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Case Presentation: A 65 year old woman was referred for worsening exertional dyspnea , primary examination revealed a stable patient, Unilateral edema was noted with decreased sloshing of the left calf. The electrocardiogram showed a sinus tachycardia and the transthoracic echo revealed the presence of multiple intra cardiac masses (RA, RV, LV), with pericardial thickening and minimal pericardial effusion. Cardiac MRI showed a pericardial thickening extending to the basal vessels associated with several masses at the expense of the lateral wall of the RA, RV, LV. An attempted echo-guided biopsy of the mediastinal lymphadenopathy was performed but the patient did not support the procedure.

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Discussion: Primary cardiac tumors are extremely rare. Examining the 22 series autopsy data, Lam KY(4)found 0,2% as a prevalence of primary cardiac tumors. The median age of the patients is 63 years. The symptoms of PCL are non specific. Diagnosis is delayed due to varied clinical presentations and limitations in diagnostic tools. Echocardiography shows uneven hypo echoic masses, and CT provides excellent anatomic assessment of cardiac lymphoma involvement. Cardiac MRI is the preferred imaging modality for cardiac masses because of its superior soft tissue characterization. The definitive diagnosis is based on the cytology of pericardial effusion or taking a trans-thoracic biopsy. Treatments of cardiac lymphomas can include chemotherapy, radiotherapy, surgery and even autologous stem cell transplantation, Subsequently our patient developed a sudden hemodynamic instability suspecting a serious pulmonary embolism despite the thrombolysis as well as the resuscitation measures, the patient died.

Conclusion: Although cardiac lymphoma is very rare, it should be considered in patients with an intracardiac mass. Its early diagnosis is essential because of the aggressive nature of these tumors.

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I. Introduction

Cardiac lymphoma is one of the rarest tumors involving the heart. Cardiac lymphomas are infiltrative, intramural, and epicardial lesions that may be singular or multiple, and the pericardium is involved in about a third of cases(1).

It is a subset of non-Hodgkin's lymphoma, characterized by poor outcomes and very often unrecognized entity due to its polymorphous and non-specific clinical symptomatology.

The preferential location of the cardiac lymphoma is rather the right-sided cardiac chambers, especially the right atrium, and it is often multifocal(2). The superior vena cava (SVC) is involved in up to 25% of cases(3), extracardiac lymphoma sites should be looked for during diagnostic evaluation, namely the mediastinum and bone marrow, in order to distinguish between primary and secondary cardiac lymphoma.

Echocardiography and MRI are the diagnosis methods of choice for this location. Histological confirmation is sometimes difficult to obtain in most living patients.

The tumor is rapidly fatal unless diagnosed and treated in time but the prognosis of this localization is often reserved in the short term and the response to chemotherapy is rarely complete and lasting.

II. Case Presentation

A 65 year old woman without cardiovascular risk factors or specific pathological history, was referred in the cardiology department of CHU Ibn Rochd Casablanca, for worsening exertional dyspnea and orthopnea. The examination on admission revealed a stable patient, the blood pressure was 122/67 mm Hg, the oxygen saturation was 95%, the pulse rate was 100 beats per minute, the respiratory rate was 19 per minute and the body temperature was 36.5C.

The cardiovascular examination was unremarkable. The peripheral ganglionic areas were free. Unilateral edema was noted with decreased sloshing of the left calf.

Biochemical investigations were within normal range, however a thrombocytopenia in the completed blood count was noted at 145000.

The electrocardiogram showed a sinus tachycardia and the transthoracic echo revealed the

presence of multiple intracardiac masses (RA, RV, LV), with pericardial thickening and minimal pericardial effusion (Fig 1).

Thoraco-abdominal-pelvic CT showed a massive pulmonary embolism and a very strong suspicion of lymphoma with lymph node and cardiac location(Fig2).

Cardiac MRI showed a pericardial thickening extending to the basal vessels associated with several masses at the expense of the lateral wall of the RA, RV, LV. The MI venous Doppler ultrasound shows extensive deep vein thrombosis to the iliac vein.

An attempted echo-guided biopsy of the mediastinal lymphadenopathy was performed but the patient did not support the procedure, subsequently our patient developed a sudden hemodynamic instability suspecting a serious pulmonary embolism despite the thrombolysis as well as the resuscitation measures, the patient died.

III. Discussion

Primary cardiac tumors are extremelyrare. Examining the 22 series autopsy data, Lam KY(4)found 0,2% as a prevalence of primary cardiac tumors. Primary cardiac lymphoma(PCL), on the other hand, accounts for less than 2% of all resected primary cardiac tumors and 0.5% of extranodal lymphomas atautopsy(2)

There are two definitions of primary cardiac lymphomasome authors consider the absence of lymphoma apart from pericardial sac, confirmed by a complete autopsy (5)while the others accept lymphoma as primaryto the heart if the tumor is in the pericardium(6)

Many authors reported that this disease is more common in the elderly age. The medianage of the patients is 63 years(3). The reported male to female ratio is 3/1(7), our patient was 64 years old.

