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SECONDARY CARDIAC TUMORS: ABOUT TWO CLINICAL CASES AND REVIEW OF THE LITERATURE

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Abstract: Secondary cardiac tumors remain a severe and relatively rare pathology but belong to a rapidly expanding field of cardio oncology. They constitute a highly heterogeneous group of pathologies whose clinical manifestations vary according to tumor size and location. These tumors are typically associated with advanced stages of the primary tumor and therefore have a poor prognosis.

We report 2 cases of secondary cardiac tumors, the 1st a cardiac metastasis of a primary mediastinal sarcoma and the 2nd a uterine leiomyosarcoma. In these 2 cases, we will review the clinical presentation of cardiac metastases and discuss the mode of dissemination, depending on the primary location. Through a review of the literature, we also take stock of the evolutionary and therapeutic modalities of cardiac metastases.

Index Terms - Cardiac metastases, leiomyosarcoma, mediastinal sarcoma, cardiac tumors, echocardiography.

I. Introduction

Although secondary tumors of the heart are 40 to 100 times more frequent than primary tumors, they are rare and generally represent a very advanced stage of the cancerous disease. [1, 2] In autoptic series, secondary cardiac localizations are found in 1-10% of patients who die of cancer [2]. Tumors that give rise to cardiopericardial metastases include lung cancer, breast cancer, melanoma, kidney cancer and thymoma. Other tumors such as cancer of the uterus, stomach, rectum, liver and lymphomas rarely give rise to cardiac metastases.[2]

Adenocarcinomas are the most commonly described histological type (36%), the others being rarer [1]. Clinical expression is generally delayed, inconsistent and non-specific.

Diagnosis is based on clinical, epidemiological and imaging modalities (Transthoracic echocardiography (TTE), Transesophageal echocardiography (TEE), CT, MRI and PET-CT), with no systematic need for biopsy. Cardiac invasion is generally associated with a poor prognosis, and treatment is difficult.

Treatment depends on tumor type and origin and may involve chemotherapy or palliative measures.

We report 2 cases of secondary cardiac tumors, the 1st a cardiac metastasis of a primary mediastinal sarcoma and the 2nd of a uterine leiomyosarcoma.

Through these 2 clinical cases, we discuss the clinical presentation of cardiac metastases, the mode of dissemination according to the primary localization and, through a review of the literature, we take stock of the evolutionary and therapeutic modalities of cardiac metastases.

II. OBSERVATIONS:

Case1:

57-year-old female patient with no cardiovascular risk factors who presented with an altered general condition and underwent a CT scan objecting to a mediastino-pulmonary mass.

The patient underwent thymectomy. Histological examination of the surgical specimen was inconclusive. Six months later, the evolution was marked by the appearance of subcutaneous nodules whose anatomopathological examination of their biopsy objectified an undifferentiated pleomorphic sarcoma of grade || (according to the FNCLCC).

A second chest CT scan (Figure 1) (Figure 3) revealed a mediastinal mass interposed between vascular structures: anterior to the pulmonary artery, posterior to the descending aorta, superior to the aortic arch and inferior to the heart cavities; it was isodense and heterogeneous on spontaneous contrast, measuring 6.7x4.8x5.58 cm with passive atelectasis of the right pulmonary hemifield and appearance of a nodular formation in the soft tissues of the right hypochondrium of secondary appearance.

Transthoracic echocardiography (TTE) (Figure 2) showed a mass attached to the lateral wall of the mobile left atrium measuring 40x14mm, prolapsing the mitral valve and the base of the left ventricle during diastole, non-obstructive, with a second echogenic mass measuring 06x05mm adherent to the atrial side of the large mitral valve, suggesting a secondary localization of his mediastinal sarcoma.

Thoracic MRI showed the left paracardiac mediastinal mass in heterogeneous hypersignal T2, moderately enhanced after gadolinium injection. This mass arrived medially in intimate contact with the pericardium opposite the left ventricle (LV), with a pericardial defect measuring 13mm, suggesting pericardial infiltration by the mediastinal mass.

After multidisciplinary consultation involving oncologist, radiologist, thoracic surgeon, anatomopathologist and cardiologist, the diagnosis of a primary mediastinal sarcoma with secondary metastasis has been retained.

Chemotherapy (CT) was indicated, but unfortunately the patient died before the end of treatment.

Case 2:

A 55-year-old patient with no cardiovascular risk factors consulted the gynecology department for pelvic pain and metrorrhagia. Pelvic ultrasound and MRI showed a polymyomatous uterus with satellite pelvic adenopathy and a permeating parietal nodule. Hysterectomy was indicated. Pathological examination of the total hysterectomy specimen with right lymph node dissection revealed a high-grade undifferentiated uterine stromal sarcoma infiltrating more than 50% of the myometrium with lymph node metastasis. A confirmatory immunohistochemical study concluded that it was a leiomyosarcoma.

