Lymphoma of the Right Atrium and Ventricle

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Primary cardiac tumors are extremely rare, with an autopsy incidence ranging from 0.001% to 0.03% [1]. Seventy-five percent of the primary tumors are benign; myxomas are the most frequent lesions and account for more than half the cases. The most prevalent primary malignant tumors are cardiac sarcomas, of which angiosarcoma is the most common in adults (37%) [2]. Conversely, cardiac lymphomas are extremely rare and affect mainly immunosuppressed individuals. They are mostly non-Hodgkin B cell type and preferentially involve the right atrium, with invasion of adjacent structures and large pericardial effusion [3]. Primary cardiac tumors present with one or more of the following: intracardiac obstruction, systemic embolization and constitutional symptoms, and are traditionally diagnosed by echocardiogram and computed tomography (CT) scan. However, in the last few years the role of cardiovascular magnetic resonance imaging (CMRI) in identifying and characterizing these tumors has emerged.

We present here the case of an elderly woman with shortness of breath in whom echocardiography revealed a large mass occupying the right atrium and ventricle. This mass had a typical appearance on CT and CMRI supporting the diagnosis of angiosarcoma, although pathologically it was diagnosed as diffuse large B cell lymphoma and was treated accordingly with chemotherapy.

PATIENT DESCRIPTION

An 88 year old woman presented to our hospital with shortness of breath, leg edema and weakness of 2 weeks duration. Her past medical history was positive for ischemic heart disease and hyperlipidemia. On admission, her vital signs were normal, jugular venous pressure was elevated, lungs were clear, heart sounds were distant, and she had significant leg edema without hepatosplenomegaly or lymphadenopathy.

Complete blood count and biochemical blood tests were normal except for mild hyponatremia and mildly elevated cholestatic liver function tests and lactate dehydrogenase (LDH) levels. Tumor markers (CEA, CA, 19-9, CA 72-4) were normal except for CA-125 which was mildly elevated (77 mg/ml, normal range 0–35). An electrocardiogram (ECG) demonstrated sinus rhythm with low amplitude complexes.

Echocardiography showed mild pericardial effusion, mildly dilated right atrium and ventricle filled by a vacuolated non-homogeneous texture mass with a dense consistency (atrial part 4.5 x 4.0 cm, ventricular part 7.5 x 3.5 cm), causing a significant obstruction of right ventricular inflow. Mid-ventricular systolic pressure gradient was estimated at 12 mmHg. The pericardial fluid was tapped and cytology examination revealed irritated mesothelial cells. No cells suspicious for malignancy were found. CT scan of the chest, abdomen and pelvis demonstrated the mass in the right heart invading the coronary sinus, as well as bilateral adrenal masses. CMRI demonstrated an iso-intense mass in T1 and steady-state free precession (SSFP) located between the inferior part of the right atrium and the upper part of the right ventricle, impeding the blood flow between them and from the inferior vena cava. In T2 it was non-homogenous with a hyper-intense center and iso-intense (compared to the myocardium) peripheral areas. With contrast it was centrally enhanced in first-pass perfusion and significantly enhanced by gadolinium [Figure 1].

These findings were thought to be typical for angiosarcoma due to the location, early hyper-bright heterogeneous gadolinium enhancement in T1, as well as heterogeneous appearance with hyper-intense center in T2. However, biopsy obtained from the atrial part of the mass demonstrated fragments of tissue infiltrated by a diffuse lymphoproliferative process composed of large atypical lymphoid cells with vesicular nuclei and variably prominent nucleoli. Mitotic figures and apoptotic bodies were seen. The malignant cells stained strongly with immunohistochemical stain for CD20 and for lymphocyte common antigen (LCA) but did not stain with CD3. More than 95% of the malignant cells stained with the proliferation marker Ki67. A diagnosis of diffuse large B cell lymphoma was made. The patient was treated with irradiation without chemotherapy because of her age, with good response.

