A Rare Clinical Presentation Caused by Atrial Myxoma: Right Heart Failure

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Abstract

Primary heart tumours are rarely encountered. Myxomas, which are included in this group of tumours, are the most common primary benign neoplasms of the heart and approximately 75-80% of them are found in the left atrium. In our case, a 72-year-old female patient, who had complaints of shortness of breath, pain in the chest and swelling in the legs that increased in the last three months. We present a case with a myxoma in the right atrium with a dimension of 80x60x15 mm attached to the interatrial septum. According to the echocardiographic examination, it clinically shows signs of right heart failure, almost completely obstructs the right atrium and extends to the right ventricle. Ascending aortic aneurysm was also detected on the echocardiography. Despite the fact that the right atrial mass obliterating the cardiac chambers was removed by surgery, the valve pathology caused by the lesion did not regress.

Key words: Atrial Myxoma, Right Heart Failure, Tricuspid Valve Insufficiency, Dyspnea.

INTRODUCTION

The majority of heart tumours (75%) are benign and the most commonly seen heart tumour is atrial myxoma (1). Most myxomas develop in the left atrium, but rarely they may develop in the right atrium (2). Large myxomas typically remain clinically silent until they reach a significant size, or they simply cause non-specific symptoms such as fatigue and palpitations. However, giant myxoma in the right heart cavity is a rare clinical presentation (3, 4). Clinically significant embolic events are much less common in patients with right atrial myxoma compared to those with left atrial myxoma (5). Additionally, myxomas may lead to syncope and sudden death due to tricuspid valve obstruction (6). Echocardiography is the standard diagnostic test and surgical approach is the curative treatment (7).

CASE REPORT

72-year-old female patient presented with complaints of shortness of breath, oedema in the ankles and pain in the chest that increased within the last three months.
Physical examination revealed no pathological findings other than a third degree systolic murmur in the tricuspid valve and bilateral pretibial oedema. The patient’s vital signs were stable at the initial evaluation. There was no comorbid disease other than hypertension in the medical history of the patient. Cardiothoracic index was increased on the postero-anterior (PA) chest radiography and normal sinus rhythm was detected in the electrocardiogram. The echocardiographic examination revealed normal left ventricular dimensions, but inferior wall was found to be hypokinetic. Ejection fraction was found to be 42% in the evaluation performed with the modified Simpson method. Left atrium size was measured at the upper limit. Aortic valve had a senile structure, and 2nd degree aortic insufficiency was detected. Mitral valve structure was thick, coarse and 1st degree mitral insufficiency was also detected. Diameter of the ascending aorta was measured to be 5.3 cm. The right heart cavities were large and had severe tricuspid insufficiency. In the interatrial septum, there was a mass with a dimension of 78x47 mm attached to the right atrium (Figure 1-2-3). It was determined that the mass obstructed most of the right atrium in the ventricular systole, extended to the right ventricle to the tricuspid valve during the diastole, obstructed the valve and resulted in severe tricuspid valve insufficiency. Although there was severe insufficiency in the tricuspid valve, no gradient increase was observed and systolic pulmonary arterial pressure (sPAP) was measured as normal. Right coronary artery system and left coronary artery system were found to be normal in the coronary angiography. Surgical procedure was planned for the patient. The rib cage was opened with median sternotomy during the surgery. A right atriotomy was performed to remove the mass with a dimension of 80x50 mm attached to the secundum septum compatible with atrial myxoma (Figure 4). The ascending aortic dilatation was then repaired. In the pathological examination, a gray-white bright patchy hemorrhagic tissue with a dimension of 80x60x15 mm was reported as myxoma due to its spindle shape and stellate appearance (Figure 5). No mass was detected in echocardiography in the postoperative 6th month follow-up. The patient had grade 3 tricuspid valve insufficiency and jet velocity was measured as 3.7m/s. No change was observed in the patient’s right ventricle diameters. At the last evaluation, the patient had no symptoms.
DISCUSSION

Tumours found in the heart are less than 1% in the general population. The familial form, which usually occurs sporadically, is encountered as Carney syndrome. Atrial myxomas constitute more than 50% of benign cardiac tumours (8). Although they appear between the ages of 30 and 60 on average, they most often start to show symptoms around 50 years of age (7). In our case, the symptoms appeared in a patient in her 70s. Symptoms and indications of cardiac tumours may not be clear and vary depending on the localization of the mass. Cases may present with a clinical picture that is similar to the findings of heart failure as a result of failure and obstruction of the valves in intracardiac tumours. As in right heart failure, jugular venous fullness, ascites and oedema may be observed.

The diagnosis of myxomas is simple and does not require special imaging methods other than two-dimensional echocardiography (7). In echocardiography, the tumour can be differentiated from normal anatomic structure with smooth lines. In addition, pericardial effusion may also be observed (9). However, pericardial effusion was not identified in our case. If the results of two-dimensional transthoracic echocardiography are unclear, transesophageal echocardiography might be required (10). Once diagnosed, surgical excision is recommended to prevent embolism and sudden cardiac death. As the recurrence rate after surgical excision is high, echocardiographic follow-up is recommended. Even though the recurrence of myxomas resected surgically is typically within the first 4 years, recurrence was also reported after 14 years (7). Pathologies caused by right atrial myxomas in the tricuspid valve after surgical excision have been reported to be completely reversible (11). However, in our case, significant tricuspid insufficiency continues even after six months. Atrial myxomas that reach large dimensions should be treated with surgical resection. Even though valvular pathologies caused by right atrial giant myxomas are reversible, it was permanent in our case. Thus, this should be considered in interventions of valvular pathologies during surgery.
Declarations
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References