CLINICAL CASE - TEST YOURSELF Neuro Imaging

Multiple cerebral lesions in a 45-year-old male with a history of a resected cardiac myxoma

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PART A

A 45-year-old male with a background history of a resected left atrial myxoma two months ago, was referred for further investigation after an epileptic episode. There was no history of trauma or fever. MRI of the brain was performed.

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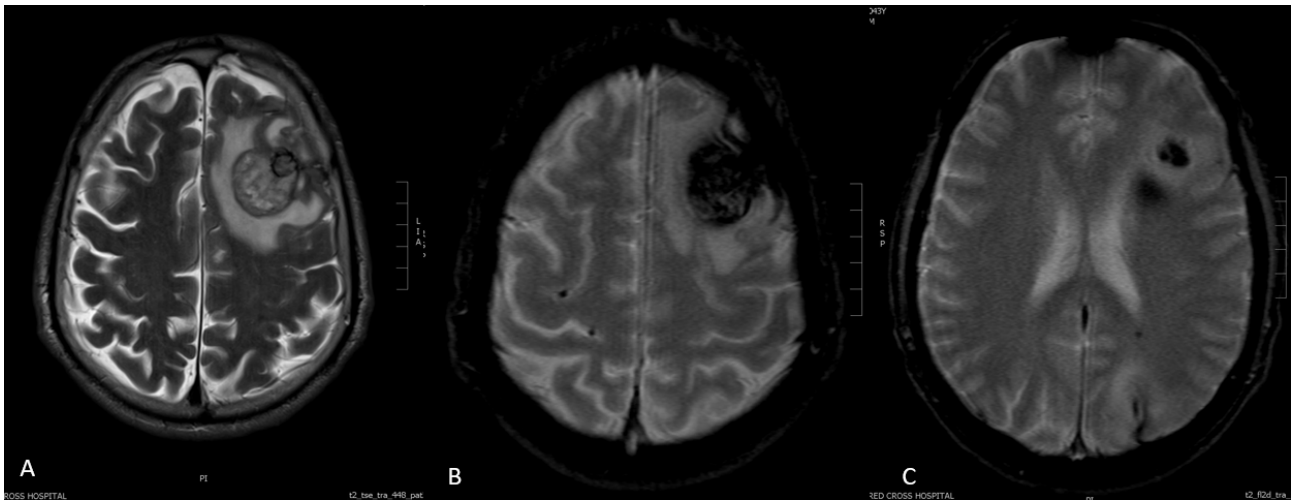

Image 1. *Axial T2-W image, SWI image (B,C).*

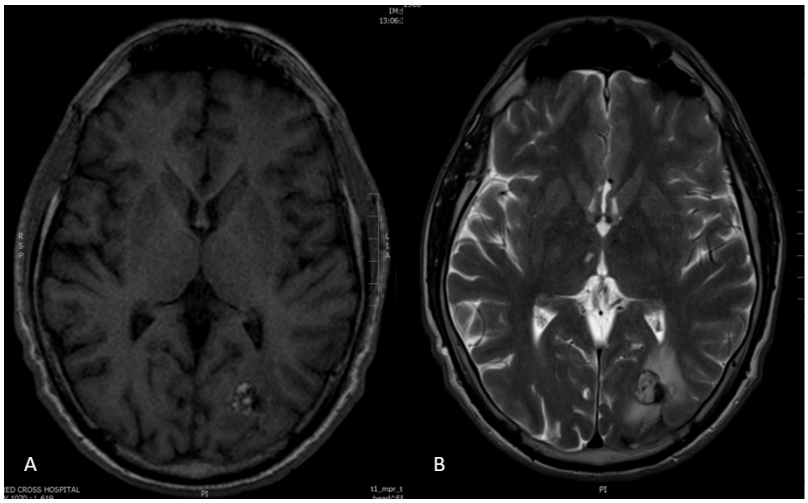

Image 2. *Axial T1-W image (A) , T2-W image (B).*

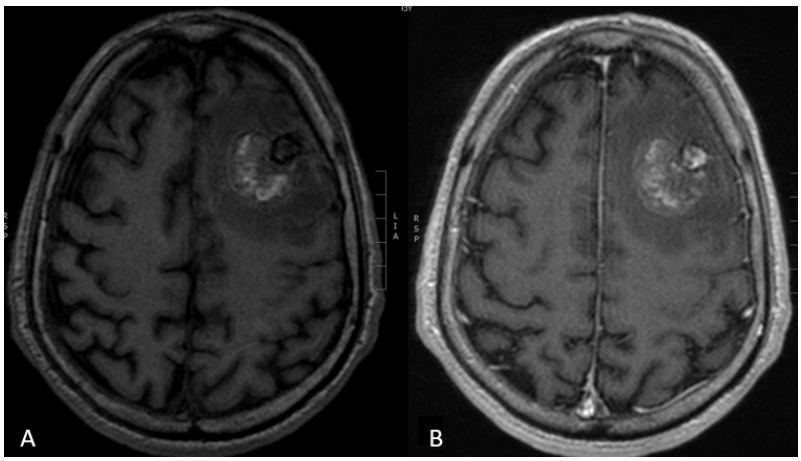

Image 3. *Axial T1-W images before (A) and after (B) contrast administration.*

PART B

Diagnosis: Myxomatous brain manifestations secondary to cardiac myxoma.