As part of the extension work-up, a thoraco-abdomino-pelvic CT (TAP CT) and a Positron emission tomography (15FDG PET) were performed.

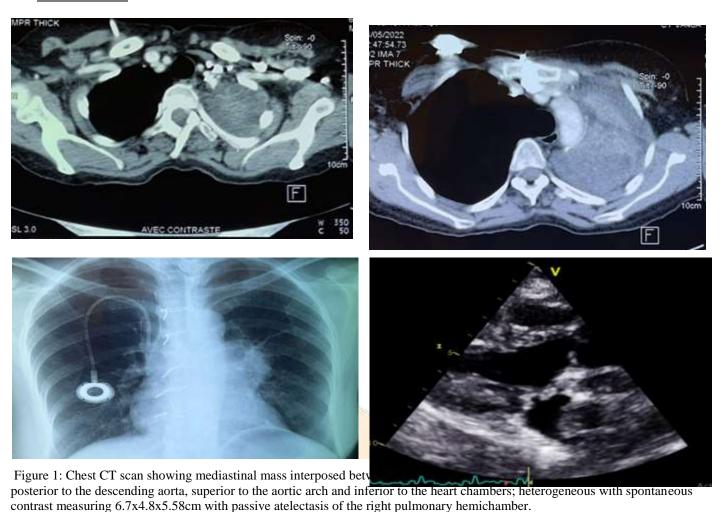
15FDG PET revealed a voluminous hypermetabolic cystic mass with an ametabolic center, located suprapubic and paravesical, associated with multiple hypermetabolic pulmonary parenchymal and subpleural nodules disseminated to both lung fields, with a secondary appearance.

TAP CT showed two tumor masses in the left anterior abdominal wall (Figure 5), pulmonary lesions suggestive of a secondary location (Figure 6) and a mass in the left atrium (LA) measuring 31x23 mm long axis.

With the onset of tachycardia and dyspnea, and the appearance of an intra-LA mass on the TAP scan, a TTE was performed (Figure 4), revealing a large, heterogeneous, fixed LA mass with a septal and inferior implantation base, measuring 30x23mm, raising the possibility of secondary cardiac metastasis with circumferential, moderately abundant pericardial effusion and fibrin deposition.

The patient underwent adjuvant CT but unfortunately died.

The first case:



The second case:

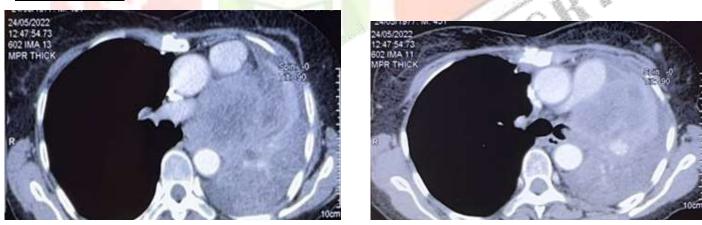
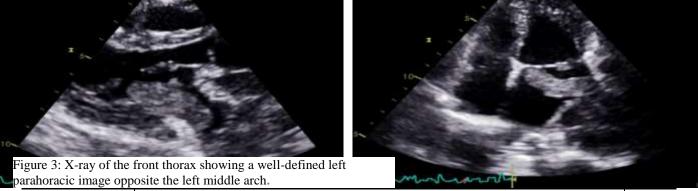
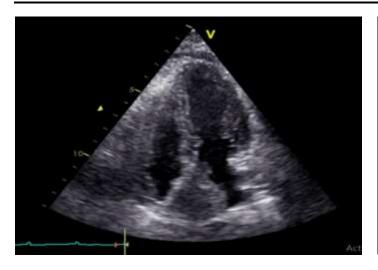


Figure 2: Transthoracic echocardiography (TTE) images showing a mass attached to the lateral wall of the mobile left atrium measuring 40x14mm prolapsing the mitral valve and the base of the left ventricle during non-obstructive diastole, with a second echogenic mass measuring 06x05mm adherent to the atrial side of the large mitral valve, all suggesting a secondary localization of mediastinal sarcoma.





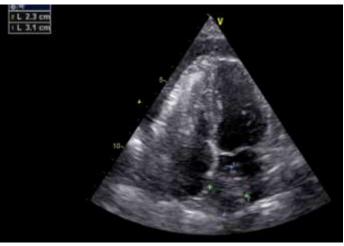
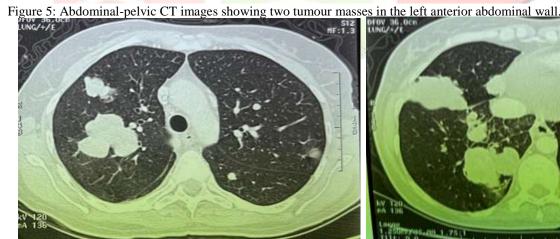


Figure 4: Trans-thoracic echocardiography images showing a large, heterogeneous, fixed mass in the left atrium, based on septal and inferior implantation, measuring 30x23mm, suggesting cardiac metastasis.







