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A GIANT RIGHT ATRIAL MYXOMA

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ÖZET

Dev Sağ Atrial Miksoma

Sağ atrial miksomalar literatürde nadir rastlanılan vakalardır. Bizim vakamız, 3 loblu, bir lobu sağ ventrikül içinde bulunan, kum saati şeklinde bir dev sağ atrial miksomaydı. Boyutları 18 x 7 x 5 cm., ağırlığı ise 170 gramdı. Literatürde bu kadar büyük boyutlarda ve ağırlıkta bir başka sağ atrial miksomaya rastlayamadığımızdan bu vakayı yayınlamayı uygun bulduk. Konu ile ilgili literatür vaka ışığında tartışıldı.

Anahtar Kelimeler : Miksoma, atrial miksoma

SUMMARY

Right atrial myxomas have rarely been reported. It was described as a giant hourglass shaped right atrial myxoma consists of three lobes, one of them had protruded into the right ventricle. Its sizes were 18 x 7 x 5 cm. and weight was 170 g. It was not possible to find another myxoma as big as this one in the literature, so we thought to report it. We reviewed and discussed the sections of the literature involving the subject.

Key Words : Myxoma, atrial myxoma

INTRODUCTION

Right atrial myxoma is a rare primary cardiac neoplasm accounting for 25 percent of atrial myxomas (1,2). Myxomas which occur in the left or right atrium comprise approximately 50 % of all primary cardiac neoplasms (3,4). It is generally agreed that myxomas are true neoplasm (5,6,7). Myxomas occur predominantly in women and famillial tendency has been reported (8). Some authors suggested a dominant mare of inheritence whereas another group supposes a familial incidence is due to an autosomal recessive trait (9,10). The initial clinical diagnosis is usually of mitral stenosis, right heart failure or a systemic illness and the cardiac mass is diagnosed later by echocardiography (11).

Two dimensional echocardiography is reliable and accurate method of diagnostic cardiac tumors (6,12).

CASE REPORT

A 24 year-old man was referred for evaluation because of suspected right heart failure. For two years he complained of fatigue on exercise. On examination, he has a sinusal tachycardia (120 beats/min), blood pressure of 120/70 mm Hg and mild jugular venous engorgement. On auscultation, splitting of the first heart sound, a low frequency mid-diastolic murmur at the apex (tumor plop) and a grade 2/6 holosystolic murmur were heard in the lower left parasternal area. There was no variation in the murmur with position and respiration. The lungs were clear, the liver was 3 cm palpated at the mid-clavicular line and peripheral edema were noticed.

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Routine blood tests and urinalysis were normal except that the sedimentation rate was 65 mm/h. 2-D echocardiography showed a large, three lobated, mobile right atrial mass attached by a peduncle to the anterior wall of the right atrium, just top of the outflow tract of the inferior vena cava. One of the lobes was in the right ventricle both systole and diastole. The other two lobes in the right atrium protruded during diastole into the inflow tract of the right ventricle, almost completely obliterating the valve orifice. It was clearly seen that both the right atrium and ventricle had enlarged (Figure 1).

