An unusual cause of spontaneous hemothorax: cardiac angiosarcoma

Spontan hemotoraksın nadir bir nedeni: Kalp anjiyosarkomu

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Summary– Angiosarcoma, the most common primary malignant neoplasm of the heart in adults, usually presents as pericardial effusion or right-sided heart failure. Rupture of an angiosarcoma-infiltrated cardiac chamber as a cause of hemothorax is very rare in the literature. In this report, we describe a 34-year-old male patient, who presented to emergency service with sudden chest pain and dyspnea. The diagnostic work-up revealed spontaneous right-sided hemothorax and a large right atrial (RA) mass with suspicious atrial perforation. An urgent surgery showed a vascularized irregular RA mass invading the parietal pericardium and pleura and a perforation of the RA free wall. Histopathologic examination confirmed the diagnosis of angiosarcoma, and the patient was subsequently referred for radiotherapy and chemotherapy.

Özet– Yetişkinlerde kalbin en sık görülen primer malign neoplazmı olan anjiyosarkom genellikle perikart sıvısı veya sağ kalp yetersizliği ile karşımıza çıkar. Anjiyosarkom ile enfiltre bir kalp boşluğunun yırtılarak hemotoraksa neden olması literatürde oldukça nadirdir. Bu yazıda, acil servise ani göğüs ağrısı ve dispne ile başvuran 34 yaşındaki erkek hasta sunuldu. Tanısal araştırmalar hastada spontan sağ hemotoraks ve sağ atriyumda büyük bir kitle olduğunu gösterdi, atriyum perforasyonu düşünüldü. Öncelikli cerrahide sağ atriyumda pariyetal perikarda ve plevraya yayılan düzensiz damarsal kitle ve sağ atriyum serbest duvarında yırtılma görüldü. Histopatolojik inceleme ile anjiyosarkom tanısı kondu ve hasta radyoterapi ve kemoterapi için yönlendirildi.

Angiosarcoma is the most common primary malignant neoplasm of the heart in adults.[1] It is a highly aggressive tumor with early local and systemic spread. Clinical presentation is most often that of pericardial effusion or right-sided heart failure;[2] however, unusual presentations are also reported. In this report, we describe a male patient with a right atrial (RA) angiosarcoma, who presented with spontaneous right-sided hemothorax.

CASE REPORT

A 34-year-old male presented to emergency service with sudden chest pain and dyspnea. His vital signs were normal except for mild tachypnea. Since the physical examination and chest X-ray suggested pleural effusion (Fig. 1a) and a hemorrhagic sample was aspirated, contrast- enhanced computerized tomography (CT) was performed. This scan revealed a right-sided pleural effusion with contrast leakage into the pleural space and a suspicious RA mass with contrast uptake (Fig. 1b). After tube drainage of 2000 cc blood, the patient was transferred to cardiology for further evaluation. Cardiac auscultation revealed normal heart sounds. His electrocardiogram was normal. Laboratory investigations were remarkable for anemia. Transthoracic echocardiography demonstrated a large mass in the RA free wall with a broad-based attachment and protrusion into the RA cavity. Lateral to the RA, there was an echo-free space, which was surrounded by an echodense irregular structure (Fig. 1c,

Abbreviations:

RA Right atrial
CT Computerized tomography

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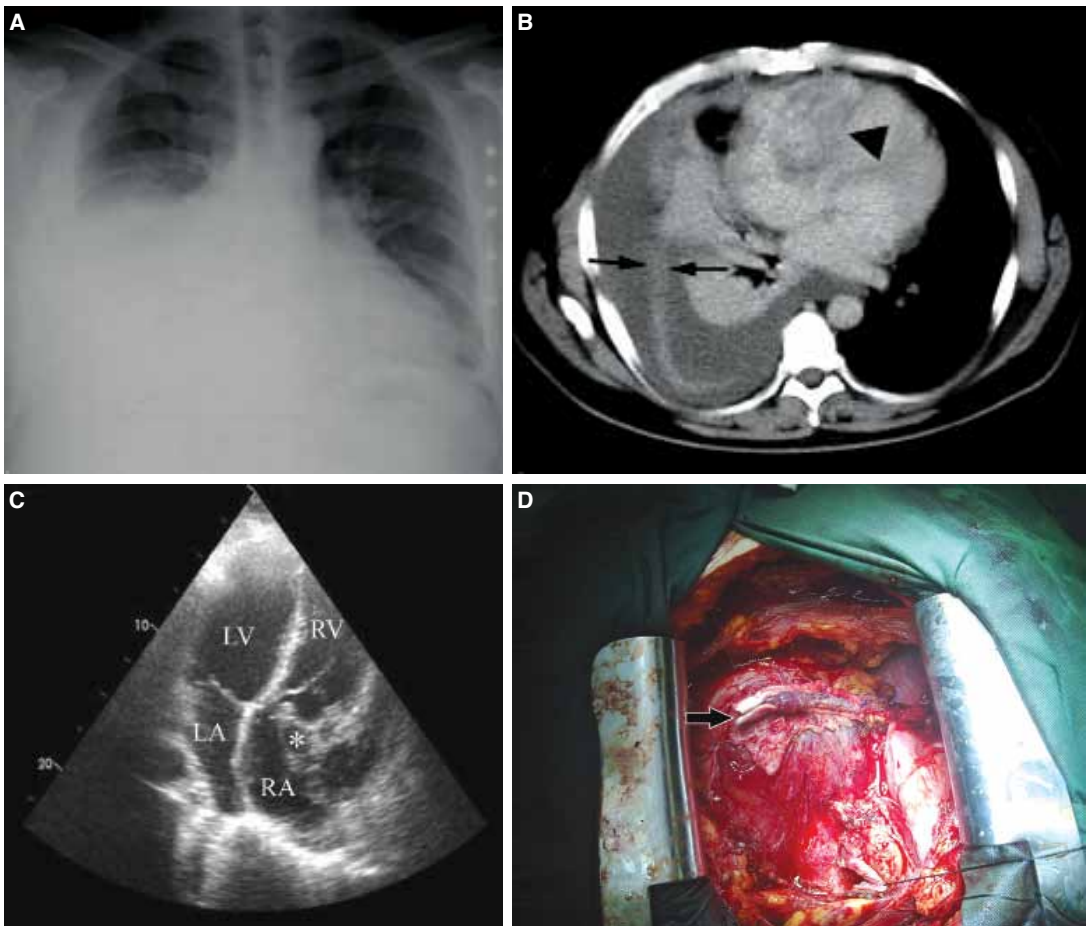

Figure 1. **(A)** Chest X-ray showing a large pleural effusion. **(B)** Contrast-enhanced CT revealed a right-sided pleural effusion with contrast leakage into the pleural space (arrows) and a suspicious right atrial mass with contrast uptake (arrowhead). **(C)** Apical four-chamber view showing an irregular mass (asterisk) in the right atrial free wall with protrusion into the cavity. **(D)** Intraoperative view showing repaired site of the right atrium (arrow). RA: Right atrium; RV: Right ventricle; LA: Left atrium; LV: Left ventricle.

Video 1*). In light of the hemothorax history, this image was interpreted as perforation of the RA wall with bleeding into the right pleural space. In the tricuspid inflow window, invasion into the tricuspid annulus and basal segment of the right ventricular wall was present. Color Doppler revealed blood flow within the mass (Video 2*).

An urgent surgery was performed to restore the integrity of the RA. Sternotomy showed a vascularized irregular mass, which invaded the right parietal pericardium and right pleura. A perforation of the RA free wall next to the appendage was found. Histologic examination of a frozen section revealed a type of malignant mesenchymal tumor. Under cardiopulmonary bypass, it was attempted to resect the mass completely, but extensive tumor infiltration of the RA and right ventricle precluded total resection. After partial resection of the mass and involved RA, the RA was repaired (Fig. 1d). Histopathologic examination confirmed the diagnosis as angiosarcoma. The patient was subsequently referred for radiotherapy and chemotherapy. At the five-month follow-up, he remained on chemotherapy.

DISCUSSION

Spontaneous hemothorax is generally caused by lung malignancies, anticoagulant medications, vascular ruptures (aortic dissection, arteriovenous malformations), pulmonary infarctions, and hematologic abnormalities.[3] Rupture of an angiosarcoma-infiltrated cardiac chamber as a cause of hemothorax is very rare in the literature.[4]

Primary cardiac angiosarcoma is more common in men, with a peak incidence between 30-50 years.[1,2] It originates from the RA almost exclusively. It usually presents with dyspnea, chest pain, hemoptysis, and syncope. Sometimes, an abrupt event may occur as a first clinical sign, as in our case. Metastases mainly to the lungs, lymph nodes, bone, pleura, and brain are found in many of patients at the time of diagnosis. Extensive tumor infiltration of the myocardium and infiltration or direct extension into the pericardium are other common properties. Echocardiography is usually the first imaging modality for the diagnosis, but CT, magnetic resonance imaging, and positron emission tomography are particularly helpful to delineate the extent of tumor infiltration and to assess for metastases.

Treatment of angiosarcoma is controversial due to the poor prognosis.[1,2] Surgical resection is indicated when no evidence of metastases exists and when myocardial resection is reparative. Although extensive tumor infiltration into the myocardium precluded total resection in our patient, restoration of the RA integrity and even incomplete resection of the mass may provide substantial symptom-free survival.[5] Chemotherapy, radiotherapy, immunotherapy, and cardiac transplantation have been used as therapeutic alternatives, but in most of the patients they do not prolong survival.

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

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Anahtar sözcükler: Tanı, ayırıcı; kalp tümörleri; hemanjiyosarkom/komplikasyon; hemotoraks/etiyoloji; sarkom/tanı.