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Primary Cardiac Lymphoma Revealed by Dyspnea in an Immunocompromised Patient: About a Case Observed at the Sylvanus Olympio University Hospital in Lomé (Togo), and Review of the Literature

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Abstract

Cardiac tumors are rare and dominated by benign tumors. However, the frequency of primary malignancies appears to increase with HIV / AIDS infection. We report the case of primary cardiac lymphoma revealed by dyspnea in an immunocompromised. This is a 61-year-old patient, followed for HIV 2 infection, on antiretroviral therapy, hospitalized for exertional dyspnea which gradually worsened over six months, associated with palpitations and faintness. The admission exam noted irregular heart sounds, right heart failure, and tricuspid rolling. The echocardiogram showed a tumor mass in the right ventricle, prolapsing into the right atrium through the tricuspid valve, an intrapericardial tumor mass with moderate effusion. Magnetic resonance imaging confirmed the tissue character of the right intraventricular mass and the pericardium. Symptomatic medical treatment resulted in functional improvement. Cardiac surgery was performed, with resection of the right intraventricular and pericardial tumor. The pathological examination of the surgical specimens concluded in large cell B lymphoma. The patient died 5 months later with a picture of deterioration in general condition.

Keywords: *Cardiac tumors, Primary lymphoma, Sub-Saharan Africa.*

Introduction

Cardiac tumors are rare and dominated by benign tumors. Primary tumors are exceptional among malignant tumors, but their frequency seems to increase with HIV / AIDS infection. According to the classification of tumors by the World Health Organization (WHO), heart tumors are extremely rare with a frequency varying between 0.0017% and 0.03% [1]. Approximately 25% of primary cardiac tumors have features of malignancy and of these 95% are sarcomas and only 5% are lymphomas. [2] A review of autopsy series over a 20-year period from 1972 to 1991 revealed an incidence of primary tumors of 0.056% versus 1.23% for secondary tumors [3]. In Africa, few data are found on cardiac masses, especially in the sub-Saharan region where a single study on cardiac masses in Côte d'Ivoire found a prevalence of 1.5 ‰ over a period of 25 years (1985-2010) with 6.8% primary and secondary lymphomas [4]. We report a case of cardiac lymphoma revealed by dyspnea in an HIV / AIDS immunosuppressed.

Clinical Observation

61-year-old patient, hospitalized on March 02, 2018 for exertional dyspnea which progressively worsened over six months until stage IV of the NYHA, associated with palpitations and faintness, without fever or signs of tuberculous impregnation. He has been monitored for HIV2 infection with well-managed antiretroviral (ARV) treatment for 8 months. He has no known history of heart disease. His clinical examination on admission had objectified: a deterioration of the general condition, an irregular pulse with an average frequency of 90 beats per minute (bpm), a respiratory rate of 18 cycles per minute without signs of struggle, oxygen saturation 88% in ambient air

with a blood pressure of 130/96 mmHg in both arms. Cardiovascular examination noted spontaneous turgor in the jugular veins. Harzer's sign was positive with congestive hepatomegaly, hepato-jugular reflux. The heart sounds were muffled, irregular with a xiphoid diastolic rolling. The lung and other examinations were normal.

The electrocardiogram (ECG) showed a basal sinus rhythm of 92 / min, microvoltage with monomorphic left delay-type ventricular extrasystoles, right atrial hypertrophy and biventricular hypertrophy. (Figure 1)

