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Case report

Brain manifestations secondary to auricular myxoma ☆,☆☆

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ABSTRACT

Cardiac myxoma is the most common benign tumor of the heart. In most cases, it is in the left atrium, which can generate neurological embolization; that can manifest as an ischemic event, aneurysm formation and less frequently as brain metastases. This is a case report of a 56-year-old male patient with cerebral embolisms secondary to a left cardiac myxoma. In these patients, the role of neurologic imaging is to detect the firsts complications and avoid secondary complications.

Introduction

The cardiac myxoma is the most common benign tumor of the heart, accounting for 83% of benign tumors. The most frequent location is in the left atrium, 90% of the cases arise from the interatrial septum expanding to the atrium followed by the mitral valve, right atrium, right ventricle and left ventricle [1–3]. Patients usually present symptoms caused by an obstruction in the mitral valve, constitutional symptoms or embolic manifestations [4].

Although histologically it is a benign lesion; this tumor has an embolization potential that could be related to tumor mobility rather than its size. Clinical manifestations vary according to location, and since it is usually located in the left atrium; systemic and neurological embolization can occur in 50% of patients, with neurological complications being associated in 12% of patients. The most frequent manifestation is the cerebral ischemic infarction followed by cerebral aneurysms and cerebral metastasis [5,6].

Case report

A 56-year-old male patient was admitted to a local clinic for an oppressive chest pain and dyspnea. Initially, an echocardiogram was performed, reporting a rounded mobile mass in the left atrium adhered to the interatrial septum (Fig. 1). Surgical

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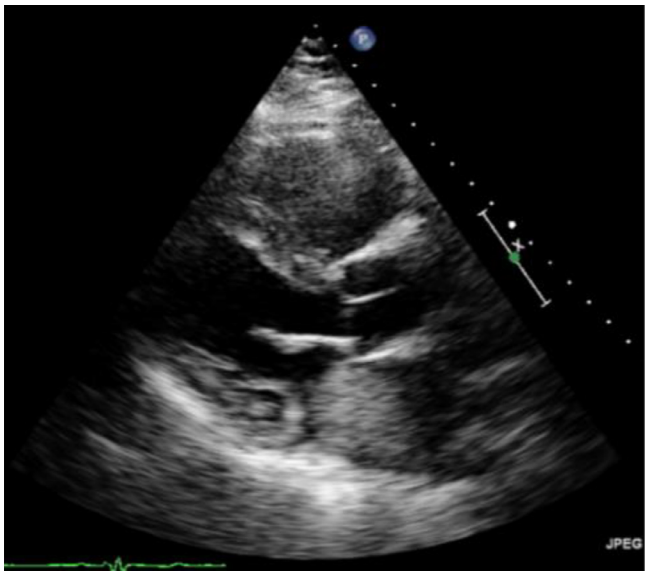

Fig. 1 – Rounded mobile mass in the left atrium adhered to the interatrial septum.

resection of the mass was performed, with histopathological report of cardiac myxoma.

The patient came back 5 months later, due to a sudden onset of decreased strength in the right upper limb, headache and disorientation. Initially, a simple brain computed tomography was requested, in which multiple hyperdense lesions were found, rounded, well defined, subcortical in location, in both cerebral hemispheres, with acute bleeding and moderate vasogenic edema surrounding them (Figs. 2A and B). The study was supplemented with a brain MRI with contrast finding multiple focal lesions, heterogeneous in all sequences, rounded, of various sizes, distributed in both cerebral hemispheres, which followed vascular pathways with areas of recent and old hemorrhage, with vasogenic edema and mild local compressive effect (Figs. 3A-D). A CT angiogram (Fig. 4) was performed and it excluded aneurysmal formations. A brain biopsy of one of the lesions was performed, with a histopathological report of hemorrhagic infarction secondary to cardiac myxoma embolization.

Discussion

Primary cardiac tumors are rare, with an incidence ranging from 0.02% to 2.8% of the general population. Three quarters of these tumors are benign (75% of cases), 83% of them are myxomas.

The cardiac myxoma is a slow proliferating tumor that originates from mesenchymal subendocardial cells, being more frequent in women between the third and sixth decade of life. In most of the cases, it presents as a sporadic lesion (93%), however, it can be a component of the Carney Complex, an autosomal dominant syndrome characterized by irregular pigmentation in the skin, myxomas, endocrine overactivity, testicular tumors and Schwannomas [1,7–9]. The most frequent location is in the left atrium followed by the mitral valve, right atrium, right ventricle, and left ventricle [3].

In most patients with myxoma, the clinical presentation is according to 3 mechanisms: pulmonary or systemic embolism, mitral valve obstruction and/or constitutional systemic symptoms. The most common initial symptom is generated by the obstruction of the mitral valve, including palpitations, dyspnea and heart failure, that have been reported in 63% of cases. Forty-five percent of patients present neurological symptoms, with cerebral ischemic stroke being this the most common manifestation [5,10].

