

A rare case of cardiac tumor: Malignant fibrous histiocytoma

Nadir bir kardiyak tümör olgusu: Malign fibröz histiyositoma

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Abstract

Cardiac tumors are generally benign; however, they can rarely be malignant. Malignant fibrous histiocytoma is a type of sarcoma. Cardiac malignant fibrous histiocytomas are clinically and histologically confused with atrial myxomas. In this case report, we present a patient who was administered to the hospital with non-specific complaints, determined to have an intracardiac mass and subsequently diagnosed with malignant fibrous histiocytoma, which shortly recurred in the form of intracardiac masses after postoperative chemotherapy.

Keywords: Cardiac tumor, Malignant fibrous histiocytoma, Operative therapy, Chemotherapy, Recurrence

Öz

Kardiyak tümörler genellikle benign karakterdedir, fakat nadir de olsa malign karakterde seyredebilir. Malign fibröz histiyositoma bir sarkom türüdür, klinik ve histolojik açıdan atrial miksoma ile karışır. Biz bu vaka takdiminde, non-spesifik şikayetlerle başvuran bir hastada intrakardiyak kitle tespit edilmesinin sonrasında Malign Fibröz Histiyositoma tanısı alan ve cerrahi sonrası kemoterapi altında kısa sürede intrakardiyak kitleler şeklinde nüks yaşanan olguyu sunuyoruz.

Anahtar kelimeler: Kardiyak tümör, Malign fibröz histiyositoma, Cerrahi tedavi, Kemoterapi, Nüks

Introduction

Cardiac tumors are more easily diagnosed with developing imaging techniques. They are generally benign; however, they can rarely be malignant. Primary cardiac tumors are quite rare with an incidence around 0.0017–0.019%, and secondary heart tumors are significantly more common [1,2]. These cardiac masses can be asymptomatic or cause a wide range of symptoms that may even result in sudden death [3]. In this case report, we present a patient who was administered to the hospital with non-specific complaints, determined to have an intracardiac mass and subsequently diagnosed with malignant fibrous histiocytoma, which shortly recurred in the form of intracardiac masses after postoperative chemotherapy.

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Case presentation

A 61-year-old female patient who did not suffer from any chronic disease presented to the cardiology department of a medical center with the complaints of back pain, chest tightness, and palpitations which started following an upper respiratory tract infection. Echocardiography revealed moderate mitral insufficiency and a hyperechogenic mass (25x22 mm) in the posterior leaflet of the mitral valve, after which the patient was referred to our clinic. The physical examination did not reveal any pathology other than 2/6 mitral regurgitation murmur at the mitral focus. The echocardiography indicated the maximum pulmonary artery systolic pressure was 45 mmHg with moderate mitral and tricuspid regurgitation. We determined a hyperechoic mass (28x15 mm) adhering to the posterior leaflet of the mitral valve. Furthermore, we determined a mean diastolic pulmonary gradient of 17 mmHg which we attributed to the mitral stenosis caused by the mass on the mitral valve. We planned a transesophageal echocardiography (TEE) to more clearly evaluate the mass. The TEE revealed a mass (42x22 mm) in the left atrium adhering to the posterior region that extended towards the mitral valve, suggestive of a myxoma (Figure 1). There was moderate mitral insufficiency, but the myxoma was thought to affect mitral valve functions and cause mitral regurgitation. After the evaluation of the heart team, the patient was decided to undergo tumor resection and mitral valve replacement. The patient was preoperatively evaluated with a PET/CT scan to determine any other primary tumor or metastases. There only was focal pathological 18F-FDG retention in the right atrioventricular region as per the initial findings. The pathological examination of the excised surgery material confirmed the malignant fibrous histiocytoma diagnosis. Medical oncologists decided on chemotherapy after relevant examinations. The patient underwent a follow-up PET/CT scan to evaluate tumoral activity after the fourth round of chemotherapy. This examination revealed increased focal FDG retention similar to the metallic hyperdensity in the left atrium adjacent to the pulmonary trunk, and increased FDG retention in multiple hypodense mass lesions in the liver. The patient was subsequently referred to our clinic for cardiac evaluation. The apical 4-chamber (A4C) view of the echocardiogram showed a solid mass (25x20 mm) in the left ventricle that infiltrated the septum with thin pedicles. Also, there was a solid mass (20x17 mm) extending over the right ventricular tricuspid lateral valve (Figure 2). The image also revealed the functional prosthetic metal valve at the position of the mitral valve. The patient underwent cardiac MRI to confirm the solid lesions. The MRI results affirmed the masses that were detected by echocardiography (Figure 3). The patient was evaluated by the cardiovascular department once more. The mass was evaluated as malignant due to rapid progression. It was decided that the patient should be closely followed up with medical treatment, and reevaluated in case of impaired cardiac function. The patient's consent was obtained for the clinical presentation.

