Cardiac Tamponade Caused by a Large Lymphoma Involving the Pericardium and the Right Heart Chambers

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Abstract

We report a case of a cardiac tamponade, presenting with dispnea, caused by a giant mediastinal mass with invasion of pericardium and right heart chambers. The heart is frequently the site of metastases of various malignant tumors. Most cases are clinically silent and are undiagnosed in vivo. Primary cardiac neoplasms are rare and occur less commonly than metastatic disease. Cardiac tamponade is a frequent consequence of cardiac involvement of mediastinal neoplasms. Unfortunately in the setting of mediastinal malignant mass and of cardiac metastatic cancer all management is palliative due to the usual presentation at a late stage of the disease.

Introduction

Although the frequency of cardiac metastasis in autopsy reports is about 25%, cardiac involvement in patient’s metastatic tumor disease is underestimated [1]. Melanoma, breast cancer and lung cancer are the most frequent primary tumors. According to the preferential metastatic pathway of the malignant disease pericardium, myocardium and endocardium can be differently involved. The majority of cardiac metastases are small and silent but sometimes they can become evident by signs such as pericardial effusion arrhythmias, ventricular outflow tract obstruction and peripheral embolization [2]. We report a case of a large mediastinic lymphoma involving the pericardium and the right chambers of the heart.

Case Report

A 69 years old Caucasian woman with hypertension, diabetes and history of a previous Hodgkin lymphoma was admitted to the emergency department with progressive shortness of breath and tachycardia. Physical examination showed a tachycardia (122 beats/minute) with normal blood pressure (110/60 mmHg); serial 12-lead electrocardiogram showed sinusal tachycardia and low voltage in both precordial and peripheral leads. The troponin I level was elevated (peak 0.15 ng/ml); the other haematological and biochemical parameters were within in normal ranges. A transthoracic echocardiogram showed a normally contracting left ventricle; the pericardial space was filled with fluid causing echocardiographic signs of cardiac tamponade (Figure 1a). The patient then underwent to a pericardiocentesis with drainage of 110 cc of yellow liquid. All the haematological, bacteriological and cytological analysis were within in normal ranges.

A transesophageal echocardiography showed an additional structure adhered to the right atrium and inhomogeneous thickening of the interatrial septum (Figure 1b). Chest Computed tomography scan (CT) showed the presence of hypo-dense tissue in the right paracardiac area, due to an invading lesion originating from the right chambers and the left atrium of the heart (Figure 1c). These findings were confirmed by a subsequent cardiac magnetic resonance (MRI) that documented a mass (6.1 cm x 6.2 cm x 6.1 cm) of solid homogenous tissue; this mass appeared iso–to hyper–intense on T2–w TSE images and in-homogeneously iso–to hyper–intense on T1–w TSE images. The mass was localized at the free wall of the right atrium invading the atrio-ventricular groove and coronary sinus and extending up to the crux until the lower wall of the left atrium and the inferior, septal and posterior wall of left ventricle. Furthermore the mass caused a sub occlusion of the right atrioventricular pass (Figure 1d).

The patient was then referred to cardio–thoracic surgery to perform a pleuro–pericardial window; during this intervention pericardial biopsy was made and microscopic examination showed a B–cell lymphoma with heart localization. The patient underwent a positron emission tomography (PET) that showed a pathological store of marker (18F-FDG) at heart level, mainly on right chambers, compatible with aggressive/malignant lymphoma (Figure 2). The patient was referred to our oncology department and she started a cytostatic chemotherapy (R-CHOP).

Figure 1: (a) First transthoracic before pericardiocentesis. (b) Transesophageal echocardiography showed an additional structure attached to the right atrium and inhomogeneous thickening of the interatrial septum. (c) Cardiac computer tomography the presence of hypodense tissue in right paracardiac area, into the right chambers of the heart, into left atrium. (d) Cardiac magnetic resonance confirming tumor extending from right atrium to tricuspid valve and left atrium.

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Discussion

Cardiac lymphomas are usually part of disseminated disease and up to 20% of patients with disseminated non-Hodgkin lymphomas have evidence of cardiac involvement at autopsy. Primary cardiac lymphomas in immunocompetent patients are an uncommon malignancy accounting for 1.3% of primary cardiac tumours and 0.5% of extra-nodal lymphomas [3]. Cardiac lymphomas manifest as an ill-defined, infiltrative mass usually best depicted with MRI because of its superior soft tissue contrast. Atrial location is typical with infiltration of atrial or ventricular walls. Lymphomas may present several different characteristics on imaging; they may have high or low signal on MRI, muscle's or lower on CT scan and may show increased or decreased contrast enhancement. So that, in addition to standard imaging techniques as echocardiography, MRI and CT they usually require nuclear medicine for their non-invasive assessment [4]. Cardiac lymphomas span the spectrum of B cell proliferations and include follicular centre cell lymphomas, immunoblastic lymphomas, diffuse large cell lymphomas and Burkitt lymphoma.

Most cases are clinically silent and are undiagnosed in vivo, although possible presenting symptoms include dyspnoea, cough, anterior thoracic pain, pleuritic chest pain or peripheral oedema depending on the site and extent of the lesions. The location of the tumor may also play a crucial role in the prognosis determining its resectability, therefore imaging studies have a crucial role in making the therapy plan. Nevertheless unlike other heart tumors treatment is not primarily surgical but includes anthracycline-based chemotherapy and anti-CD20 treatment. Chemotherapy has been typically used alone or combined with radiotherapy [5].

As mentioned echocardiography (trans-thoracic and trans-esophageal), CT and MRI are complementary investigations, and are all used in the evaluation of all cardiac lesions. Transthoracic echocardiography is an appropriate initial investigation which can give information on the size, location, shape and mobility of the tumor; it also has the best spatial and temporal resolution of the cardiac imaging modalities, providing excellent anatomic and functional information [6]. Transesophageal echocardiography can confirm the anatomical localization, quantify baseline valvular stenosis or regurgitation, and guide cannulation [7]. Magnetic Resonance Imaging (MRI) and ECG gated computed tomography (CT) is useful diagnostic tools providing additional aids in detecting metastases and determining the resectability of the mass. MRI has the highest soft tissue contrast of the imaging modalities, which makes it the most sensitive technique for detection of tumor infiltration. CT has a high degree of tissue resolution and can help with the characterization of all the cardiac tumors [8]. A very
invasive cardiac lymphoma as described is a rare finding and from our knowledge there are only few reports in the literature.

Unfortunately in the setting of cardiac tumors all management is palliative and our patient presents at a late stage of the disease when the prognosis is extremely poor.

References