The embolization is due to the detachment of the friable and spongy component of the cardiac myxoma into the circulation. The impact is in between 30% and 50% of cases and is related to the handling of the myxoma during surgery or secondary to the turbulence of the blood flow where they are located [6,7,11,12].

The site of the embolism depends on the location of the myxoma. Tumors located on the right-side lead to pulmonary embolisms or unusually embolisms at the systemic level when exist an intracardiac communication. In contrast, tumors located on the left side (more frequent) produce systemic embolism, including the brain, eyes, kidneys and coronary arteries [8,11].

The intracranial manifestations after the embolization are of 3 types: infarcts, aneurysms and brain metastases.

Infarcts: If the emboli remain intraluminal occluding the light, either by embolization of tumor particles or thrombotic material covered with tumor cells [13].

Aneurysms: They usually are presents as multiple lesions, most of them are fusiform. The most common location is in the middle cerebral artery and its branches [9,10].

Regarding to the formation of these aneurysms, it has been raised 3 mechanisms:

1. Infiltration of cerebral vessels by myxoma cells, through the vasa vasorum generating destruction of the architecture of the arterial walls and weakening of the subintimal tissue like the mechanism of mycotic aneurysms.
2. The tumor cells produce vascular occlusion and perivascular lesions with subsequent scarring and pseudoaneurysm formation.
3. The tumor cells produce destruction of the architecture of the arterial wall and subsequent dilatation, forming the aneurysm by direct transendothelial invasion [12].

Since the growth of this type of cells is slow, the embolic episode and aneurysm formation are usually separated by a significant amount of time [13].

Brain metastasis

They are rare, there are only a few cases reports, although the brain is the main site of metastasis of these tumors. They are secondary to an embolization of the tumor cells that transgress the vessel wall and leads to seeding in the brain parenchyma [9,12].

The tendency to metastasize has been related to the overproduction of chemokines CXC, interleukin-8 and oncogenes related to tumor growth produced by myxoma cells. Additionally, a role for interleukin-6 in the development of brain

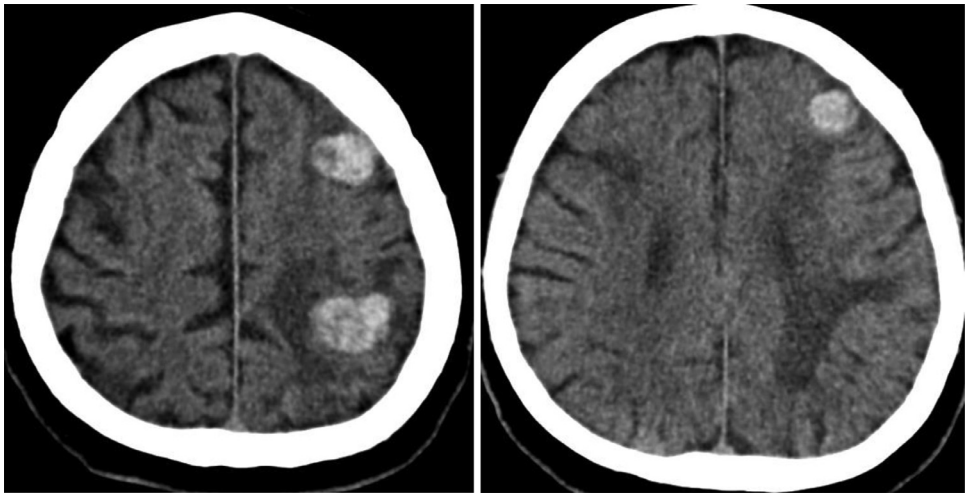

Fig. 2 – (A-B) Simple brain computed tomography (axial) with multiple hyperdense lesions well defined, subcortical in location with moderate vasogenic edema surrounding them.

