Case study

Cardiac localizations of Hodgkin lymphoma: a case report

Introduction:

Cardiac involvement during Hodgkin lymphoma is rare and is often seen at a late stage of the disease. The diagnosis of cardiac localization is difficult to make while the patient is alive, Transthoracic echocardiography is used to identify cardiac involvement in most cases, but other imaging techniques such as CT or MRI are usually used. Histological evidence is essential for diagnosis and the prognosis of these localizations remains guarded despite advances in the treatment of lymphomas in general.We report a case of cardiac localization of Hodgkin's lymphoma with a pericardial effusion.

Case Report Summary:

Mr. L. A, 42 years old, followed for Hodgkin lymphoma in complete remission for 3 years, presented to the emergency room for dyspnea .

On admission, the patient was conscious, heart rate was100 bpm, BP=129/80 mmHg, Cardiac auscultation objective a regular rhythm with muffled heart soundswithout signs of right heart failure. The ECG showed a sinusal tachycardia at 100 bpm, with a diffuse micro-voltage and negative T waves in the anteroseptal.

The TTE and CT scan allowed to orientate the etiological diagnosis a by revealing the presence of an voluminous mass in the right ventricle (RV), It was adherent to the free wall of the RV which it infiltrated, without a clear boundary between the healthy myocardium and the tumoral process. It extended into the RV outflow tract with the presence of a second mass in the pulmonary tract and in the trunk of the pulmonary artery C associated with a large circumferential compressive effusion, PET-FDG exploration was performed and itwas in favor of a lymphomatous dissemination at the mediastinal level.

After the six course chemotherapy, the myocardial tumor infiltration and pericardial effusion regressed; the general condition improved.

Discussion:

Hodgkin's disease is now considered a special type of B lymphoma, characterized by the proliferation of large cells called Reed-Sternberg cells within a characteristically reactive lymphoid tissue.

Cardiac lymphomas, whether primary or secondary, are rare; their clinical manifestations are infrequent and the diagnosis of the cardiac localization is rarely made during the patient's lifetime.

MRI is a relevant technique to study a cardiac mass because of its high signal resolution. high signal resolution. The CT scan offers a less good tissue contrast but because of its high spatial resolution, it allows to better specify the anatomical relationships of the lesion (coronary arteries, pericardium). These 2 examinations can complement each other's, and allow guided biopsies to be performed in order to confirm the histological nature of cardiac masses.

The 18-FDG PET scan plays an important role in the initial diagnoses and also for the evaluation of the treatment effectiveness.

Treatment usually involves a protocol of the chemotherapy and/without radiotherapy with a generally good evolution.

Final considerations:

Most cases of cardiac lymphoma are aggressive, progress rapidly and often have life-threatening complications such as heart failure and arrhythmia. It is therefore important to diagnose and treat them quickly. Moreover, the prognosis is poor because no clear treatment guidelines have been established.

Keywords: Hodgkin's lymphoma, pericardial effusion, cardiac involvement, dyspnea

Introduction:

Cardiac involvement during Hodgkin lymphoma (HL) is rare and is often seen late in the course of the disease. The diagnosis of cardiac involvement is difficult to make during the patient's lifetime. This is certainly due to the insidious character of this disease which rarely expresses itself in a noisy way and whose clinical symptoms are not specific. We report an observation of Hodgkin's lymphoma with cardiac involvement illustrating a mode of discovery such as dyspnea associated with pericardial effusion.

Clinical case:

Mr. L. A, 42 years old, followed for a scleronodular Hodgkin's disease since 2011, treated three times by chemotherapy with obtaining a remission respectively of 9 years, 6 years, and 3 years in complete remission since 3 years, presents to the emergency room for a dyspnea NYHA stage III with episodes of nocturnal paroxysmal dyspnea. This symptomatology evolved in a context of apyrexia, asthenia and weight loss. The clinical examination on admission found a conscious patient with a heart rate of 100 beats/min and a blood pressure of 129/80 mmHg. Cardiac auscultation objective a regular rhythm with muffled heart sounds without signs of right or left heart failure. The pleuropulmonary examination was normal. There was no peripheral adenopathy or splenomegaly. The EKG showed a regular sinus rhythm at 100 beats per minute, with a diffuse micro-voltage and negative T waves in the anteroseptal.

