

Resection of a giant right atrial angiosarcoma invading the atrioventricular groove

Atriyoventriküler oluşu tutan dev sağ atriyal anjiyosarkom rezeksiyonu

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ABSTRACT

Cardiac angiosarcomas are the most commonly seen primary malignant tumors of the heart. They usually present abruptly with non-specific symptoms after being asymptomatic for a long period. Metastases are frequently present at the time of diagnosis, which limit the indication of surgical resection to the minority of patients. In this report, we describe a 28-year-old male case with a non-metastatic giant primary right atrial angiosarcoma, who underwent successful surgical excision of the tumor and reconstruction of the right atrium with a bovine pericardial patch.

Keywords: Angiosarcoma; cardiac tumors; obstruction syndrome; superior/inferior vena cava.

Cardiac angiosarcomas are the most common primary malignant tumors of the heart.^[1] They usually present abruptly with non-specific symptoms after being asymptomatic for a long period.^[1] Metastases are usually present at the time of diagnosis, which limit the indication of surgical resection to the minority of patients.^[2] In this report, we describe a case with non-metastatic giant primary right atrial angiosarcoma, who underwent successful surgical excision of the tumor and reconstruction of the right atrium with a bovine pericardial patch.

CASE REPORT

A 28-year-old man was admitted to our emergency department with worsening shortness of breath

ÖZ

Kardiyak anjiyosarkomlar, kalbin en sık görülen primer malign tümörleridir. Genellikle uzun bir süre asemptomatik seyrettikten sonra, nonspesifik semptomlar ile aniden ortaya çıkar. Sıklıkla tanı esnasında, cerrahi rezeksiyon endikasyonunu hastaların küçük bir kısmı ile sınırlayan metastazlar bulunur. Bu yazıda, başarılı bir şekilde tümörün cerrahi olarak rezekte edildiği ve sağ atriyumun sığır perikard yaması ile rekonstrüksiyonunun yapıldığı, metastatik olmayan dev primer sağ atriyal anjiyosarkomlu 28 yaşında erkek bir olgu sunuldu.

Anahtar sözcükler: Anjiyosarkom; kalp tümörleri; tıkanma sendromu; superior/inferior vena cava.

and bilateral lower limb edema for one month. His medical history was non-specific. Chest X-ray showed typical flask-shaped cardiomegaly (Figure 1a) and transthoracic echocardiography confirmed the presence of moderate to severe pericardial effusion with a giant heterogeneous mass occupying most of the right atrial cavity. The mass was protruding through the tricuspid valve into the right ventricle and extending from the superior to the inferior vena cava (Figure 1b). Chest computed tomography (Figure 1c) and cardiac magnetic resonance imaging (Figure 1d) confirmed the giant tumor in the right atrium, which was most probably angiosarcoma. Further examinations revealed bilateral lower limb deep vein thrombosis, ascites, congested liver, high liver enzymes, superior vena



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Figure 1. (a) Chest X-ray showing the flask shape cardiomegaly typical for large pericardial effusion. (b) Apical 4 chamber echocardiogram showing the same huge mass filling almost the whole right atrium. (c) Preoperative contrast-enhanced computed tomographic scan shows an irregular low-attenuation mass (7.5x6.8 cm) in the right atrium. (d) Cardiac magnetic resonance imaging showing same huge mass in the right atrium invading the adjacent pericardium with small extension to the right ventricle.

cava obstruction symptoms, and mildly impaired kidney function. He had worsening symptoms and his overall condition was deteriorating during his admission to the coronary intensive care unit. Based on a multidisciplinary approach, the decision was made for cardiac surgery for surgical debulking of the tumor to relieve severe symptoms and to repair of the right atrium.

The giant tumor (11x12x14 cm), which was invading most of the atrial wall and extending to atrioventricular groove up to the right coronary artery, was resected in a single mass (Figure 2b). The right atrial defect was repaired by a large bovine pericardial patch (Figure 2a). Suturing the pericardial patch to the atrioventricular groove was done with interrupted sutures under direct visualization of the right coronary artery to avoid its compression. His postoperative course was uneventful and his symptoms improved dramatically following surgery. Histopathological examination of the resected right atrial mass was reported as a high-grade angiosarcoma (Figure 2c, d). The patient was discharged two weeks after surgery and he received the first cycle of chemotherapy

after one month from surgery (first cycle of three, weekly/paclitaxel 175 mg/m²). A total of six cycles of chemotherapy followed by radiotherapy to the site of the tumor was planned.

The patient was uneventful for seven months after discharge. However, he died later in the oncology hospital due to distant metastases to the lungs, liver, and bone.

