

# MYXOMA ORIGINATING FROM RIGHT VENTRICULAR APEX OBSTRUCTING VENTRICULAR INFLOW

## SAĞ VENTRİKÜL GİRİŞ YOLUNU TIKAYAN SAĞ VENTRİKÜL APEKSİNDEN KÖKEN ALAN MİKSOMA

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*SUMMARY: Unusual case of right ventricular myxoma producing right-sided inflow obstruction in a 67-year-old female patient is described. Transatrial en bloc tumor excision with a partial normal ventricular wall at its attachment was performed.*

*Key Words: Right Ventricular Myxoma, Right Ventricular Mass.*

### INTRODUCTION

Myxomas are benign tumors of the heart and they mostly originate from the left atrium (1). Right ventricular occurrences are rarely reported in the literature. Sporadic cases of right ventricular myxomas have been reported (2, 3). Cardiac myxoma usually presents as a benign pedunculated tumor, but sometimes exhibits malign behavior by producing right-sided inflow or outflow obstruction. We report an unusual case of ventricular myxoma originating from the right ventricular apex with the clinical findings of right ventricle inflow obstruction.

### CASE REPORT

A 67-year-old female patient was referred to our department with a preliminary diagnosis of right ventricular mass. She had dyspnea, palpitations and fatigue on exertion that started 3 months before admission. She was afebrile, pulse rate was 84 beats per min, respiratory rate was 16 breaths per min, and BP was 110/70 mm Hg.

*ÖZET : 67 yaşında bayan hastada sağ ventrikül çıkım yolunu daraltan ender bir sağ ventrikül miksoma olgusu anlatılmıştır. Tümör yapıştığı yerden transatrial olarak parsiyel ventrikül duvarı ile en-blok çıkartılmıştır.*

*Anahtar Kelimeler: Sağ Ventrikül Miksoma, Sağ Ventriküler Kitle.*

With regards auscultation, a grade 3/6 holosystolic murmur was best heard along the left sternal border. The liver was palpable 4-6 cm below the right costal margin. Chest roentgenogram showed mild cardiomegaly and chronic obstructive pulmonary disease findings. Electrocardiogram demonstrated right-axis deviation. The erythrocyte sedimentation rate was 25 mm in the first hour, and the remaining laboratory data were normal. Transthoracic and transesophageal echocardiograms showed a mobile 5x4 cm right ventricular mass (Fig. 1) originating from the apex, and prolapsing into the tricuspid valve. There was no mass lesion seen in the other cardiac chambers. Doppler echocardiographic examination revealed mild tricuspid regurgitation. Her coronary angiography showed no abnormalities. The patient underwent elective surgery on December 12, 2001. The heart was exposed through midline sternotomy. After the institution of cardiopulmonary bypass in the standard fashion



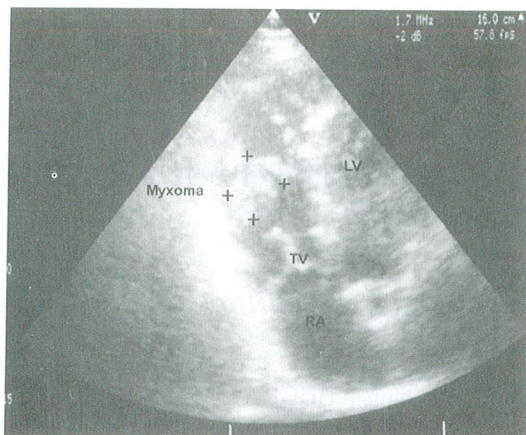


Fig. 1: Transthoracic echocardiogram in the four-chamber view showing an irregular, echo dense right ventricle myxoma below the tricuspid valve (RA=right atrium, LV=left ventricle, TV=tricuspid valve).

using two separate venous cannulae, a right atriotomy was performed. A 5x4 cm lobulated myxomatous mass originating from the right ventricular apex was seen through the tricuspid valve (Fig. 2). It was excised en bloc with a partial normal ventricular wall at its attachment (Fig. 3). The tricuspid valve was examined by filling the right ventricle with saline solution. No additional reconstructive procedures on the tricuspid valve were necessary. Histology of the resected specimen showed myxoid matrix containing polygonal stellate spindle cells. The postoperative course of the patient was uneventful and she was discharged after 1 week.