The TTE showed a pericardial effusion which was circumferential, of great abundance, compressive without significant respiratory variations, with thickening of the two pericardial sheets and the presence of an echogenic, voluminous mass in the right ventricle (RV), It was adherent to the free wall of the RV which it infiltrated, without a clear boundary between the healthy myocardium and the tumoral process. It extended into the RV outflow tract with the presence of a second mass in the pulmonary tract and in the trunk of the pulmonary artery (figure 1 and 2). Biology showed an inflammatory syndrome. LDH is elevated to 800 U/l (N < 290 U/l).

The thoracic Angio scan shows lymphomatous mediastinal adenopathy associated with two masses of the RV and the trunk of the pulmonary artery with invasion and abundant pericardial effusion and bilateral pulmonary micronodules (figure 3).

PET-FDG exploration is in favor of a lymphomatous dissemination at the mediastinal level: at the level of the known cardiac mass which is intensely pathological hypermetabolic (SUVmax=26) with a diffuse and intensely pathological hypermetabolic infiltration of the anterior mediastinum (SUV max=21) measuring 147x62x115 mm and an intensely pathological hypermetabolic lymph node

magma under the carina (SUV max=10.3) measuring 56x18x46 mm. With no other pathological hypermetabolic focus detectable on the rest of the examination (Figure 4,5,6).

It is therefore a LH with mediastinal and cardiac localization. CHOP chemotherapy (cyclophosphamide, doxorubicin, vincristine and prednisone) was administered as an emergency. After the 6th course of chemotherapy, we witnessed a clear regression of myocardial tumor infiltration and pericardial effusion (Figure 7 and 8), with improvement of the general condition.

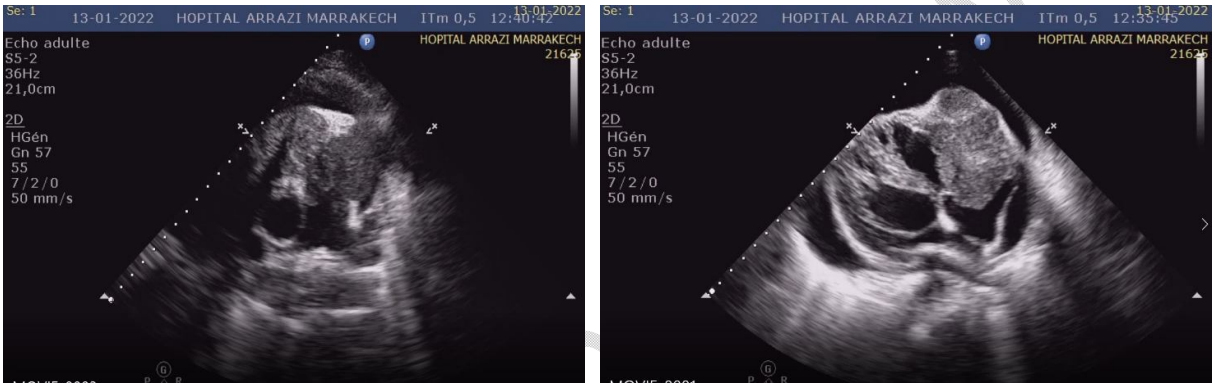


Figure 2: TTE showing an echogenic, bulky, poly-lobed mass infiltrating the free wall of the RV with a circumferential pericardial effusion of great abundance

