MALEFICIENT'S HORN LIKE APPEARANCE MYXOMATOUS FUSIFORM ANEURYSM OF THE RIGHT MIDDLE CEREBRAL ARTERY: AN EVIL CAUSED BY THE ATRIAL MYXOMA

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ABSTRACT:
Atrial myxoma is the most common neoplasm of the heart which consist of 50% of the total cases reported and tend to seed peripherally after operation(1). When the dissemination occurred in the brain, patient normally presented with stroke symptoms due to tumour embolism. However, cerebral aneurysm due to metastatic deposit after atrial myxoma incision is rare.

In a study by M. Anvari et al showed that the most common presentation of atrial myxoma is shortness of breath (63%), followed by chest pain (37%) and neurological symptoms, which is mostly related to stroke (26%)(2). In some cases, patient presented with constitutional symptoms like loss of weigh and appetite and some with anaemic symptoms(2). Size of the lesion mainly related to the local symptoms due obstruction of the blood flow in the heart, but the rate of dissemination is depending on the mobility of the lesion(3). Histologically, mayo clinic has divided the tumour based on its gross anatomy into solid and papillary type. The solid type is normally larger and causing obstructive symptoms, however papillary type is the one that tends to embolise peripherally(4).

Atrial myxoma can be diagnosed by echocardiogram and mostly located within the left atrium, specifically within the fossa ovalis in 75% of the cases (3). In some cases, MRI is needed to confirm the diagnosis because sometimes thrombus, vegetation or primary lymphoma of the heart might mimic the tumour. In a case of brain dissemination, the stroke due to embolization can be detected via plain CT brain as it will show multifocal infarction predominantly at the cortimedullary junction which is not specific to the arterial territory. As the dissemination occurred at the vessel wall, it can lead to aneurysm formation which can be detected by plain CT brain and confirmed by MRI or cerebral angiogram. We are reporting a case of multiple fusiform cerebral aneurysm 9 years after left atrial myxoma operation.

Keywords: Cerebral artery aneurysm, atrial myxoma

CASE REPORT

This is a case of 53 years old lady with previous history of stroke in 2011. She regained full neurological resolution after 1 year of rehabilitation. Echocardiogram done after 1 month noted left atrial myxoma and confirmed by CT pulmonary artery. CTPA shows large hypodense mass occupying the left atrium measuring 4.5 x 4.3 x 4.7cm (AP x W x CC) causing left atrial dilatation and significant narrowing of mitral valve opening (Figure 1). The mass is successfully operated, and patient recovered very well after that.

She presented again 8 years later with intermittent right upper limb numbness associated with headache. Otherwise, there was no weakness, nausea or vomiting and no constitutional symptoms. On examination, power is normal bilateral upper and lower limb, normal reflex and negative cerebellar sign. Plain CT brain showed hyperdense tubular structure at the right sylvian fissure and left ambient fissure with multiple calcifications in the brain. MRI brain was ordered and demonstrated multiple fusiform aneurysms involving right MCA and left PCA.

Patient was then subjected for cerebral angiogram to confirm the diagnosis. Cerebral angiogram which was done 1 month after that showed 2 fusiform aneurysms located at the distal M1 segment of right MCA and P2 segment of left PCA. No contrast blush seen. Patient was stable throughout the procedure. Case was discussed in Neurology conference with primary team and decided for conservative management.

DISCUSSION

Atrial myxoma is a benign cardiac tumour which tend to cause peripheral embolism. Normally, the diagnosis was made either as a sequel of embolization phenomenon in the brain or due to local effect within the heart. Systemic embolization to the brain is normally associated with stroke, however aneurysm is also reported but rare. In this patient, the development of multiple cerebral aneurysms occurred after 9 years of operation.

