Case Report

Extranodal natural killer/T cell lymphoma with cardiac and abdominopelvic nodular deposits: Unique presentation on 18-fluorodeoxyglucose positron emission tomography computed tomography scan

ABSTRACT

Cardiac lymphoma as secondary extension of non-Hodgkin's lymphoma is described in 30% of autopsies; however, a few cases are diagnosed *in vivo*. Primary lymphoma of the heart and pericardium is rare, accounting for 1.3% of primary cardiac tumors and only 0.5% of extranodal lymphomas at autopsy. We present a unique case of 68-year-old man diagnosed with extranodal lymphoma with cardiac and abdominopelvic nodular deposits on 18-fluorodeoxyglucose positron emission tomography-computed tomography scan findings.

Keywords: 18-fluorodeoxyglucose positron emission tomography-computed tomography scan, abdominopelvic nodular deposit, cardiac lymphoma, natural killer/T cell lymphoma

INTRODUCTION

Extranodal lymphomas are usually found in the stomach, intestines, nasopharynx, skin, bones, thyroid gland, breasts, lungs, testes, and brain. Primary cardiac lymphoma (PCL) is rare, although reports on the frequency of secondary lymphoma spreading to the heart state a range of 16%–28%.^[1,2] We present a case of CL, in whom the initial diagnosis was probably missed before the patient's pacemaker surgery performed for complete heart block which is one of the clinical presentations of these patients with importance of staging 18-fluorodeoxyglucose positron emission tomography-computed tomography (¹⁸FDG PET-CT) scan.

CASE REPORT

A 68-year-old man with a history of idiopathic thrombocytopenic purpura (postsplenectomy and on treatment) was treated with antibiotics and antipyretics for pyrexia of unknown origin (PUO). He further underwent pacemaker placement for complete heart block. After an interval of 1½ months, he presented with fever with chills

and two-dimensional (2D) echo color Doppler revealed large echo dense mass in AV groove measuring 22.6 cm² and a left atrial mass possibly myxoma/sarcoma with mild to moderate pleural effusion for which he was referred for a whole body ¹⁸FDG PET-CT scan. The scan demonstrated a large area of irregular uptake involving soft tissue nodular masses in interatrial septum and left atrium extending into mitral valvular region, and possibly tricuspid valvular region, extending in interventricular septum and further into aortopulmonary recess, also extending along posterior wall of right atrium superiorly between region of pulmonary

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trunk and superior vena cava (maximum standard uptake value [SUVmax] 12.2) with irregular linear uptake along region of aortic arch [Figures 1, 2a and b]. There was mild pericardial effusion. Multiple nodular soft tissue deposits [Figure 3a and b] were seen in the abdominopelvic region which were predominantly at the end of the vessels. Nodular lesion seen measuring approximately 1.8 cm × 1.5 cm medial to left adrenal gland abutting left renal vessels (SUVmax 8.9), lesion abutting left renal pelvis (SUVmax 8.6), near left posterior upper abdomen wall (SUVmax 8.3) and bilateral iliac fossa (SUVmax 5.4). There was a focal uptake seen in thickening involving abdominal aorta at the level of renal

vessels (SUVmax 7.4) and also a curvilinear focal uptake in sacral neural foramina on the left side (SUVmax 5.6).

CT-guided biopsy from a deposit near the left posterior upper abdominal wall was suggestive of non-Hodgkin's lymphoma (NHL; peripheral natural killer/T-cell lymphoma). The cells showed strong cytoplasmic positivity for CD3, CD2, CD56, T1a, and negative for CD5, CD7, CD4, CD8, CD30, NK1. Epithelial membrane antigen was negative, and Epstein-Barr virus RNA was equivocal. The final diagnosis was NHL stage IV (heart and abdomen). He received 1 cycle of cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy, after which he succumbed to death due to complication secondary to low platelet counts.

DISCUSSION

Cardiac tumors are classified as primary benign or malignant tumors that arise from the heart, or as secondary metastatic tumors that invade the heart. Primary cardiac tumors occur with a low incidence. It is estimated that secondary metastatic tumors are a hundred times more common than primary cardiac lesions.