DISCUSSION

Malignant cardiac tumors are usually rare and fatal.^[3] Angiosarcomas, as in our case, are highly aggressive, locally invasive and may likely to cause obstructive manifestations.^[3] The mean survival is usually less than a year.^[4] The origin of this tumor is usually the right atrium and typically presents insidiously with non-specific signs and symptoms.^[5] Pericardial effusion and possibly tamponade are often present on admission.^[6] Complete removal of the tumor and cardiac reconstruction may be the only solution to offer those patients symptomatic relief and improved quality of life.^[7] Management should mainly encompass chemotherapy and radiotherapy,

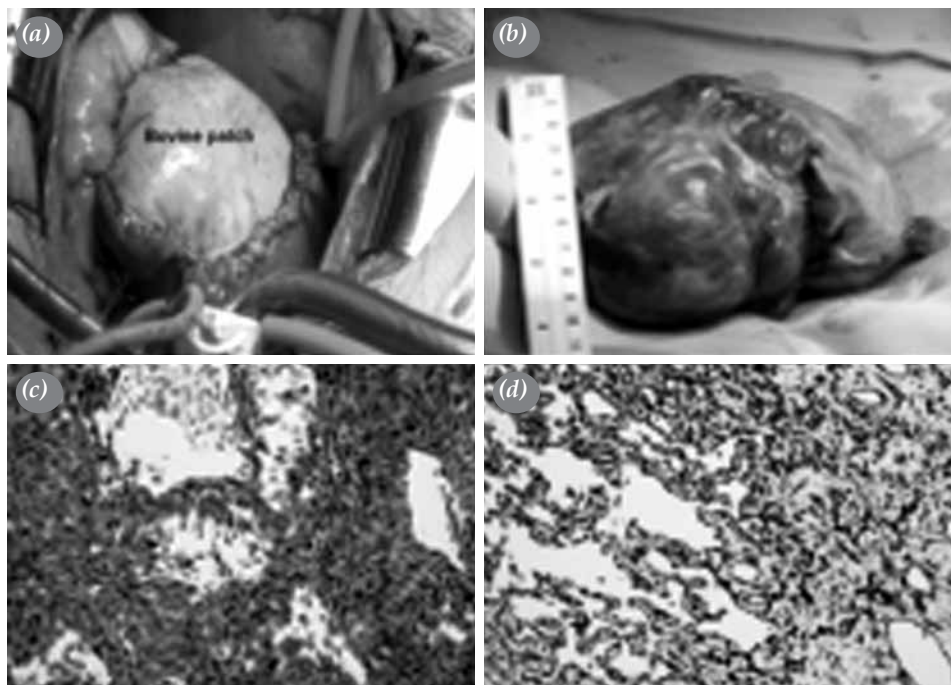


Figure 2. (a) Intraoperative view after excision of the tumor and repair of the right atrial defect with bovine pericardial patch. (b) Macroscopic photograph; resected giant tumor of primary right atrial angiosarcoma, tumor size: 11x12x14 cm. (c) Microscopic pathology photographs of the tumor with histologic photomicrograph (H-E x 400); the tumor consists of spindle-shaped cells with pleomorphic dark nuclei with multiple anastomosing vascular spaces. Areas of hemorrhage and necrosis can also be found. (d) Immunohistochemistry photomicrograph, which confirmed the diagnosis of angiosarcoma.

as metastatic involvement is frequent at the time of presentation.^[8] Complete or palliative resection with adjuvant therapy improves the quality of life and may increase the length of survival for less than a year.^[9] If the surgical margins are involved with the tumor or disease progression, these patients may be considered for heart and lung transplantation, although there are still concerns on the tumor growth following treatment

with immunosuppressive agents.^[10] The following table compares some cases of angiosarcomas with our case (Table 1):

Review of the literature reveal that primary cardiac angiosarcomas have a tendency to occur in the middle age and are more common in males than females.^[10] In a study of 24 patients with angiosarcomas, there were 14 men and 10 women (M/F: 1.4:1) with a mean

Table 1. Comparison between previously published cardiac angiosarcoma cases and our case

Author	Journal and year of publication	Age/Sex	Presenting symptom	Size of the tumor after its resection (cm)	Metastasis before surgery	Survival in months
Chen SW et al. ^[6]	Tex Heart Inst J 2012	33/M	SOB and tamponade	6x5.8x3.5	No	14
Park WK et al. ^[7] (Case 1)	Korean J Thorac Cardiovasc Surg 2012	31/M	SOB and tamponade	5x6	No	5
Park WK et al. ^[7] (Case 2)	Korean J Thorac Cardiovasc Surg 2012	37/M	SOB, syncope and tamponade	4x5	No	-
Bouma et al. ^[8]	Journal of Cardiothoracic Surgery 2011	50/F	SOB, shoulder pain and pericardial effusion	5.1x4.4	No	-
<i>Our case</i>	-	28/M	SOB, DVT, Pericardial effusion, severe obstructing symptoms	11x12x14	No	7

SOB: Shortness of breath; DVT: Deep vein thrombosis.

age of 42.2 years (range 20 to 68 years).^[5] Most of the angiosarcomas were located in the right atrium, particularly the lateral wall of the right atrium sparing the septum in most of the cases.

On the other hand, the most unique findings of our case are as follows: First, it is the largest cardiac angiosarcoma reported to date, which was completely resected surgically. Second, our case was diagnosed on the first day of admission, which highlights the importance of using new imaging modalities, including cardiac magnetic resonance imaging. Third, surgical treatment was unavoidable, since the patient was very symptomatic and the obstructive manifestations of the tumor were advanced with an evidence of superior and inferior vena cava obstruction.

In conclusion, although the prognosis of patients with cardiac angiosarcomas is very poor, multidisciplinary approach, including surgery, can be helpful to improve the quality of life and survival of these patients. Optimal imaging studies are essential before the decision for surgical resection is made. Of note, right atrial reconstruction with a bovine pericardial patch is possible, even following a wide resection of the atrial wall.

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