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Diffuse involvement of the heart and great vessels in primary cardiac lymphoma

Aysel Aydin Kaderli[1]*, İbrahim Baran[1], Ozlem Aydin[1], Murat Bicer[2], Tayyar Akpinar[3], Fahir Ozkalemkas[4], Dilek Yesilbursa[1], and Sumeyye Gullulu[1]

[1]Department of Cardiology, Uludag University Faculty of Medicine, 16059 Gorukle, Bursa, Turkey; [2]Department of Cardiovascular Surgery, Uludag University School of Medicine, Bursa, Turkey; [3]Department of Nuclear Medicine, Uludag University School of Medicine, Bursa, Turkey; and [4]Department of Hematology, Uludag University School of Medicine, Bursa, Turkey

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Primary cardiac lymphoma (PCL) is an extremely rare disorder. In this report, a 57-year-old male with diffuse large B-cell lymphoma involving the heart and great vessels is presented. Trans-thoracic echocardiography was the first modality used to establish the diagnosis. Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) showed diffuse increased metabolic activity of the heart walls and hypermetabolic lesions occupying cardiac chambers in some areas. The patient underwent systemic chemotherapy, and after 13 days, a marked regression of the tumour mass was evident based on echocardiographic examination. After completing six R-CHOP chemotherapy treatments, PET imaging was planned to control the residual mass, but the patient was intubated due to pneumonia that developed after the sixth chemotherapy session and subsequently died due to sepsis.

Keywords Primary cardiac lymphoma • Echocardiography • Positron emission tomography • Heart block

Primary cardiac lymphoma (PCL) is extremely rare and is usually B-cell non-Hodgkin's type when present, accounting for 1% of all primary cardiac tumours and 0.5% of extranodal lymphomas;[1] however, it is more common in immunocompromised patients.[2] PCL remains asymptomatic until it produces a mass effect by obstructing cardiac chambers and great vessels or until it causes pulmonary or systemic embolization, complete atrioventricular (AV) block, and cardiac tamponade.[3] Its rarity and heterogeneous clinical presentation make the diagnosis difficult. Prognosis is poor due to diagnostic delay and the relevance of the site of the disease.[4] In this report, the case of a 57-year-old male with diffuse large B-cell lymphoma involving the heart and great vessels is presented.

Case report

A 57-year-old male with a history of diabetes mellitus was referred from another institution because of presyncope and pericardial effusion. He presented with mental confusion, weight loss, dyspnoea, dizziness, and presyncope with effort for 20 days. On admission, he had a body temperature of 37.2°C, a blood pressure of 110/74 mmHg, and a heart rate of 48 bpm, and lung fields revealed rales in the lower third section. Heart sounds were arrhythmic and bradycardic on auscultation. His erythrocyte sedimentation rate was 17 mm/h, complete blood count showed normocytic anaemia, and lactate dehydrogenase (666 IU) and C-reactive protein (6.63 mg/dL) were elevated.

Electrocardiography (ECG) indicated atrial fibroflutter with episodes of first-degree and complete AV block. Trans-thoracic echocardiography (TTE) revealed hyperechogenic masses, especially at the interatrial septum, right atrium free wall, right atrioventricular junction, anterior wall of the right ventricle, and posterior of the mitral annulus. The tumour circumferentially surrounded the ascending aorta and pulmonary artery. The pericardium was thickened and there was mild pericardial effusion (*Figure 1A*). No abdominal or mediastinal lymph nodes exceeding physiological size were detected, and no masses were found via computerized tomography (CT) scan; bone marrow examination was normal. Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) imaging was performed using a hybrid scanner (Biograph-6, Siemens Medical Systems, Erlangen, Germany). Whole-body images were obtained 60 min following injection of 185 MBq FDG using the three-dimensional

* Corresponding author. Tel: +90 2242951663; fax: +90 2242951626. *E-mail address*: ayselkaderli@yahoo.com, aakaderli@uludag.edu.tr