Even though the actual relationship of atrial myxoma and cerebral aneurysm is not fully understood, few theories were suggested for the formation of fusiform aneurysm in the brain. The first theory is due to embolization of the myxomatous cell to the intracranial vasculature that led to inflammation and subsequently scarring. This leads to hemodynamic changes and pseudoaneurysm formation in a later life. This change occurred slowly over times. In this patient, she developed fusiform cerebral aneurysm only after 9 years of atrial myxoma incision (5).

Second theory is due to infiltration of the vasa vasorum of the intracranial artery by the embolised tumour, thereby destroying the architecture of arterial walls. This is however not really accepted as intracranial artery lack of vasa vasorum as compared to extracranial artery, however it can be seen with the presence of artherssclerotic plaque(6). In fact, there is no report that showing extracranial artery involvement in the case of disseminated atrial myxoma.

The myxomatous aneurysm can be fusiform or saccular, however in Sabolek M et al, 91% of the aneurysm are found to be fusiform (7), as seen in our patient. Intracranial arteries that are commonly involved based on frequency are middle cerebral artery (74.2%) with slight laterality toward the right side, followed by anterior cerebral artery (13%), cerebellar arteries (7%), posterior cerebral artery (5%) and the least is basilar artery (1%) (7). Most of the reported case shows that the aneurysm normally located within the distal branches of the vessel(3). In our case, the right MCA aneurysm located at the distal M1 and PCA aneurysm at the distal P3 segments.

Another theory is due to overproduction of interleukin 6 within the tumour itself lead to inflammatory infiltration of the cerebral vessel wall and formation of aneurysm as described by Koo et al(8). He suggested that IL-6 may promote

invasion of myxomatous cells which can degrade the extracellular matrix as well as the wall integrity. Hence, the level of IL-6 serum can be used to monitor the time course of aneurysm formation.

In terms of imaging, this saccular aneurysm can be seen in plain CT in which it will appears as hyperdense tubular structure normally within the fissure following course of the arteries. Sometimes it might be associated with calcifications as seen in our case. The hyperdensity is due to accumulation of myxoid matrix in the aneurysmal wall (1). In MRI, myxomatous aneurysm sometimes appears as flow void in T1W/T2W due to fast flow of blood within. Contrast enhancement may be seen as the result of enhancing tumoral tissue within the arterial wall(1).

Apart from that the macroscopic appearance of the atrial myxoma can also be evaluated to predict the possibility of tumoral embolization. M Anvari et al describe that papillary type has higher incidence of embolization compared to solid type(2). This is as a result of an irregular or friable villous surface of the tumour. In other had solid type is mainly associated with obstructive symptoms and less systemic embolization is observed.

In term of treatment, myxomatous aneurysm of the cerebral arteries is normally treated conservatively as normally it is fusiform in appearance without neck. However, there has been reported a case of saccular aneurysm of the PICA and coiling was successfully done(5). The theory that suggests tumour infiltration into the wall causing aneurysm led to question whether chemotherapy might be useful as a method of treatment. However, results of doxorubicin therapy were equivocal (4). Once the diagnosis of atrial myxoma is established, it should be completely removed because this will minimize the risk of tumour cell embolization or metastases, however it does not eliminate the risk of delayed aneurysm formation.

CONCLUSION

We concluded that even though histologically, atrial myxoma is a benign tumour, the consequences of the tumour embolized must be taken seriously which may lead to stroke or myxomatous aneurysm of the cerebral arteries. We suggest that monitoring of serum interleukin 6 after atrial myxoma operation is useful to detect any recurrence or cerebral arteries aneurysmal formation due to systemic emboli.