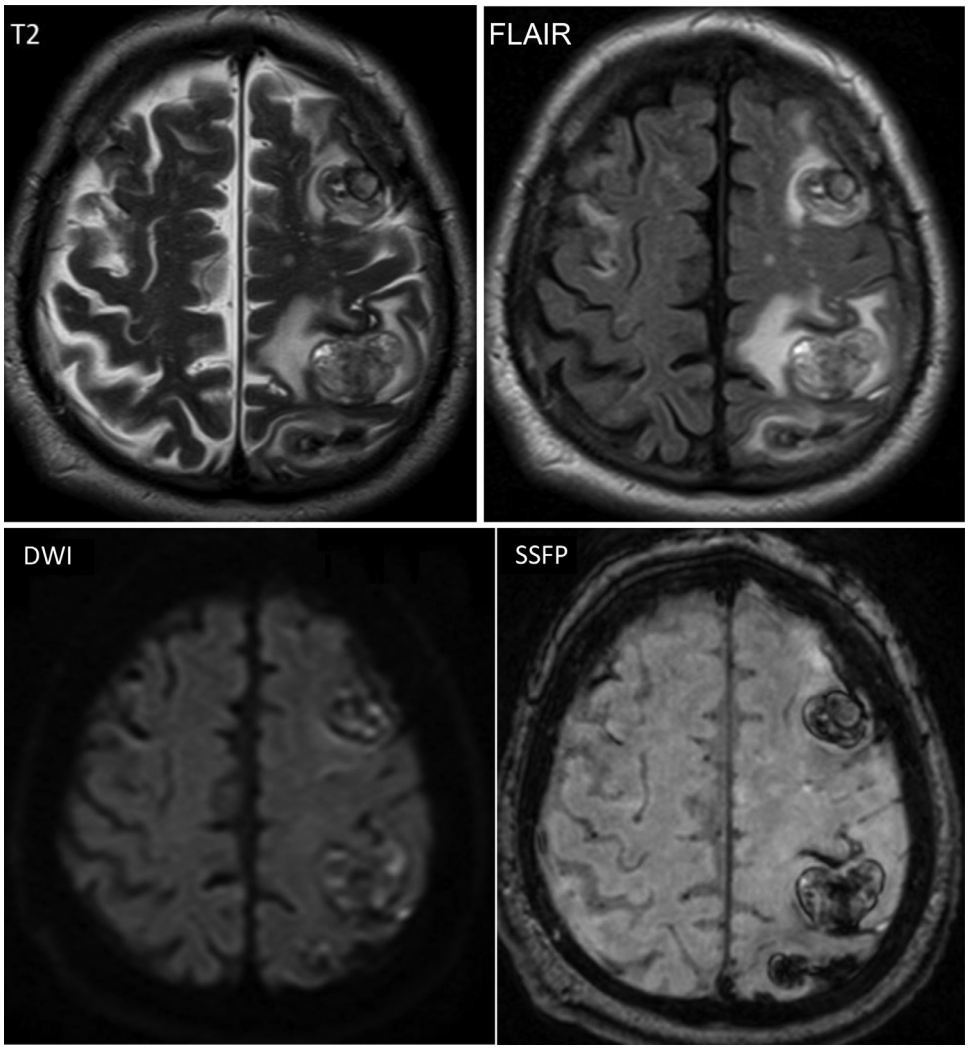

Fig. 3 – (A, B, C, D) Brain magnetic resonance with contrast finding multiple focal lesions, heterogeneous in all sequences which followed vascular pathways with areas of recent and old hemorrhage.

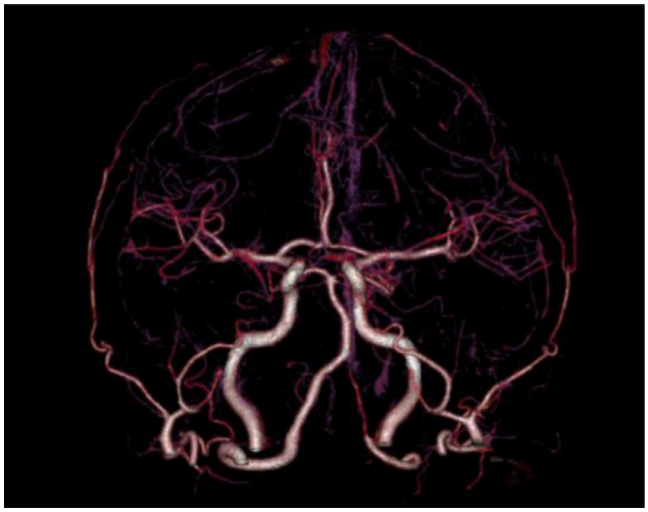

Fig. 4 – CT angiogram excluded aneurysmal formations.

metastases has been suggested, since the adhesion of myxomatous cells to brain endothelial cells could mediate by them [1,2,6,9].

Typically, these lesions are multiple, hemorrhagic, located in the frontoparietal region. They are usually identified at the time of the initial diagnosis of the myxoma or a few months after the diagnosis. However, cases of diagnosis of metastasis have been reported 3 years after the onset of symptoms. The natural history of these metastases is extremely variable, they are usually stable for years, completely asymptomatic, but can also manifest with neurological deficit [1,11,13].

The main differential diagnoses when multiple hemorrhagic lesions are found in the brain are metastases secondary to other neoplasms such as choriocarcinoma, melanoma or renal and thyroid carcinoma [9,14].

Regarding treatment, surgery is appropriate for diagnostic purposes in cases of 1 or 2 isolated brain metastases. Radiation therapy has had very good results in small, multiple lesions; palliative chemotherapy could be administered to patients with multiple brain metastases to obtain a longer period without recurrence [1,11].

In conclusion, the role of routine images in the detection of late neurological complications of cardiac myxomas has not been established, they are usually done when there are already symptoms, so we suggest that cardiac myxoma patients undergo brain MRI to detect and treat complications early.

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