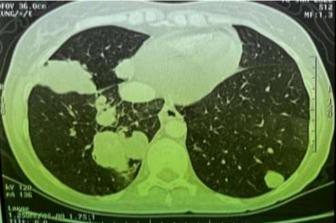


Figure 6: Chest CT scan showing pulmonary lesions suggesting a secondary location of the uterine tumour.

III. DISCUSSION

Secondary cardiac tumors are rare and often represent the final stage of cancerous disease.

The most common primary tumors are lung, breast, melanoma, kidney and thymoma, while uterine, stomach, rectal and lymphomas rarely metastasize to the heart [2].

Secondary cardiac invasion occurs through lymphatic or hematogenous extension, or directly by contiguity (mediastinal tumor), as in the case of our patient with mediastinal sarcoma. Involvement of the various cardiac tunics (endocardium, myocardium, epicardium, pericardium), and thus the topography of lesions, is related to the mode of dissemination of the primary neoplasia. Hematogenous dissemination may result in nodular myocardial involvement linked to emboli in the coronary network (melanoma, sarcomas) or, more rarely, endocardial involvement.

Mediastinal lymphatic extension is the most frequent pathway, with pericardial involvement found in lung, breast and hematological neoplasia [2].

Extension via the pulmonary and systemic venous networks may occur via the pulmonary veins, as in 2nd case, or via the superior vena cava in bronchial cancer, or via the inferior vena cava in certain cancers of subdiaphragmatic origin (kidney or liver carcinomas and sarcomas)[3].

Renal, testicular and hepatocellular cell carcinomas can sometimes extend into the inferior vena cava and progressively develop in the right atrium. Lung tumors can invade the heart directly through the pulmonary veins. This is a rare condition, particularly when the lung tumor is small and located far from the heart [3]. This was the case in our patient, whose right pulmonary metastasis led to left atrial metastasis via the pulmonary veins.

In the series by M. Krime, we noted more frequent involvement of the LV in pulmonary neoplasia, which is rather in favour of involvement by contiguity, and more frequent involvement of the RV in ENT and pelvic neoplasia, which follows the pathophysiological hypothesis of hematogenous dissemination [4].

The clinical presentation of cardiac metastases is usually silent, as in the case of our two patients. Diagnosis remains difficult because of the silent symptomatology and must be evoked in the presence of unusual cardiac clinical manifestations in a patient with neoplasia.

Lipothymia and syncope are also a frequent presentation of left-sided cavity tumors, and are the expression of obstruction to blood flow, usually through the mitral valve.

On the other hand, these secondary tumors often remain asymptomatic, and are often diagnosed during extension studies of the initial disease or found at autopsy.

Invasion of the left heart can be a life-threatening event, leading to a number of complications such as pulmonary venous outflow obstruction[5], cardiac tamponade [6], ventricular arrhythmias [7], complete atrioventricular block[8], left ventricular outflow obstruction[9] and myocardial infarction [10].

The discretion of clinical signs and the non-specific nature of simple paraclinical examinations justify the interest of other non-invasive imaging techniques. The diagnosis of cardiac metastases is generally made by echocardiography, CT scan as in our 2 patients, or even MRI or, more recently, PET-scanner.

TTE is essential for diagnosis and characterization of the cardiac mass. TEE confirms the anatomical site, the valvular involvement and the possibility of tumor resection.

On MRI, there is no specific aspect of these cardiac invasions, whose secondary nature is suggested by the neoplastic or metastatic context and the multiplicity of lesions. Secondary tumors frequently appear hyposignal T1, hyper-signal T2 with more or less marked enhancement. In cases of disseminated disease, biopsy of the cardiac lesion for diagnostic purposes is not performed [2].

The therapeutic decision is determined by the extent of the disease. When the heart is the only metastatic site, excisional surgery may be attempted [2]. In the 1st case, excisional surgery was not performed, given the location of the mediastinal mass, which was interposed between the vascular structures.

Resection is usually incomplete in the case of metastatic cardiac disease, as opposed to benign tumors or even primary malignancies. Heart transplantation is never indicated for cardiac metastases.

In metastatic forms, systemic chemotherapy remains a therapeutic option, adapted to the primary tumor[2].

The prognosis for cardiac metastases is generally poor in the short term, and treatment is usually only palliative, with an average survival of no more than 04 months according to the literature [2].

IV. CONCLUSION

The clinical diagnosis of cardiac metastasis should be made when cardiovascular symptoms appear in patients with known neoplasia. Despite advances in treatment, cardiac metastases are still the least understood of all metastases and remain highly controversial in terms of oncological treatment. Cardiac invasion is generally associated with a poor prognosis, but earlier diagnosis and multidisciplinary management could improve survival.

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