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Case Report

An atypical presentation of an atypically localized cardiac myxoma

Atipik klinik ile tespit edilen atipik yerleşimli kardiyak miksoma

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ABSTRACT

Atrial myxomas are the most common benign primary cardiac tumors that can lead to many complications as defined in literature. Although the majority occur in the left atrium and attached to interatrial septum, they can arise from any cardiac chamber. Here we report the case of a 55-year-old woman whom was referred to our outpatient clinic for etiological diagnosis of unilateral transient loss of vision. Transesophageal echocardiography revealed a mass that was suspected as cardiac myxoma arising from the posterior wall of the LA in the vicinity of the left superior pulmonary vein. During the surgical procedure cardiac mass was removed totally and the pathological examination confirmed the diagnosis as cardiac myxoma. In patients with transient ischaemic symptoms but without atrial fibrillation echocardiography should be performed to diagnose of potential mass in left atrium.

Keywords: atrial myxoma, cardiac tumor, echocardiography

ÖZ

Atriyal miksomalar, en sık karşılaşılan benign primer kardiyak tümörlerdir. Literatürde tanımlandığı üzere birçok komplikasyona yol açmaktadır. Çoğunlukla solda atriyumda olsa da atriyum ve interatriyal septuma bağlı olarak, herhangi bir kardiyak boşlukta ortaya çıkabilir. Bu olgu raporunda 55 yaşındaki bir kadın hastamızı sunduk. Kardiyoloji polikliniğine tek taraflı geçici görme kaybı etyolojisinin araştırılması amacıyla danışıldı. Transözofageal-ekokardiyografide kardiyak miksoma olarak şüphe edilen bu kitlelenin cerrahi esnasında sol atriyum posteriyor duvar, sol üst pulmoner ven çevresinden kaynaklandığı tespit edildi ve kitle tamamen çıkartıldı. Patolojik inceleme sonucunda tanı kardiyak miksoma olarak doğrulandı. Geçici iskemik semptomları olan ancak atriyal fibrilasyonu olmayan hastalarda sol atriyumda potansiyel kitle tanısını koymak için ekokardiyografi yapılmalıdır.

Anahtar Kelimeler: atriyal miksoma, kardiyak tümör, ekokardiografi

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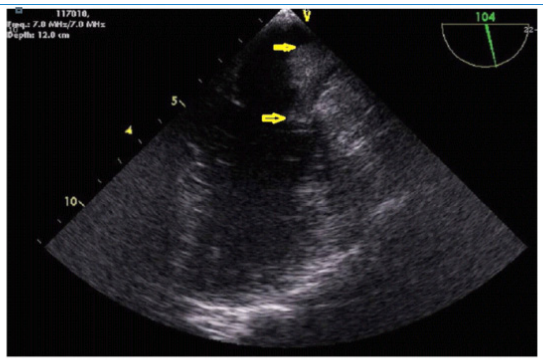
Introduction

Cardiac myxomas are the most frequent benign intra cardiac tumors that can lead to many embolic complications as described in literature [1]. Atrial myxomas are related to systemic embolisation in around 30 to 40 % of cases [2]. Temporary loss of vision caused by intracardiac myxomas embolisation have been observed rare in the literature [3].

Case report

Here we report the case of a 55-year old woman who was referred for etiological diagnosis of unilateral transient loss of vision by an ophthalmologist. She had a history of diabetes mellitus. Physical examination revealed blood pressure 100 / 60 mmHg. Heart rhythm was regular at 66 bpm. The patient had no prior history of heart murmur, syncope, shortness of breath, or chest pain. Further physical examination revealed a soft grade 2/6 systolic murmur at the left sternal border.

Atrial fibrillation was not observed in a 24-hour rhythm holter recording. After a mass was seen in transthoracic echocardiography (TTE), transesophageal echocardiography (TEE) was performed and revealed a 3.8x2.8 cm sized, unclear site of attachment (Figures1a,1b and1c).



Figüre 1a: Traneosephagial echocardiographic findings. A mass with irregular margin in the left atrium and unclear site of attachment (in midesophageal long-axis view showing) Laa left atrial apendicis ,LV left ventricle

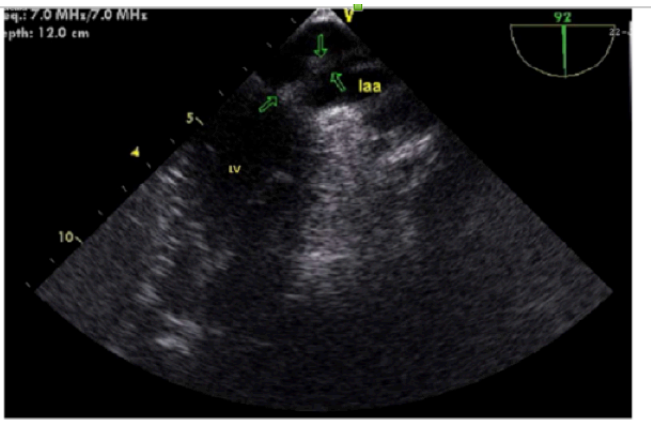


Figure 1b: midesophageal long-axis view (LA left atrium, LV left ventricle,)