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FIGURE LEGENDS:

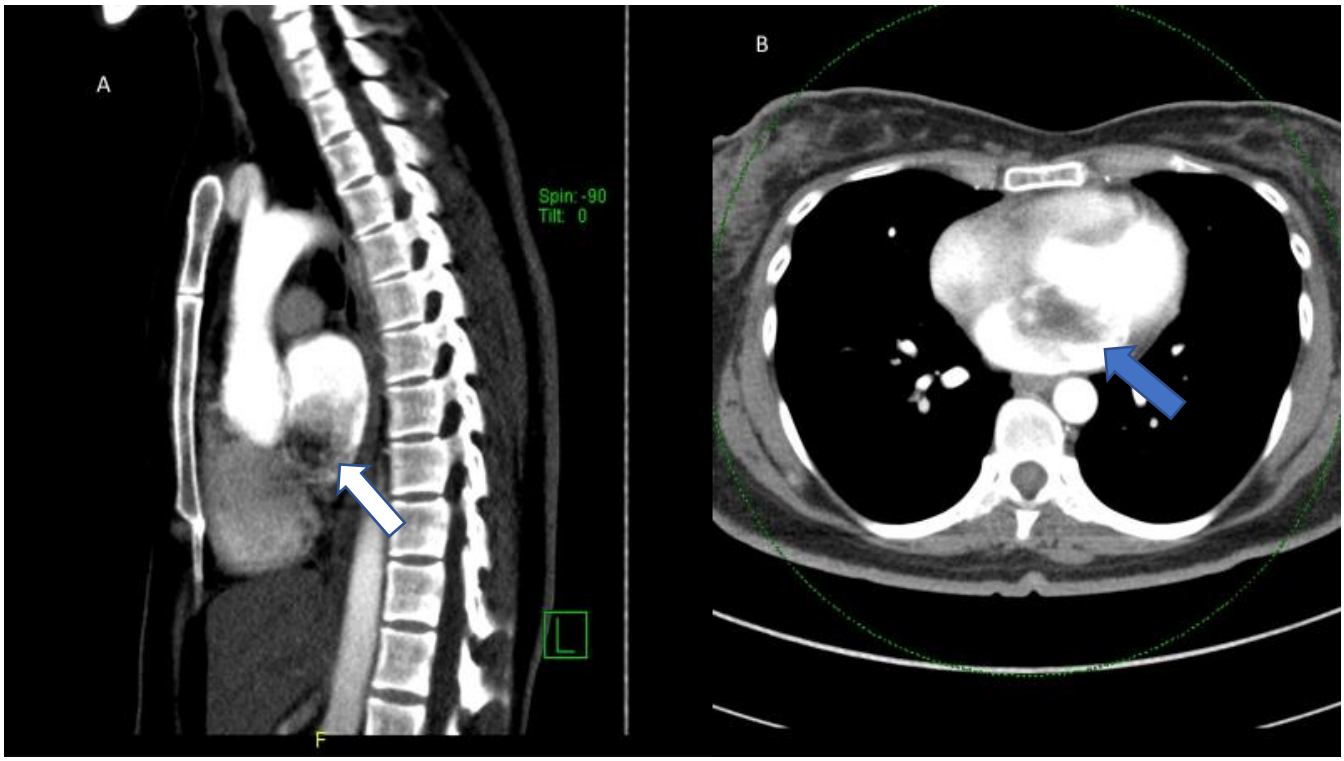

Figure 1A: Reconstructed sagittal image of CTPA showing hypodense mass at the right atrium occupying >50% of right atrial volume (white arrow). B: Axial images showing the mass in the left atrium causing left atrial dilatation and narrowing the left atrial outflow tract (blue arrow). This will lead to reduction of the left ventricular end diastolic volume and subsequently cardiac output.

