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A Rare Case of Undifferentiated Pleomorphic Sarcoma with Central Necrosis in the Right Atrium

Sağ Atriyumda Nadir Görülen Santral Nekrozlu İndeferansiye Pleomorfik Sarkom

ABSTRACT

Primary cardiac tumors, which are uncommon types of tumors, can be presented with a variety of clinical signs and symptoms, depending on their location. We present a case of a 57-year-old female patient with a severe right-sided heart failure. Examination using 2-dimensional transthoracic and 3-dimensional transoesophageal echocardiography detected a large, oval, tumor-like formation within the right atrium, which compromised the blood flow from the superior and inferior vena cava. It appeared to have an irregular echo-free space in its central part, probably due to necrosis. Thoracic multi-slice computed tomography revealed a heterogeneous, expansive, tumor-like mass in the right atrium, with signs of bleeding in its center. Although there were no signs of metastatic dissemination, it could not be excluded that the tumor-like mass originated outside of the heart. The patient underwent surgical resection of the tumor. The surgery was accompanied with bleeding complications that developed due to the central necrosis with local infiltration. During the postoperative period, severe systemic inflammatory response syndrome developed and the patient died. Pathologists diagnosed undifferentiated pleomorphic cardiac sarcoma for which the prognosis is usually poor. The median survival of patients with this type of diagnosis is less than 1 year, even with surgical resection and further adjuvant therapy.

Key words: Undifferentiated pleomorphic sarcoma, right atrium, right heart failure

ÖZET

Nadir tümör tipleri olan primer kardiyak tümörler, bulundukları yere bağlı olarak çeşitli klinik belirti ve semptomlarla karşımıza çıkabilir. Bu yazıda, ciddi sağ kalp yetmezliği olan 57 yaşındaki bir kadın hastayı sunmaktayız. 2D-transtorasik ve 3D-transözofageal ekokardiyografi kullanılarak yapılan incelemede, sağ atriyumda superior ve inferior vena kavadan gelen kan akışını olumsuz etkileyen büyük, oval, tümör benzeri bir oluşum saptandı. Muhtemelen nekroza bağlı olarak oluşumun orta kısmında düzensiz bir boşluğa sahip olduğu görülmüştür. Torasik çok kesitli bilgisayarlı tomografide sağ atriyumda heterojen, büyük, tümör-benzeri, merkezinde kanama bulguları olan bir kitle saptandı. Metastatik yayılma belirtisi olmamasına rağmen, tümör benzeri kitlenin kalp dışından kaynaklandığı dışlanamadı. Tümöre cerrahi rezeksiyon uygulandı. Ameliyatta lokal infiltrasyonlu santral nekroza bağlı gelişen kanama komplikasyonları gelişti. Postoperatif dönemde ciddi sistemik inflamatuar yanıt sendromu gelişti ve hasta kaybedildi. Patolojik olarak, prognozu genellikle kötü olan indiferansiye pleomorfik kardiyak sarkom teşhisi koydu. Bu tip tanılara sahip hastaların medyan sağkalımı, cerrahi rezeksiyon ve ileri adjuvan tedavi ile bile 1 yıldan azdır.

Anahtar Kelimeler: İndiferansiye pleomorfik sarkom, sağ atriyum, sağ kalp yetmezliği

Primary malignant cardiac tumors are infrequent, with an incidence of 0.02%.[1] Only 25% of them are malignant sarcomas and are differentiated according to their histological features.[2] These tumors could cause either left- or right-sided heart failure, embolic episodes, conduction abnormalities, and a variety of clinical signs and symptoms,[3] depending on their exact location in the cardiac chambers. Cardiac sarcomas are high-grade, clinically aggressive neoplasms associated with poor clinical prognosis.[1-3] In this case report, we present a case of a cardiac undifferentiated pleomorphic sarcoma in the right atrium.i

Case Report

57-year-old female patient was admitted to the Military Medical Academy of the University of Defense, Belgrade, Republic of Serbia, due to severe signs and symptoms

