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A Rare Case of Primary Cardiac Lymphoma Presenting with Acute Myocardial Infarction: to Be Happy or to Be Sorry?

Akut Miyokart Enfarktüsü ile Başvuran Nadir Bir Primer Kardiyak Lenfoma Vakası: Üzülmeli mi? Sevinmeli mi?

CASE REPORT
OLGU SUNUMU

ABSTRACT

Among primary malignant tumors of the heart, primary cardiac lymphomas are extremely rare. Early diagnosis is crucial in primary cardiac lymphoma cases as its non-specific symptoms often lead to delayed diagnosis and poor prognosis. In this case report, we presented a challenging case of primary cardiac lymphoma that was noticed during echocardiography of a patient admitted with acute myocardial infarction. A 32-year-old man was admitted to the emergency department with acute anterior ST-elevated myocardial infarction. His angiogram revealed an acute occlusion in the proximal left anterior descending artery with otherwise normal coronary arteries. After the total occlusion was passed with a guidewire, only a dense thrombus was observed. Therefore, an embolic source was suspected. Echocardiography revealed a giant mass (6 cm × 2.5 cm) attached to the interatrial septum. The patient was referred to early surgery for the resection of the mass. Histopathology and immunohistochemistry of the resected specimen demonstrated B cell non-Hodgkin lymphoma. Positron emission tomography and computerized tomography showed no lymph node and organ involvement. Two weeks after surgery, he was discharged and referred to the hematology department for chemotherapy. After 6 cycles, the positron emission tomography scan showed no abnormal accumulation indicating complete remission 7 months later. The clinical course of the patient was favorable for 1 and a half years. Acute myocardial infarction may be a manifestation of a rare entity such as primary cardiac lymphoma and an embolic source should always be considered. This is a case of pathologically diagnosed and successfully treated primary cardiac lymphoma.

Keywords: Acute myocardial infarction, embolism, primary cardiac lymphoma

ÖZET

Kalbin primer malign tümörleri arasında primer kardiyak lenfomalar oldukça nadirdir. Spesifik olmayan semptomları sıklıkla gecikmiş tanıya ve kötü prognoza yol açtığından, primer kardiyak lenfoma vakalarında erken tanı çok önemlidir. Bu olgu sunumunda, akut miyokart enfarktüsü ile başvuran bir hastanın ekokardiyografisi sırasında fark edilen bir primer kardiyak lenfoma olgusunu sunduk. 32 yaşında erkek hasta, akut anterior ST yükselmeli miyokart enfarktüsü ile acil servise başvurdu. Anjiyografide, proksimal sol ön inen arter tam tıkalı iken diğer koronerler tamamen normal idi. Kılavuz tel ile total oklüzyon geçildikten sonra sadece yoğun trombüs izlendi. Bu nedenle embolik bir kaynaktan şüphelenildi. Ekokardiyografide interatriyal septuma yapışık dev bir kitle (6 × 2,5 cm) saptandı. Hasta kitle rezeksiyonu için erken cerrahiye sevk edildi. Rezeke edilen örneğin histopatolojisi ve immünohistokimyası, B hücreli Hodgkin olmayan lenfoma ile uyumlu geldi. Pozitron emisyon tomografisi ve bilgisayarlı tomografide lenf nodu ve organ tutulumu görülmedi. Ameliyattan iki hafta sonra hasta taburcu edildi ve kemoterapi için hematoloji bölümüne sevk edildi. Altı kür kemoterapi sonrası, yedinci ay kontrolünde PET taramasında anormal bir birikim görülmedi ve tam remisyon kabul edildi. Bir buçuk yıl sonraki kontrolde de hastanın klinik durumu iyiydi. Akut miyokart enfarktüsü, primer kardiyak lenfoma gibi nadir bir antite ile ortaya çıkabilir, bu nedenle enfarktüste her zaman embolik kaynak da düşünülmelidir. Bu vaka, başarılı cerrahi ve kemoterapi uygulanan, patolojik tanısı olan bir kardiyak lenfoma hastasıdır.