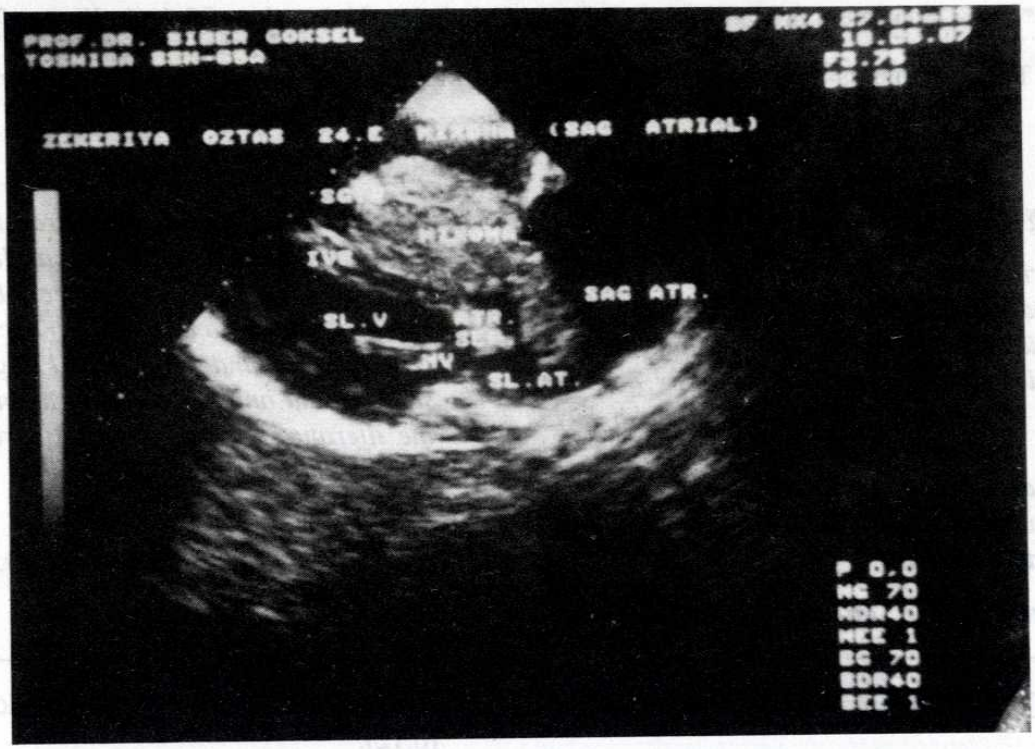

Figure 1. The preoperative doppler study of the patient

At surgical exploration, a large (17x8x5 cm), pedunculated tumor was remowed from the right atrium (Figure 2). Surgical findings were confirmed with echocardiography. The tumor had grown from the right atrial wall by a 1.5 cm peduncle. It was excised with its peduncle and a cuff of normal tissue to prevent relapse. The tricuspid valve was seen as intact. The endothelial defect was closed with primary stures. Microscopic examination confirmed the diagnosis of myxoma.

After surgery the patient become asymptomatic and diuretic was given only. He was dicsharged 11 days after operation without any complications. Recurrence was not seen up to now although postoperative controls continued for three years. In postoperative family screen there were no pathologic finding in any person. Postoperative echocardiogram was normal (Figure 3).