The symptoms of PCL are nonspecific. It can manifest as a heart rhythm disturbance, episodic syncope, vena cava superior syndrome, respiratory distress (7)or even as a restrictive cardiomyopathy (7–9) However, the most common symptoms are dyspnea, constitutional complaints(fever, chills, sweats and weight loss), chest pain, heart failure and pericardial effusion (7). In our case, the patient presented with dyspnea and worsening orthopnea.

Diagnosis is therefore often a great challenge as patients present with a wide spectrum of clinical presentation, generally depending on the site of involvement of the heart. They may involve the right chambers of the heart, with RA being the most commonly affected. These patients can develop pulmonary embolism, A-V block and disturbances in hemodynamics, but there are cases were only the left heart was involved(10). It can concern all three layers of the pericardium, myocardium, and endocardium(11), If PCL involves the pericardium, effusion or constrictive pericarditis may develop.(11) PCL-related cardiac wall rupture has also been described(12). The cardiac MRI of our patient showed a pericardial thickening extending to the basal vessels associated with several masses at the expense of the lateral wall of the RA, RV, LV, associated to a massive pulmonary embolism.

Diagnosis is delayed due to varied clinical presentations and limitations in diagnostic tools. In this regard, several diagnostic techniques can be used: ECG can objectify Several electrocardiographic abnormalities, typically an atrial arrhythmia or atrioventricular block ranging from first to third degree.(3),(13) More unusual cases show left or right bundle branch block or ventricular arrhythmia(3). Sudden death due to cardiac arrhythmia may even be the initial presentation of cardiac lymphoma.(3)(14)

Erythrocyte sedimentation rates and / or C-reactive protein (CRP) may be elevated (15)Information regarding lactate dehydrogenase levels is controversial because some authors have detected increased serum levels, Leukocytosis and neutrophilia can be found in some cases of cardiac lymphoma (16) ; our patient had only a minimal thrombocytopenia at 145 000 .

Regarding imaging, Chest radiography remains limited for the detection of heart tumor, it can sometimes show non-specific signs such as pleural effusions or cardiomegaly. Transthoracicechocardiography remains a ring stone for evaluation of heart disease and in many cases it remains the first-line imaging method, it is a good noninvasive diagnostic tool that can detect pericardial effusion and the presence of the tumor(17).

Echocardiography shows uneven hypo echoic masses with poor mobility, stiffness, wide basement, irregular shape, and boundary

However, complementary assessment with cross-section a limaging is now essential for further characterization of the tumor and the extent of involvement, CT provides excellent anatomic assessment of cardiac lymphoma involvement, primarily due to high isotropic spatial and temporal resolution.

It benefits from fast acquisition time and is a primary alternative in imaging patients with known contraindications to MR, On CT, cardiac lymphoma classically presents as a variably or poorly enhancing mass typically arising in the right side of the heart with similar prevalence of right atrial and ventricular involvement(12).

Cardiac MR is the preferred imaging modality for cardiac masses because of its superiorsoft tissue characterization, high temporal resolution, multiplanar imaging capabilities, and unrestricted field of view(18). Recently, FDG-PET was used to detect the primary effusion lymphoma of the pericardium(19).

The definitive diagnosis is based on the cytology of pericardial effusion or taking a transthoracic biopsy.(7) Cardiac lymphomas are usually B-cell

neoplasms, most frequently DLBCL, followed by follicular lymphoma and Burkitt lymphoma(20).

The management of cardiac lymphoma is varied. Treatments differ and include chemotherapy, radiotherapy, surgery and even autologous stem cell transplantation.(21,22)

Surgeryis usually palliative and there is no evidence that it improves survival because it is difficult to resect the tumor completely. Most of the authors suggest that PCL should be considered a systemic disease and this treatment should always include chemotherapy(3).

The prognosis for patients with either primary and secondary heart lymphoma is unfortunate, at recent analysis of PCL and treatment results (cohort of 128 patients treated with chemotherapy, surgery, radiotherapy or chemotherapy /combined radiation) demonstrated a median overall survival of about12 months(3)the main prognostic factors are Initial non-specific signs and symptoms, rapid progression of cardiac involvement, and late diagnosis. (23,24).

IV. Conclusion

Cardiac lymphoma manifests as singular or multiple intramural masses with infiltrative margins and a strong predilection for the right heart, Echocardiography is often the initial imaging modality, but CT and MR imaging provide superior soft tissue contrast and anatomic information. Although cardiac lymphoma is very rare, it should be considered in patients with an intracardiac mass. Its early diagnosis is essential because of the aggressive nature of these tumors.

