

# Primary Benign Schwannoma of the Left Atrium

Selim Aydın<sup>1</sup>, Yücesin İsmail Arslan<sup>2</sup>, Aybanu Gökçen Tuğgun<sup>2</sup>, Fikri Yapıcı<sup>2</sup>, Fırat Hüsni Altın<sup>1</sup>

<sup>1</sup>Diyarbakır Region Training and Research Hospital, Clinic of Cardiovascular Surgery, Diyarbakır, Turkey

<sup>2</sup>Dr. Siyami Ersek Chest, Heart and Vascular Surgery Training and Research Hospital, Clinic of Cardiovascular Surgery, İstanbul, Turkey



## ABSTRACT

Schwannoma of the heart is considered as the most rare tumor in non-myxomatous tumors of the heart. A 64 year-old male was admitted with the complaint of dyspnea for 1 year. Transthoracic echocardiography revealed severe degenerative mitral insufficiency. Perioperative transesophageal echocardiography revealed a mass in the left atrium. Complete resection of the tumor and mitral ring annuloplasty was performed. Histopathology showed a benign schwannoma. Schwannomas are one of the cardiac masses which may be encountered in patients undergoing open heart surgery due to other cardiac pathologies.

**Key Words:** Schwannoma; cardiac tumor; transesophageal echocardiography

## Sol Atriyumun Primer Benign Schwannomu

### ÖZET

Kardiyak kökenli schwannom kalbin nonmiksomatöz tümörleri arasında en nadir görülenlerdendir. Altmış dört yaşında erkek hasta bir yıldan beri olan nefes darlığı şikayeti ile başvurdu. Transtorasik ekokardiyografide dejeneratif ileri mitral yetmezlik saptandı. Perioperatif transözofageal ekokardiyografide sol atriyumda kitle görüldü. Tümör tamamen rezekt edildi ve mitral ring anuloplasti yapıldı. Histopatoloji sonucu benign schwannom olarak geldi. Schwannomlar diğer kardiyak patolojiler için açık kalp cerrahisi yapılan hastalarda da saptanabilecek kardiyak kitlelerdendir.

**Anahtar Kelimeler:** Schwannom; kardiyak tümör; transözofageal ekokardiyografi

## INTRODUCTION

Among the non-myxomatous tumors of the heart, schwannomas are extremely rare pathologic entities<sup>(1)</sup>. Schwannoma or neurilemoma is a nerve sheath tumor, which consists of Schwann cells. Neurogenic tumors can be benign and primary considering the origin or sometimes malign and metastatic<sup>(2)</sup>.

## CASE REPORT

A 64 year-old male was admitted with complaint of dyspnea of one year duration. The patient did not have other cardiovascular symptoms and his medical history was unremarkable.

Physical examination revealed a 2/6 systolic murmur in the apex, which expands to the axilla. Routine blood tests, cardiac enzyme levels, ECG and chest x-ray were normal. A transthoracic echocardiography revealed severe degenerative mitral regurgitation. An angiogram demonstrated normal coronary arteries and left ventricular function. Mitral valve surgery was planned. Perioperatively, a 2x2 cm mass was detected by transesophageal echocardiography in left

atrium in the operating room (OR) setting (Figure 1). Following median sternotomy, cardiopulmonary bypass was instituted by the usual fashion and left atrial incision was made in Sondergaard's groove. The origin of the mass was assessed. Mass was localized on the left atrium wall, superior to the right upper pulmonary vein. Complete resection of the mass with the adjacent atrial wall was performed and the defect was repaired with 4/0 polypropylene suture. Mitral ring annuloplasty was performed using a 29 mm Carbomedics annuloflex annuloplasty ring.

Histopathology showed a benign schwannoma originating from the left atrium. The mass consisted of an encapsulated ovoid tumor, measuring 2.5x2.x1 cm, with yellow and brown nodular lesions on cross sections. Microscopic sections revealed spindle cells with eosinophilic cytoplasm and elongated nuclei with minimal mitotic activity and no hypercellularity and no anaplasia (Figure 2A). Immunohistochemistry analysis showed the tumor cells were positive for S100 and negative for CD34, SMA and desmin (Figure 2B). Apart from acute atrial fibrillation, which occurred and treated medically in postoperative day two, the postoperative

