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JCIMCR OPEN ACCESS Journal of Clinical Images and Medical Case Reports

ISSN 2766-7820

Case Report

Open Access, Volume 3

Recurrent stroke associated with brain metastasis due to a cardioembolism secondary to left atrial Sarcoma: Case presentation and review of the literature

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Received: Sep 13, 2022

Accepted: Oct 12, 2022

Published: Oct 19, 2022

Archived: www.jcimcr.org

DOI: www.doi.org/10.52768/2766-7820/2110

Abstract

Primary cardiac tumours are rare; 75% of primaries are benign mesenchymal, mostly myxomas. Primary sarcomas are the second most frequent type. Only a few hundred primary cardiac sarcomas have been reported, most diagnosed by autopsy. Sarcomas of the intima are polypoid mesenchymal masses anchored to a vessel or the cardiac wall. To our knowledge, there is a unique report of primary cardiac Sarcoma complicated with cerebral infarction and cerebral metastatic disease. Metastases show a preference for lung, liver, and mediastinum, but the brain is usually a sanctuary. We describe a female in her forty's with recurrent stroke associated with a cardiac sarcoma, also presenting brain metastases, who died a year after starting the problem by pulmonary thromboembolism.

Keywords: Cardiac tumour; Sarcoma; Stroke.

Background

Primary tumours of the heart are rare. About 75% of primary cardiac tumours are benign mesenchymal tumours, most of which are myxomas. Although very rare, primary sarcomas of the heart are the second most common type of primary cardiac tumours [1]. Currently, only a few hundred primary cardiac sarcomas have been reported, most of which are based on autopsy series [2]. Intimal sarcomas are malignant mesenchymal tumours originating in the intima as polypoid masses anchored to a vessel or the cardiac wall. They have a thrombus-like appearance [3,4]. There is a wide variety of possible clinical presentations. They appear by four distinct mechanisms: obstruction of blood flow and interference with valvular function; local invasion leading to arrhythmias or pericardial effusion; embolic phenomena; or systemic symptoms. All mechanisms may overlap and present in different combinations, resulting in a variable clinical picture [5]. Given the fragility of the tumour, patients often experience systemic embolisms, which may be the initial form of presentation [6]. The site of metastases shows a preference for the lung, liver, and mediastinum, but the brain appears to be an uncommon site of metastases [7].

Citation: Marcela GV, Davalos RP, Jorge VC, Cázarez RAM, Ildefonso RL, et al. Recurrent stroke associated with brain metastasis due to a cardioembolism secondary to left atrial Sarcoma: Case presentation and review of the literature. J Clin Images Med Case Rep. 2022; 3(10): 2110.

Clinical case

A woman in her 40s presented in ER in late May 2020 with sudden dizziness, followed by confusion and bradypsychia that resolved within minutes. However, it made left hemiparesis evident. She had a history of arterial hypertension (losartan 50 mg PO qDay). She denied tobacco, alcohol or illicit drug use and another relevant history. On examination, the heart was rhythmic and without murmurs. She had only mild left facial and body hemiparesis on neurological examination with brachial predominance, symmetrical muscle stretch reflexes ++, and bilateral plantar flexor response. There were no other significant data. A cranial MRI showed an acute infarct in the right middle cerebral artery territory, mainly in the right frontal operculum (Figure 1). Routine blood tests, including TSH, lipid profile and SARS-CoV2, were all normal.

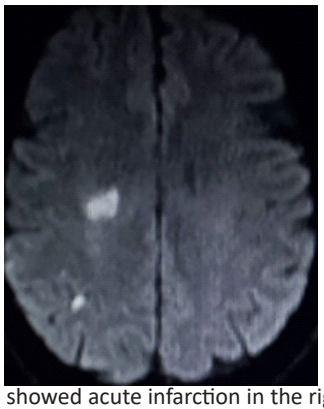

Figure 1: MRI showed acute infarction in the right middle cerebral artery, mainly involving the right frontal operculum.

A 24-hour Holter and transthoracic echocardiogram were reported without significant alterations. CT angiography was also irrelevant. The immunological and prothrombotic laboratory profile was also negative. In the following month, he presented sudden dysarthria. A new MRI showed ischemic lesions in the right middle cerebral artery territory (Figure 2).

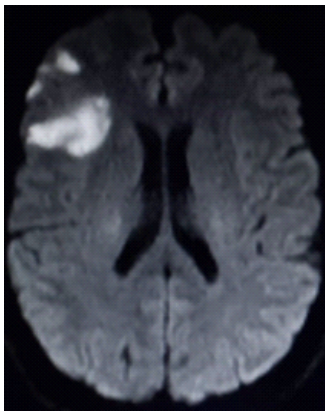

Figure 2: MRI shows a new small ischemic lesion within the watershed left middle cerebral artery territory.

A new transthoracic echocardiogram showed a 9 x 2 cm mass originating in the atrial roof, causing severe mitral valve stenosis and moderate insufficiency (Figure 3).

A new event was clinically present, and a new MRI showed infarct areas in the contralateral hemisphere (Figure 4).

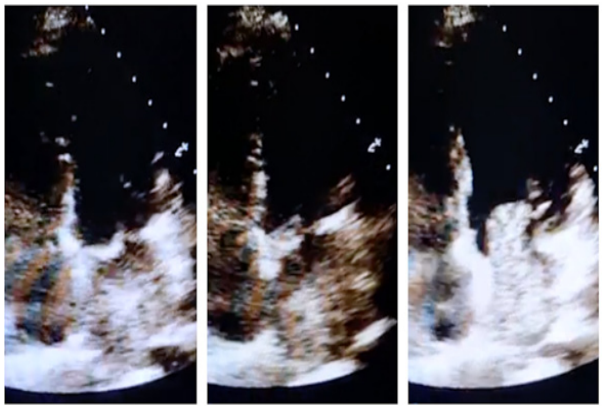

Figure 3: A transesophageal echocardiogram shows a 9 cm2 mass lesion originating in the atrial roof, causing severe mitral valve stenosis and moderate insufficiency.

