Primary Atrial Lymphoma in a Patient with Dyspnea

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Abstract

Background: Primary cardiac lymphoma is extremely rare.

Case report: In this care report, we present the case of a 46-year-old man with primary cardiac lymphoma involving left atrium and interatrial septum, presenting as dyspnea palpitation and irregular heart beat. The diagnosis was obtained by transthoracic echocardiography and surgical biopsy with subxiphoid approach which revealed diffuse large B-cell non-Hodgkin's lymphoma, CD 20+. After 8 courses of chemotherapy, the patient achieved complete remission. After 4 months, however, he developed exertional dyspnea. Right atrial recurrent lymphoma extension was diagnosed. He is currently under external radiotherapy treatment.

Conclusion: patients with primary cardiac lymphoma have a very poor prognosis.

Keywords: primary, atrial lymphoma, dyspnea

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Introduction

Primary cardiac neoplasms are rare [1]. A wide range of presenting symptoms can be manifested depending on the tumor location, pattern of growth, and histologic type. Occasionally, patients present with embolic stroke. This manner of presentation is usually associated with tumors growing in the heart's chambers. Lesions most commonly presenting as cardiac mural masses include myxoma, sarcoma, metastatic lesions, and mural thrombus. [2]

Disseminated cardiac lymphoma is more common than primary cardiac lymphoma, and it has been reported to account for 15% of all the cardiac and pericardial metastases in non-AIDS series [3]. Patients may present with intractable congestive heart failure, pericardial effusion, and cardiac arrhythmia [4-6] or cardiac tamponade. [7] Patients usually have nonspecific symptoms; however, rapid progression of cardiac dysfunction can occur after these symptoms. The most common gross appearance is nodular or polypoid masses which predominantly involve the pericardium with variable myocardial infiltration. Histologically, these are diffuse aggressive lymphomas usually of small noncleaved or immunoblastic types. [4] Patients with mechanical obstruction may benefit from surgical resection.[8] Primary cardiac lymphomas (PCLs) make a small fraction of primary cardiac malignancies. Reported tumors have always been invasive with nodular or diffusely infiltrative patterns of growth [1, 10].

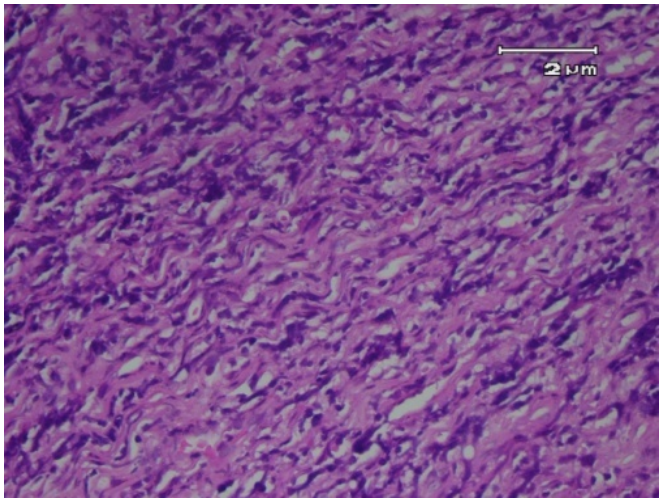

Figure 1: *Malignant round cells, with vesicular nuclei and prominent nucleoli. Apoptotic cells and mitoses were numerous (H&E stain) ×2µm*

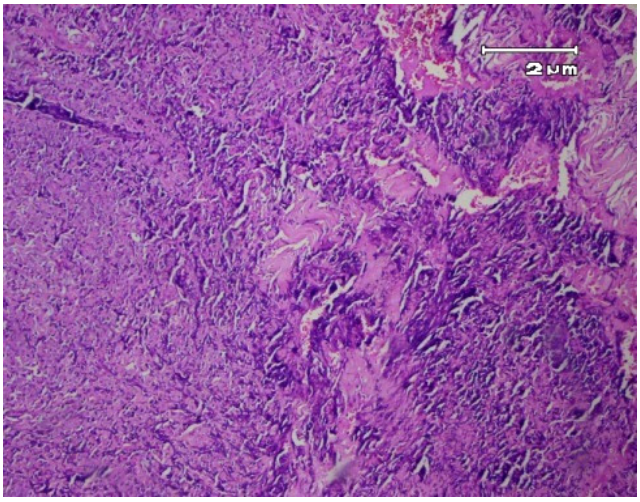

Figure2: *Diffuse infiltration of malignant lymphoid cells×2µm*

Case Report

A 46-year-old man was transferred to our institution from a regional hospital due to a sudden deterioration in his clinical condition. He had been treated there because of dyspnea on exertion, palpitation, arrhythmia and a large pericardial effusion without the evidence of tamponade. The routine laboratory investigations (including HIV test) were negative.

On admission to our department, a physical examination showed tachypnea, a heart rate of 115 beats per minute, and an arterial pressure of 145/90 mm Hg. Cardiac examination revealed an II/VI holosystoloic murmur.The resting ECG showed sinus tachycardia rhythm and P pulmonale. No evidence of venous thrombosis was observed. There were no significant laboratory findings.

