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Case Report

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CARDIAC SARCOMA MASQUERADING AS MELANOMA: A CASE REPORT

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ABSTRACT

Primary cardiac sarcomas are a rare clinico-pathological entity difficult to diagnose and with a poor prognosis. A high degree of suspicion is required in young patients with a cardiorespiratory symptoms. This case tend to show the insidious nature and the aggressiveness of this pathology in a young patient of 24 years with a transthoracic echocardiogram showing a right ventricular mass, cervical and mediastinal lymphadenopathy at CT scan and a histopathological analysis in favor of melanoma.

INDEX TERMS: Cardiac Sarcoma, melanoma, myxomas, surgery.

1- INTRODUCTION

Primary heart tumors are rare, 75% are myxomas and only 25% are malignant with sarcomas being the most common histological type but remaining rare due to a general incidence of 0.0017% and 0.019%, thus rarely encountered with only a few cases having been described in literature.[1-3]

Symptoms are non-specific and depend on the chambers and the cardiac structures involved, so the diagnosis relies mainly on image modalities such as echocardiography, computed tomography, coronary angiography and magnetic resonance imaging.[1]

The urgency of diagnosis is set to avoid lethal cardiac complications such as arrhythmias, ischemia due to obstruction of coronary arteries, heart failure, sudden cardiac death, metastasis or delay in management.

Despite surgical resection and chemotherapy, prognosis remains poor with a median survival of 6–12 months.[7-8-9]

2- CASE

A 24 years old woman with no past medical history, non-smocking habits presented to the emergency department with a 6 month history of worsening palpitations, chest tightness and progressive dyspnea on exertion (NYHA Class II-III), all evolving in a context of fatigue lethargy and unencrypted weight loss, however no syncopal episodes reported.

Physical examination revealed BP 125/80 mmHg, sinus tachycardia of 120 b.p.m, an oxygen saturation of 95% on room air without supplemental oxygen, a respiratory rate of 22 breaths/ minute.

Examination revealed no peripheral edema and a non-elevated jugular venous pressure. Cardiac auscultation revealed a II/VI pan-diastolic murmur with no additional sounds and no right ventricular heave. The chest was resonant to percussion, but breath sounds were decreased in the right pulmonary base.

A thoracic scan revealed a mass of the right ventricle hypodense of 42mm not enhanced after injection of the contrast product and a low abundance pleural effusion on the right with pulmonary collapse.

A biopsy was done on a cervical adenopathy and identified it as a metastasis of a melanoma.

A specialized dermatological examination was carried out revealing only begnin like nevus.

A complement of the CT scan of the entire body was done and no other metastasis were revealed.

3 cycles of Dacarbazine were done with no clinical and radiological benefits noted.

In the meantime a Cardiac MRI showed the increase in the size of the cardiac mass.

After a multidisciplinary assessment, the cardiac surgical removal of the mass was planned by median sternotomy.

Due to the parietal infiltration The surgical resection of the mass was incomplete. The histological examination

showed a high-grade sarcoma with spindle cells (anti-HMB45 positif and anti-melanA positif).

After the rehabilitation from surgery, ifosfamide monotherapy was started but the general condition of the patient got worse and she died a week after her second cycle and 6 month from the diagnosis.

3- DISCUSSION

Primary cardiac sarcomas are malignant neoplasms deriving from pluripotent mesenchymal cells and confined to the heart, the molecular histogenesis is poorly known and there are no specific genetic mutations reported.[1]

The median age at diagnosis of cardiac sarcoma is 40–50 years.[7]

Studies suggest a male predisposition.[10]

It typically occur in the right atrium and rarely in the left. They often infiltrate into the septum and other structures of the heart resulting in significant outflow obstruction thus are very aggressive.[2.3]

In majority of cases, cardiac tumors can be silent; however, when symptoms occur, they are determined based mainly on the size and anatomical location of the tumor.[4] They classically present with signs of congestive heart failure depending on the obstructing atrium. Right heart being the prefered localisation it can mimick the symptoms of right sided heart typically dyspnea,syncope,. Most times, these sarcomas can be confused with benign conditions such as myxoma which are more common tumors of the left atrium.[6]

Most cardiac sarcoma are diagnosed by thansthorasic echocardiography, eventually followed by a presurgical transoesophageal echocardiography.[11,12]

A full body computer tomography and MRI are used for searching any signs of loco-regional and distant invasion.

In addition, MRI better defines the lipomatous, myxoid and thrombotic composition of intracardiac muscle.[12]

Cardiac surgery is the cornerstone of the treatment and is possible in about 33% of these cases, the chemotherapy is used for neo- and adjuvant purposes. Radiation therapy, a still debated issue due to high cardiac toxicity it can induce and is usually performed after surgery.[12.13]

Our patient rapidly deteriorated given a significant poor prognosis due to the extent of the mass coupled with the misinterpretation of the histologic diagnosis. The delay in the initiation of the right regimen of chemotherapy, the aggressive nature, and the incomplete resection of the tumour all conducted to the rapid decline. Early surgical resection with chemotherapy and or radiotherapy (even if the use of the later is controversial and can cause cardiac dysfunction) may had help improved the survival of our patient.

Auto-transplantation is also an option to be reserved for those patients in whom metastases have been excluded through a multidisciplinary evaluation. This approach has been developed to overcome the technical difficulties associated with the resection of left heart tumours, often hard to treat surgically, it would facilitate complete resection (R0) and avoid the immunosupprive treatments. However the perioperative mortality can reach 15%.[13]

CONCLUSION

Today more than ever, the need for more studies concerning the management of this rare entity is primordial and necessary specially the advanced and metastatic setting because the majority of cases are discovered at these stages given the non specific symptoms.

But more importantly this case call us to be alert and to keep the possibility of this diagnosis in mind when dealing with a young patient with cardio-respiratory signs.

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