Anahtar Kelimeler: Primer kardiyak lenfoma, embolizm, akut miyokart enfarktüsü

Primary cardiac tumors are extremely rare, with an autopsy incidence ranging from 0.001% to 0.03%.[1] Among primary malignant tumors of the heart, primary cardiac

Dilay Karabulut, M.D.[1]
Gülsüm Türkyılmaz, M.D.[2]

[1]*Department of Cardiology, Bakırköy Dr. Sadi Konuk Training and Research Hospital, İstanbul, Turkey*
[2]*Department of Cardiovascular Surgery, Bakırköy Dr. Sadi Konuk Training and Research Hospital, İstanbul, Turkey*

Corresponding author:
Dilay Karabulut
✉ dilay_karakozak@hotmail.com

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lymphomas (PCL) are extremely rare, too.[2] Almost all cardiac lymphomas are mostly non-Hodgkin B cell type which originates on the right side of the heart, especially the right atrium. Epicardial and pericardial infiltration with pericardial effusion is typical.[3] Early diagnosis is crucial in PCL cases as its non-specific symptoms often lead to delayed diagnosis and poor prognosis.[4]

In this case report, we presented a challenging case of PCL that was noticed during echocardiography of a patient admitted with acute anterior myocardial infarction (AMI).

Case Report

A 32-year-old man was admitted to the emergency department with new-onset typical chest pain. He had no family history, diabetes, and hypertension. He had only the record of smoking as a risk factor, and he also had a history of stomach bleeding 1 year before. On the physical examination, heart rate was 90 bpm and blood pressure 120/75 mm Hg. Pulmonary sounds were clear with no crackles, and heart sounds were regular with no murmur or gallop.

The initial electrocardiogram showed anterior ST-segment elevation with reciprocal changes in the inferior leads, so the patient was taken for emergency coronary angiography. His angiogram revealed an acute occlusion in the proximal left anterior descending artery with otherwise normal coronary arteries (Figure 1, Video 1*). After the total occlusion was passed with the guidewire, thrombolysis in myocardial infarction (TIMI)-1 flow was achieved. There was no calcification, plaque, or dissection, but only dense thrombus was observed. Therefore, an embolic source was suspected. Balloon angioplasty and stenting were performed, and the procedure was finished with complete revascularization.

The patient was referred to the coronary care unit for post-infarction management. Transthoracic and transesophageal, and 3-D echocardiography revealed a pedunculated giant mass (6 × 2.5 cm) in the left atrium attached to the interatrial septum fossa ovalis region and protruding to the left ventricle in diastole (Figure 2-4, Video 2-4*). Notably, this mobile mass prolapsing into the left ventricle was suspected to be myxoma. Also, left ventricular ejection fraction (40%) was impaired with anteroseptal mid, septum mid, and apical hypokinesis. There was mild mitral and tricuspid regurgitation and no pericardial effusion. Chest radiograph revealed cardiomegaly with no pleural effusion. The patient was referred to early surgery for the resection of the mass.

Histopathology and immunohistochemistry examination of the resected specimen demonstrated B cell non-Hodgkin lymphoma. After the histopathological diagnosis of the specimen obtained after surgery indicated the presence of lymphoma, positron emission tomography (PET) and computerized tomography (CT) were performed. They showed no lymph node and organ involvement. Two weeks after surgery, he was discharged in stable condition and referred to the hematology department for chemotherapy. Cyclophosphamide, vincristine, and prednisone therapy were initiated. After 6 cycles were administered, a PET scan performed 7 months after surgery showed no abnormal accumulation indicating complete remission. The clinical course of the patient was favorable for 1 and a half years.

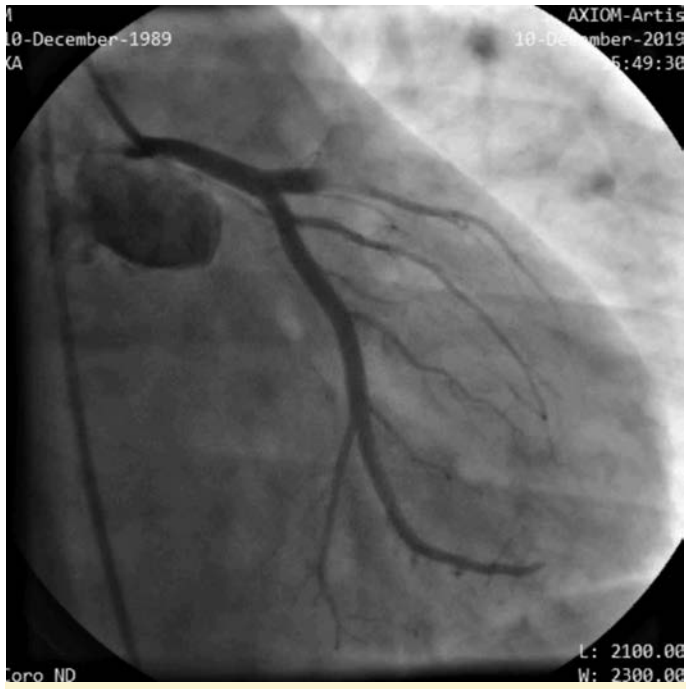

Figure 1. Angiography showed acute occlusion of the proximal left anterior descending artery with otherwise normal coronary arteries. There was no calcification, plaque, or dissection, but only dense thrombus was observed.

