efficacious treatment paradigms for this patient population.

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