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### **Središnja medicinska knjižnica**

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Primary cardiac lymphoma presenting as atrial flutter and total heart block

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## **Abstract**

Primary cardiac lymphoma is extremely rare. We present a case of 70-year old man with primary cardiac lymphoma involving interatrial septum, presenting as atrial flutter and total heart block. The diagnosis was obtained by echocardiography guided transvenous endocardial biopsy which revealed diffuse large B-cell non-Hodgkin lymphoma, CD 20 +. After six courses of immunochemotherapy the patient achieved a complete remission. After two months he developed a series of epileptic attacks. Intracerebral lymphoma extension was diagnosed. Two cycles of high dose of methotrexate and cranial irradiation were applied resulting in a second complete remission.

## **Key words**

Non-Hodgkin lymphoma, Heart neoplasms, Heart block, Echocardiography, Chemotherapy

## Introduction

Primary cardiac lymphoma (PCL) is a non-Hodgkin lymphoma (NHL) confined to the heart or pericardium or NHL in which the bulk of the tumor is located in the heart (1, 2). PCL is extremely rare; it represents 1.3% of all primary cardiac tumors, 1% of all lymphomas and 0.5% of extranodal lymphomas. It occurs more commonly in immunocompromised patients, particularly in association with human immunodeficiency virus infection. Usual presenting symptoms are chest pain, congestive heart failure, pericardial effusions and arrhythmias. Although occasionally successful in establishing the diagnosis, pericardial or pleural fluid evaluation alone can lead to misdiagnosis. Diagnostic cytologic samples were obtained in only 67% of patients with a PCL (3). Biopsy of cardiac tissue is mandatory because 75% of all myocardial tumors are benign. The prognosis is generally poor but prolonged survival has been reported after chemotherapy. Chemotherapy is the only effective treatment of PCL. Standard therapy protocols are CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone) or CNOP (mitoxantrone instead of doxorubicin)\_(2, 4). Radical surgery is discouraged.

## Case report

A 70-year-old immunocompetent man with chronic hepatitis B was transferred to our institution from another hospital because of atrial flutter and total heart block. He was admitted to the other hospital because of a 3-month history of low grade fever and arthralgias. No infective cause was diagnosed. On admission his temperature was 37.6 C, blood pressure 160/85 mmHg, heart rate 50 bpm, lungs were clear on auscultation, cardiac examination revealed normal heart sounds without murmurs, gallops or rubs. ESR was accelerated, CBC showed normocytic anemia, LDH and CRP were elevated. Transaminases were mildly elevated. ECG indicated atrial flutter with episodes of total AV block. Transthoracic echocardiography (TTE) revealed a signal intense mass at the atrial septum (figure 1). Transesophageal echocardiography (TEE) confirmed the presence of an avascular tumor measuring 5.2x2.5 cm involving interatrial septum, bases of mitral and tricuspid valves and aortic root (figure 2). The valves were morphologically and functionally normal. Thoracic and abdominal CT scan revealed a tumorous mass originating from the interatrial septum and expanding between mediastinal great vessels. Maximal mediastinal tumorous mass diameter was 3.6 cm while mediastinal lymph nodes were only mildly enlarged, measuring 1.8 cm in their greatest diameter. Peripheral and infradiaphragmatic lymph nodes were not enlarged. TTE guided transvenous endocardial biopsy revealed a diffuse large B-cell NHL, CD 20 +. Bone marrow biopsy was unremarkable. Immunochemotherapy with rituximab (genetically engineered anti-CD20 antibody), cyclophosphamide, doxorubicin, vincristine and prednisone (R-CHOP) was given. TTE after three days showed significant tumor regression. TTE and CT scan after three cycles showed no signs of tumor but a reduction of left ventricular ejection fraction (LVEF) to 45-50%. The cardiotoxic drug doxorubicin was switched to the less cardiotoxic mitoxantrone and three more cycles of R-CNOP were applied. Therapy was complicated with pneumonia and febrile neutropenia despite G-CSF prophylaxis. Evaluation after six courses of treatment confirmed the complete remission. Mild LVEF reduction and atrial flutter persisted, there were no signs of total AV block. Two weeks later the patient was admitted because of epileptic attacks. Brain CT scanning showed a left

frontal tumor mass consistent with intracerebral extension of lymphoma. He received two cycles of high-dose methotrexate and cranial irradiation resulting in a second complete remission. Eighteen months after diagnosis and 11 months after the end of treatment the patient remains in complete remission with moderate encephalopathy.