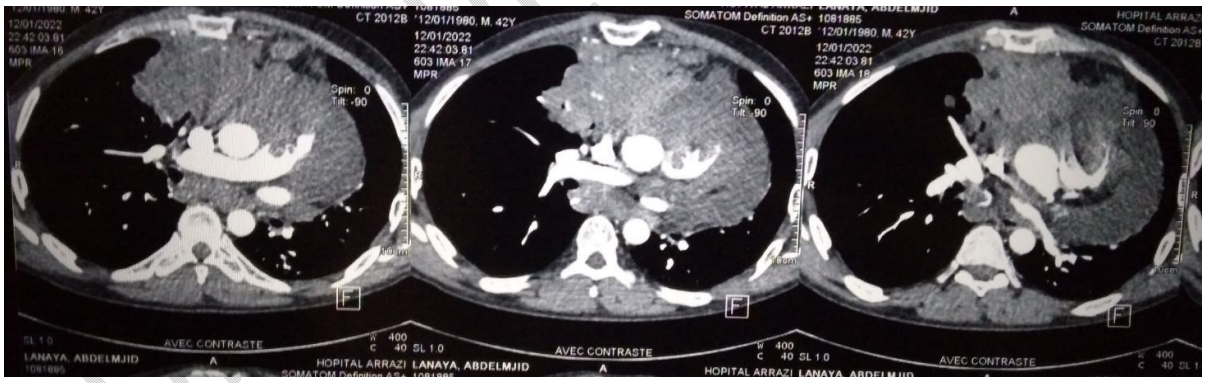


Figure 3: Thoracic angioscan showing two masses of the RV and pulmonary artery trunk with extensive pericardial invasion and effusion

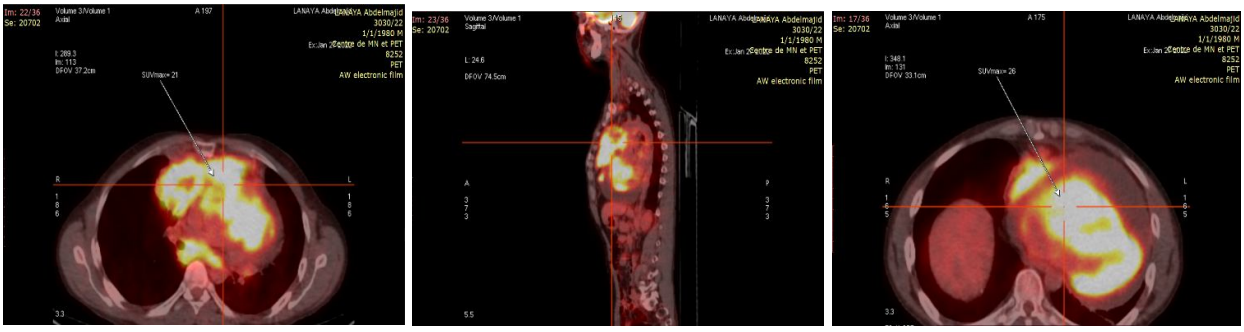
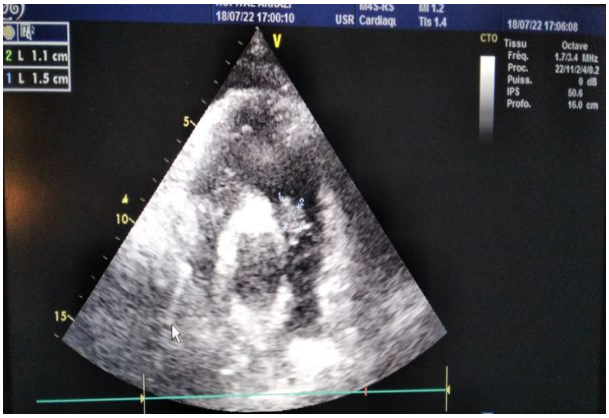


Figure 4, 5 and 6: PET-FDG showing lymphomatous dissemination at the mediastinal level: at the level of the known cardiac mass with a diffuse infiltration of the anterior mediastinum and a subcarinal lymph node magma.



