

CASE REPORT

Successful resection and reconstruction of primary cardiac lymphoma

Primer kardiyak lenfoma: Başarılı rezeksiyon ve dekonstrüksiyon

Barçın Öz cem, M.D.,¹ Hatice Soner Kemal, M.D.,² Özlem Balcıođlu, M.D.,¹
Hanife Özkayalar, M.D.,³ İlhan Sanisođlu, M.D.¹

¹Department of Cardiovascular Surgery, Near East University Hospital, Nicosia, Cyprus

²Department of Cardiology, Near East University Hospital, Nicosia, Cyprus

³Department of Pathology, Near East University Hospital, Nicosia, Cyprus

Summary– Primary cardiac lymphoma (PCL) is one of the rarest tumors of the heart. The most common type is diffuse, large B-cell lymphoma. Most often, the right atrium and the right ventricle are involved, and if not diagnosed and treated in time, it can be fatal. In this case, a female patient underwent an urgent operation for a large, infiltrative, right atrial mass. Extensive resection of the lateral walls of both atria and the interatrial septum as well as reconstruction were performed successfully. The pathological evaluation suggested PCL. The aim of this case is to raise awareness of this disease and to highlight clinical and surgical approaches.

Özet– Primer kardiyak lenfoma, kalbin en nadir tümörlerinden olup, en sık rastlanan tipi yaygın büyük B-hücreli lenfomadır. Sıklıkla sağ atriyum ve sağ ventrikül ile ilişkili olup, tanı ve tedavinin gecikmesi halinde ölümcül olabilmektedir. Bu yazıda, sağ atriyumda infiltratif görünümde yoğun kitle nedeni ile acil olarak ameliyat edilen olgu sunuldu. Her iki atriyumun yan duvarları ve atriyumlar arası septum geniş çapta kesildi ve ardından rekonstrüksiyon uygulandı. Bu olgu sunumunun amacı, hastalığın farkındalığını artırmak, klinik ve cerrahi yaklaşımı vurgulamaktır.

Tumors and thrombi are the most common types of cardiac masses. Primary cardiac lymphoma (PCL), the most common type being diffuse large B-cell lymphoma, is one of the rarest tumors of the heart.⁽¹⁾ Early diagnosis and treatment is important, since it can be fatal.

Presently described is a case of infiltrative PCL in a patient who had successful extensive resection of the lateral walls of the atria and the interatrial septum in addition to reconstruction.

CASE REPORT

A 52-year-old female patient was referred to our institution with progressive dyspnea at rest for 10 days. On physical examination, she was pale, hypotensive (blood pressure: 80/50 mmHg), and pulse rate was

105 bpm. There were no physical signs of systemic disease. Electrocardiogram showed sinus tachycardia. Transthoracic echocardiography (TTE) revealed a huge, right atrial mass that almost completely obliterated the right atrial cavity, allowing minimal flow from the tricuspid valve. Global pericardial effusion measuring approximately 2.0 cm at end diastole was seen, causing the right ventricle and the left atrium to collapse during systole. A chest computer tomography scan was performed to further define the borders of the mass (Fig. 1a). Based on these results and hemodynamic instability, the patient was scheduled for emergency cardiac surgery. Preoperative laboratory findings indicated a hemoglobin level of

Abbreviations:

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|-----|----------------------------------|
| CPB | Cardiopulmonary bypass |
| PCL | Primary cardiac lymphoma |
| TOE | Transesophageal echocardiography |
| TTE | Transthoracic echocardiography |

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Correspondence: Dr. Hatice Soner Kemal. Yakın Dođu Üniversitesi Hastanesi, Kardiyoloji Anabilim Dalı, Lefkoşa, Kıbrıs. Tel: +90 392 - 444 05 35 e-mail: kemal.hatice@hotmail.com

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10 g/dL, hematocrit of 32.9%, white blood cell count of $9.85 \times 10^3/\mu\text{L}$, and normal kidney and liver function test results. The erythrocyte sedimentation rate was high at 60 mm/hour.

Following induction of anesthesia, transesophageal echocardiography (TOE) was performed and confirmed a huge, heterogeneous, broad-based mass in the right atrium, with invasion into the interatrial septum and the right atrial free wall (Fig. 1b). There were no other cardiac abnormalities and tricuspid valve movements were normal with no outflow obstruction. Through a median sternotomy, approximately 1000 cc hemorrhagic pericardial effusion was drained and sent for cytological and microbiological analysis. After heparinization, bicaval cannulation was applied and cardiopulmonary bypass (CPB) was initiated. Under total CPB, a classic right atriotomy was performed with right atrial incision, and a huge, irregular mass filling the right atrium cavity was observed. The mass seemed to originate from the intra-atrial septum, thickening the septal wall, and extending to the right atrial cavity and the right and left atrial lateral walls. The mass was removed with the thickened septum and the left and right atrial lateral walls (Fig. 2a). The mass was irregular, soft, gelatinous, and fragile, like jelly, and the septal segment had calcified, necrotic areas (Fig. 2b). Bovine pericardium and a Dacron (E. I. du Pont de Nemours and Company, Wilmington, DE, USA) patch were used for the reconstruction of the atrial walls and the interatrial septum, respectively. After the de-airing procedure, the patient was weaned from CPB and the sternum was closed.