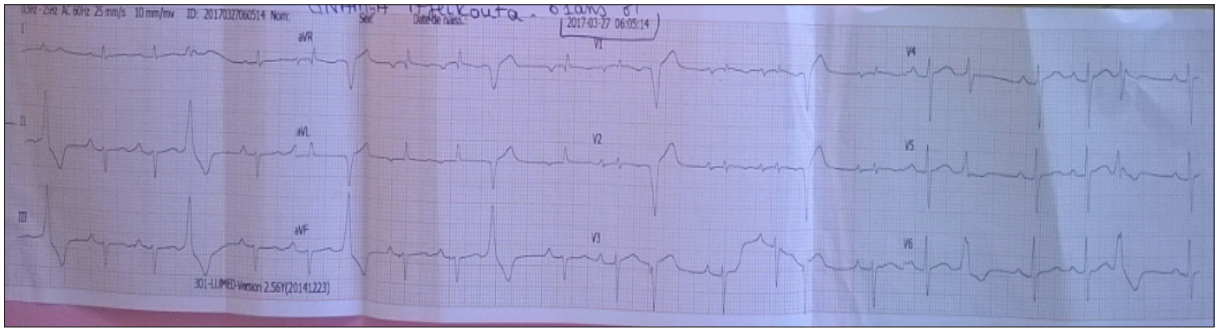

Fig1. *EKG showing left-delayed ventricular extrasystoles.*

The chest x-ray showed cardiomegaly at the expense of the right heart chambers with CTI of 0.58 and normal lung parenchyma.

Echocardiography made it possible to demonstrate a pedicled mass sitting in the right ventricle measuring, 70 mm x 45 mm x 37 mm, tissue-like and infiltrating the adjacent walls, and including the lateral leaflet of the tricuspid valve, prolapsing through the tricuspid orifice, thus obstructing in diastole, the filling of the right ventricle. The mitral filling flow was a relaxation abnormality with no ultrasound evidence of adiastole. We did not observe an associated intracavitary thrombus. Left ventricular ejection fraction (LVEF) and the tricuspid annular plane systolic excursion (TAPSE) were normal (Figure 2). We also noted an anterior pericardial mass of 59 mm x 33.8 mm located at the height of the first, associated with a moderate, non-compressive pericardial effusion. A transesophageal ultrasound was not accessible for technical reasons.

Magnetic resonance imaging confirmed the tissue character of the intracardiac mass, heterogeneous with the pericardial mass, and moderate pericardial effusion (Figure 3).

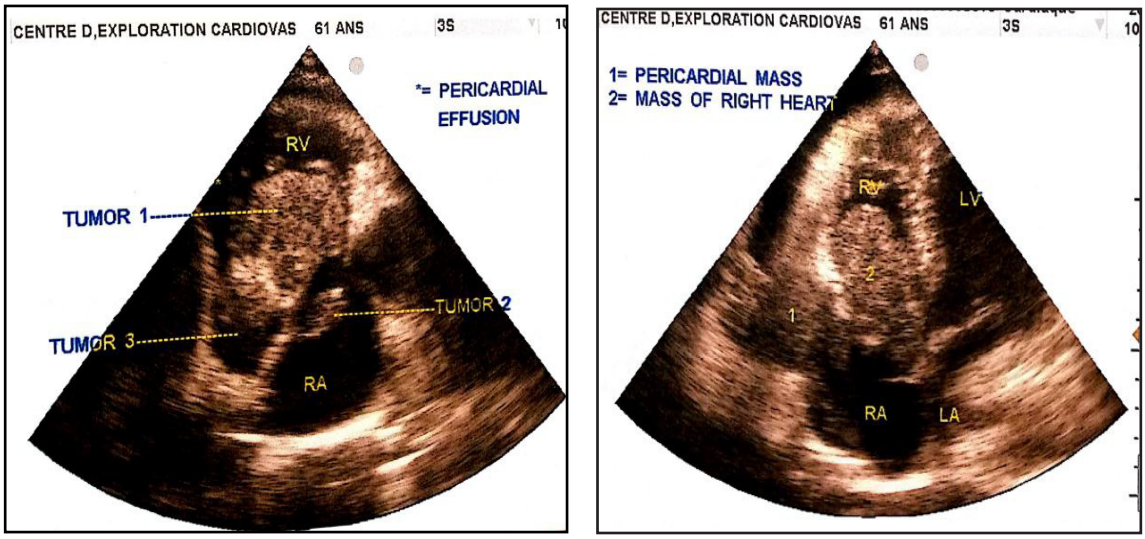

Fig2. *Apical view in two-dimensional mode, centered on the right cavities. Pedicled tumor in the right ventricle (measuring 70 mmx45 mmx 37 mm) and prolapsing into the opening of the tricuspid valve. Intra pericardial tumor measuring 59 mmx33,8 mm with moderate pericardial effusion*