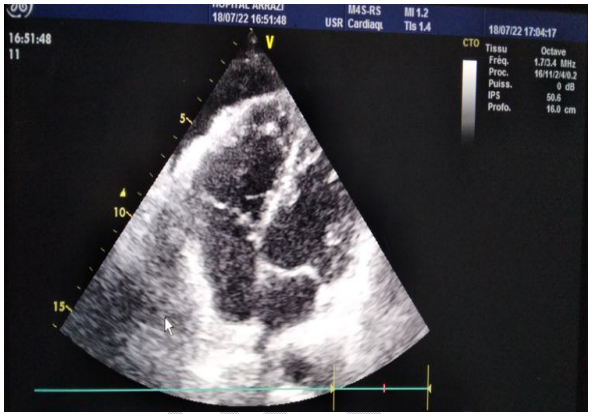


Figure 7 and 8: TTE showing clear regression of myocardial tumor infiltration and pericardial effusion

Discussion:

Cardiac metastases are relatively rare events however much more common than primary cardiac tumors [1,2,3]. The most common tumors associated with cardiac metastasis are lung and breast carcinomas [1]. Other rarer tumors have a high propensity to metastasize to the heart: lymphomas, melanomas and extracardiac sarcomas [1,4].

Cardiac involvement in Hodgkin lymphoma is rare and is often seen late in the course of the disease. Cardiac lymphomatous localization can occur either by hematogenous or lymphatic dissemination or by contiguity from an intrathoracic lymphomatous tumor [5,6]. All cardiac structures can be affected, with a clear predilection for the pericardium and myocardium.

Lymphomas are most often reported in single case studies [7,8]. Although cardiac lymphomas frequently occur in weakened areas, susceptible to opportunistic infections, immunocompromised, notably by infection with the human immunodeficiency virus (HIV), or in transplanted subjects [9,10], it is not uncommon for cases of lymphoma to be observed in immunocompetent subjects [11], which is the case in our patient.

Cardiac lymphomas are silent in more than 90% of cases [1,6]. Their clinical manifestations are infrequent, discrete, or even absent, which explains the discrepancy between the frequency of anatomical involvement (at autopsy) and the rarity of the clinical description of these localizations. When they do occur, the signs are often not very specific (dyspnea, chest pain, pericardial effusion, arrhythmia, cardiac arrest, pulmonary embolism) [1], which leads to an underestimation of their frequency. The diagnosis of cardiac localization is difficult to make during the patient's lifetime.

Cardiac involvement is frequently found at autopsy of patients who have died of lymphoma. These are most often secondary locations of systemic HL rather than primary cardiac lymphoma [12,10].

The electrocardiographic changes observed are not specific but can draw attention to cardiac involvement, especially in secondary forms. They may be conduction or rhythm disturbances, electrical signs suggestive of pericardial effusion (repolarization disturbances with micro-voltage) as in our patient's case, or signs of myocardial infarction. It is therefore important to check the electrocardiogram regularly in any patient with HL and to perform a cardiac echocardiogram if there is any doubt.

Trans thoracic echocardiography (TTE) is the basic examination for the detection of cardiac involvement of HLs, which may present differently depending on the tunica involved [13,14,15]. Most commonly, it is in the form of fixed or mobile, often polylobed, tumor vegetations located on the valvular or endocardial endocardium, more commonly in the right than in the left cavities [16,17]. The tumor mass can be single or sometimes multiple and involve the three tunics of the heart. Pericardial involvement is the most frequent and is very often translated by a more or less abundant pericardial effusion, sometimes responsible for tamponade and often associated with a tumor infiltration of this tunica [17,18]. Other aspects of cardiac involvement are much less suggestive: these are mainly the infiltrating forms of the myocardium, which are rarer and present in the form of a thickening, most often localized, giving a shiny appearance to the pathological myocardium and responsible for an alteration in segmental kinetics [19]. These different aspects of cardiac involvement may coexist in the same patient.

Echocardiography (ETT) also remains a simple and non-invasive means for monitoring the response to treatment of these cardiac localizations. However, it can sometimes be misinterpreted [20,21], hence the interest of transesophageal cardiac echocardiography (TEE), which allows a more detailed analysis of cardiac structures.

