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Address for Correspondence: Dr. Yakup Ergül,
İstanbul Mehmet Akif Ersoy Göğüs Kalp ve Damar Cerrahisi
Eğitim ve Araştırma Hastanesi,
Çocuk Kardiyoloji/Elektrofizyoloji Bölümü,
İstanbul-Türkiye
Phone: +90 212 692 20 00/4019
E-mail: yakupergul77@hotmail.com - yakupergul77@gmail.com
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A rare cause of dyspnea: Left atrial angiosarcoma

 Ebru İpek Türkoğlu,  Oğuz Yavuzgil¹
Department of Cardiology, İzmir Kemalpaşa State Hospital; İzmir-Turkey
¹Department of Cardiology, Faculty of Medicine, Ege University;
İzmir-Turkey

Introduction

Primary malignant cardiac neoplasms are extremely rare tumors. In the present report, a case of a patient with a large car-

diac mass in an unusual location is described after obtaining his and his relative's approval.

Case Report

A 64-year-old male patient was admitted to the cardiology clinic with dyspnea that increased over 1 month. The patient suffered from no other known condition. Electrocardiography showed sinus rhythm, heart rate of 85/bpm, and blood pressure of 134/84 mm Hg. His laboratory parameters and NT-proBNP level were within normal limits (NT-proBNP=121 pg/mL, upper limit for adjusted gender and age <210 pg/mL). A transthoracic echocardiogram showed a large (4.6×5.4 cm) mass originating from the left atrium (Videos 1 and 2). Thorax computed tomography confirmed a 4.8×5.4 cm mass with smooth borders located at the inferior site of the left atrium (Fig. 1, 2). Two radiologists reviewed CT images and the mass was considered extracardiac, with central low-density characteristics, probably a complex cystic lesion such as a bronchogenic cyst or an esophageal duplication cyst. A transesophageal echo confirmed a probable extracardiac mass as well. Preoperative cardiac MRI and PET-CT were not performed because the initial diagnosis did not reveal a malignant tumor. Coronary angiogram showed normal coronary arteries and a mass image with arterial blood supply from the distal circumflex artery. Thereafter, the patient underwent cardiac surgery for removal of the mass. Because the mass was considered extracardiac, the first surgical cut was left anteroateral. Hence, the surgical exploration was compatible with an intracardiac mass originating from the left atrial basis; sternotomy was performed and surgery was continued under cardiopulmonary bypass. The 6.5×5.5×4 cm left atrial mass