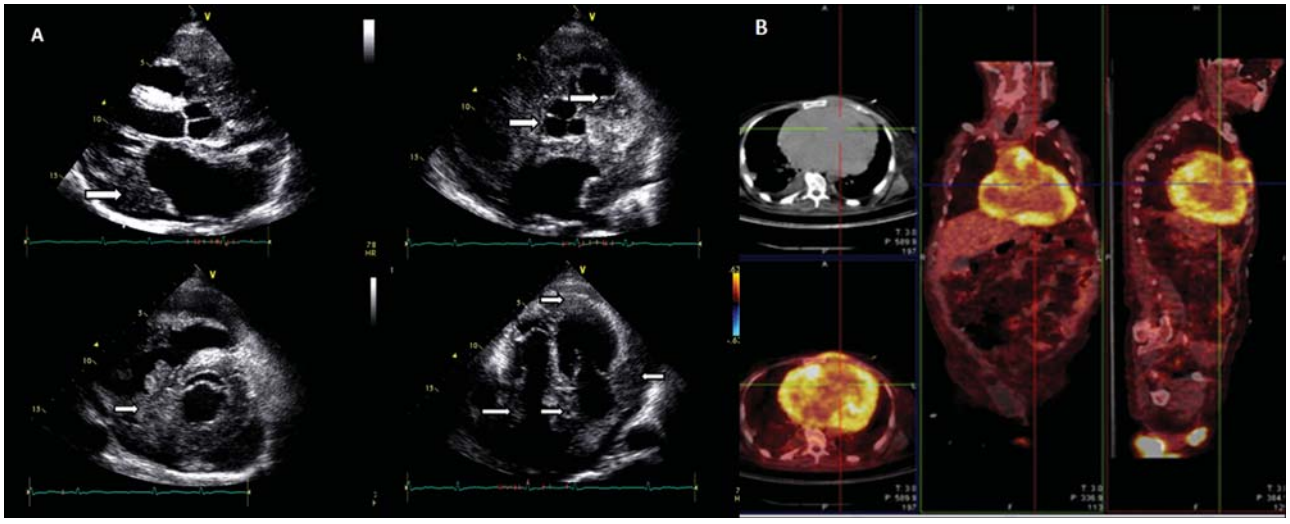

Figure 1 (*A*) Trans-thoracic echocardiography revealed hyperechogenic masses, especially at the interatrial septum, right atrium free wall, right atrioventricular junction, anterior wall of the right ventricle, and posterior of the mitral annulus. The tumour circumferentially surrounded the ascending aorta and pulmonary artery (white arrows). (*B*) Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography images showed diffuse increased metabolic activity of the heart walls and hypermetabolic lesions occupying cardiac chambers in some areas.

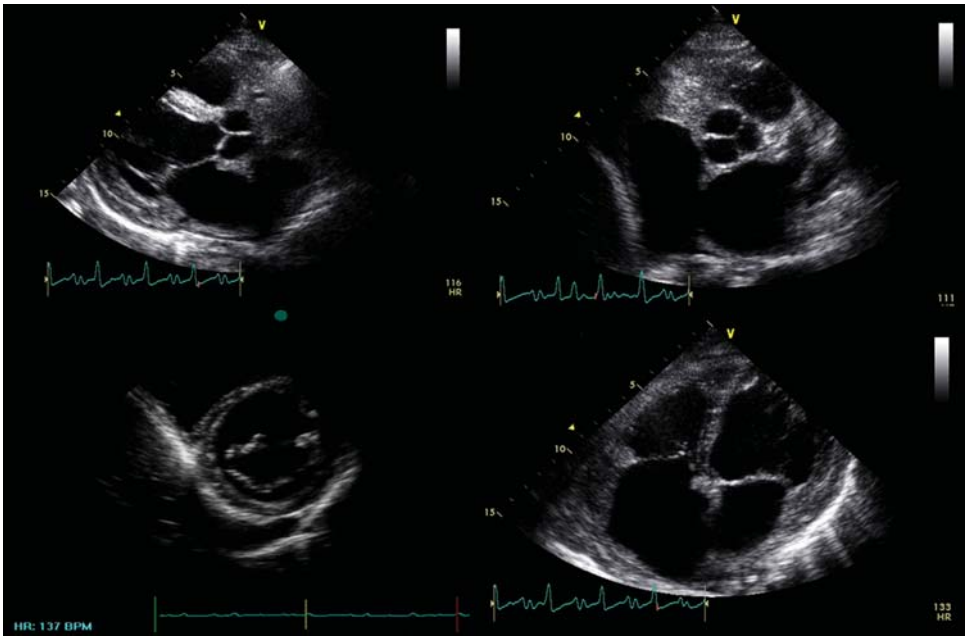

Figure 2 There was marked regression of tumour mass at the echocardiographic examination after 13 days of R-CHOP treatment.

acquisition method. Images showed diffuse increased metabolic activity of the heart walls and hypermetabolic lesions occupying cardiac chambers in some areas (*Figure 1B*). No other areas of pathological FDG uptake were observed in the rest of the body.

