

An Atypically Localized Atrial Myxoma: A Case Report

Bilgehan Erkut, MD,¹ Cevdet Ugur Koçogulları,² Sakir Arslan, MD,³ Fuat Gündoğdu, MD,³
Hikmet Koçak, MD,¹ Yahya Islamoglu, MD³

¹Department of Cardiovascular Surgery, Faculty of Medicine, Atatürk University, Erzurum, Turkey;

²Department of Cardiovascular Surgery, Faculty of Medicine, Afyon Kocatepe University, Afyon, Turkey;

³Department of Cardiology, Faculty of Medicine, Atatürk University, Erzurum, Turkey

ABSTRACT

Atypical cardiac myxomas are rare occurrences and may present with a variety of clinical manifestations depending on the location and morphology. A 46-year-old woman had a 4 × 3 × 2-cm myxoma originating from the superior wall of the left atrium, found by echocardiography and multislice tomography. The tumor was successfully treated by surgical excision. The resected tumor was a well-defined encapsulated mass with a narrow-base stalk originating from the right wall of the left atrium in between the right upper and lower pulmonary vein. The patient recovered without complication and was discharged 6 days after the operation. At 1-year follow-up, echocardiography revealed normal cardiac function without recurrence in terms of mass. Although up to 80% of myxomas are localized in the left atrium, of which 75% involve in the interatrial septum, it should not be forgotten that myxomas can appear in an atypical localization, as occurred in our case.

INTRODUCTION

Primary cardiac neoplasms are rare entities, with an incidence of approximately 0.001% to 0.03% in unselected patients at autopsy. Three quarters of the tumors are benign. Nearly half of the benign heart tumors are myxomas, and most of the rest are lipomas, papillary fibroelastomas, and rhabdomyomas. In adults, approximately 25% of primary cardiac tumors are malignant, and one third of these tumors are angiosarcomas [MacGowan 1993].

Intracardiac myxoma is the most common tumor of the heart with an estimated incidence of 0.5 per million populations per year [Keeling 2002]. Although they are benign mesenchymal and usually polypoid myxomatous or pedicle tumors, they may cause sudden death because of intracardiac obstruction or embolism.

Cardiac myxomas generally originate in the left atrium (75%) attached via a stalk to foramen ovale in the interatrial septum; however, these tumors may also arise from atypical sites such as the right atrium (15%-20%), atrial appendage, anterior and posterior wall, left or right ventricle, and on the valves [MacGowan 1993; Keeling 2002; Darwazah 2006].

We report here an extremely rare case of a myxoma originating from the right wall of the left atrium and determined haphazardly, which was diagnosed and located using transesophageal echocardiography (TEE), multislice tomography (MSCT), and histopathological analysis.

CASE REPORT

A 46-year-old woman was referred to our hospital for the evaluation of angina. This symptom had appeared for 6 months and was not associated with physical activity. The blood pressure of the patient was 120/80 mmHg, pulse rate was 90 beats per minute without deficit, respiratory rate was 22 per minute, and body temperature was 36.2°C. Auscultation did not reveal any murmur. No abnormal neurological symptoms or signs were found.

Diagnostic Work-up

Blood biochemistry revealed normal serological findings for liver and kidney function. Chest radiography revealed a normal cardiothoracic rate and bilateral lung zone. We performed echocardiography and coronary angiography because the patient had a history of angina pectoris. The coronary angiography of the patient was normal except for slow coronary flow. Transthoracic echocardiography (TTE) revealed a normal range for left ventricular function, but the left atrium was minimally distended (44 mm) and contained a mobile mass. TEE was then performed the next day. There was a heterogeneous mass that was an echogenic tumor and seemed to originate from the right wall of the left atrium free interatrial septum (Figure 1) and was swinging during every cardiac beat. Because of the localization, this mass did not cause mitral inflow obstruction or regurgitation. Although this mass was thought to be a myxoma, explorative thorax and abdomen MSCT were performed the same day to eliminate the possibility of it being a cardiac metastatic mass. By this explorative procedure, the exact localization and nature of the mass

Received December 7, 2006; received in revised form February 5, 2007; accepted February 8, 2007.

Correspondence: Bilgehan Erkut, MD, Atatürk Bulvarı, Eda Apartmanı, Palandoken Tıp Merkezi Ustu, Kat: 3, No: 3, 25080 Erzurum, Turkey; 90-533-745-10-06; fax: 90-442-316-63-40 (e-mail: bilgehanerkut@yahoo.com).