CASE REPORT
OLGU SUNUMU

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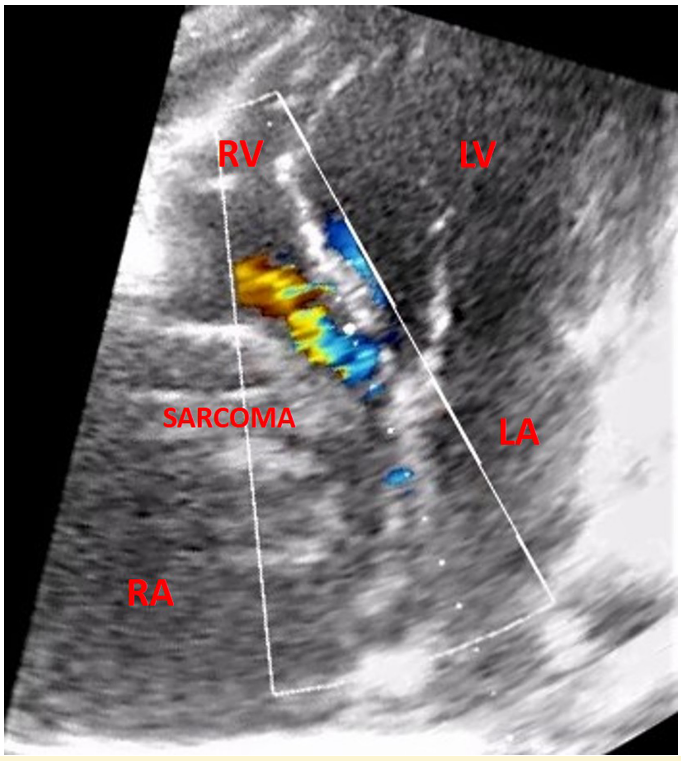

Figure 1. Apical four-chamber view in transesophageal echocardiography. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

of right-sided heart failure. Three months before this event, she experienced progressive fatigue, shortness of breath, and bilateral lower extremity edema. She was hospitalized at another local hospital because of pericardial effusion, which regressed after pharmacological therapy was administered. At this point, any malignancy or systemic tissue disease had been excluded based on the results of the brain, abdominal, and chest thoracic multi-slice computed tomography (MSCT) scanning. At the time of admission to our institution, the patient had severe weakness, dyspnea, jugular venous distension, and peripheral edema, while electrocardiogram findings were considered normal. In addition, 2-dimensional (2D) transthoracic echocardiography was performed and it revealed a large (6.7 × 4.9 cm in size), oval, tumorous, soft tissue formation in the right atrium, which compromised the blood flow from the vena cava (VC) inferior and across the tricuspid valve, with a peak gradient of 35 mmHg. Left and right ventricles had preserved standard dimension and function, without any signs of pericardial effusion (Figure 1). Furthermore, 3-dimensional (3D) transesophageal echocardiography (TEE) examination confirmed a large, tumorous formation (8.2 × 7.9 cm in size), which originated from the right atrial lateral wall and almost occupied it, compromising the blood flow. Discrepancies refaridng the dimension of the mass between the 2D and 3D echocardiography techniques probably originate from a different position of a probe and modality of echocardiography. Within the tumor (Figure 2), an irregular, echo-free space was detected, possibly arising due to necrosis.

Thoracic MSCT detected a heterogeneous, tumorous, expansive mass (19 × 8 × 8 cm in size) in projection of the right atrium,