## Discussion

Atrial flutter and complete heart block are uncommon presentations of PCL. The rarity and heterogeneous clinical presentation of PCL make the diagnosis difficult so that it is frequently not made during lifetime of the patient. In our case the diagnosis was established and treatment started within three months after the onset of symptoms, while the patient was still in relatively favorable clinical condition. We believe that his lymphoma was of primary cardiac origin because the bulk of the tumor was located in the heart and it had peculiar extension pattern which followed the great vessels, instead of the usual mediastinal nodal spread. Although fever, increased ESR, CRP, LDH and anaemia point to a more diffuse disease the patient had also chronic hepatitis B with permanent elevation of liver transaminases. We believe that mild mediastinal lymphadenopathy was secondary to intrathoracic propagation of primary cardiac lymphoma. Since the biopsy of the atrial septum carries a much higher risk of cardiac perforation than the biopsy of the left ventricle, the procedure was performed in the cardiac catheterization laboratory under simultaneous echocardiography guidance. The first cycle of chemotherapy must be given with caution because of a high risk of cardiac rupture during rapid tumor regression (5). Intracranial progression of PCL, as seen in our patient, seems quite frequent. This can be explained by the seeding of lymphoma cells from intracardiac or intravascular tumor parts. Since drugs used commonly for first-line treatment of lymphoma do not penetrate the blood-brain barrier, an intracerebral relapse should not be interpreted as indicating chemorefractoriness of the tumor. Indeed, our patient responded favorably to high-dose methotrexate and brain irradiation, modalities used for treating primary cerebral lymphomas (6). Unfortunately, he developed encephalopathy, a common complication of brain irradiation in his age group.



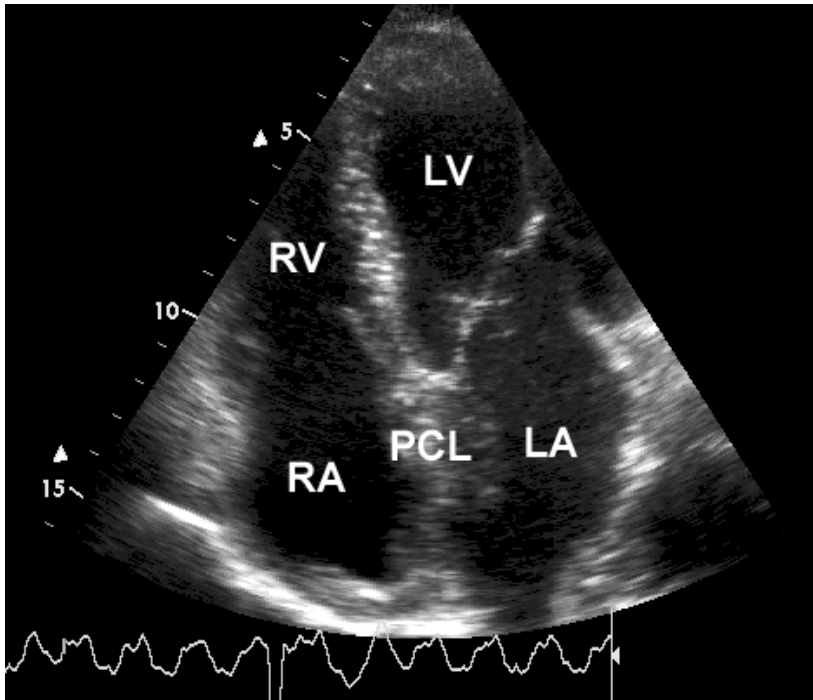
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## Figure legends

**Figure 1.** TTE (apical four chamber view) demonstrates PCL in interatrial septum.

LA indicates left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.



**Figure 2.** TEE demonstrates PCL in interatrial septum.

LA indicates left atrium; LV, left ventricle; RA, right atrium.