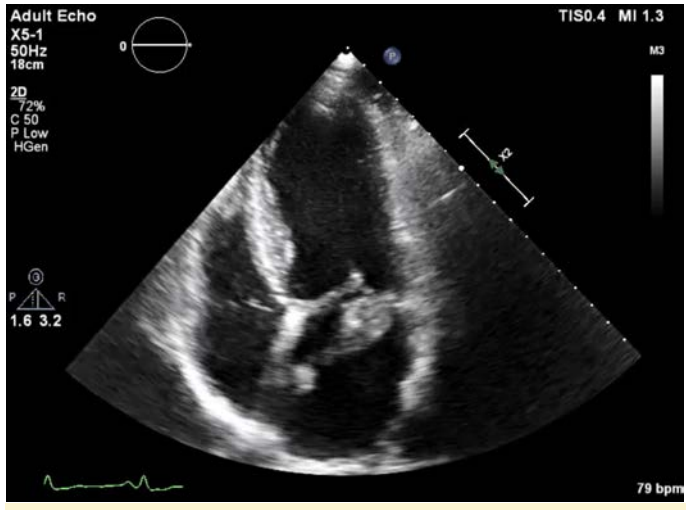

Figure 2. Transthoracic echocardiography revealed a pedunculated giant mass (6 × 2.5 cm) attached to the interatrial septum fossa ovalis region and protruding to the left ventricle in diastole.

ABBREVIATIONS

AMI	Acute anterior myocardial infarction
CT	Computerized tomography
MRI	Magnetic resonance imaging
PCL	Primary cardiac lymphomas
PET	Positron emission tomography
STEMI	ST elevation myocardial infarction
TIMI	Thrombolysis in myocardial infarction

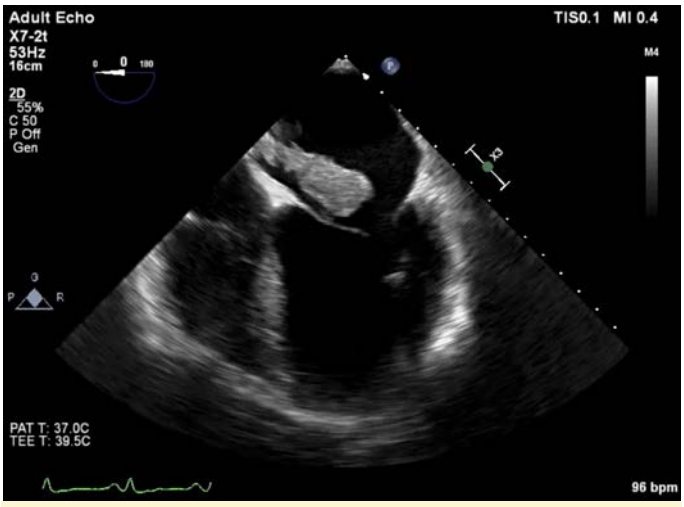

Figure 3. Transesophageal echocardiography revealed a pedunculated giant mass (6 × 2.5 cm) attached to the interatrial septum fossa ovalis region and protruding to the left ventricle in diastole.

Discussion

Primary cardiac lymphoma is a rare malignant cardiac tumor, which is especially found among immunocompromized and elderly individuals, and usually affects the right chambers of the heart and/or pericardium.[5] In our case, the tumor was limited within the left atrial chamber and there was no pericardial effusion and no extra involvement in the other body sites, as proved by CT and PET imaging.

Patients usually present with symptoms related to tumor extension and/or embolus (obstruction, arrhythmia, pulmonary or systemic embolism, conduction disturbances, fever, weight loss, and cardiac tamponade.[6,7] Our patient was admitted with angina due to anterior ST elevation myocardial infarction (STEMI), and he had no complaint of fever, night sweats, and chronic cough.

