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Simultaneous Occurrence of a Large Asymptomatic Prolapsing Left Atrial Myxoma with a Cutaneous Squamous Cell Carcinoma

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ABSTRACT

Synchronous myxoma of the heart and other malignancies are extremely rare. We report a case of a 64-year-old man who had a large left atrial myxoma that obstructed the mitral valve, as well as an unrelated, coexistent cutaneous squamous cell carcinoma in the sacral area. During the preoperative evaluation for non-cardiac surgery, the tumor was diagnosed coincidentally by echocardiographic examination. Echocardiography findings were consistent with a large left atrial myxoma originating from the posterior wall and prolapsing into the left ventricular cavity through the mitral valve, causing mitral stenosis. The mass was successfully completely excised. Histologic examination of the mass confirmed the diagnosis of cardiac myxoma. We report a casual echocardiographic finding of a left atrial myxoma that obstructed the mitral valve outflow tract, and an unrelated, synchronous cutaneous squamous cell carcinoma in the sacral area.

INTRODUCTION

Metastatic lesions to the heart are much more common than primary lesions. Myxoma is the most common benign cardiac tumor in adults [Nuno 2001]. If a neoplasm of another organ is found at the same time, it is difficult to differentiate the cardiac tumor from a metastasis. Isolated metastasis of cutaneous squamous cell carcinoma to the heart has not been described. Myxomas rarely remain asymptomatic, especially if they are large. The patients may manifest one or more of the classic triad of symptoms: hemodynamic obstruction, embolism, and constitutional effects [Waller 1995]. Synchronous myxoma of the heart and other malignancies are extremely rare. We report a patient with a left atrial myxoma and a squamous cell carcinoma (SCC) who consequently underwent surgical treatment procedures with a good outcome.

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CASE REPORT

A 64-year-old man was admitted to the Department of Reconstructive Surgery with a diagnosis of cutaneous squamous cell carcinoma with localization in the sacral area. Local institutional review board approval was obtained, and the patient provided informed consent. Preoperative evaluation included cardiologic consultation. The patient's medical history included no cardiac and systemic disorders. Upon physical examination, the patient was healthy, with a blood pressure of 130/80 mmHg and a heart rate of 82 beats/min. On cardiac auscultation, loud S1 and a diastolic murmur of grade 1-2/4, maximal at the lower left sternal border, was audible. All other systems were normal, except the cutaneous tumor in the sacral area.

Hemoglobin level was 14 g/dL, total WBC counts was 6500/mm^3, and erythrocyte sedimentation rate was 14 mm/hr. Other hematologic and biochemical laboratory results were within normal limits. The electrocardiogram (ECG) was normal and the chest x-ray showed mild cardiomegaly. Transthoracic (TTE) and transesophageal echocardiography (TEE) showed a left atrial mass (5.1 × 3.0 cm) originating from the posterior wall and prolapsing into the left ventricular cavity through the mitral valve, causing mitral stenosis (Figure 1, A). During diastole, the mass prolapsed through

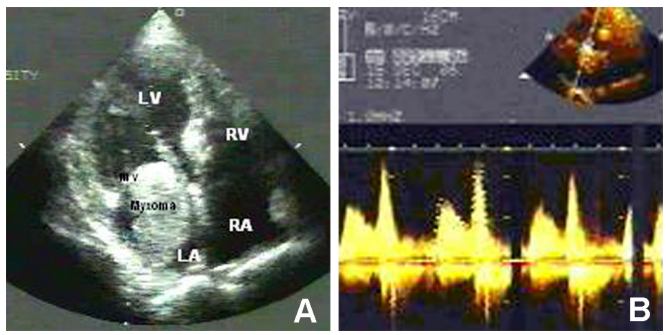

Figure 1. A, Transthoracic apical four-chamber recording of a large left atrial myxoma with significant left and right atrial and right ventricular dilation. Myxoma arising from the posterior wall. The tumor prolapses through the mitral orifice in the left ventricular cavity. LA indicates left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. B, Continuous wave Doppler revealed moderate left ventricular inflow obstruction.

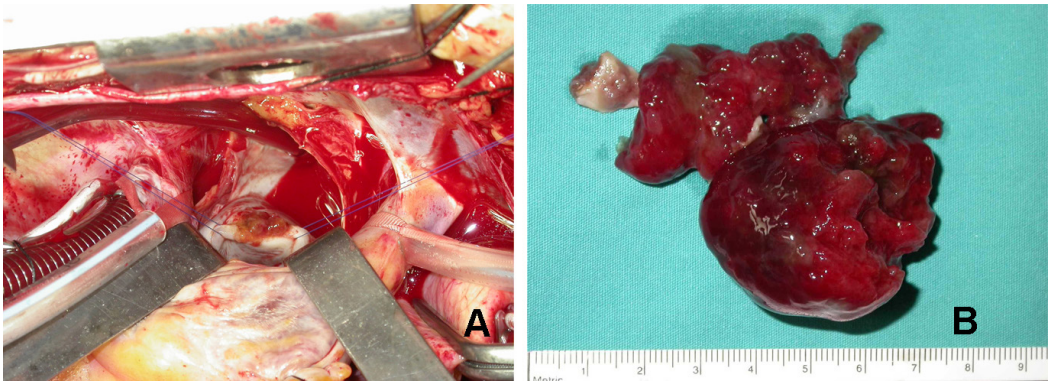

Figure 2. A, Surgical view of the myxoma through the left atriotomy. B, Macroscopic view of the operative specimen.

