

Primary Cardiac Lymphoma

Primary cardiac lymphoma (PCL) is a non-Hodgkin's lymphoma that occurs only in the heart. It is a very rare condition and is reported to be increasingly diagnosed pre-mortem. We present the case of an 87 year-old woman referred with a two-month history of dyspnea and peripheral edema. The transthoracic echocardiogram (TTE) and the computed tomography (CT) of the chest revealed an 8 cm right atrial (RA) mass. The tumor was excised, and the patient had a favourable immediate postoperative outcome.

The patient, with a 2-month history of shortness of breath worsening and increased leg edema was referred from a local hospital with diagnostic images suggesting a RA mass. Clinical examination, except for the peripheral edema, was unremarkable. Blood screening tests showed a GFR of 35 ml/min. The TTE revealed a large RA mass not resembling a thrombus and without extension into the inferior vena cava (IVC). This was confirmed by a CT scan, showing that the mass was 8 cm in size and had a broad attachment extending from the roof of the RA to the interatrial septum (IAS) (Figure 1). There was no other intrathoracic or abdominal pathology.

Despite her age, surgical removal of the tumor was justified, due to its obstructive nature and the symptoms it caused.

During surgery, the tumor did not appear to be a myxoma. Full resection was not possible due to invasion of the RA wall. The IAS was resected and closed with a CorMatrix patch. During the first few hours after the operation, the patient developed sudden



Fig. 1. Image confirming cardiac tumoral mass (see text for description).

ventricular fibrillation and cardiac arrest, requiring a new urgent sternotomy. There was no evidence of tamponade or bleeding, and after internal cardiac massage and a single internal defibrillation, the heart recovered sinus rhythm.

No cause for this event could be found. The patient made a satisfactory recovery, with only acute kidney injury but maintaining good diuresis. She was transferred to her local hospital on the 13th postoperative day, for the oncology team to decide on further treatment.

Histology results indicated a diffuse type B cell lymphoma.

Primary cardiac lymphomas are rare. The rate of occurrence is less than 1.5% of all primary cardiac tumors. The most common clinical presentations are heart failure, pericardial effusion and atrioventricular block. The histological type in immunocompetent patients is diffuse large B cell-type in more than 80% of cases. In immunocompromised patients there is a high chance of much more aggressive cell types.

In general, the prognosis is poor, although patients are very sensitive to chemotherapy. Median survival is 12 months. The preferred localization is the right atrial or ventricular wall (92% of cases), followed by the left atrium and ventricle.

The diagnostic approach is made by TTE, magnetic resonance and CT scan of the chest, abdomen and pelvis. CT scan is particularly useful to exclude other sites of disease.

The main differential diagnoses are myxoma, other cardiac tumors, or thrombus formation.

Surgery is usually indicated in the presence of an intracardiac tumor, although resection is usually incomplete due to the invasive characteristic of the PCL.

Once the diagnosis is confirmed by histopathology, chemotherapy is the best treatment option. Cases of RA wall perforation have been reported caused by tumor lysis syndrome after chemotherapy.

Primary cardiac lymphomas are very rare, but they are currently diagnosed more frequently pre-mortem. Absence of extracardiac diseases is associated with moderate survival improvement, but overall, prognosis is very poor.

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Secondary Cardiac Plasmacytoma

Malignant cardiac tumors are rare and, among them, cardiac plasmacytoma is extremely uncommon.

We present the case of a 68-year-old woman with multiple myeloma (MM) under chemotherapy treatment, who was admitted to the emergency department due to a 3-month history of episodes of dyspnea and symptoms suggestive of a heart condition. A PET-CT scan had previously revealed multiple hypermetabolic masses mainly in the right chambers, consistent with an active cancer process. A magnetic resonance imaging (MRI) was performed for better anatomical mapping of the interference of these masses with cardiac motility.

In addition, the purpose of this work is to present a brief update of this uncommon heart condition, and to highlight the usefulness of imaging methods, especially PET-CT scan and MRI, which have been shown to be valuable for proper diagnosis and follow-up.

