Case Report of a Left Atrial Myxoma Associated with Carney's Syndrome

Olgu Sunumu; Carney Sendromu İlişkili Sol Atriyal Miksoma

Carney's Syndrome and Left Atrial Myxoma

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Özet

Primer kardiyak neoplaziler, metastatik tümörlere oranla oldukça nadir görülürler. Primer kardiyak tümörlerin %70-%80'i benign miksomalardır. Miksomalar; kist ve mikroabse oluşumu, embolizasyon, senkop ve ani ölüm gibi komplikasyonlara yol açabilirler. Kardiyak miksomalar nadiren, primer nodüler adrenal kortikal hastalık, meme fibroadenomu, testis tümörleri, jigantizm ya da akromegali ile seyreden pituiter adenomaları ile birliktelik gösterebilirler. Bu duruma Carney sendromu adı verilir. Biz burada, Carney sendromu ile birliktelik gösteren ve sol atriyal miksoma tanısı ile ikinci kez ameliyat edilen bir vaka takdim etmekteyiz.

Anahtar Kelimeler

Carney Kompleksi; Miksoma; Pigmentasyon Bozuklukları

Abstract

Primary cardiac neoplasms are very rare as compared to metastatic tumors. 70% to 80% of them are benign myxomas. Complications of myxomas include cyst and microabscess formation, embolization, syncope and sudden death. Rarely, cardiac myxomas are associated with primary nodular adrenal cortical disease, mammary fibroadenomas, testicular tumors or pitiutary adenomas with gigantism or acromegaly known as Carney's syndrome. We present a patient with a left atrial myxoma who underwent reoperation associated with Carney's syndrome.

Keywords

Carney Complex; Myxoma; Pigmentation Disorders

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Introduction

Myxomas are the most frequent benign intracardiac tumors located mostly at the left atrium (75%) [1]. Myxomas mostly originate from the fossa ovalis. Other sites of origin are; the mitral annulus, mitral valve, aortic valve or the inferior vena cava [2]. It is known that myxomas can also cause embolisation, infection, syncope and suddent death [3]. Recurrence rates reported for cardiac myxomas are 4% to 7% for sporadic cases and 10% to 21% for familial cases [4]. In this case report, we present a patient who was operated for recurrent familial myxoma (Carney Syndrome).

Case Report

53 year old male patient was admitted with complaints of dyspnea and palpitations. He had a past medical history of hyperlipidemia, smoking and operation due to left atrial myxoma 15 years ago. His sister had been operated for myxoma aswell. There was a hyperpigmentation and nevus on the face and on the neck. On physical examination, there was a pansystolic murmur heard at the mesocardiac focus. The electrocardiography (ECG) was normal. The echocadiography revealed a left regular margined atrial mass (myxoma?) of 3.5x 2 cm diameter originating from the interatrial septum. A diagnosis of recurrent familial myxoma was made. Blood sample results for T3, T4, TSH, cortisol levels were within the normal ranges. Coronary angiography was performed to evaluate the patency of coronary artery disease. The patient had a %40 stenosis of the first diagonal branch of the left anterior descending artery. He was then referred to our cardiovascular surgery clinic for excision of the left atrial myxoma. The decision for operation was made for the left atrial myxoma associated with Carney's syndrome. The operation was performed under cardiopulmonary bypass, mild hypothermia using crsytalloid cardioplegia. A left atriotomy was done and the left atrial myxoma was visulalized (Figure 1). It originated from the interatrial septum. The myxoma was totally excised and the material was sent to the pathology department for microscopic investigation. The pathology report revaled a left atrial mass compatible with an atrial myxoma. The patient was discharged at the sixth postoperative day without any complication.

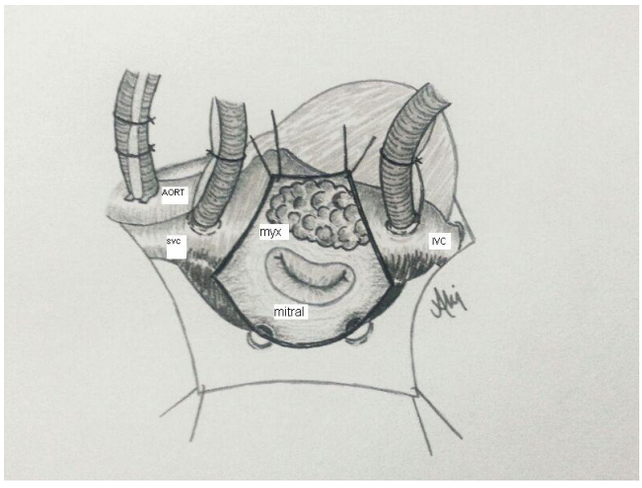

Figure 1. Illustration of intraoperative view of the left atrial myxoma.

Discussion

Myxomas are the most frequent primary heart tumors. The incidence of myxomas following autopsy series was reported as % 0.017 and % 0.028 (1). They are mostly located at the left atrium (%75). Surgical resection is the only way to treat myxomas and patients should be referred to surgery as soon as possible. Myxomas can present at any age group but occur more often between the 3rd and the 6th decades of life [5] as is the case with our patient who was 53 years. Familial forms, which are more frequently diagnosed in younger individuals, constitute 10% of all myxomas and have autosomal dominant transmission [6]. Familial cardiac myxomas are characteristically seen in atypical locations and have a high recurrence rate. First-degree family members should be screened and followed carefully. Recurrence rates reported for cardiac myxomas are 4% to 7% for sporadic cases and 10% to 21% for familial cases [4]. Carney's complex, as described by J.A. Carney, is characterized by the association of cutaneous pigmentation, fibromyxoid tumors of the skin, myxomas of the heart, endocrine overactivity, and autosomal dominant inheritance [5,7].

Our patient presented here had a hyperpigmentation and nevus on the face and on the neck. By the guidance of the physical examination findings and the familial history of the left atrial myxoma, the patient was diagnosed as Carney's sydrome and was treated succesfully by surgery.

Competing interests

The authors declare that they have no competing interests.

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