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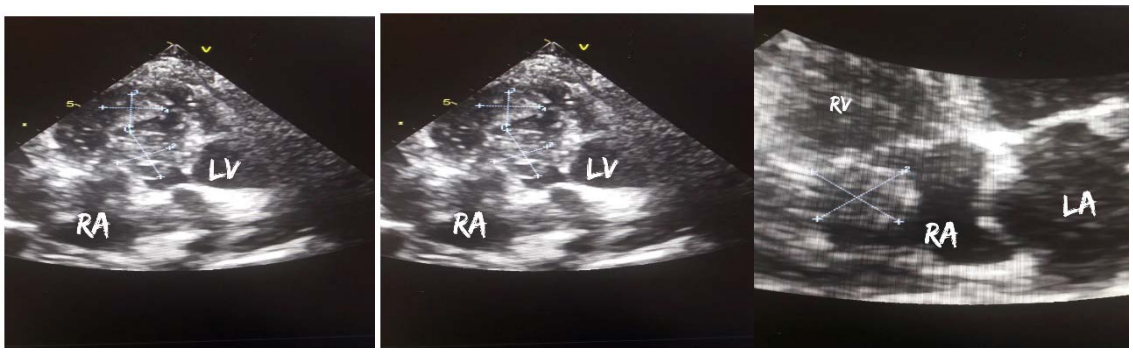

Figure 1: Subcostal section showing the mass lining the lateral wall of the RA and RV.

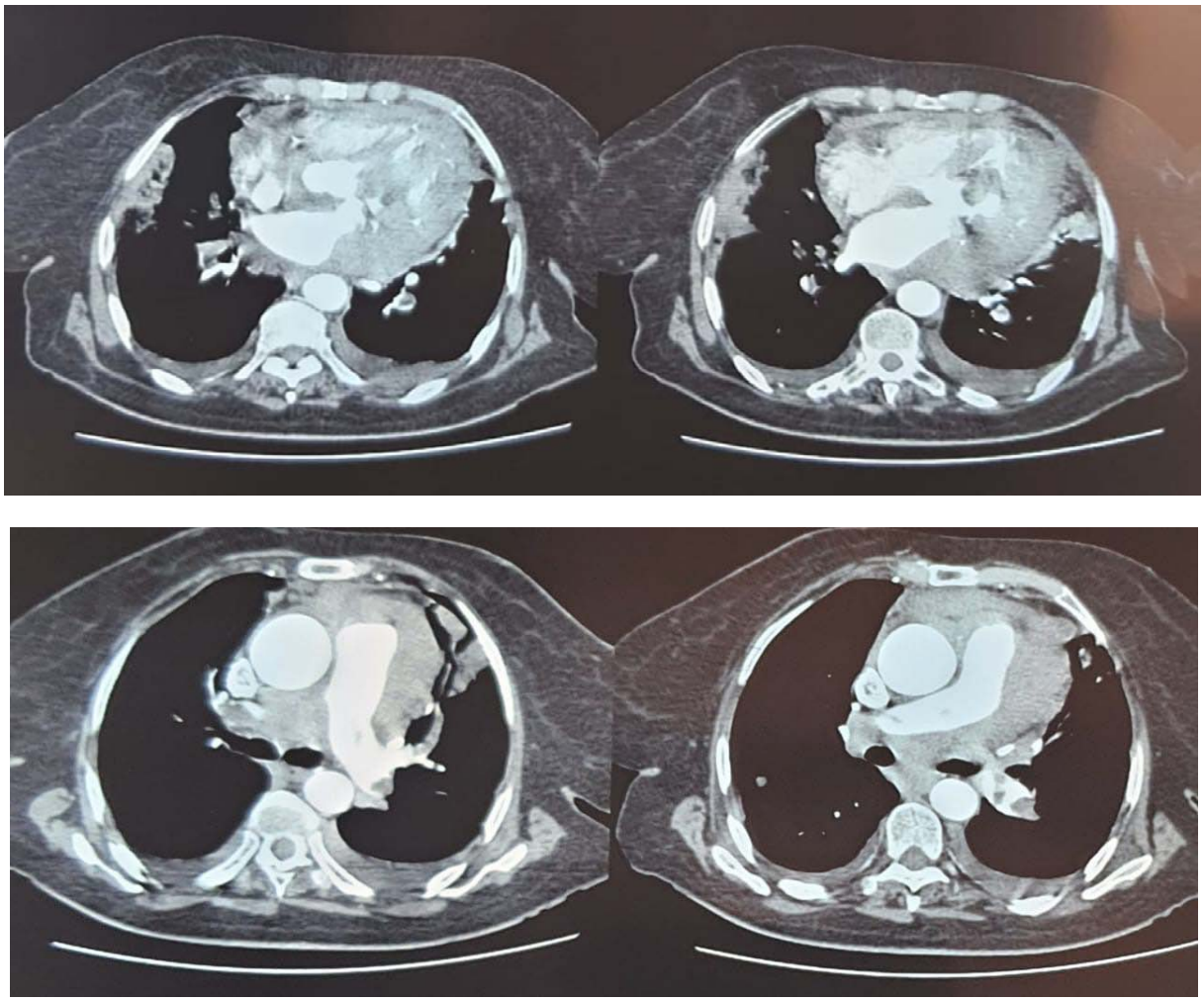

Figure 2: Thoracic CT: Massive pulmonary embolism with strong suspicion of lymphoma with ganglionic and cardiac localization.