



A Rare Case of Large Left Atrial Myxoma Associated with Carney Syndrome

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Received 15 March 2013; Accepted 23 September 2013

Abstract

Carney Syndrome (CS) is an autosomal dominant multiple neoplasia syndrome that includes cardiac, endocrine, cutaneous, and neural tumors. Cardiac myxomas can be seen in the course of CS. A 46-year-old female patient was admitted to our clinic with palpitation, cough, and exertional dyspnea. Physical examination revealed pigmented lesions, especially scattered in the neck, chest, and extremities. Echocardiography revealed a left atrial mass, 6 × 4 cm in size, originating from the interatrial septum that was protruding into the left ventricle through the mitral valve during diastole. The tumor was excised from the interatrial septum with its pedicle. The patient was discharged at the fifth postoperative day without any complication. Cardiac myxomas are a cornerstone of this disease since cardiac manifestations and related complications account for the majority of mortality seen in the course of CS.

J Teh Univ Heart Ctr 2014;9(4):191-193

This paper should be cited as: Kocabeyoğlu SS, Özyüksel A, Çetin E. A Rare Case of Large Left Atrial Myxoma Associated with Carney Syndrome. *J Teh Univ Heart Ctr 2014;9(4):191-193.*

Keywords: Myxoma • Carney complex • Multiple endocrine neoplasia

Introduction

Carney Syndrome (CS) is an autosomal dominant multiple neoplasia syndrome that includes cardiac, endocrine, cutaneous, and neural tumors. Although the age at presentation may vary, CS is usually diagnosed in young patients, predominantly in female gender. Cardiac myxomas seen in the course of CS are most often encountered in the left atrium. Extra-cardiac manifestations include pigmented skin lesions, cutaneous myxomas, adrenal cortical disease, myxoid mammary fibroadenoma, and testes tumors in male patients. Pituitary adenoma, melanotic schwannomas, and thyroid disease may be encountered in lower percentages.¹

In this case report, we describe a middle-aged female patient who underwent surgery with the diagnosis of a large left atrial myxoma associated with CS.

Case Report

A 46-year-old female patient was admitted to our clinic with palpitation, cough, and exertional dyspnea. Physical examination revealed pigmented lesions, especially scattered in the neck, chest, and extremities. The thyroid gland was mildly enlarged on palpation, but thyroid ultrasonography did not reveal any nodular or diffuse enlargement. A mild-moderate diastolic left-sided murmur was notable. She had

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been followed up with fibroadenomas located at the upper quadrant of the right breast. Echocardiography revealed a left atrial mass, 6 × 4 cm in size, originating from the interatrial septum that was protruding into the left ventricle through the mitral valve during diastole (Figure 1). Peak pulmonary artery pressure was measured to be 65 mmHg, which was accompanied by mild to moderate tricuspid regurgitation. Biochemical and hormonal parameters as well as thyroid hormone levels were within normal limits, except for mildly elevated hepatic enzymes (Serum glutamic oxaloacetic transaminase [SGOT] = 55 IU/l, Serum glutamic pyruvic transaminase [SGPT] = 66 IU/l, serum cortisol = 10 µg/dl, insulin like growth factor [IGF]-1 = 202 ng/dl, growth hormone [GH] = 5 ng/dl, and oral glucose tolerance test with 100 gr glucose: fasting blood glucose level = 110 mg/dl; 1st hour blood glucose level = 170 mg/dl; 2nd hour blood glucose level = 153 mg/dl and 3rd hour blood glucose level = 143 mg/dl).

Genetic analysis of the patient (especially the PRKAR1A gene) could not be accomplished because of the lacking laboratory facilities in a newly established medical center. The patient was operated on with the diagnosis of left atrial myxoma. Following aortic and bicaval cannulation, cardiopulmonary bypass was established. Under mild hypothermia, the aorta was cross-clamped and the heart was arrested with cold blood cardioplegia. Left atriotomy was performed. A large encapsulated soft and lobulated left atrial myxoma, 6 × 4 cm in size, was attached to the interatrial septum with a broad base. The tumor was excised from the interatrial septum with its pedicle (Figure 2). The defect in the interatrial septum was primarily sutured. Weaning from cardiopulmonary bypass was uneventful. Pathological examination confirmed the diagnosis of myxoma. Postoperative echocardiography revealed mild tricuspid regurgitation with a peak pulmonary artery pressure of 30 mmHg. The postoperative course was uneventful and the patient was discharged at the 5th postoperative day without any complication.



Figure 1. Left atrial myxoma (white arrow) seen on echocardiographic evaluation – four chamber view

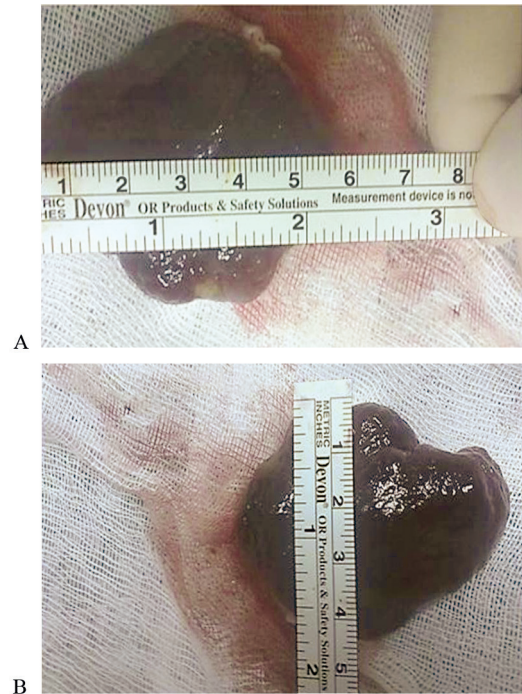


Figure 2. A & B: Excised large atrial myxoma measuring 6 × 4 cm

Discussion

We described a middle-aged patient with left atrial myxoma associated with concomitant clinical manifestations, which are components of CS: pigmented skin lesions and fibroadenomas of the breast. Other possible manifestations of the disease were screened except for genetic analysis.

The overall lifespan of patients with CS is decreased, and cardiac causes of death are the most common among other etiologies.² Age at presentation may vary from birth to a median age of 20 years. Our patient's age was higher, which may reflect the late onset of the disease as well as late admission to a medical center. Since more than half of the cases are familial, application of rigorous screening protocols are suggested for the first-degree relatives of the affected patients.³ Our patient's first-degree relatives were screened for the cardiac and other manifestations of CS. The recurrence rate of the myxomas encountered in the course of CS is higher than that of the sporadic cases and this is an important issue since it may require multiple surgical interventions.^{4,5} Postoperative short and long-term follow-up controls are crucial in these patients.

Conclusion

CS is a complex genetic multiple endocrine neoplasia syndrome with various components relating to different organ systems. Different clinical properties are well known



and diagnostic criteria are defined concerning the large variety of these manifestations. Cardiac myxomas are a corner stone of this disease since cardiac manifestations and related complications account for the majority of the mortality seen in the course of CS.

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