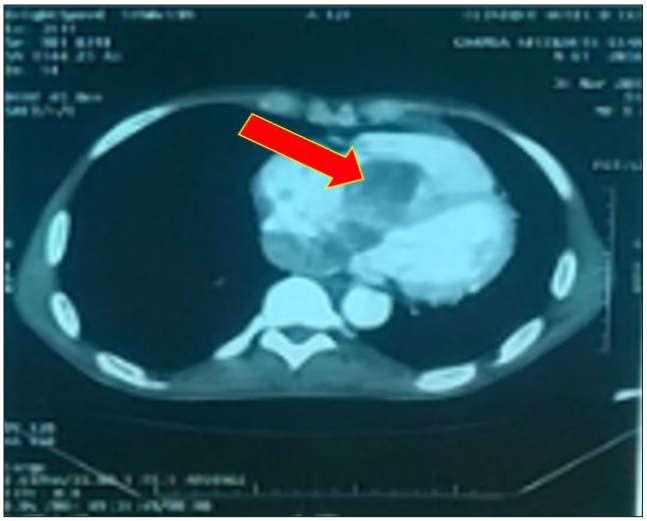

Fig3. *Magnetic resonance imaging (MRI) : right intra-ventricular and pericardial tumor.*

Cardiac surgery was performed and consisted of excision of the right intraventricular tumor (Figure 4) and pericardial**.** Pathologic examination revealed histologic evidence for primary cardiac type B large cell lymphoma. (Figure 5)

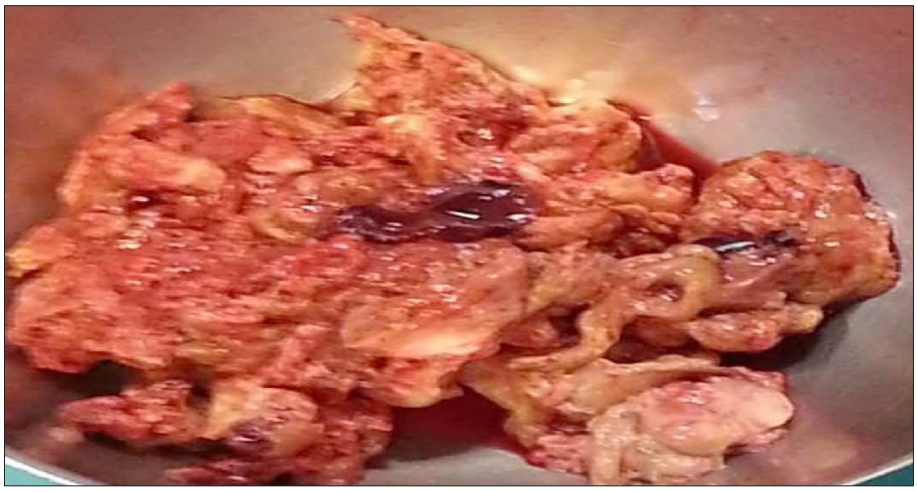

Fig4. *Macroscopic aspect of the operative part of the right intra-ventricular tumor*

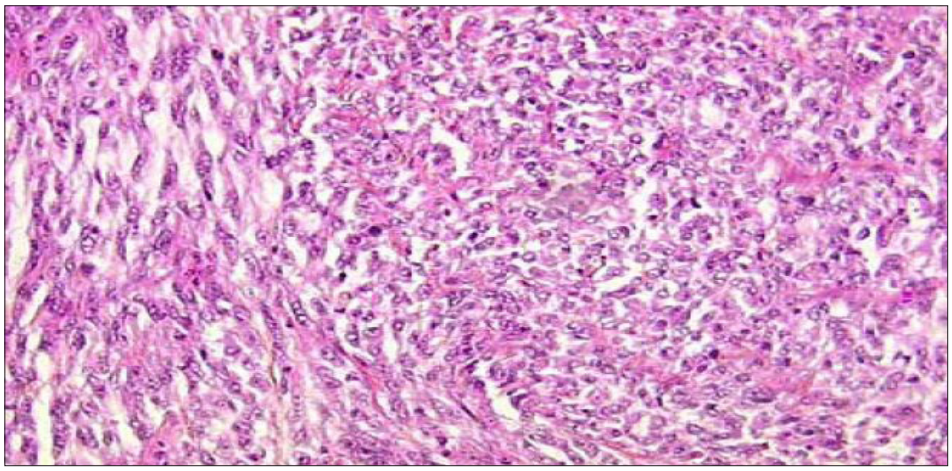

Fig5. *Microscopic appearance: giants cells B lymphoma*

The blood count was normal with a hemoglobin level of 14g / dL, a normal blood count with a CD4 lymphocyte count of 495 / µL. the rest of the biology noted thrombocytopenia at 59000 / mm3, creatinemia was at 17mg / L, hypocalcemia at 82mg / L, and hyperchloremia at 113mEq / L. There was no biological sign of a primary non-cardiac tumor: total prostate specific antigen (PSA), alpha-foeto-protein, Australian antigen (Ag HBS), electrophoresis of serum proteins were normal.