the mitral valve, resulting in a turbulent color flow doppler pattern. Continuous wave Doppler revealed moderate left ventricular (LV) inflow obstruction, with calculated mean and peak pressure gradients of 6.1 mmHg and 20 mmHg, respectively. The antegrade mitral flow was affected with signs of obstruction (Figure 1, B). The left ventricle sizes were normal. But, the left atrium, right ventricle (4.3 cm), and right atrium (4.2 cm) were enlarged, and they were associated with mild tricuspid regurgitation.

Coronary angiography was performed. Coronary arteries and systolic left ventricular function were normal. The patient was subsequently transferred to a cardiothoracic unit for surgical removal of the tumor. Surgical exploration of the left atrium revealed a 5×3.5×3 cm dull, purplish mass attached to the posterior wall (Figure 2, A). The mass was attentively and successfully completely excised (Figure 2, B). Histologic examination of the mass confirmed the diagnosis of cardiac myxoma. Four weeks after the removal of the myxoma, the patient underwent an operation for the treatment of squamous cell carcinoma. The excised tumor was 6 mm in depth and poorly differentiated. These findings were associated with a relatively bad prognosis and higher metastasis rates. Unfortunately, the patient died 2 years after surgery due to complications of his metastatic disease. No curative treatment was possible.

DISCUSSION

Synchronous myxoma of the heart and other malignancies are extremely rare [Nuno 2001]. To our knowledge, this is the first report in the literature of a cardiac left atrial myxoma and a synchronous cutaneous squamous cell carcinoma.

There are several clinical manifestations in patients with myxoma. The clinical presentation of a cardiac myxoma basically depends on the location of the tumors, and varies with their size, shape, and also factors of the patient [Waller 1995]. The most common site of attachment is the atrial septum at the fossa ovalis, but rare cases are reported of attachment to the left atrial wall [Oliveira 2010]. The most common clinical presentation relates to obstruction of blood flow. The left-sided cardiac myxomas may present with signs of mitral stenosis and insufficiency, and right-sided tumors with dyspnea, syncope, and distention of neck veins. They may also lead to multiple emboli in the systemic or pulmonary circulation, depending on their localization. Atrial myxomas may impair ventricular filling to such an extent that cardiac output becomes significantly reduced and incapable of further augmentation with stress. In rare cases, large myxomas can remain asymptomatic if the tumor growth is very slow [Meng 2002]. Our case, in spite of having an unusually large left atrial myxoma, which led to right chambers and left atrium enlargement, was without symptoms. It is possible that this patient's lack of symptoms may have been the result of a gradual adaptation to a more sedate lifestyle.

Metastatic lesions to the heart are much more common than primary lesions. Myxoma is the most common benign cardiac tumor in adults [Nuno 2001]. Although metastatic basal cell carcinoma and melanoma of the heart have been more frequently documented, metastatic cutaneous squamous cell carcinoma—a particularly rare, especially isolated metastasis of cutaneous squamous cell carcinoma to the endocardium—has not been described [Kypson 2007]. More commonly, metastatic SCC involving the pericardium originates from the lung and uterine cervix [Boukhalil 2006].

Simultaneous occurrence of atrial and pleural myxomas with pulmonary squamous cell carcinoma has rarely been reported [Hasleton 1989, Nakanishi 2007]. However, there is no established association between cardiac myxomas and cutaneous squamous cell carcinoma. A review of the literature did not reveal a similar occurrence. In our case, the preoperative evaluation revealed sacral cutaneous squamous cell carcinoma and a solitary tumor in the left atrium, but it could not be established whether the cardiac tumor was a primary lesion, part of a more complex syndrome, or a metastasis from the skin.

The present case demonstrates some interesting features. First, the usual site of attachment of a myxoma is in the area of the interatrial septum. Our case showed left atrial myxoma originating from the posterior wall. It is rare to find a myxoma attached at the posterior left atrial wall. Second, various clinical signs and symptoms produced by cardiac myxomas have

been reported; however, asymptomatic large cardiac myxomas of the left atrium, as described in this patient, are very rare. Third, it is important that myxoma and squamous cell carcinoma coexist at the same time.

In this article we report a casual echocardiographic finding of a left atrial myxoma that obstructed the mitral valve outflow tract along with an unrelated, coexistent cutaneous squamous cell carcinoma in the sacral area. It is not possible to make any biological connection between these tumors. However, more reported cases in the future will increase the impact of our case in the literature.

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