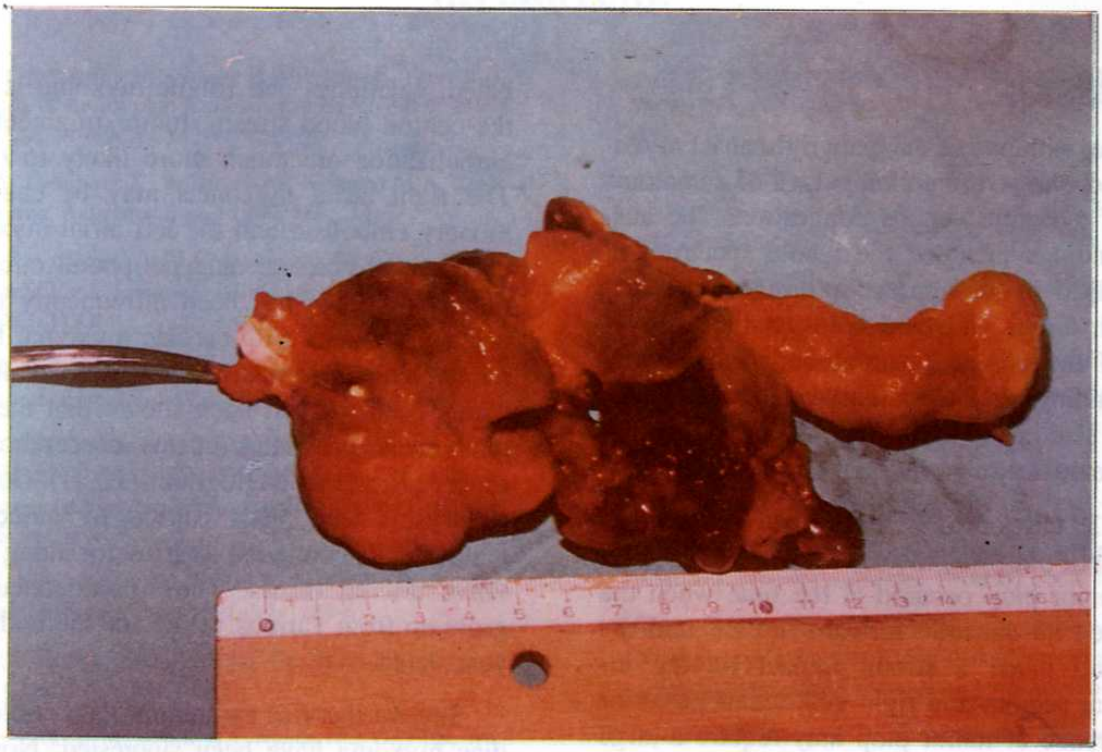

Figure 2. The postoperative photography of the giant right atrial myxoma

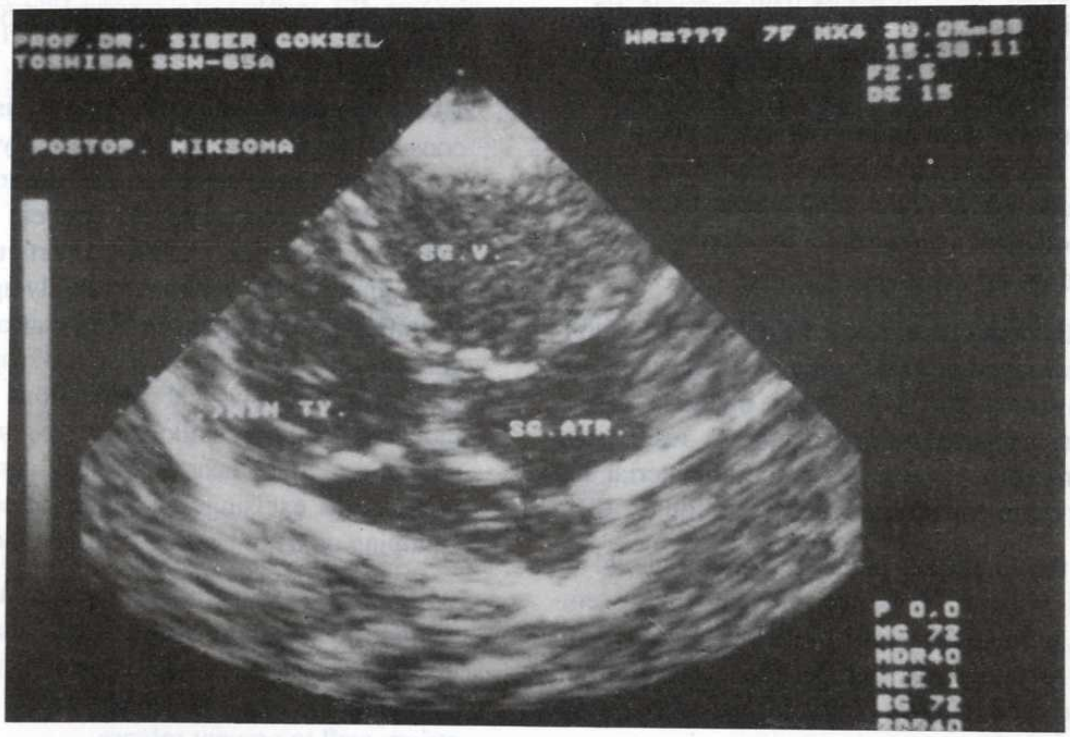

Figure 3. The postpreoperative doppler study of the patient

DISCUSSION

This case, with one of the giant right atrial myxomas reported, showed remarkable lack of symptoms and lack of systemic venous congestion. The auscultatory findings included the whole spectrum of signs described in such cases; splitting of the first heart sound, tumor plop and holosystolic murmur. Absence of the classic postural variation in auscultatory findings in our patient was probably related to the size of the tumor that obstructed the tricuspid valve orifice, regardless of body position.

Among 39 cases of right atrial myxoma, the presence of tumor plop was mentioned in 12 (31 %) (2, 13, 14). Tumor plop occured at the time when the mass stopped its diastolic forward motion into the ventricle and made a strong impact on the interventricular septum and right ventricular posterior wall. Occurence of tumor plop may require a large mass or long enough tumor stalk to allow the impact of the mass of the ventricular wall (1).

Clinical findings frequently shown are fatigue on exercise, loss of weight, fever, malaise, swelling and pain in joints. Anemia, Raynoud's phenomena, leucocytosis, increased eryhtrocyte sedimentation rate, hyperglobulinemia often recorded in series of patients. Fatigue on exercise and increased sedimentation rate were present at our patient. Such symptoms and signs have been explained as an unspecific responce to necrotizing tumor tissue or as an immunological response to the foreign protein represented by the myxonma (15,16).

Two dimensional echocardiography is a noninvasive technique of great value in the diagnosis of cardiac myxoma (6,7,17). Other methods of diagnosis are catheterization, magnetic resonnance imaging, microscopic study of embolic material and perceiving during another cardiac operation (7, 18).

When the diagnosis made, operation should be performed without delay to prevent tumor embolization, pseudoaneurysm and cardiac failure. Embolization probably does not readily occur in solid, capsular, coated myxomas. However, when a lobated, gelatinous and fragile myxoma is located in the central blood stream, tumor fragmentitaion and embolization are much more likely to occur (19). The right atrial myxomas may be cause of pulmonary embolism and the left atrial myxomas may be cause of cerebral. and peripheral ones. Massive tumor embolism has been infrequently reported to occur in patients with cardiac myxoma. More commonly, repeated embolization of small tumor fragments occur. It had been shown that the myxomas may occur pseudoaneurysms of cerebral and coronary arteries by angiograms (20,21). Occosionally, aneurysms have been caused by embolic myxomatous cells continued to grow for along time to involve arterial media. Thus, pseudoaneurysm may grow up from various vessels of the body for some years later.

Several theories explaining then etiology of cardiac myxoma have been suggested. Now it is generally accepted that the tumor arises from undifferantiated non malignant cells, particularly apt to be present in the fossa ovalis region of the atrial septum. A number of factors yet remain to be explained; with regard to both generalized constitutional symptoms and the effect on plasma proteins (20, 21).

At operation tumor consistency is an important consideration, since the gelatinous myxoma is tumor type most likely to embolize intraoperatively. In these cases precautions to prevent intraoperative tumor embolization should be particularly stressed. The tumor quality cannot be readily appreciated preoperatively, but a lobated tumor at angiography or echocardiography should arise the suspicion of a fragile myxoma. Surgical precautions to avoid intraoperative tumor embolization consist of not manipulating the heart during cannulation and prefentially excising the tumor after aortic crossclamping on a nonbeating cardioplegic heart (19).

During the operation, appropriate precautions should be taken to avoid fragmentation and embolization of myxomatous material. The tumor should be excised extensively with a cuff of normal atrium wall to prevent relapse.

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