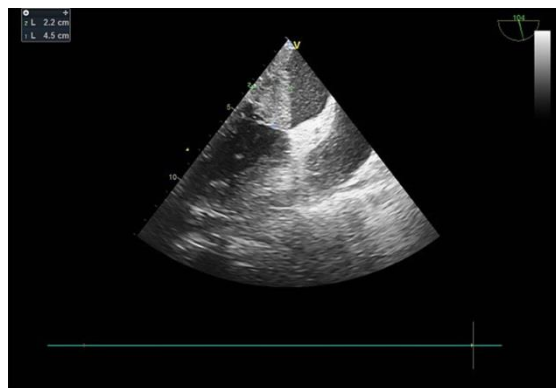


Figure 1: Transesophageal echocardiogram showing mass (42x22 mm) in the left atrium adhering to the posterior region that extended towards the mitral valve, suggestive of a myxoma



Figure 2: The apical 4-chamber view of the echocardiogram showing a solid mass (25x20 mm) in the left ventricle that infiltrated the septum with thin pedicles and a solid mass (20x17 mm) extending over the right ventricular tricuspid lateral valve

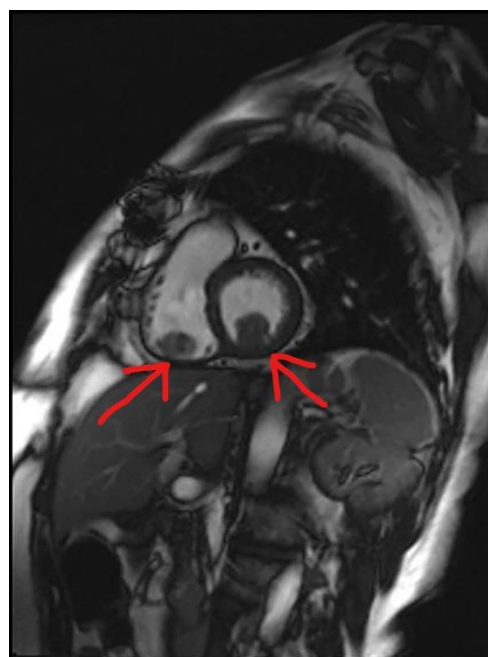


Figure 3: MRI showing masses in the left and right ventricle

Discussion

Malignant fibrous histiocytoma is a type of sarcoma that typically occurs in the extremities and the trunk. There are less than 100 reported cases of primary cardiac malignant fibrous histiocytomas in the literature [4]. The reported patients are aged between 14 and 77 years [5]. Cardiac malignant fibrous histiocytomas are clinically and histologically confused with atrial myxomas [6,7]. The first cardiac malignant fibrous histiocytoma case was reported in 1978 [8]. The majority of the

recently reported cases were associated with rapid prognosis in the form of recurrence, local infiltration, and distant metastasis, and eventual death. Complete surgical excision is the only treatment that has been shown to prolong survival [9]. However, even with complete resection, the recurrence is common in most patients with different sources reporting a median survival of 6 to 12 months [10]. Adriamycin and doxorubicin have been shown to improve survival outcomes compared to other drugs [11]. Radiation significantly reduces the incidence of local recurrence [12,13]. One study analyzed 42 cardiac sarcoma patients who received treatment between 1988 and 2013, and found that a multimodal therapy (any combination of surgery, radiotherapy and chemotherapy) was associated with better survival than surgery alone [14]. Patients with a cardiac mass should be examined in detail to determine surgical requirements. Patients with postoperative recurrence should be evaluated in terms of cardiac functions instead of mass involvement. In addition, the physician should consider medical follow-up together with surgical intervention.

Conclusion

In this case study, we report a rare case of malignant fibrous histiocytoma. We conclude that these cases must be carefully monitored for recurrence and metastasis. A multidisciplinary approach including cardiovascular and oncology specialists is crucial for determining the optimal treatment.

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