

A case of cavernous hemangioma in the right ventricular outflow tract

Sağ ventriküler çıkım yolu yerleşimli bir kavernöz hemanjiyom olgusu

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Primary cardiac tumors are very rare and myxomas constitute most of them. Hemangiomas are even rarer. As most of cardiac hemangiomas are asymptomatic, they are generally incidentally detected in postmortem examination or imaging studies. Echocardiography is the preferred imaging modality for the clinical diagnosis in symptomatic patients, while the definitive diagnosis is based on histopathological examination. Treatment is surgical excision of the tumor. As there is a risk of recurrence for incompletely resected tumors, particularly, postoperative follow-up is necessary. In this article, we report a rare cardiac cavernous hemangioma case with remarkable features including symptomatic profile of the patient, localization of the tumor in the right ventricular outflow tract and its macroscopic appearance mimicking myxoma. The patient is followed uneventfully.

Key words: Cardiac; cavernous; hemangioma; ventricular.

Cardiac hemangioma is a rare tumor which comprises approximately 2% of all primary cardiac tumors.^[1] It is made up of capillary or cavernous vascular channels and since it is usually asymptomatic, it is generally coincidentally detected via autopsy, echocardiography, computed tomography (CT), or magnetic resonance imaging (MRI). However, due to its localization and dimensions, symptoms such as arrhythmia, tachycardia, atypical chest pain, and dyspnea are possible, and in isolated cases, it has even been known to cause pericardial effusion, congestive heart failure, embolic events, and even sudden death. Echocardiography is the preferred method for diagnosing cardiac hemangioma,

Primer kardiyak tümörler oldukça nadir olup, bunların birçoğunu miksomalar oluşturur. Hemanjiyomlar ise daha nadirdir. Kardiyak hemanjiyomların çoğu asemptomatik olduğundan, genellikle otopside ya da görüntüleme incelemeleri sırasında tesadüfen saptanırlar. Semptomatik olgularda ekokardiyografi klinik tanı için tercih edilen bir yöntem olmakla birlikte, kesin tanı histopatolojik inceleme ile konulur. Tedavi, tümörün cerrahi olarak eksizyonudur. Özellikle tam olarak çıkarılamayan tümörlerde nüks riski bulunduğu için, ameliyat sonrası takip gereklidir. Bu yazıda, hastanın semptomatik oluşu, tümörün sağ ventriküler çıkım yolu yerleşimi ve miksoma ile karışan makroskopik görünümü gibi dikkat çekici özellikleri olan nadir bir kardiyak kavernöz hemanjiyom olgusu sunuldu. Hasta sorunsuz takip edilmektedir.

Anahtar sözcükler: Kardiyak; kavernöz; hemanjiyom; ventriküler.

with CT and MRI being used as complementary imaging techniques if necessary. Surgical removal of the tumor is the recommended treatment, and patients need to be followed up for recurrence.

Herein, we present a case of cavernous hemangioma localized in the right ventricular (RV) outflow tract in a female patient with complaints of left chest pain and dyspnea. It was detected by both echocardiography and angiography and was then treated surgically. The patient was discharged uneventfully and is currently being followed-up to ensure that there is no recurrence.



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CASE REPORT

A 48-year-old female patient visited a cardiology clinic with complaints of left chest pain and dyspnea was diagnosed as having a cardiac mass via echocardiography and angiography. She was then admitted to our cardiovascular surgery clinic for surgical treatment. The patient's physical examination and chest X-ray did not reveal any noteworthy findings, and she had normal sinus rhythm on electrocardiography. Her blood tests were within normal ranges, but hyperlipidemia was present. Under standard cardiopulmonary bypass (CPB), the tricuspid valve was explored through a right atriotomy, and a 2x2 cm tumor giving the impression of myxoma was excised from the RV outflow tract (Figure 1). In the macroscopic examination, the material was revealed to be a polypoid tumor measuring 2 cm in diameter with a smooth surface, and the cut surface contained patchy bleeding areas. A microscopic examination demonstrated that the tumor was composed of vast vascular channels lined with a single-layered row of endothelial cells that were anastomosing with each other. A few of these contained thrombus, and the stroma showed fibrosis in patches, leading to the diagnosis of cavernous hemangioma (Figure 2). The patient, who did not have any postoperative problems, was then discharged with a schedule for periodic follow-ups. No findings related to a relapse have been observed thus far in 36-months follow-up period.

