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Primary cardiac angiosarcoma of the right atrium: a rare entity presenting with an atrial arrhythmia

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Introduction: Primary cardiac tumors are rare entities and 75% are benign. Angiosarcoma is the most common malignant primary cardiac tumor.

We report the case of cardiac angiosarcoma presenting with an atrial arrhythmia.

Clinical case: A 39-year-old female patient with no past medical history presented to the emergency department with heart palpitations and atypical chest pain.

Electrocardiogram on admission showed atrial flutter with a heart rate of 153 beats per minute.

Laboratory analysis were performed showing elevated D-dimer levels (2210 ug/L).

A thoracic CT scan was performed, which ruled out pulmonary embolism, but showed multiple pulmonary nodules and a right atrial (RA) mass measuring 48 mm that could correspond to a thrombus or neoplasia.

The patient was admitted in the Cardiology ICU of our hospital and was started on beta-blocker and amiodarone with conversion to sinus rhythm. Additional exams were performed:

- Transthoracic echocardiogram (TTE) revealed an heterogenous 32,6 x 17,7 mm mass in the lateral wall of the RA with an adherent mobile mass near the tricuspid valve with 28 mm diameter (possible adherent thrombus).

- Cardiac magnetic resonance imaging confirmed a RA tumor with invasion of the atrial free wall and compression of the superior vena cava.

Due to the unclear etiology of the RA mass, ultrasound-guided intracardiac biopsy was performed. Pathological examination revealed spindle cell proliferation, consistent with the diagnosis of angiosarcoma. Immunohistochemical staining was positive for Vimentin, CD34 and CD31, with 70% Ki67 expression.

Later on, the patient developed melena with significant drop of hemoglobin levels, requiring daily red blood cell transfusions and anticoagulation had to be stopped.

The patient was transferred to the Internal Medicine ward and thoracic-abdomen-pelvis staging computed tomography (CT) scan showed a significant increase in the number of pulmonary nodules, bilateral ovarian masses, 4 hepatic nodules and ileum metastization.

During hospitalization, the patient developed right leg deep venous thrombosis and thoracic CT scan revealed bilateral pulmonary embolism.

After improvement of the clinical status, palliative chemotherapy was started and the patient was discharged, maintaining regular outpatient follow-up in the Oncology Department for 1 month.

Cardiac angiosarcoma generally presents in a late stage of the disease with metastatic involvement. When surgical treatment is not possible, despite agressive chemotherapy, the prognosis remains poor.

Abstract P229 Figure. Echocardiogram: right atrium mass