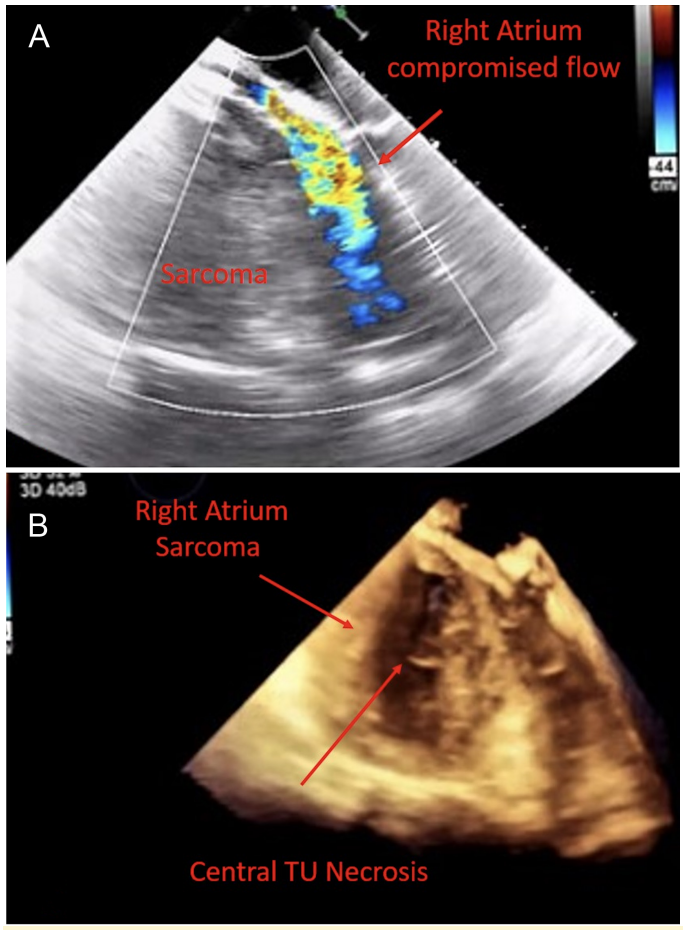

Figure 2. (A) Two-dimensional transesophageal echocardiography (TEE) of the right atrium shows sarcoma compromising normal flow and (B) three-dimensional TEE of the right atrium shows central tumor (TU) necrosis.

with signs of active bleeding in its central part. Signs of tumor dissemination were detected using the MSCT. Indeed, cardiac magnetic resonance imaging indicated a mass (12 × 9 × 10 cm in size) adjacent to the lateral right atrial wall and basal lateral right ventricular wall. The patient underwent almost complete of the tumor surgical resection, with bleeding complications due to its local invasiveness and central necrosis (Figure 3). Surgical

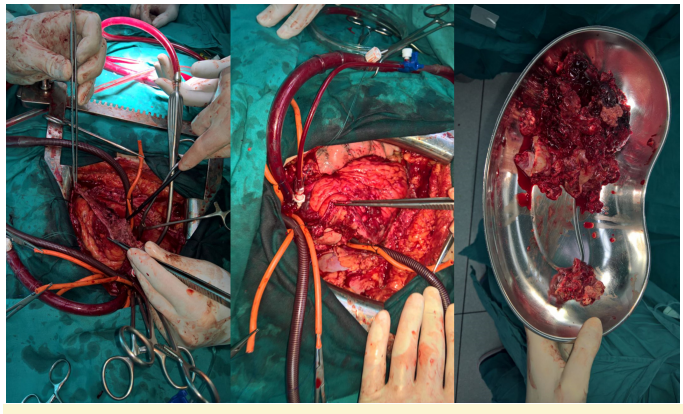

Figure 3. Surgical material after resection.

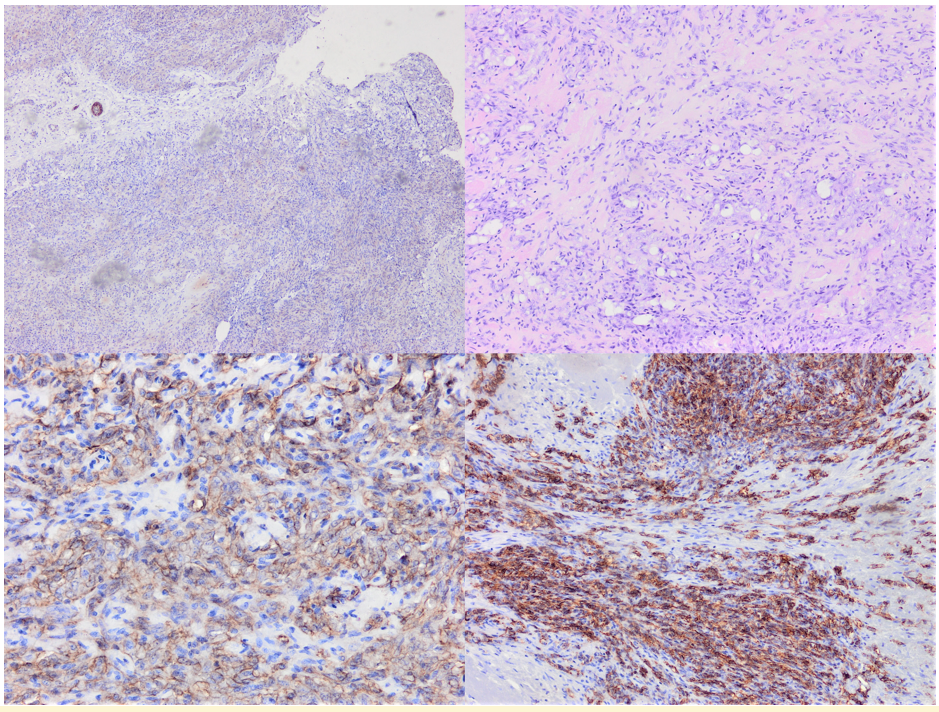

Figure 4. Histopathological findings of undifferentiated pleomorphic sarcoma.

findings confirmed the oval-shaped tumor in the right atrium (8.0 × 7.5 cm in size). Pathohistological analysis revealed spindle-shaped and oval tumor cells, diffusely arranged in the tumor tissue. Mitosis and pathological mitosis of the nucleus showed that the tumor cells had a low degree of differentiation and a high grade of malignancy. Undifferentiated pleomorphic sarcoma was established as the definitive pathological diagnosis (Figure 4).