Brain MRI of our patient demonstrated multiple intraparenchymal lesions spread through both hemispheres with high signal on T1W images and intermediate signal on T2W images. The lesions showed heterogenous gadolinium enhancement, with areas of necrosis, and were surrounded by important perilesional oedema. Some of the lesions demonstrated a peripheral dark signal intensity rim on T2W images, probably due to accumulation of iron and hemosiderin from recurrent hemorrhages. SWI also revealed multiple punctate areas of hemosiderin in both hemispheres. During hospitalization, biopsy of one of the lesions was performed which revealed the myxomatous origin of the lesions. The final diagnosis of the patient was intracranial manifestations associated with left atrial myxoma.

Cardiac myxoma, a typically benign and slow-growing tumor originating from mesenchymal cells of the endocardium, can present with a variety of clinical syndromes beyond cardiac events [1]. Although histologically benign, this tumor has an embolization potential which is associated with the increased mobility of the tumor. Cardiac myxoma has also been associated to the autosomal dominant syndrome called Carney's complex, which is characterized by the triad of spotty pigmentation, myxomas and endocrine overactivity.

Neurological events have been reported in approximately 20-30% of patients with atrial myxoma, including headache, limb or body weakness, numbness and new-onset seizures [2,3]. Typically affecting younger patients, these complications can be caused by distant emboli that typically result in cerebral infarcts [4]. However, after acute myxomatous emboli, there are another two potential complications aside from infarcts: myxomatous emboli may invade the vessel walls leading to aneurysm formation or they may form space-occupying metastatic lesions in the brain parenchyma [5].

Brain myxomatous lesions are typically detected either at the same time or within a few months after the diagnosis of cardiac myxoma. However, cases of metastatic myxomatous brain lesions have been reported even 3 years after the diagnosis. These lesions are typically multiple, hemorrhagic with a tendency to appear in the frontoparietal region. Other sites of systemic myxomatous emboli of left atrium myxoma include the kidneys, bones and soft-tissues [6,7]. These metastases can be attributed to the dislodgement of components of the atrial myxoma in the systemic circulation. The main differential diagnosis of multiple intraparenchymal lesions with a high T1 signal and low T2 signal are hemorrhagic metastases and multiple cerebral cavernous malformations. Hemorrhagic brain metastases typically originate from melanoma, renal cell carcinoma, thyroid carcinoma and choriocarcinoma and less commonly from lung, breast and hepatocellular carcinoma [8].

The exact process behind the formation of aneurysms secondary to cardiac myxomas remains unclear. One hypothesis is associated with vascular and perivascular damage due to large emboli leading to scarring and subsequent pseudoaneurysm formation [9]. A second hypothesis involves infiltration of myxoma cells into cerebral blood vessels via the vasa vasorum, resulting in destruction of the arterial wall, similar to the pathophysiology of mycotic aneurysms [9]. The last suggested cause is the neoplastic process, where authors have observed myxomatous cells within the aneurysmal vessel. It is has been suggested that these neoplastic cells cause destruction of the arterial wall architecture and subsequent dilatation, forming the aneurysm by direct invasion of the endothelium [10]. Aneurysms due to cardiac myxomas are typically fusiform, multiple and tend to appear at peripheral vessel bifurcations distal to the circle of Willis.

Due to the rarity of these cases, there are no precise guidelines for the management of myxomatous