Histopathological findings revealed diffuse myocardial infiltration with large pleomorphic, atypical tumor cells. The tumor cells showed a positive immu-

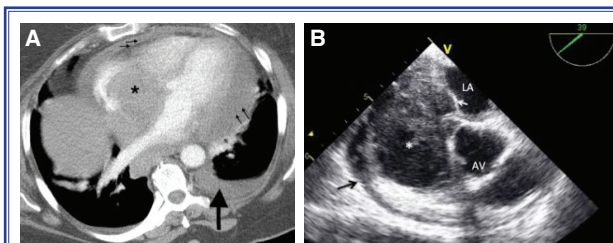


Figure 1. (A) Chest computer tomography showing the right atrial mass (asterisk), pericardial effusion (thin arrows), and pleural effusion (thick arrow). (B) Intraoperative transesophageal echocardiography.

Asterisk: Mass; AV: Aortic valve; Black arrow: Pericardial effusion; LA: Left atrium; White arrow: Interatrial septum.

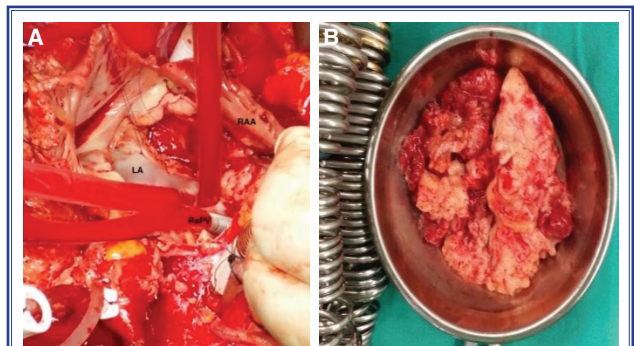


Figure 2. (A) Intraoperative view. The interatrial septum and the left and right lateral atrial adjacent walls were removed. (B) Macroscopic view of the removed mass.

LA: Left atrium; RAA: Right atrial appendix; RsPV: Right superior pulmonary vein.

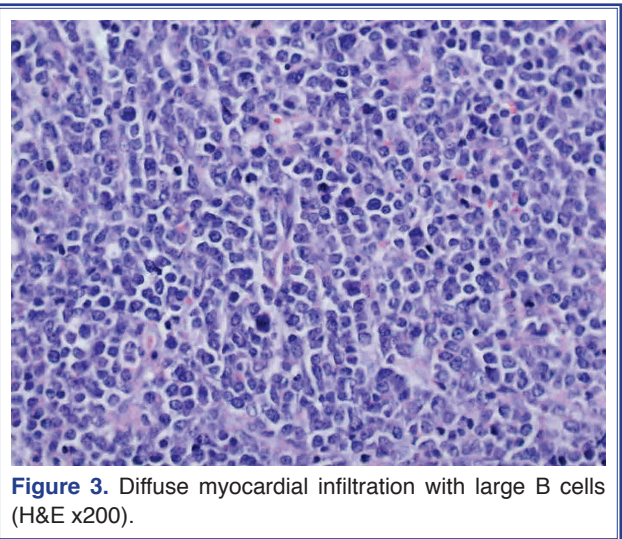


Figure 3. Diffuse myocardial infiltration with large B cells (H&E x200).

nohistochemical expression for CD20, CD79a, CD10, and bcl-6, but negative for cytokeratin, smooth muscle actin, Melan-A, bcl-2, and T-cell markers (CD3, CD5, CD30), conforming to PCL (Fig. 3). Operational margins were all clear.

After 2 weeks, the patient was discharged in good condition and referred to the oncology department for staging and further treatment. Postoperative echocardiography showed normal right heart function with normal blood flow.

DISCUSSION

This case report describes PCL that was highly infiltrative to the interatrial septum and atrial chambers. Primary cardiac tumors are extremely rare, and only 1.3% of malignant tumors are PCL.^[1]

PCL tumors can manifest, as any cardiac neoplasm can, as a thrombus formation and peripheral embolism, with arrhythmias, heart failure, pericardial effusion, sudden cardiac death, exertional dyspnea, or with systemic symptoms, such as weight loss and fever.^[2] In our case, the patient was in a difficult state, with hypotension and tachycardia present due to pericardial effusion, and very severe dyspnea. PCL usually occurs in adults, with a male-to-female ratio of 2:^[3] and often involves the right side of the heart. The involvement of the right atrium and epicardial and pericardial infiltration with pericardial effusion is typical.^[3]

In this case, TTE and hemodynamic state established the diagnosis and urgency of surgical treatment. But TOE was extremely important and valuable in providing information on localization, clearly displaying the attachment and anatomical relationship of the tumor, and guiding the surgeons' resection. In such complicated and urgent cases, TOE should be performed prior to sternotomy in order to plan the surgical approach and safely perform the excision.

It is known that surgical resection of PCL is often difficult and incomplete, and it is recommended to be reserved for patients with life-threatening hemodynamic compromise. Complete remission is achieved in fewer than 60% of cases.^[4] In our case, hemodynamic instability urged operation. It is necessary to diagnose PCL as early as possible and to use intensive treatment with new chemotherapy drugs in order to achieve better outcomes.^[5] One treatment modality without the other is not enough for complete treatment. Oncological staging procedures should be similar to those used for systemic lymphomas, as heart involvement is usually secondary rather than the only location. PCL usually affects older, immunocompetent patients, but all should be tested for HIV, since extranodal and heart involvement is more common in HIV-related lymphomas.^[6]

In conclusion, we report a case of PCL in a patient who had successful extensive resection of the tumor

and reconstruction, and was then referred for further treatment.

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Keywords: Cardiac tumor; diffuse large B-cell lymphoma; primary cardiac lymphoma.

Anahtar sözcükler: Kardiyak tümör; diffüz büyük B-hücreli lenfoma; primer kardiyak lenfoma.