Introduction of additional, standard chemotherapy was considered because of its demonstrated beneficial effects. Unfortunately, the patient developed severe systemic inflammatory response syndrome during the postoperative period, and died due to multiple organ dysfunction.

Discussion

Echocardiography and 3D TEE are imaging techniques that are valuable for the diagnosis of intracardiac masses (myxoma, thrombus, and infected thrombus). However, they poorly differentiate between different types of cardiac masses.[4] Definitive diagnosis can only be made based on the pathohistological analyses and findings. Features of masses detected by using non-invasive imaging techniques that are suggestive of malignancy include ill-defined margins of the mass, dimensions larger than 5 cm, tissue heterogeneity, broad attachment site, and right heart-sided tumors.[5] Sarcomas account for only 10-20% of all primary cardiac tumours.[6] These malignancies develop from the pluripotent mesenchymal cells of the ventricles, atria, or pericardium depending on their specific subtype. Undifferentiated pleomorphic sarcomas account for more than one-third of all malignant cardiac tumors,[7] but only 5% of them are soft tissue tumors.[8,9] In this case report, we presented a rare case of primary, cardiac undifferentiated, pleomorphic sarcoma, which had originated from the lateral wall of the right atrium. Undifferentiated pleomorphic sarcoma of the right side of the heart is described in only a few cases previously and predominantly originates from the left side of the heart.[10-15] We suppose that its first manifestation was a pericardial effusion, for which the patient was hospitalized for the first time, but it was unrecognized at that moment. Three months later, the patient experienced rapid progression of symptoms and a massive tumor with central necrosis in the right atrium was detected upon the patient's second hospitalization. The tumor compromised the blood flow in VC inferior and VC superior and across the tricuspid valve. It resulted in prominent signs and symptoms of right heart failure. Positron emission tomography/computed tomography was not carried out due to the fact that the patient's clinical state represented an emergency. Metastatic dissemination was not detected, in contrast to the previous reports.[10,15] According to one small retrospective study, nearly 50% of patients with cardiac sarcoma had some evidence of metastatic disease at the time of disease presentation.[13] Primary cardiac sarcomas are associated with an increased risk of metastases, especially brain metastases, which are associated with higher mortality,[14] but there are also reports of tumor dissemination to other locations in the body.[15] Most studies with a multidisciplinary approach and addition of adjuvant chemotherapy did not conclude that the chemotherapy changed the course of the disease, but there are several reports of the improved survival rate following the chemotherapy use.[15] As the disease is very rare, there is a lack of data regarding the chemotherapy success from the randomized trials, which makes an individual and multidisciplinary approach to each patient even more significant.

We presented a case of cardiac sarcoma with extremely rapid growth of the tumor and its local invasion to the surrounding structures, but without distant metastases. It should be

emphasized that cardiac sarcomas have a rapidly progressive course, with non-specific symptoms, which remain unrecognized until they affect hemodynamics.[11,12,15,16] Yadsar et al[11] in their case report described the onset of symptoms (e.g., dyspnea, cough) 7 days before admission to a hospital. Aksu et al[12] mentioned the appearance of non-specific symptoms 2 months before being admitted to a hospital. Estrada et al[10] presented a patient who underwent mitral valve annuloplasty 9 months before another admission to a hospital, at which point a tumor was detected. Winchester et al[17] examined the reports of 319 patients and showed that recurrences and metastases occurred in 14.1% and 7.8% of the cases, respectively.

An overall conclusion is that the best therapy for patients with this type of tumor is complete surgical resection of the tumor. Unfortunately, complete surgical resection is often not possible, due to its extensive invasion within the surrounding cardiac structures. Previous reports indicate that the majority of the patients with this diagnosis have a median survival rate of less than 1 year.[6,9] Therefore, an individual and multidisciplinary approach should be applied for these patients and rapidly growing nature should be kept in mind.

Ethics Committee Approval: Ethics Committee of "General hospital "Prim.dr. Abdulah Nakas" (Approval No: 555-70/21).

Informed Consent: The authors certify that they have obtained all appropriate patient consent forms. In them, the patient's family has given consent for images and other clinical information to be reported in the journal. The patient's family understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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