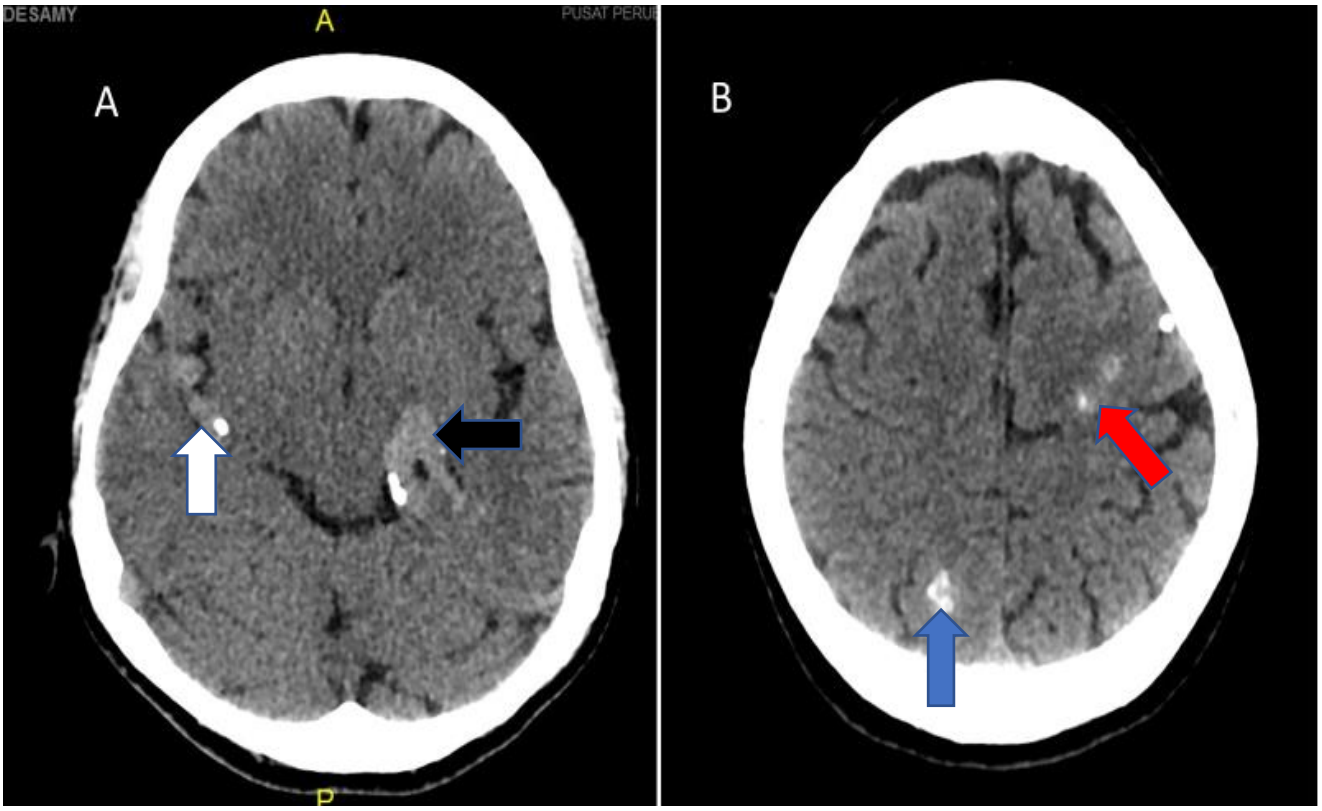

Figure 2A: Plain CT Brain in axial view noted hyperdense tubular lesion at the right sylvian fissure (white arrow) and left ambient cistern (black arrow) adjacent to the medial left temporal lobe associated with peripheral calcification. B: Scattered calcifications in both cerebral hemispheres involving right parietal (blue arrow) and left frontal lobe (red arrow).