A transthoracic echocardiogram indicated left atrium enlargement, large vegetation in mitral valve, filling three fifths of the left atrial cavity. Further evidence for the existence of this mass was the presence of a filling defect on the color Doppler echocardiogram, showing the location of the mass with the surrounding color flow. The mass showed a small degree of motion, but no free-floating movement suggesting a mobile thrombus was observed. A transesophageal echocardiogram was performed. A huge non-homogenous, non-encapsulated irregular mass which virtually filled the left atrial cavity and caused external pressure of the right upper pulmonary venues was found. The mass also infiltrated the right atrium causing external pressure to superior and inferior vena cava. Extention to pericardial space was also found.

Surgical operation was performed with subxiphoid approach. Pericardium was opened, large tumoral mass was seen and biopsy was performed; 200 cc fluid was drained from the pericardial cavity. Cytologically, the infiltrate was composed of cells with high nuclear-cytoplasmic ratios and round to lobulated vesicular nuclei, containing multiple prominent nucleoli (Figure 1).

Immunohistochemical findings were compatible with the diffuse large B cell lymphoma. Malignant cells stained positively with antibodies directed against CD20. CD3 was negative. Further staging workup by physical examination, imaging studies, and bone marrow biopsy showed no other sites of the disease. There was no clinical history of recurrent infections or autoimmune disorders. The patient was diagnosed with primary cardiac large B-cell lymphoma and received 8 cycles of cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy. The patient showed no evidence of the disease for 4 months. After 4 months, however, he developed exertional dyspnea. Transesophageal echocardiography showed a small mass in the right atrial wall, fixed, smooth and sessile. No mass was observed in the left atrium elsewhere. Right atrial recurrent lymphoma extension was diagnosed.

Discussion

Primary cardiac lymphoma is an extremely rare and rapidly evolving malignancy, arising from the right heart in the majority of the cases. Its incidence is higher in immunocompromised patients. Pathognomonic clinical presentation has not been

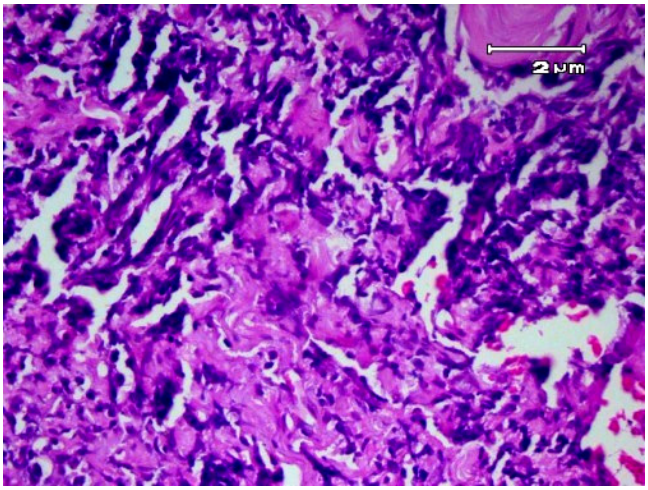

Figure 3: *Diffuse infiltration of malignant lymphoid cells×2µm*

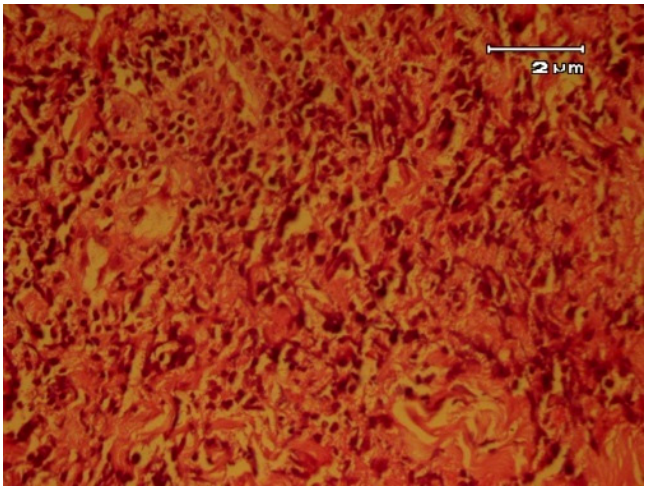

Figure 4: *CD20 Positivity in malignant cells×2µm*

described, but unresponsive heart failure, precordial pain, rhythm abnormalities, and pericardial effusion-tamponade are the most common features. Its inconsistent presentation, along with its rarity, makes the diagnosis of this condition difficult. [11] In view of the fact that early therapy seems to offer the only chance for a cure, in the presence of clinical or imaging evidence of cardiac mass or unexplained refractory pericardial effusion, aggressive diagnostic procedures may be indicated in order to obtain specimens for cytologic or histologic examination. This particularly applies to immunocompromised patients. Transesophageal echocardiography, ECG-gated MRI, and gallium-67 uptake can often be helpful in clarifying the diagnosis [12].

A number of examples of intracavitary lymphomatous masses associated with an invasive component have been reported. Proctor et al describe a case of an intracavity lymphoma tumor mass that mimicked cardiac myxoma on echocardiography. A polypoid PCL has also been reported to simulate prosthetic mitral valve thrombus on echocardiography.[13] Two cases of fatal pulmonary tumor embolus arising from a PCL of the right atrium are reported. Wargotz et al describe a patient presenting with central nervous system complaints and radiographic lesions of the left cerebral hemisphere. At autopsy, the sole site of lymphoma involved the posterior walls of the atria. It is worthwhile noting that in each of these reported cases, the intracavitary mass was associated with an invasive component.[14] Our patient had similar presenting signs and symptoms to other reports. Overall, these patients have a very poor prognosis; and after 4 months our patient experienced a recurrence.

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