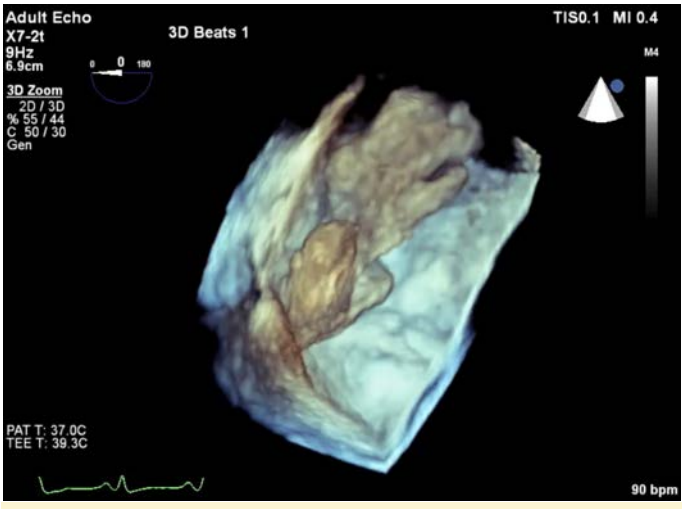

Figure 4. 3-D Transesophageal echocardiography revealed a pedunculated giant mass (6 × 2.5 cm) attached to the interatrial septum fossa ovalis region and protruding to the left ventricle in diastole.

Cardiac tumors are among the possible embolic sources of coronary embolization and can present as AMI.[8] Embolic causes must be suspected in patients who have otherwise healthy coronary arteries. Systemic embolization is more frequently associated with polypoid tumors with an irregular and friable surface and tumors that prolapse into the ventricle.[8]

Petrich et al[4] combined the data from all available cases of primary cardiac lymphoma in the literature up to 2011. In contrast to our case, they reported that only 6% of the patients had an embolic event, and there were no cases of coronary embolization.

It is important to perform the echocardiography in all patients presenting with AMI before beginning medical treatment since this is the only way to avoid the use of potentially harmful anticoagulant agents that increase the risk of embolization in the case of PCL either by lysis of accumulated thrombi or by hemorrhage and release of small fragments. In the case presented, PCL was of the polypoid type with an irregular and brittle surface, prolapsing through the mitral valve. Despite these characteristics and the tumor's large size, no obstructive symptoms or peripheral embolic phenomena had been documented before the current admission. We thought that AMI might be related to coronary embolism due to lymphoma because of the young age of the patient and the absence of any risk factors other than smoking.

Among the diagnostic tools, a transthoracic echocardiogram should be considered as the first approach for cardiac tumors. Although echocardiography is the mainstay in clinical practice for the detection of cardiac tumors, it is inadequate for distinguishing between tumors and thrombi because of its limited tissue characterization. Cardiac CT and magnetic resonance imaging (MRI) are useful for differentiating not only between tumors and thrombi but also between benign and malignant tumors.[9] In our patient, because the mass was large and protruded into the left ventricle, malignant involvement was observed, and no extra focus was detected in PET CT, so the patient was referred to surgery without performing MRI.

Management of cardiac lymphoma is varied. The treatment of primary cardiac lymphomas includes standard chemotherapy for B-cell lymphomas, combined with surgical resection, radiation therapy, or both. The prognosis for patients with either primary or secondary cardiac lymphoma is usually poor. A recent analysis of PCL and treatment outcome patterns demonstrated a median overall survival period ranging from 1 to 22 months.[4] Therefore, early diagnosis is critical. Our patient had no recurrence and careful follow-up is ongoing.

With this case report, we aim to highlight the fact that a diagnosis, as standard in daily clinical practice of AMI may be a manifestation of a rare entity such as PCL and an embolic source should always be considered in cases of AMI. The absence of atherosclerotic coronary lesions or thrombi is compatible with the high rate of recanalization seen in emboli from PCL, as in our patient. We also emphasize the fundamental importance of echocardiography in this case, which was performed early and enabled a correct diagnosis and immediate referral for the only treatment that could prevent a potentially poor prognosis.

This is a case of pathologically diagnosed and successfully treated primary cardiac lymphoma. In this case, we wanted to underline that coronary embolism should be considered in young patients who had AMI without risk factors and that we should consider malignancy among possible causes.

*Supplementary video files associated with this article can be found in the online version of the journal.

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