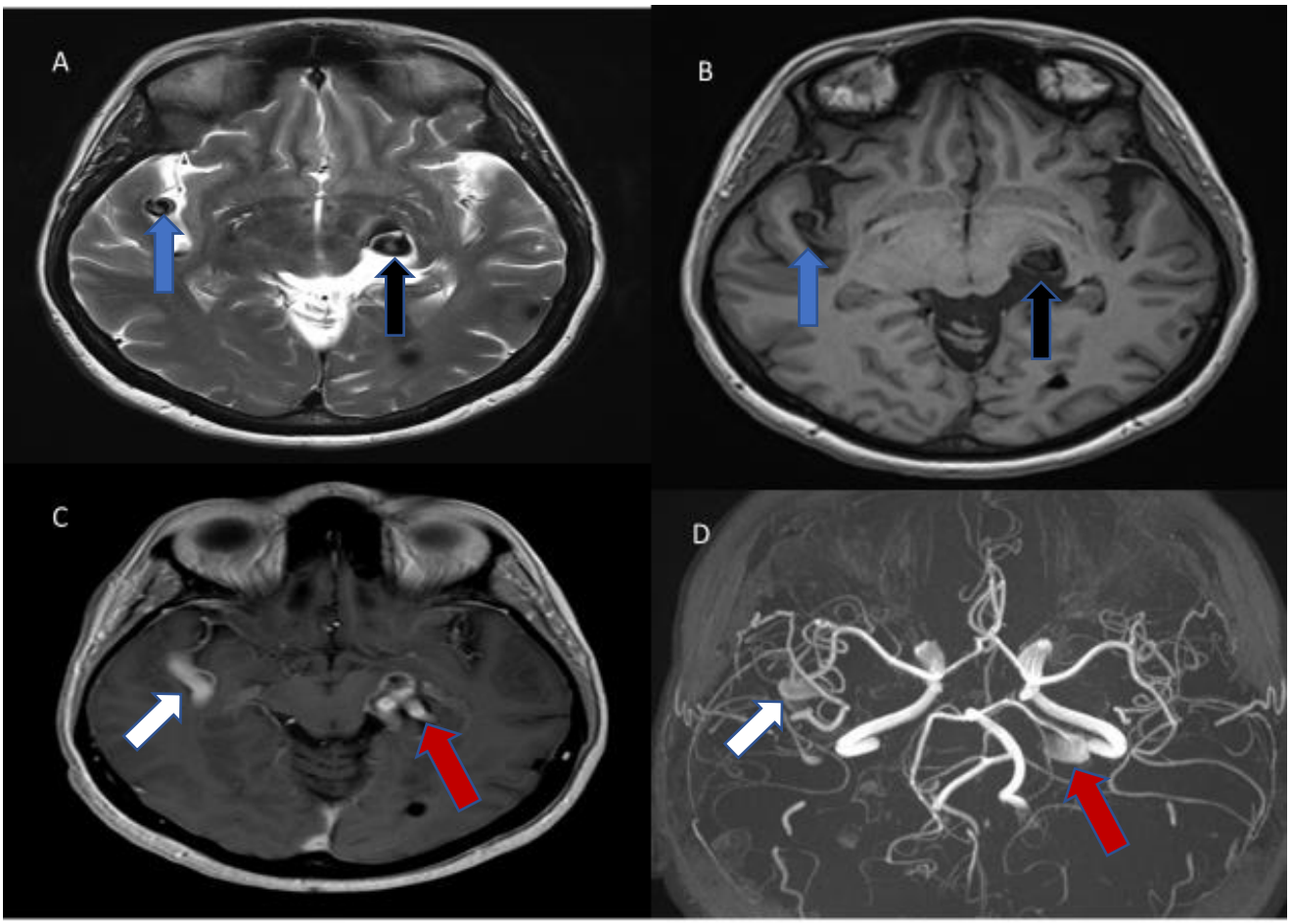

Figure 3: Axial images of MRI brain in T1W(A) and T2W(B) demonstrated signal void at the right sylvian fissure (black arrow) and left ambient cistern (blue arrow). There are also multiple blooming artefacts in SWI in keeping with calcifications as seen in previous CT scan. Figure 3C and 3D: MRA images demonstrated a large fusiform aneurysmal dilatation of M2 segment of right MCA within the right sylvian fissure (white arrow). There is also similar appearing fusiform aneurysm at the P2 segment of left PCA within the ambient cistern (red arrow). No evidence of thrombosis or restricted diffusion in DWI/ADC to suggest acute infarction.

