Massive pericardial effusion as the primary manifestation of high-grade malignant lymphoma

Yüksek dereceli habis lenfomanın esas bulgusu olarak ciddi perikart efüzyonu

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Pericardial effusion might be the first presentation of various pathologies including malignant tumors. Massive pericardial effusion as the primary manifestation of high-grade malignant lymphoma is a very rare condition. A 53-year-old woman presented with progressive dyspnea of one week history. Physical examination showed venous distention of the neck veins and diminished heart sounds. The chest X-ray demonstrated increased cardiothoracic index. Transthoracic and transesophageal echocardiographic examinations showed massive pericardial effusion without any other pathology. Hematologic and biochemical tests showed only anemia. The patient underwent pericardiocentesis. Pericardial adenosine deaminase test and cultures were negative. Cytopathologic examination of the fluid showed huge lymphocytes and highly atypical lymphoid cells consistent with high-grade malignant lymphoma (non-Hodgkin's lymphoma). Immunohistochemical analysis showed positivity for leukocyte common antigen. No other primary origin could be determined.

Key words: Lymphoma, non-Hodgkin/diagnosis/pathology; pericardial effusion.

Perikart efüzyonu habis tümörler de dahil çeşitli patolojilerin ilk bulgusu olabilir. Yüksek dereceli habis lenfomanın esas bulgusu olarak yaygın perikart efüzyonu ise çok nadir bir durumdur. Elli üç yaşında kadın hasta, bir haftadır var olan ilerleyici nefes darlığı ile başvurdu. Fizik muayenede boyun damarlarında gerginlik gözlendi; kalp sesleri zayıflamıştı. Göğüs radyografisinde kardiyotorasik indeks artmış bulundu. Transtorasik ve transözofageal ekokardiyografi incelemelerinde yaygın perikart efüzyonu dışında başka bir sorun görülmedi. Hematolojik ve biyokimyasal incelemelerde sadece anemiye rastlandı. Hastaya perikardiyosentez yapıldı. Perikardiyal adenozin deaminaz testi ve kültür sonuçları negatif bulundu. Perikart sıvısının sitolojik incelemesinde, yüksek dereceli habis lenfoma (non-Hodgkin lenfoma) ile uyumlu dev lenfositler ve ileri derecede atipik lenfoid hücreleri saptandı. İmmünhistokimyasal değerlendirmede lökosit ortak antijeni pozitif bulundu. Hastada başka bir primer kaynak bulunamadı.

Anahtar sözcükler: Lenfoma, non-Hodgkin/tanı/patoloji; perikart efüzyonu.

There are multiple etiologies for massive pericardial effusion. On this list, malignant pathologies such as lung and breast cancers are not uncommon. Although lymphomas are a common underlying etiology for pleural effusions, pericardial effusion due to malignant lymphomas are rare in adults.[1] In addition, massive pericardial effusion as the primary manifestation is an extremely rare condition.

CASE REPORT

A 53-year-old female was admitted to our emergency unit with progressive dyspnea of one week history. Her past medical history was unremarkable. Vital signs were stable with systolic/diastolic blood pressures of 130/85 mmHg and heart rate of 90 bpm. Physical examination showed venous distention of the neck veins and diminished heart sounds. Arterial blood gas analysis was unremarkable. The chest X-ray demonstrated increased cardiothoracic index with normal lung fields. The patient was referred to the echocardiography laboratory for identification of any cardiac pathology including heart failure, valvular dysfunction, aortic dissection, or pericardial effusion. Transthoracic echocardiography showed massive pericardial effusion with a swinging heart and no other pathology including a cardiac mass. Transesophageal

Received: February 5, 2009 *Accepted:* March 23, 2009

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echocardiography showed similar findings. For identification of the etiology, further laboratory tests were performed. Complete blood count revealed anemia (hemoglobin 10.6 g/dl) suggesting a chronic disease. White blood cell and platelet counts were normal. Peripheral blood smear showed no abnormality. Blood chemistry and thyroid function tests were also in normal limits. Anti-HIV test was negative. Serologic markers such as high-sensitivity CRP, rheumatoid factor, anti-nuclear antibody, and anti-dsDNA were all negative. Tumor markers of CEA, CA 19-9, CA 125, beta-HCG, and alpha-fetoprotein were negative. For diagnostic and therapeutic purpose, pericardiocentesis was performed. A total of 1,000 ml of hemorrhagic fluid was withdrawn. Pericardial adenosine deaminase test was negative. Aerobic and anaerobic cultures of the fluid were negative. Cytopathologic examination of the fluid showed huge lymphocytes with hyperchromatic nuclei and highly atypical lymphoid cells consistent with high-grade malignant lymphoma (non-Hodgkin's lymphoma) (Fig. 1). In addition, immunohistochemical analysis showed positivity for leukocyte common antigen. Thoracoabdominal computed tomography scans failed to reveal the primary origin of the pathology. The patient was referred to the department of hematological oncology for further diagnosis and treatment.

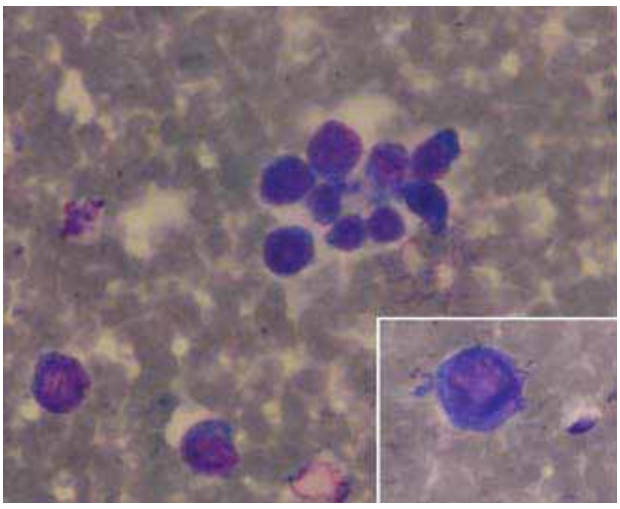

Figure 1. Atypical lymphoid cells (May-Grünwald-Giemsa stain; x100) and the inset showing a sample cell with the same staining (x200).

DISCUSSION

Pericardial fluid accumulation in patients with lymphoma might be due to direct involvement of the pericardium and/or hematogenous spread. In general, other organ-system involvement is seen in effusive lymphomas, and primary effusive lymphomas are very rare. Primary effusive lymphoma is a distinct clinicopathologic entity with an exclusive or dominant involvement of serous spaces without a detectable solid tumor mass.[2] It is generally seen in AIDS patients and is extremely rare in HIV-negative patients.[3,4] Diagnosis is generally made by cytopathologic examination of the pleural fluid. Morphologically, the neoplastic cells are large, have round-to-irregular nuclei, prominent nucleoli, and varying amounts of cytoplasm that is occasionally vacuolated. The appearance of the cells may be immunoblastic, plasmablastic, or anaplastic.[5] Despite various therapies including chemotherapy, radiotherapy, and surgery, the prognosis is poor. Patients with malignant lymphomas and massive pericardial effusion are treated palliatively with pericardiocentesis.

In conclusion, pericardiocentesis is necessary in patients with massive pericardial effusion for therapeutic and diagnostic purposes. The fluid should be investigated cytopathologically since pericardial effusion can be the primary manifestation of malignant lymphomas.

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