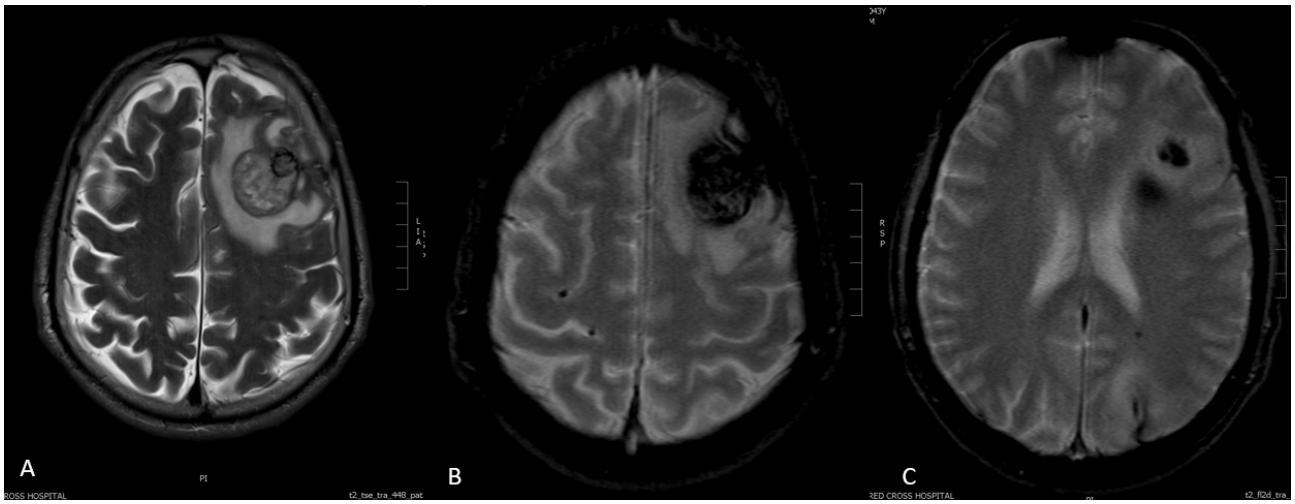

Image 1. *Axial T2-W image (A) revealing intracerebral lesions in the left frontal lobe with hemosiderin deposits and perilesional odema. SWI image (B,C) demonstrating punctuate areas of hemosiderin in both hemispheres.*

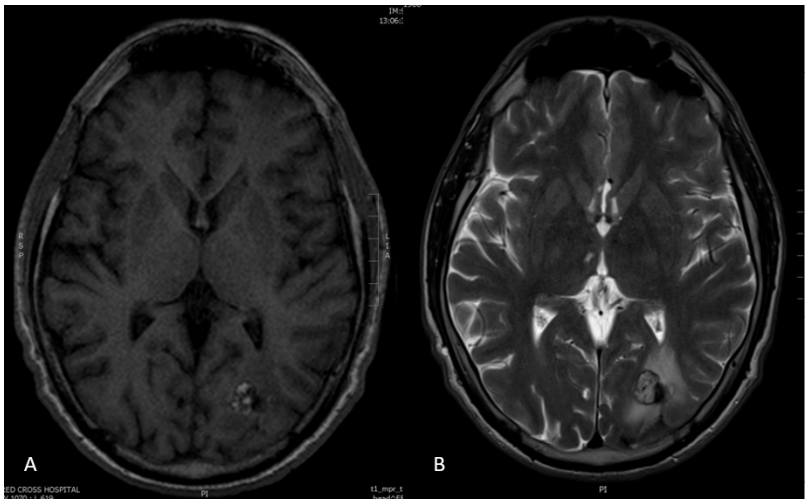

Image 2. *Axial T1-W image (A) showing another lesion in the right occipital lobe with a high signal on T1-W image and a peripheral dark rim on T2-W image (B).*

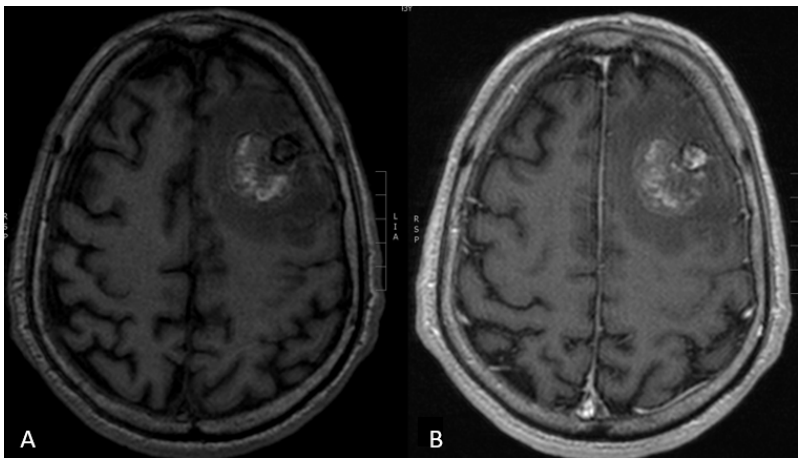

Image 3. *Axial T1-W images before (A) and after (B) contrast administration demonstrating the high signal onT1-W image and the heterogeneous enhancement of the lesion.*

brain lesions secondary to cardiac myxomas. Most authors suggested that surgery might be appropriate if only one or two lesions are identified or if a lesion causes significant mass effect. In cases of multiple intraparenchymal lesions radiotherapy or chemotherapy might be appropriate in order to obtain a larger period of time without recurrence [9].

In conclusion, as demonstrated in our case, long-term follow up of patients with a history of atrial myxoma is recommended. Atrial myxoma, although benign, can lead to some life-threatening complications associated with embolic fragments of the tumor. For patients experiencing embolic cerebral infarction associated with atrial myxoma, especially if myxomatous emboli are confirmed in another location, CTA or MRA should also be considered along. Apart from assessing the presence of myxomatous intracranial aneurysms, this particular group of patients should be closely monitored for the possible development of metastatic myxomatous lesions. **R**

KEY WORDS | Brain, cardiac myxoma, myxomatous lesions, hemorrhagic lesions

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