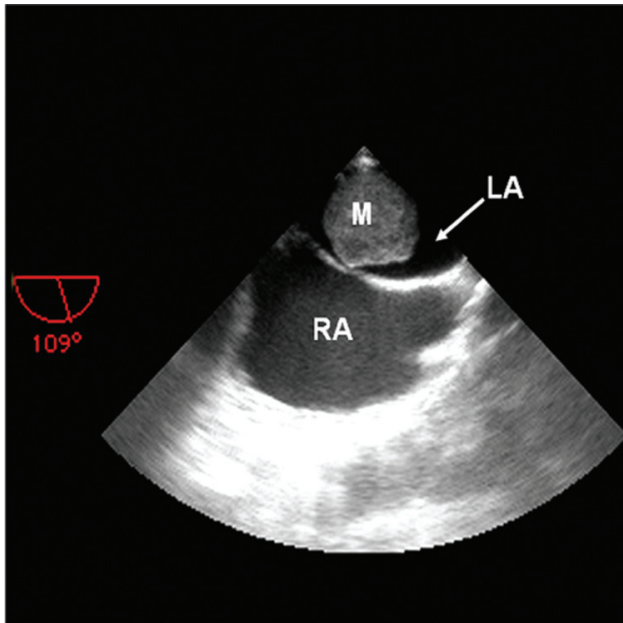


Figure 1. Transesophageal echocardiogram shows a heterogeneous mass adhering to the right wall of the left atrium free septum. RA indicates right atrium; LA, left atrium; M, myxoma.

would be determined. MSCT revealed that a narrow-base stalk was attached near the right upper pulmonary vein in the left atrium (Figure 2).

Operative Technique

The patient was transferred to our surgical department for operation. Under extracorporeal circulation with mild systemic hypothermia and cold cardioplegic arrest, the left atrium was incised along the interatrial groove and we found a myxoma occupying the left atrium.

A well-defined encapsulated gelatinous mass ($4 \times 3 \times 2$ cm) with a narrow-base stalk was attached 1 cm inferior away from the edge of the left atrial incision, that is, it was attached between the right upper and lower pulmonary veins in the right wall of the left atrium (Figure 3A). The mass was resected en bloc with its pedicle and normal atrium tissue with wedge resection of the left atrium wall by starting from the left atrium incision. The wall defect and left atrium incision was directly sutured continuously, and the left atrium was closed. The tumor weighed 138 g and measured $4 \times 3 \times 2.5$ cm in size (Figure 3B).

Postoperative Course

The postoperative course was uneventful. The duration in intensive care unit was 1 day. The patient was discharged on the 6th postoperative day. On pathologic examination, the yellow and shiny fibro adipose material was defined as a myxoma with $4 \times 3 \times 2.5$ -cm dimensions. At 6 months follow-up, the patient was free of complications. There was no residual

mass in the left atrium, and she had good left ventricle function in echocardiography.

DISCUSSION

A left atrium myxoma was first described in a postmortem examination in 1845; however, the introduction of echocardiography has greatly facilitated the antemortem diagnosis of this tumor. Although it is rare, myxoma is the most common cardiac neoplasm in adults [Chitwood 1992; Kamiya 2001]. About 75% of myxomas originate in the left atrium, 15% to 20% in the right atrium, and only 3% to 4% in the ventricle [MacGowan 1993]. Most myxomas arise from the interatrial septum at the border of the fossa ovalis. However, myxomas can be found in atypical localizations, arising from the posterior or anterior left atrial wall or atrial appendage, in order of declining frequency. As seen in our case, the myxoma was attached between the right upper and lower pulmonary veins in the right wall of the left atrium.

As the mass originated from a place other than the septum in the left atrium, we were led to consider a clinical diagnosis other than myxoma. Thrombus was thought to be among the alternative diagnoses as well as other cardiac malign and benign masses (such as angiosarcoma, papillary fibroelastoma, and lipomas) and metastatic cardiac masses. Cardiac thrombus was not considered because the patient had no pathology concerning the mitral valve, atrial fibrillation, and the mass was not settled on the auricula. The former diagnoses were not ruled out because papillary fibroelastoma are mostly small masses and originate from the ventricular or aortic surface of the aortic valve, lipomas mostly originate in the interatrial septum or mitral or tricuspid valve, and angiosarcomas generally originate from the right atrium or pericardium [Sugeng 2004]. Moreover, as this could be a metastatic cardiac mass, thorax and abdominal MSCT was performed and no origin-causing metastasis was found. It was highly probable that the mass was an atypically localized myxoma. The diagnosis of the mass was confirmed by peroperative macroscopic appearance and was supported by the postoperative pathological analysis.