COMMENT

Among primary cardiac malignant tumors, lymphomas are extremely rare and affect mainly immunosuppressed individuals. Fewer than 60 cases of immunocompetent patients have been reported since 1945 [1]. As mentioned above, they are mostly non-Hodgkin B cell type and preferentially involve the right atrium, with invasion of adjacent structures and large pericardial effusion [3]. The distinction between primary cardiac lymphoma and secondary cardiac involvement in systemic lymphoma

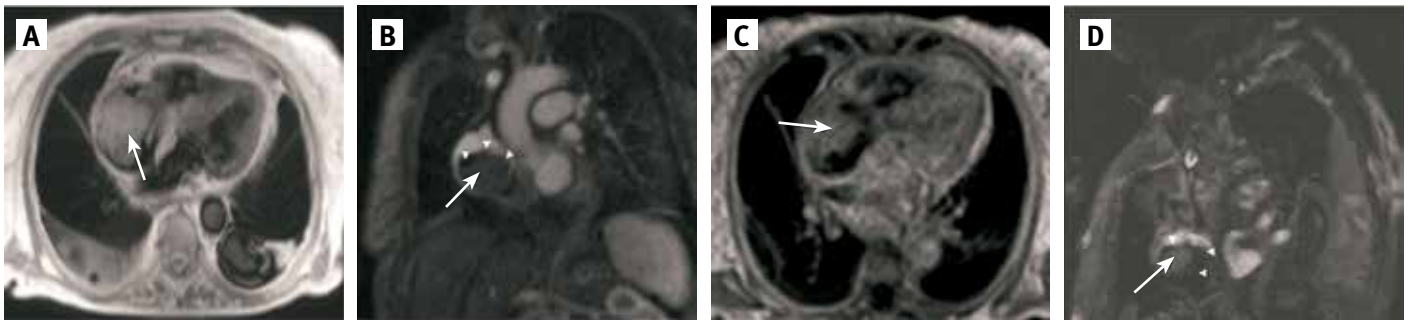

Figure 1. [A] Axial T2-weighted black blood image showing a large mass in the right atrium, brighter than normal myocardium (arrow)

[B] First-pass T1-weighted gradient echo showing central enhancement (arrow) with peripheral sparing (white arrow heads) of the mass, suggesting highly vascularized tissue

[C] T1-weighted black blood turbo spin echo four-chamber view post-gadolinium with central non-homogenous enhancement of the mass (arrow)

[D] Short axis delayed enhancement showing central retention of contrast (black arrow) material in the mass with peripheral sparing (white arrow heads) suggesting either a clot or necrotic tissue

is based on the absence of lymphoma outside the pericardial sac as well as bulk of the neoplasm within the pericardium or lymphomatous cardiac infiltration at the time of the initial diagnosis. The techniques mainly used for diagnosis of cardiac masses include transthoracic and transesophageal echocardiogram, CT scans and, during the last few years, CMRI. The latter enables optimal assessment of the location, functional characteristics and soft tissue features of the mass and may therefore possibly provide an accurate identification [4]. However, the case presented above emphasizes the need for biopsy despite the characteristic appearance on different imaging modalities in order to achieve an accurate definitive diagnosis that will determine the appropriate treatment.

The imaging methods most frequently used for the diagnosis and evaluation of cardiac masses are echocardiography, CT and CMRI. Angiosarcomas appear on CMRI as a lobular broad-base mass located in the right atrium. They are iso-intense on T1-weighted sequences and heterogeneous on T2-weighted sequences. On T1-weighted fast spin-echo (FSE) sequences, they are typically low signal, and T2-weighted FSE images typically show increased signal and central areas of hyper-intensity, consistent with hemorrhage and necrosis, and areas of moderate signal intensity in more peripheral regions. Enhancement after administration of gadolinium and the delayed enhancement are strong and heterogeneous due to the high vascularity of the tumors [4]. Lymphomas usually appear as lobular large masses located in the right atrium with either diffuse infiltration of the right ventricle or multiple nodules and are accompanied with pericardial effusion. They are iso-intense on T1-weighted sequences and hyper-intense on T2-weighted sequences. They may appear iso-intense or hypo-intense relative to myocardium on both T1 and T2-weighted FSE sequences. Enhancement after administration of gadolinium is heterogeneous with high delayed enhancement [4].

In conclusion, the typical appearance on MRI can help in the diagnosis of heart malignancies. However, this case highlights the need for biopsy to achieve an accurate definitive diagnosis despite the characteristic appearance using different imaging modalities.

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"I hate with a murderous hatred those men who, having lived their youth, would send into war other youth, not lived, unfulfilled, to fight and die for them; the pride and cowardice of those old men, making their wars that boys must die"

Mary Roberts Rinehart (1876-1958), American crime novelist

"Think for yourself and question authority"

Timothy Leary (1920-1996), American psychologist and writer known for advocating psychedelic drugs