#### DISCUSSION

Myxomas are neoplasms of endocardial origin. The tumor usually projects from the endocardium into the cardiac chamber. The cells giving rise to the tumor are considered to be multipotential mesenchymal cells that persist as embryonal residues during septation of the heart and differentiate into endothelial cells, smooth-muscle cells, angioblasts, fibroblasts, cartilage cells and myoblasts. The prevalence of myxomas in the atrial septum is therefore understandable (4, 5).

Myxomas occur in all age groups but are particularly frequent between the third and sixth

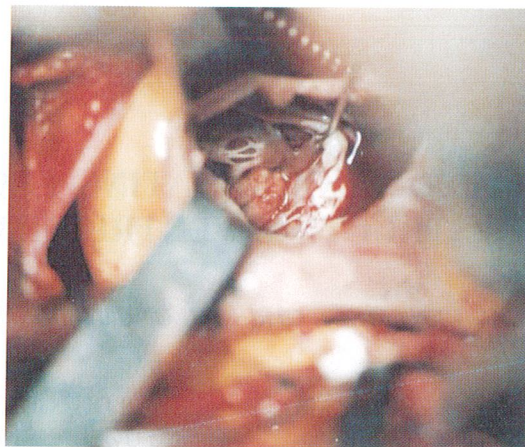


Fig. 2: Right atrial view of right ventricular myxoma.



Fig. 3: Right ventricle myxoma (fresh specimen).

decades of life (6). Diagnosis of myxoma is difficult until the patient has significant symptoms. The clinical features of myxomas are determined by their location, size and mobility. General symptoms may be caused by arterial embolization of myxoma or the space occupying nature of the tumor leading to intracardiac obstruction and constitutional symptoms.

Echocardiography, computed tomography, and magnetic resonance imaging are important non-invasive diagnostic tools. The diagnosis of myxoma is usually made by two-dimensional echocardiography, and transesophageal echocardiography provides accurate information



in respect of size, exact location, morphological characteristics and point of attachment (7).

Hemodynamically, the motion of the myxoma back and forth, called the "wrecking ball effect", may obstruct both the inflow of the right ventricle and the orifice of the tricuspid valve during the systole and may damage leaflets of the tricuspid, leading to regurgitation (8).

Specifically, right ventricular myxomas may produce single or multiple pulmonary emboli (9), pulmonary valve obstruction (3), right ventricle failure (10) and even syncope (11).

Clinically evident embolic events are uncommon. Nevertheless, there have been reports of the embolization of tumor fragments into the pulmonary arteries with subsequent pulmonary hypertension, and also of lethal fulminant pulmonary embolism in cases of right atrial or right ventricular myxoma (12).

Our careful review of the literature using Medline showed that 102 cases of myxoma originated from the right ventricle. Nineteen cases were reported as a right-sided obstruction of the inflow or outflow. Myxomas of the right ventricle may originate from the tricuspid valve, pulmonary valve and anterior papillary muscle of the tricuspid valve and free wall of the right ventricle. Free wall attachment is more commonly found than other localizations in Medline research.

Immediate total surgical resection of the myxoma is indicated as soon as the diagnosis is made. In 1955, Crafoord reported the first successful excision of a left atrial myxoma using temporary cardiopulmonary bypass (13). The transatrial approach for total excision is the general surgical choice for all cases, but resection via right ventriculotomy can also be performed if right atrial exposure is not appropriate. Intraoperative fragmentation of the tumor and embolism must be avoided. All chambers of the heart should be inspected to rule out a multifocal tumor. Mechanical damage to a heart valve or adhesion of the tumor to valve leaflets might call for valve repair by annuloplasty or replacement with a prosthetic valve (14). The rate of operative mortality is 0 to 3% (15).

The recurrence of cardiac myxomas has been well documented, and so close echocardiographic

follow up is mandatory (16). Although cardiac myxomas are histologically benign, they may be lethal depending on their cardiac position.

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