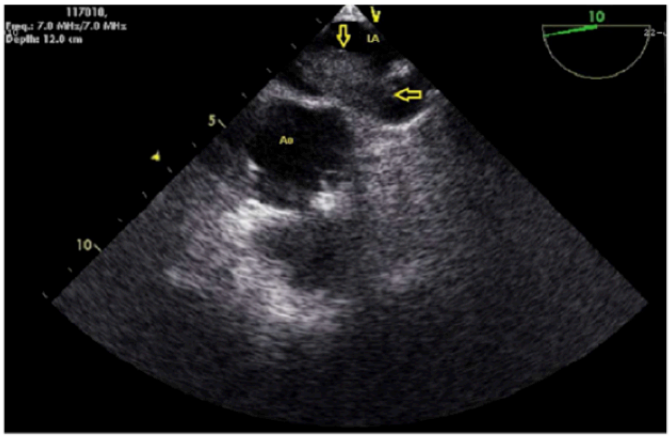


Figure 1c midesophageal short-axis view (LA left atrium, Ao aortic annulus)

It was highly mobile and occasionally extending to the mitral valve area during the diastole. TEE exhibited a large mass with wide base, located at posterior wall of the high left atrium (LA) in the vicinity of left upper pulmonary vein. At first vision was suspected this mass as thrombus causing central retinal artery emboli. So she was treated with the unfractionated heparin and we planned to recheck the size of mass in the left atrium after 7 days. TTE was performed again and we observed no change in the size of the mass.

The patient was referred to surgery. After median sternotomy and dissection of the right atrium, left atrium was explored through transeptal approach, myxomatous large mass with a gelatinous appearance was totally resected during the surgery. The mass was multilobed and a friable with a large pedicle It was attached to the posterior wall of the high left atrium in the vicinity of the left superior pulmonary vein with base of implantation of 5,0 x 3.0 cm diameter.

Macroscopically the lesion was soft, polypoid and lobulated. Microscopic examination revealed a papillary tumor in a myxoid stroma. Tumor was composed of polygonal and stellate cells. There was no mitotic activity, pleomorphism and necrosis in the tissue. (Figures 2a and 2b).

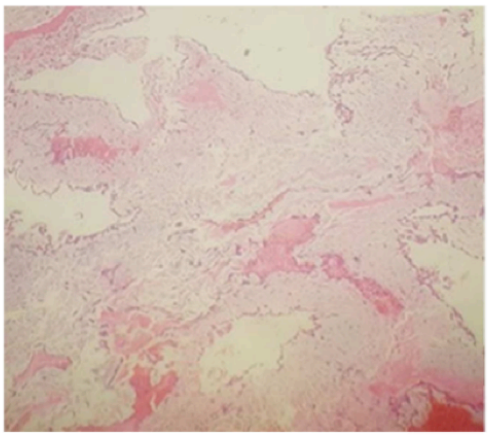


Figure 2a : Cardiac tumor. Microscopic image shows tumors cells aligning on surface surrounded with a myxoid stroma (4X magnification, H&E stained section)

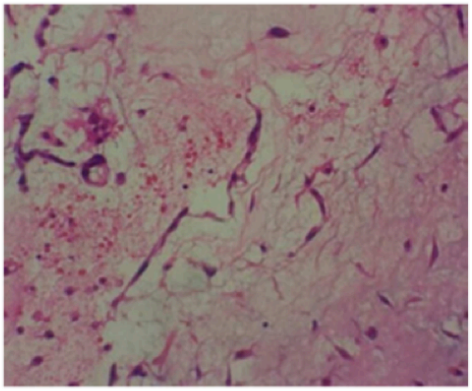


Figure 2b: Medium magnification of tumor. Necrosis and mitotic activity is absent. (40X Magnification, H&E stained section).

Due to the complete atrioventricular (AV) block after surgical procedure, a temporary pacemaker was implanted during the first postoperative day. TTE performed after surgery revealed a mild mitral regurgitation. As the sinus rhythm restored, the pacemaker was removed in the postoperative second day.

Discussion

Atrial myxomas represent approximately 50% of all cardiac tumors and nonfamilial forms occur mainly in the 5th and 6th decade of life [2]. They originate mainly from mesenchymal cells left atrium subendocardial. Although myxomas usually originate in the LA (75%) attached to fossa ovalis, these tumors may also arise from atypical sites such as right atrium (15–20%), posterior or anterior LA walls, atrial appendage, left or right ventricle, and on the valves [4,5]. Myxomas must be at the top of the differential diagnosis list in all intracardiac tumors. Although they are histologically benign, may lead to syncope, systemic embolism or sudden death [6,7].

TTE and TEE have an important role of diagnosis. However, a definitive transthoracic echocardiographic diagnosis of a left atrial myxoma is difficult when the site of origin cannot be clearly identified. According to the literature posterior wall origin of myxomas was identified in 28 cases (including our case report) researches, due to lower incidence of symptoms, posterior wall localized atrial myxomas are diagnosed later than the others [1]. Embolization to the central nervous system may result in transient loss of vision and ischemic attack, stroke, or seizure. Likewise, embolisation to retinal artery may lead to severe irreversible visual impairment, nevertheless it may cause temporary loss of vision as in our case. This is particularly noticeable in patients with diabetes mellitus, high blood pressure and elderly [8]. Any embolic event may be the only symptoms that should lead an early diagnosis [9].

As a conclusion although myxomas originating from the posterior wall of LA are extremely rare, it should be kept in mind for the differential diagnosis of a mass in LA especially in patients with transient ischaemic symptoms but without atrial fibrillation.

Declaration of conflicting interests

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