Figure 1. Pre-contrast thoracic CT cross-sectional image of the mass

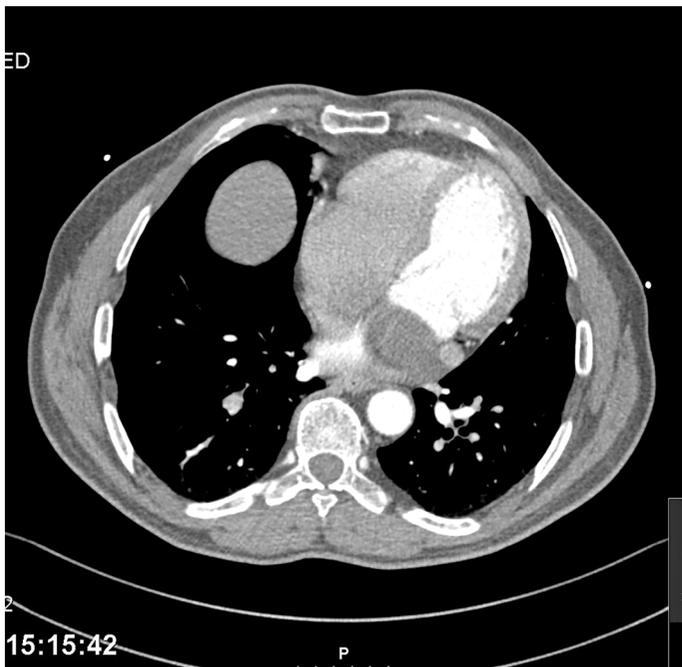


Figure 2. Post-contrast thoracic CT cross-sectional image of the mass

was removed with its capsule from the left atrium and sent to the pathology laboratory with an initial diagnosis of lipoma. The pathology report confirmed the diagnosis of left atrial angiosarcoma via immunohistochemical analysis. The tumor was found to be desmin, SMA, panCK, S100, MyoD1, Myogenin, and CD45 negative and CD31, CD34, D2-40 focal, and Ki67 (20%) positive, with malignant cells in the surgical borders. Because the initial diagnosis completely changed, the patient was referred to the oncology department. Oncological evaluation was made by both medical and radiation oncology departments. Postoperative PET-CT did not show metastasis, but due to the aggressive nature of the tumor, the patient underwent chemotherapy and radiotherapy following the oncology council's decision. The choice of treatment modality was especially difficult because of the rareness of the tumor. A paclitaxel-based chemotherapy was initiated. After completion of chemotherapy, thorax irradiation was performed by the radiation oncology department. Follow-up has been performed with PET-CT scans every 6 months. To the best of our knowledge, this was the first case of left atrial angiosarcoma reported in our country. The initial diagnosis was made in February 2017, and the patient did well 2 years following the diagnosis. Unfortunately, the patient died in the 24th month of diagnosis because of multiple brain metastases.

Discussion

Primary cardiac tumors are rare and primarily benign in nature, with the most common form being myxoma, which generally appears as a solitary mass in the left atrium. Differential diagnosis with thrombus is essential (1). The

most common form of malignant heart tumors is sarcoma (2). The incidence of primary cardiac sarcoma is approximately 0.0001% in autopsy series, which is extremely low. The most common form of sarcoma is angiosarcoma, which primarily originates from the right atrium (3-5). Although the right atrium is the most common site for an angiosarcoma, the left atrium, right ventricle and left ventricle have also been reported (5). Associated symptoms are nonspecific and generally delayed depending on the involved chamber and until the tumor reaches a certain size. The most frequent symptoms are dyspnea, chest pain, right heart failure, palpitation, and fever (5). As in our case, diagnosis is made on the basis of the pathology of the surgical specimen (6). Surgery is the treatment of choice, but radical excision is generally not possible because of the size and location of the tumor. Combination chemotherapy and radiotherapy are superior options for survival, but only in patients with radical resection. The first-line chemotherapy is an anthracycline-based chemotherapy, which is well established for cardiotoxicity. Recently, a second-line regimen with docetaxel or paclitaxel has been defined. Irradiation is recommended for prevention and management of residual tumor, but the curative dosage is generally similar to the cardiotoxic dose (5, 7). Due to its aggressive nature with increased risk of recurrence and metastasis, the expected survival is unfavorable, usually 6–12 months from the time of diagnosis (3, 4, 6). Unfortunately, only metastases at diagnosis and radical excision have a significant impact on the prognosis of cardiac sarcomas; chemotherapy, radiotherapy, age, gender, the degree of malignancy, and histological subtypes have no impact (7). Longer survival is reported in few cases and the location correlates with survival. Generally, left heart tumors appear to have a better prognosis (6). Less than a dozen cases with left atrial angiosarcoma have been reported in English literature in the past two decades (3, 4). Experience sharing is crucial because of the low number of patients with angiosarcoma in an unusual location as the left atrium.

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

Video 1. Transthoracic echocardiographic image of the mass; apical 4-chamber view

Video 2. Transthoracic echocardiographic image of the mass; apical 4-chamber view

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Address for Correspondence: Dr. Ebru İpek Türkoğlu,
İzmir Kemalpaşa Devlet Hastanesi,
Kardiyoloji Kliniği, 35730
İzmir-Türkiye
Phone: +90 532 628 34 72
E-mail: dripek73@yahoo.com
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