In some cases, echocardiography can be used to make the diagnosis, but the use of another imaging technique such as CT or MRI is usual.

The main interest of imaging, once the diagnosis of a mass has been made, is to specify whether it is a thrombus or a tumor. Once the diagnosis of a tumor has been made, the imaging must then point to an etiology: primary or secondary tumor. This characterization approach must also allow the distinction between the benign or malignant nature of the tumor, and above all to perform the topographic and extension workup to orient an appropriate therapeutic strategy once histological confirmation is obtained.

However, the sensitivity of MRI is better than that of CT, with a sensitivity of more than 90% [17,18], due to its high signal resolution [22,23], and it is currently the reference examination for the diagnosis of cardiac tumors [24]. CT offers a less good tissue contrast but, because of its high spatial resolution, it allows better precision of the anatomical relationships of a lesion. These 2 examinations can therefore be complementary and allow guided biopsies to be performed to confirm the histological nature of cardiac masses.

Cardiac lymphomas are complicated by compression and invasion of the right heart chambers. Right heart failure is thus the most frequent mode of expression reported in the literature [7,8]. The preferential localization is intra-right heart cavity [9, 11 ,8]. The incidence of pericardial effusion in lymphoma is estimated at 15% [9,8].

In our observation, pericardial effusion was the main clinical manifestation of the disease with repolarization and micro-voltage disturbances on EKG.

In addition to MRI, 18-FDG PET scans play an important role in the initial workup and are an effective examination for the follow-up of treated patients [22, 25].

The lymphomatous nature of cardiac involvement is evoked when it is associated with other peripheral or mediastinal lymph node locations. In this case, biopsy of an accessible adenopathy or mediastinal adenopathy is sufficient to confirm the diagnosis [26]. Cytological analysis of pericardial or pleural puncture fluid in case of fluid effusion allows the diagnosis of cardiac lymphoma in 67% of cases [17]. However, a negative cytological examination should not exclude the diagnosis of lymphoma and should be repeated.

The response of the cardiac localization, together with other nodal involvement, to chemotherapy may be an argument in favor of its lymphomatous origin [26].

The diagnosis of cardiac lymphoma can be difficult when the cardiac involvement is apparently isolated. In this case, endo-myocardial biopsy by peripheral venous catheterization is required [27,28]. Surgical biopsy of the cardiac tumor by thoracotomy remains difficult to perform in these often very fragile patients and should therefore remain the exception.

In principle, lymph node biopsy or cytological analysis of the effusion is sufficient to establish a diagnosis when the lymphoma is located secondarily in the heart [29]. The use of invasive procedures is not justified, and often it is the comparison of imaging studies performed before the initiation of treatment and afterwards that secondary lymphomatous cardiac involvement is retained.

The evolution of cardiac lymphomas can also be marked by the occurrence of serious embolic accidents because of a tendency to orificial enclosure or intracardiac floating of a pedunculated lymphomatous mass.

The treatment of cardiac lymphoma involves the usual chemotherapy for lymphoma, with or without radiotherapy.

The prognosis of cardiac lymphoma remains very poor, even if prolonged remissions are sometimes reported [16,18,26,30]. Indeed, most observations reported in the literature mention the early death of patients, the delay in diagnosis being surely the main cause. The prognosis can be improved by early diagnosis and management.

In our patient, we noted a regression of the mass size after 6 sessions of chemotherapy.

Conclusion:

Cardiac lymphoma is a rare entity and very often unrecognized because the clinical symptomatology is so nonspecific. Echocardiography and MRI are the diagnostic tools of choice for these localizations. However, histological confirmation is sometimes difficult to obtain in most patients.

The prognosis of these localizations is often poor in the short term because the diagnosis is very often late and the response to chemotherapy is rarely complete and durable.

Most cases of cardiac lymphoma are aggressive, progress rapidly and often have life-threatening complications such as heart failure and arrhythmia. It is therefore important to diagnose and treat them quickly. Moreover, the prognosis is poor because no clear treatment guidelines have been established.

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