CL as a secondary extension of NHL is described in 30% of autopsies;^[3] however, few cases are diagnosed *in vivo*. Of all primary cardiac tumors, lymphomas are rare, especially among immunocompetent individuals (1.3%) and only 0.5% of extranodal lymphomas at autopsy.^[4,5] PCL is a rare clinical entity. The frequency of secondary lymphoma spreading to the heart state a range 16%–28%.^[1] Curtsinger *et al.*^[6] reported 15 cases in 1989, and Nakchbandi reviewed 29 more cases in 1995. In the latter report, there were 6 cases related to AIDS. In the report of 15 cases by Curtsinger *et al.*, only 2 cases were diagnosed with PCL before death. It was indicated that most PCL cases were diagnosed at autopsy. However, because of advances in diagnostic methods, such cases are now mostly diagnosed

Figure 1: Maximum intensity projection images showing abnormal 18-fluorodeoxyglucose uptake in cardiac and abdominopelvic region

Figure 2: (a and b) Fused positron emission tomography-computed tomography and computed tomography images: Large area of irregular uptake involving soft tissue nodular masses in interatrial septum and left atrium extending into mitral valvular region, and possibly tricuspid valvular region

Figure 3: (a and b) uptake in multiple nodular soft tissue deposits in abdomen pelvis

before death.^[7] More than 80% of PCLs are noted as diffuse B-cell lymphomas, mostly with a large cell type. However, the incidence of NHL natural killer/T-cell type is scarce which was the histology of our case with prodigious abdominopelvic soft tissue deposits. Patients with PCL range in age from 18 to 77 years with equal sex distribution. The more common symptoms of cardiac involvement include dyspnea, congestive heart failure, pericardial effusion, and hemopericardium. Cardiac arrhythmias and nonspecific electrocardiographic abnormalities are also common, notably atrioventricular blocks ranging from first degree to third degree. They commonly manifest as an ill-defined, infiltrative mass in the myocardium, often with an associated pericardial effusion.

They can result from the retrograde lymphatic spread, hematogenous spread, or direct extension from adjacent masses.^[8] The most common appearance of CL is hematogenous spreading into the interatrial septum followed by infiltration of cardiac structures by contiguous lymphoma of the mediastinum. These tumor usually extend along the epicardial surfaces of the heart, primarily encasing adjacent structures including coronary arteries and the aortic root. When infiltration beyond the myocardium occurs, the right atrium is most commonly involved. It is an oncologic emergency due to its rapid evolution and advanced stage of organ infiltration. Treatment most commonly includes anthracycline-based chemotherapy and anti-CD20 treatment. Chemotherapy has been used alone or in combination with radiation therapy. Palliative surgery has been performed, mainly for tumor debulking. The prognosis for patients with either primary or secondary lymphomatous heart involvement is usually poor; late diagnosis is one of the major factors affecting the outcome.

The diagnostic modalities such as echocardiography and CT scans are less useful in the early diagnosis of cardiac lesions.^[9] In CT, the attenuation of CL may be similar to or lower than that of normal myocardium. At magnetic resonance imaging (MRI), it has the variable signal intensity

and contrast enhancement.^[10] PET-CT has gained favor as an imaging modality to assess tumor extent and response to chemotherapy and to predict the risk of disease relapse in patients with lymphoma. However, it is important to understand the normal patterns of cardiac FDG activity that can be seen in oncologic ¹⁸F-FDG PET-CT studies.^[11] These include focal and regional patterns of increased FDG myocardial activity. Focal activity can be observed in papillary muscles, the atria, the base, and the distal anteroapical region of the left ventricle. Regional increased cardiac FDG activity may be diffuse or localized in the posterolateral wall or the base of the left ventricle. Abnormal patterns of cardiac FDG activity not related to malignancy include those associated with lipomatous hypertrophy of the interatrial septum, epicardial and pericardial fat, increased atrial activity associated with atrial fibrillation or a prominent crista terminalis, cardiac sarcoidosis, endocarditis, myocarditis, and pericarditis. Knowledge of these patterns of cardiac FDG activity is important to be able to recognize malignant disease involving the paracardiac spaces, myocardium, and pericardium.

¹⁸F-FDG PET-CT may have the advantage of detecting the tumor or metastases at an early stage, and in cases such as the one described above, it may help in the differentiation between benign and malignant lesions of the heart, which is difficult with other imaging modalities. The unparalleled feature of ¹⁸F-FDG PET-CT in detecting malignant lesions due to its high hypermetabolism, helps even the smaller lesions in addition to staging extranodal lymphoma and also modify the disease management. This has high value when lesions are located in the ventricular myocardium with large intracavitary masses because it enables to differentiate neoplastic masses from thrombi. We also emphasize the fact that Integration of ¹⁸F-FDG PET-CT into lymphoma staging and monitoring facilitates the detection of cardiac involvement and the assessment of response to therapy. Serial PET has been suggested to be more accurate than MRI and echocardiography for assessing CL regression.^[12]

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Conflicts of interest

There are no conflicts of interest.

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