The patient consulted for a 3-month history of dyspnea. A control PET-CT scan revealed multiple hypermetabolic lesions located mainly in the heart and affecting the right adrenal gland, the right pelvic cavity, and the abdominal wall (with no evidence of bone hypermetabolic lesions), consistent with an active cancer process (Figure 1). In view of this condition, MRI was suggested for better anatomical discrimination, and to study the cardiac dynamics affected by these masses.

The study was performed using a Siemens Avanto 1.5T system (Siemens, Erlangen, Germany) with surface coils and ECG gating. All the images were obtained during a simple apnea. Anatomical spin-echo, STIR sequences and functional SSFP (True FISP) cine sequences were performed in 2, 4 and 3 chamber cardiac axes, and short-axis sequence from base to apex.

Image analysis showed multiple isointense, pedunculated masses with respect to the myocardium in SSFP cine-MRI and T1 sequences, behaving as hyperintense in STIR sequences.

Two pedunculated masses were identified in the interventricular septum, one of them prolapsing towards the left ventricular outflow tract during systole. Two images in the right atrium were also observed, one of them implanted in the tricuspid annulus and prolapsing towards the ventricular chamber.

STIR images (Figure 2) showed hyperintense fo-

cal areas in mid inferior, mid inferolateral, and basal anteroseptal segments, which would be associated to myocardial infiltration areas due to the tumoral process. Despite these findings, biventricular systolic function was preserved.

Diagnosis was made through leukemia and lymphoma immunophenotyping of the cardiac masses by flow cytometry, revealing a cytometric profile consistent with moderate size LNH B CD10+. These findings were consistent with plasma cells and suggestive of plasmacytoma.

Multiple myeloma (MM) is characterized by uncontrolled proliferation of plasma cells, generally restricted to the bone marrow. (1) Extramedullary MM spread occurs when malignant plasma cells form tumors (plasmacytomas) in other parts of the body outside the bone marrow, whose reported incidence in newly diagnosed MM ranges from 7% to 18% of patients, or 6% to 20% of cases during the course of the disease. (1-3)

The mechanisms of extramedullary spread in MM are poorly understood. Extramedullary spread in MM can have two different origins: direct extension from skeletal tumors when they disrupt the cortical bone; or hematogenous metastasis to distal organs. However, scientific evidence is not enough to demonstrate it. (1)

Extramedullary masses may arise as primary plasmacytomas in patients with normal bone marrow analysis, or manifest themselves in patients with confirmed diagnosis of MM (secondary plasmacytoma), the latter being more aggressive. (4-6)

According to published series, 80 to 85% of patients with extramedullary MM had plasmacytomas in muscles, tendons, fat, or digestive tract, and 15% of the cases in glands, liver, kidneys, respiratory airway, skin, or breast; cardiovascular system involvement was extremely rare or absent. (4, 5, 7) In a review of the literature, Keung et al. found 9 cases of extramedullary MM involving the heart followed by a report

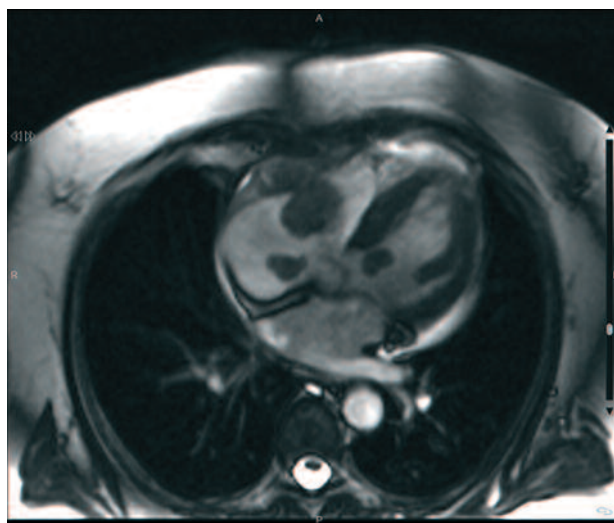


Fig. 1. See text for description.