A biopsy was performed from the periphery of the aorta via a mini anterior thoracotomy. Microscopic examination of the biopsy specimen revealed diffuse large B cell lymphoma. Biochemical examination of the serum showed hypercalcaemia (corrected Ca: 15.1 mg/dL) on follow-up. The level of parathyroid hormone (PTH) was suppressed (3 ng/L, normal, 12–72 ng/L). Saline infusion, furosemide, steroid, and calcitonin were started to ameliorate the hypercalcaemia. After the diagnosis was established, systemic

chemotherapy consisting of cyclophosphamide, doxorubicin hydrochloride, vincristine sulphate, prednisolone, and monoclonal CD20 antibody (Rituximab) (R-CHOP) was initiated. There was marked regression of the tumour mass upon echocardiographic examination after 13 days of R-CHOP treatment (*Figure 2*). After chemotherapy, no symptoms of pulmonary or systemic embolism and no signs of complete AV block were detected, and the cardiac rhythm persisted as atrial fibroflutter. After six sessions of R-CHOP chemotherapy, PET imaging was planned to control the residual mass, but the patient developed pneumonia requiring intubation after the sixth chemotherapy session and later died due to sepsis.

Discussion

Primary cardiac tumours are rare, with one-fourth of them being malignant[5] and PCLs account for 1% of primary cardiac tumours.[1] The rarity and heterogeneous clinical presentation of PCL make its diagnosis difficult. Although in this case the diagnosis was established in the first month after the onset of symptoms, PET showed diffuse involvement of the heart walls. Delay in diagnosis appears to be a major factor associated with poor outcomes in PCL patients. On the basis of published data, it is difficult to estimate the effectiveness of early diagnostic imaging techniques. TTE was the first diagnostic modality used for the diagnosis of PCL in the patient presented in this study, and it revealed the clinical significance and haemodynamic effects of the tumoural mass. A CT scan failed to show the details of myocardial and pericardial infiltration, but it provided additional information about extra-cardiac involvement.

Recent studies in the field of oncology have combined the functional imaging of PET and the anatomic imaging of CT, thereby providing an efficient method for measuring tumour metabolism and proliferation, as well for analysing anatomic structure. In addition, with PET/CT imaging, tumours can be classified as malignant based on high FDG uptake, and the precise extent of tumours can be assessed.[6] For the patient described in this study, PET/CT provided information about both the extent of cardiac involvement and the lack of extracardiac organ involvement.

In the majority of cases, PCLs have been reported to arise in the right chambers of the heart, and the atria are involved in most patients.[4] In this case, there was diffuse involvement of the heart walls and also a large mass on the interatrial septum. Atrial flutter and complete heart block are uncommon presentations of PCL.[2] The present patient's ECG indicated atrial fibroflutter with episodes of first degree and complete AV block. The AV block disappeared after R-CHOP therapy.

Hypercalcaemia develops in 15% of patients with non-Hodgkin's lymphoma at any stage.[7] There is only one reported case of a patient with PCL complicated with hypercalcaemia as a paraneoplastic syndrome.[8] In this case, the serum calcium level was measured at 15.1 mg/dL during the follow-up period, and intact PTH was suppressed.

PCL, unlike other cardiac malignancies, responds to chemotherapy. In the case described here, there was marked regression of the tumour 13 days after initiation of R-CHOP therapy without any cardiac complications. PCL is a rapidly progressing disease and should be considered as an oncologic emergency.[9] Delay in diagnosis, paraneoplastic syndromes, and tumour effects such as complete AV block appear to be major factors associated with poor outcomes in PCL patients. If there are clinical and radiological suspicions of PCL, aggressive diagnostic procedures should be applied, and therapy should be started before irreversible cardiac damage occurs.

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