DISCUSSION

The reported rate of cardiac tumors is quite variable when autopsy cases and surgically removed cases are taken into account, with the estimated frequency ranging between 0.0017 and 0.33%.^[2] Most of these cases are secondary tumors that formed as a result of metastasis or direct invasion of another tumor. Primary tumors are rarer, with 50% of these being myxomas. Hemangiomas constitute only 2% of all primary cardiac tumors.^[1]

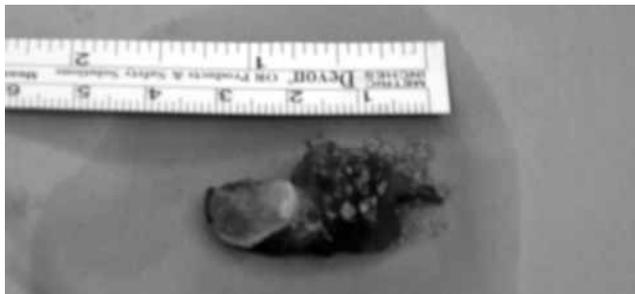


Figure 1. The macroscopic appearance of the tumor which simulated atrial myxoma.

Cardiac hemangiomas can be observed in either gender and at any age. Symptoms depend on the anatomic localization of the tumor and the cardiac hemodynamics of the patient. Symptom severity is relevant to the pressure that the tumor exerts on cardiac structures, outflow tract obstruction, pericardial effusion, and degree of congestive heart failure. This type of tumor may cause mild symptoms such as chest pain or difficulty in breathing; however, newly formed atrial fibrillation, cerebral stroke, and even sudden death have been observed, although infrequently.

The tumor is generally localized in the left ventricular (LV) lateral wall, RV anterior wall, interventricular septum, and, to a lesser degree, in the RV outflow tract.^[2,3] Development can occur in the intracavitary, intramural, or extramural regions, and it can grow to reach a diameter of 13 cm, remain stable, or even demonstrate a certain amount of involution.^[4]

Echocardiography supported by CT or MRI is a reliable method for diagnosing cardiac hemangioma, with hyperechoic, intracavitary solid masses having been previously observed. Although these tumors may render medium and high T₁-weighted images, they generally produce very dense T₂-weighted images on MRI, whereas on CT, the image of the mass is heterogenous and well-demarcated with low attenuation. Coronary angiography is an additional diagnostic test that can be used to eliminate obstructive coronary artery disease (CAD), determine tumor involvement in the coronary arteries, or reveal the conditions of feeder arteries. Although establishing whether or not a tumor is benign is possible with an

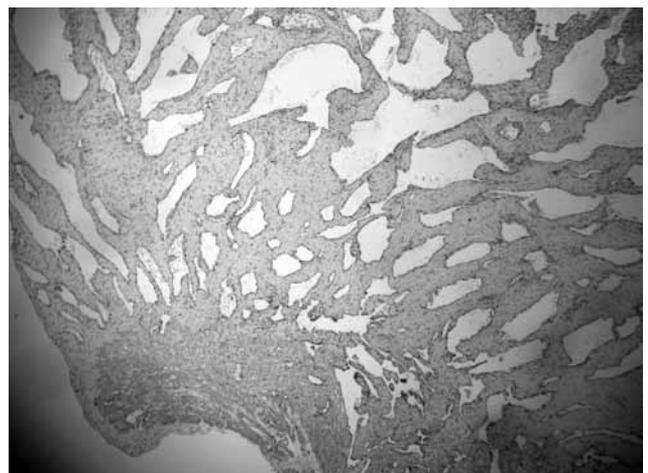


Figure 2. The tumor is composed of vast vascular channels lined by single-layered endothelial cells anastomosing with each other (H-E x 200).

MRI due to its well-demarcated and non-infiltrative characteristics, a specific diagnosis can only be made histopathologically.^[2]

Tumors have three histological types. Cavernous tumors are composed of numerous dilated, thin-walled arteries while capillary tumors are comprised of smaller arteries that resemble capillaries. In addition, arteriovenous tumors are made up of dysplastic malformed arteries and veins.^[2] Cardiac hemangiomas may contain characteristics of all three types together, and most bear fibrous and fat tissues.

In differential diagnosis, benign lesions, such as a thrombus in the heart, myxoma, lipoma, fibroelastoma, and cysts should be taken into consideration. In addition, special attention should also be paid to malignant tumors like angiosarcoma.^[1,2,5]

A complete resection of the tumor is the preferred method of treatment. Even though intracavitary and intramural hemangiomas are not excised completely, which leads to a higher risk of recurrence and ventricular arrhythmia than with cardiac hemangioma,^[6] the postoperative prognosis is still quite good for many of these patients.

In terms of its potential risks, cardiac hemangioma is a tumor that necessitates surgical removal, and patients with this type of tumor should be followed up in the postoperative period because of the risk of recurrence.

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