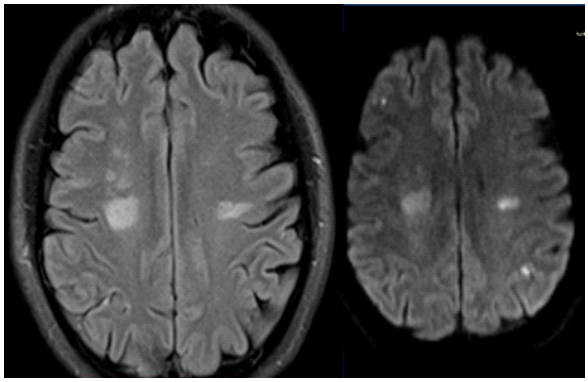

Figure 4: Secondary infarct bilateral cerebral areas showed in axial views in flair and diffusion MRI images.

The patient underwent surgery. Trans-operative findings of a tumour lesion in the left atrium invading the mitral valve and adjacent vessels and the histopathological diagnosis was a poorly differentiated intimal sarcoma was followed up by oncology. A whole-body 18F-FDG PET/CT scan was performed and was negative.

In late 2020, the patient presented for the third time with focal neurological symptoms, and a new cranial MRI showed an occupational process with perilesional oedema compatible with brain metastasis (Figure 5). Consequently, radiotherapy was started. Unfortunately, one year after starting the problem, the patient developed pulmonary thromboembolism and died.

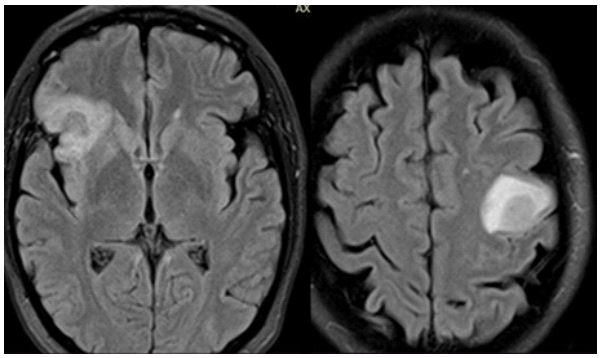

Figure 5: MRI showing a right peri-insular mass with perilesional oedema and a left frontal occupational process with perilesional oedema, both compatible with metastasis.

Discussion

Virtually all types of sarcomas in the heart have been described [10]. They are sporadic tumours. Their incidence is 0.03%, although cardiac Sarcoma is the most frequent primary tumour in up to 30% of cases [11,12]. It can occur at any age; its maximum incidence is around 40 years, affecting men and women equally, and survival is only 24 months or less after complete resection [13,14].

Its clinical presentation is varied; it may begin with a sudden onset, with positional symptoms of chest discomfort, dyspnea, orthopnea, cough, or syncope. Poor heart valve function results in heart failure, arrhythmias or obstructive symptoms, depending on the chamber affected [15].

Sun YP et al. in 2017 described the first report of a primary Sarcoma complicated with cerebral infarction and brain metastases after initial cardiac mass resection surgery. These tumours can lead to brain metastases and cerebral vascular complications such as infarcts, haemorrhages, and intracranial aneurysms [16]. Patients often suffer systemic embolisms, and the brain is a frequently affected site.

Diagnosis is with noninvasive techniques, with echocardiography (transthoracic or transesophageal) beneficial for this purpose. Cardiac CT and MRI help predict the cardiac mass's malignant nature, with features such as broad-based lesions. These lesions occupy the entire cardiac chamber, infiltrating the myocardium, pericardium or necrosis of adjacent structures and metastasis [17]. Transesophageal echocardiography is the first step in diagnosis because it is a reliable and inexpensive test showing the tumour's size, location, and fixation. MRI, CT, and PET can show the different cardiac extents and metastases present in up to 80% of patients at the time of diagnosis. A definitive diagnosis requires histologic examination [18].

The diagnosis in this patient was challenging because the first echocardiogram was reported negative, and we considered subclinical atrial fibrillation. Two new trials support evidence that prolonged cardiac event monitoring increases the detection of subclinical Atrial Fibrillation (AF) in patients with a Transient Ischemic Attack (TIA) or acute ischemic stroke. In the PER DIEM trial of patients with cryptogenic stroke, new AF was observed in 15.3 % of patients in the insertable cardiac monitor group (placed for one year) compared with 4,7% in the external loop recorder group (worn for four weeks) [19]. The STROKE-AF trial of patients with large or small vessel stroke detected new AF in 12.1% of patients in the insertable cardiac monitor group compared with 1.8% in the usual care group of dedicated external cardiac monitoring [20]. Fortunately, a second transthoracic echocardiogram was performed after the second ischemic stroke, and we made the correct diagnosis.

This presentation is interesting because of the rarity of cardiac sarcomas and the clinical presentation of a rare recurrent infarct associated with metastases. To our knowledge, there is only another report in the literature like the one we present.

The outcomes of these patients are unfavourable, with poor short-term survival. Patients treated with surgery alone have a median survival of 3 months to 1 year. Postoperative radiotherapy is effective, but its value is limited by the sensitivity of the heart to radiation, resulting in cardiomyopathy and chronic pericarditis [21].

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