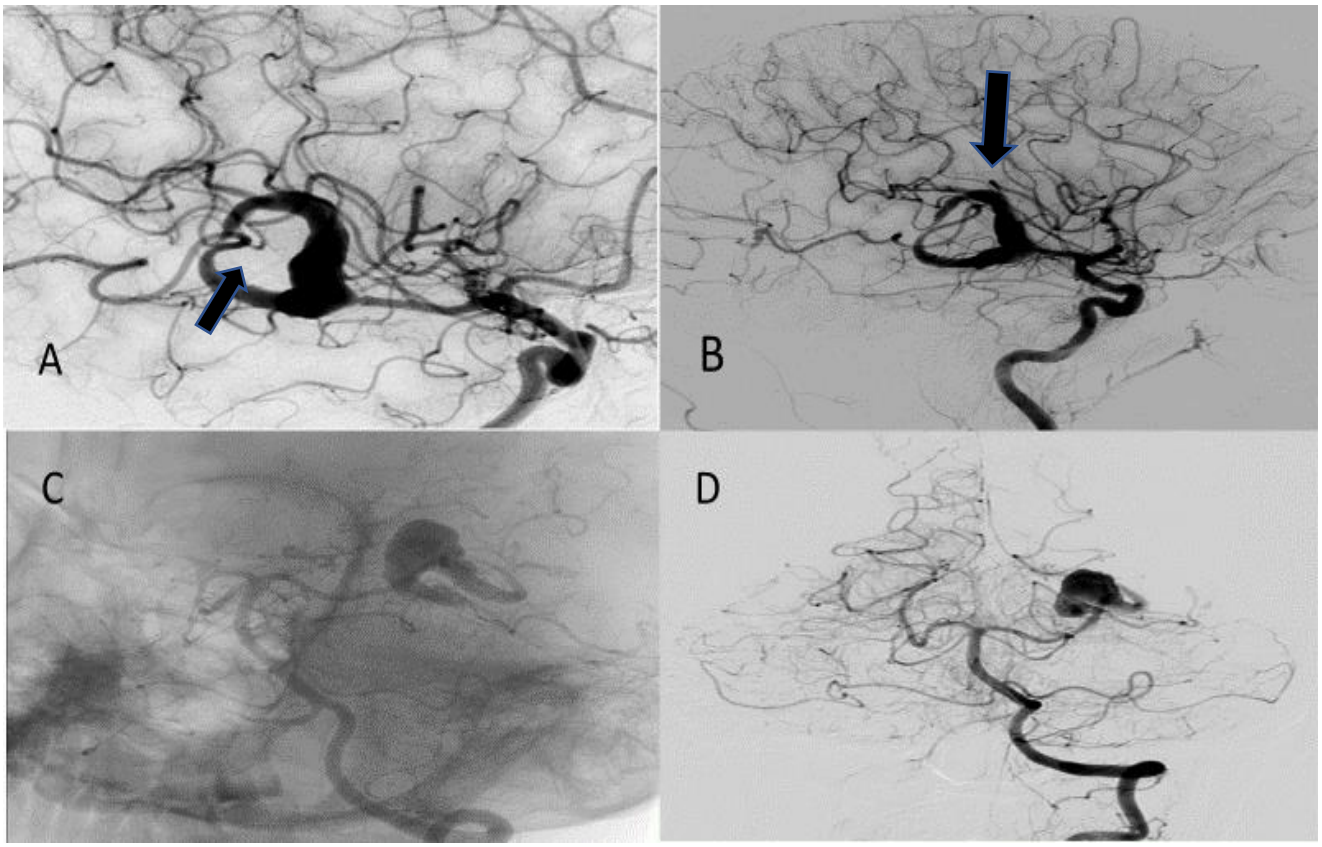

Figure 4A and B: DSA images of cerebral angiogram confirmed the presence of two fusiform aneurysm at the distal M1 segment of right MCA forming Maleficient's horn like appearance as seen in lateral oblique view and lateral view (Black arrow). Figure 4C and D: 3D reconstructed images and AP view DSA of vertebral artery showing smaller similar appearing aneurysm at P2 segment of left PCA.