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A Very Rare Case of Intra-cardiac Lymphoma Presenting as Splenic and Renal Infarcts

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Abstract

Primary lymphomas of the heart are extremely rare, accounting for less than 2% of all primary cardiac tumors. Histologic confirmation is fundamental in developing an early treatment plan, facilitating improved prognosis. Clinical presentation is heterogeneous and generally related to the affected cardiac chamber. The right atrium and right ventricle are the two most frequently involved sites [1]. Arterial and/or venous embolic events also may occur depending on the site of cardiac involvement and should prompt further investigation to confirm the diagnosis. Our patient presented with vague symptoms of abdominal pain. Evaluation revealed splenic and renal infarcts. Further investigations revealed a 4.5 x 3.5 cm left ventricular mass; multiple biopsies failed to yield a diagnosis. Intra-operative resection allowed for histologic confirmation of primary cardiac diffuse large B-cell lymphoma. Unfortunately, 2 weeks after resection her tumor had recurred. Chemotherapy with R-EPOCH resulted in complete remission after 2 cycles and remained in remission after total of 6 cycles. This case demonstrates successful treatment of an aggressive non-germinal center rare intra-cardiac lymphoma with a more aggressive regimen R-EPOCH compared to R-CHOP.

Case Presentation

A 50-year-old female presented to the emergency room with 2-day history of abdominal pain and flank pain, associated with nausea, vomiting, diarrhea and night sweats with temperatures ranging from 102-103 Fahrenheit. Computed Tomography (CT) abdomen/pelvis revealed mild colitis and multiple areas of wedge shaped infarcts in the spleen and bilateral kidneys concerning for emboli. Blood cultures were sent; IV antibiotics and heparin drip were initiated.

Over the hospital course, blood cultures were persistently negative. Formal testing for vasculitis and acquired hypercoagulable state was unremarkable. CT angiogram to identify the source of emboli revealed a left ventricular filling defect concerning for intra-cardiac thrombosis. Cardiac magnetic resonance imaging (MRI) confirmed a left ventricular mass, 4.5 x 3.5 cm concerning for sarcoma, invading the anterior wall and the anterior septum with a second focus in the posterolateral wall. Cardiothoracic surgery was consulted, however there were concerns the tumor could not be completely resected without causing irreversible damage and leaving insufficient myocardium to provide adequate cardiac function. The tumor was deemed unresectable on this basis. True cut needle biopsy of the left ventricular mass via a left anterior thoracotomy under the guidance of a trans-esophageal echocardiogram (TEE) was performed; biopsy only resulted in benign myocardial muscle with no detection of neoplasm.

Eventually resection of left ventricular tumor through a left atrial approach after detaching the anterior mitral leaflet was attempted and a large exophytic mass measuring 3 x 2 x 2 cm was performed. Surgeons were able to resect to margins with clean appearing muscle with no gross residual tumor remaining.

The procedure was complicated by sustained ventricular tachycardia requiring defibrillation, a third degree AV block, mediastinitis requiring prolonged antibiotic and 3D vascular CT angiogram revealed thinning of the intraventricular septum suspicious for possible myocardial infarction.

Pathology was consistent with intra-cardiac diffuse large B cell lymphoma with non-germinal center immunophenotype and high proliferative index of 95%. Immunohistochemistry was positive for: CD45, CD79A, CD5, BCL-6, MUM-1, negative for CAM5.2, AE1/AE3, S-100, ALK, CD10, Cyclin and CD99. CD3 stained only background T-cells. In situ hybridization was negative for EBV RNA.

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Metastatic disease was ruled out with MRI of brain and CT chest/abdomen/pelvis. Repeat scan 2 weeks post-operatively showed recurrence of the intra-cardiac mass. Treatment was initiated with R-EPOCH (rituximab, etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin). PET-CT after 2 cycles showed no evidence of FDG avid lesions. She completed 4 more cycles and a repeat PET-CT and cardiac MRI was consistent with remission [2].

Discussion

Primary cardiac lymphoma, involving only the heart and/or pericardium or as a lymphoma with the bulk of the tumor located on the heart is extremely rare in immunocompetent patients, accounting for only 1.3% of all cardiac tumors [3]. Primary cardiac tumors remain asymptomatic until it produces a mass effect when the tumor obstructs cardiac chambers and great vessels, pulmonary or systemic embolization, complete AV block and cardiac tamponade.

Cardiac involvement as an initial presentation of malignant lymphoma is a rare occurrence [4]. Secondary involvement of the heart has seen in 8.7 - 27.2% of documented clinical case of lymphoma [4-6]. Many other reports indicate that the majority of intracardiac tumors have occurred on the right site of the heart, the reason for which has yet been to be found [7,8]. This case is interesting as the patient presented with multiple organ infarcts with no cardiac signs and symptoms in spite of having a 4 x 3 cm mass situated in the left ventricle.

Echocardiography was the first non-invasive study for examining the chambers of the heart and pericardium [9] but trans esophageal echocardiography (TEE) was a more sensitive technique for assessing patients [10]. Unfortunately, TTE for this patient only reported a moderate - large pericardial effusion located mid-apical and lateral to the left and right ventricle, compressing the right ventricle apically and inferiorly. CT and MRI have advantages over echocardiography because they provide better contrast resolution, appear homogenous, iso or hypointensity on T1 weighted image and relatively hyperintensity of T2 weighted image [11]. It is thus very important in cases presenting with multiple infarcts to look for the source with more sensitive imaging modalities when the ECHO is normal.

Because of its rarity, data has been lacking to produce definitive guidelines regarding management of this entity. No definitive treatment of primary cardiac lymphoma has been established. The available literature suggested systemic chemotherapy has been the only effective therapy [12] and the majority of cases have treated with combination chemotherapy with varying results [13,14]. Addition of the anti-CD20 monoclonal antibody, Rituximab, to backbone chemotherapy regimens such as CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone) has shown good outcomes in CD20 positive tumors [2].

This case also shows that surgery is not an effective treatment for intracardiac lymphoma. Radiotherapy is another modality in treating primary cardiac lymphoma but only applicable in patients with good hemodynamics or diffuse disease. Nascimento et al. in their case series showed that patients who were treated with a combined modality of systemic chemotherapy and additional post-chemotherapeutic

radiation therapy had a better long-term progression-free survival compared to those treated with systemic chemotherapy alone [15]. Here we demonstrate a case where after a complicated diagnosis, treatment of intra-cardiac lymphoma with systemic chemotherapy led to successful treatment and remission of lymphoma.

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