The patient was put on chemotherapy but progressed to death after 5 months.

Discussion

According to Lam and al, while nearly 20% of patients with lymphomas present with cardiac metastasis, we note that cardiac lymphoma (called primary (if it only affects the heart and / or the pericardium, as in our clinical observation), represents only 1% of the primary cardiac masses [3]. In our observation, the search for the frequent cause of primary extracardiac tumors was negative and the field of immunosuppression to HIV / AIDS is a strong argument in favor of the diagnosis of lymphoma primary, as previously reported [1,4,5]. But the diagnostic confirmation was histological on biopsy specimen. Myxoma of the atrium remains the most frequent cardiac tumor of primary masses with a prevalence of 30% and 16% respectively in the studies of Shapiro in the United Kingdom [6] and of Yao and al, in Côte d'Ivoire [4] Dyspnea is the most frequent mode of presentation as in the series of Yao and al [4] and the clinical case reported by Farris and al [5]. Imaging remains essential in the aetiological diagnostic argument [4, 5, 6] especially when access to surgery is difficult. Transthoracic and transesophageal cardiac Doppler ultrasound provides essential elements that cross-sectional imaging (CT scan and / or MRI) can strengthen. Myxoma and papillary fibroelastoma appear to be solitary tumors and are more localized in the atrium for myxoma and in contact with the mitral valve for fibroelastoma [6]. In our case, the double right ventricular and pericardial localization, the invasive character on the adjacent walls, allowed a strong etiological suspicion of the malignant character and made a thrombus unlikely. The absence of any sign of tuberculous impregnation in our case and access to histology made it possible to rule out a cardiac tuberculoma as found by Anzouan-Kacou JB and al, in Ivory Coast on an immunosuppressed to HIV; with complete regression under medical treatment [7]. Most of the series [1-3, 6, 8] are autopsy cases, especially if the cardiac tumor is malignant or metastatic, which makes the prognosis poor and its right localization makes the surgical technique difficult. The clinical presentation depends on the location, size and infiltration of the malignant cardiac tumor. The evolutionary modalities of these cardiac tumors, on the physiopathological bases recalled by Shapiro and Farris [5, 6], are essentially made of: - rhythm and / or conduction disturbances which may or may not be symptomatic (palpitations, faintness as in our case, syncope),

- Hemodynamic decompensation (systolic heart failure - observed in our case, an increase in ventricular filling pressures, cardiogenic shock).
- Embolization with multiple embolisms and systemic infarctions on a hand and on the other hand pulmonary embolisms responsible for pulmonary arterial hypertension.
- Pericarditis, pericardial effusion (as in our observation), cardiac tamponade, pericardial constriction or not, when there is pericardial involvement.

The treatment of benign tumors remains surgical resection with a survival of 95% at 3 years according to Elbardissi and al study [9]. The management of malignant tumors remains dependent on the etiology (histology), whether or not they are metastatic and on the possibilities of chemotherapy and or radiotherapy. The prognosis of malignant cardiac tumors remains poor, especially in immunosuppressed to HIV / AIDS, as is the case for our patient.

Conclusion

Heart masses corresponding to malignant tumors are extremely rare. But HIV / AIDS immunosuppression appears to increase the incidence of primary cardiac lymphoma, which manifests as dyspnea with mostly right-sided heart failure. Cardiac Doppler ultrasound coupled with CT or magnetic resonance imaging provides strong arguments for the diagnosis. Histology confirms the nature of the tumor and makes it possible to consider therapeutic possibilities despite the poor prognosis. Cardiac tuberculoma and thrombus are useful differential diagnoses in sub-Saharan Africa,

while myxoma remains the most frequent benign primary tumor.

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