## Correspondence

Selim Aydın

E-mail: draydinselim@yahoo.com

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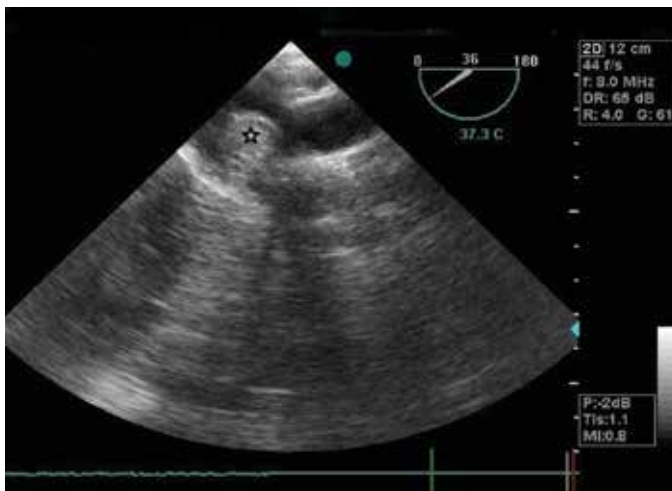
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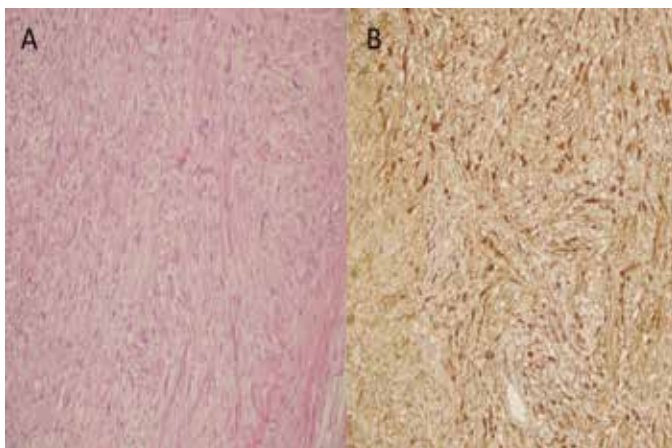
period was otherwise uneventful. Patient was discharged home on postoperative day 6. On the follow-up controls, which were performed on the 3<sup>rd</sup> and 6<sup>th</sup> month, no evidence of recurrence was present in transthoracic and transesophageal echocardiography.

## DISCUSSION

Primary tumors of the heart are found in 0.02% of routine autopsies<sup>(3)</sup>. In surgical series, about 83% of all cardiac tumors are benign in nature and 50% of these are being myxomas. Cardiac schwannoma is considered as a very rare tumor of the heart<sup>(1)</sup>. Primary cardiac schwannomas mostly originates from the nervous system of the heart. Thus, schwannomas are usually found in the right atrium and especially near the cardiac plexus but sometimes can also be found in the ventricular myocardium, atrioventricular valves and in the left atrium as in our case. Left atrium is not a common localization for this type of tumors. Early et al. previously reported a review of 14 cases of atrial schwannoma, in 9 cases; the tumor was reported to be in the right atrium<sup>(4)</sup>.



**Figure 1.** Left atrial mass is seen in the left atrium on these transesophageal echocardiography



**Figure 2.** Microscopic appearance. **A-** Much of the tumor is composed of bland spindled cells with elongated nuclei that are separated by abundant extra-cellular matrix material. (Hematoxylin and eosin; original magnification x175.) **B-** Diffuse nuclear and cytoplasmic positive staining with S 100. (S 100; original magnification x200.)

The clinical signs and symptoms related to schwannomas depend on the location and the size of the tumor. Most common presenting symptoms are dyspnea, chest pain, orthopnea, pretibial edema, syncope and arrhythmias. The cardiac function might deteriorate as a result of myocardial invasion, atrioventricular valve insufficiency and arrhythmia. Sometimes patients might be asymptomatic because of early detection and relatively small size of the tumor or might be presented with other associated cardiac lesions such as in our case.

Cardiac schwannomas can be screened by transthoracic echocardiography. Transesophageal echocardiography, magnetic resonance imaging and computed tomography are helpful for determining the location and the extension of the mass. In our report, despite the use of preoperative transthoracic echocardiography we couldn't detect a mass originating from the left atrium. However, the mass was revealed by the use of perioperative transesophageal echocardiography.

Definitive diagnosis can only be made by histologic examination. Positive immunohistochemical analysis for S-100 protein and negative immunohistochemical analysis for CD34, desmin and SMA support that the tumor originates from the Schwann cell<sup>(5)</sup>.

In case of encountering an encapsulated and well-demarcated mass in the atrium during the operation, a cardiac schwannoma might be considered. The objective of the surgery should be complete resection of the mass and repairing the defect with primary suture technique or cardiac reconstruction with autologous pericardium or synthetic patch. Reconstruction of the atrial wall might cause atrial fibrillation but can be treated medically.

The prognosis is excellent for those benign lesions with complete and margin free resection. On the other hand, even when a complete resection is possible the prognosis of a malign lesion is more challenging<sup>(6)</sup>.

In conclusion, schwannomas as well as other cardiac masses can be detected in patients undergoing open-heart surgery for other cardiac pathologies. One should bear in mind that routine perioperative transesophageal echocardiography is an extremely valuable tool in the diagnosis of such masses which cannot be demonstrated during preoperative routine evaluation.

## CONFLICT of INTEREST

The authors reported no conflict of interest related to this article.

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