TTE can generally be used to determine the location, size, shape, attachment, and mobility of a tumor. A TEE approach is particularly helpful in detecting the site of insertion and morphologic features of atrium and ventricular myxomas.

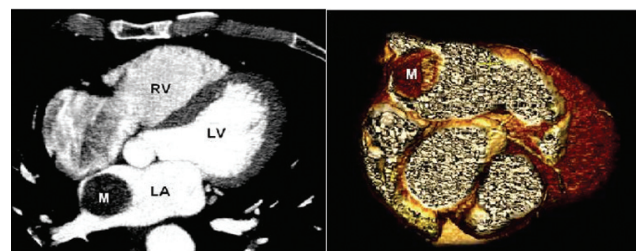


Figure 2. Multislice computed tomography view depicts a mass attached near the right upper pulmonary vein in the left atrium. M indicates myxoma; LA, left atrium; LV, left ventricle; RV, right ventricle.

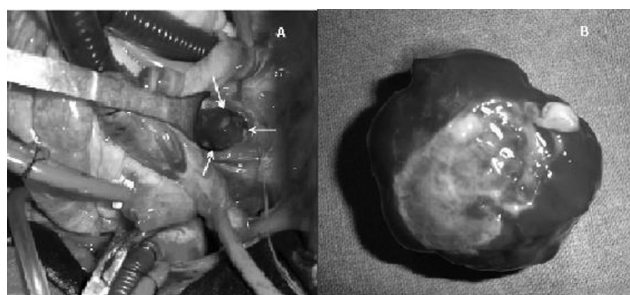


Figure 3. A, The picture obtained during surgery shows the myxoma in the left atrium free from the interatrial septum (arrows). B, The myxoma weighed 138 g and measured $5 \times 4 \times 2.5$ cm in size.

However, diagnosis can only be confirmed by histological examination of the excised tumor.

The triad of cardiac myxoma consists of obstruction of the intracardiac tract, embolism, and various constitutional manifestations. Furthermore, myxoma may cause valve obstruction, and, with future tumor growth, left ventricular outflow tract obstruction. A higher risk of embolization has also been reported and events occur in 30% to 43% of the patients. Constitutional manifestations of patients with myxoma are various and mimic those of an infection, collagen disease, or malignant disease [Chakfe 1997]. Our case revealed none of these, and laboratory findings did not include anemia or elevated C-reactive protein. The erythrocyte sedimentation rate and the level of gamma globulin, which sometimes increase, were not assessed in our case.

Cardiac myxomas may recur postoperatively, and the reason is unclear. Incomplete resection, intraoperative displacement, embolization, and multifocal genesis have been proposed as possible explanations [Chakfe 1997; Endo 1997; Terada 2000]. For these reasons, a total pedicle resection of the tumor stalk and its attachment is recommended. To avoid local recurrence from residual infiltrating myxoma tissue, we

made an extended resection through the left atrium right wall around the tumor.

Although cardiac myxomas can be diagnosed by echocardiography, atypical localization may make diagnosis difficult. As in our case, alternative diagnoses must be considered for atypically settled myxomas and the final diagnosis must be made in view of preoperative and postoperative results. As myxomas may recur and cause life-threatening complications, they must be immediately operated on, a large curative resection must be made to prevent recurrence, and patients must be routinely and periodically followed-up with echocardiography.

REFERENCES

- Chakfe N, Kretz JG, Valentin P, et al. 1997. Clinical presentation and treatment options for mitral valve myxoma. *Ann Thorac Surg* 64:872-7.
- Chitwood WR Jr. 1992. Clarence Crafoord and the first successful resection of a cardiac myxoma. *Ann Thorac Surg* 54:997-8.
- Darwazah AK, Hawari MH, Hussein IH, El-Hassan HK. 2006. Multiple left ventricular myxomas arising from the interventricular septum and ventricular trabeculae: a case report. *Heart Surg Forum* 9:E587-9.
- Endo A, Ohtahara A, Kinugawa T, et al. 1997. Characteristics of 161 patients with cardiac tumors diagnosed during 1993 and 1994 in Japan. *Am J Cardiol* 79:1708-11.
- Kamiya H, Yasuda T, Nagamine H, et al. 2001. Surgical treatment of primary cardiac tumors: 28 years' experience in Kanazawa University Hospital. *Jpn Circ J* 65:315-9.
- Keeling IM, Oberwalder P, Anelli-Monti M, et al. 2002. Cardiac myxomas: 24 years of experience in 49 patients. *Eur J Cardiothorac Surg* 22:971-7.
- MacGowan SW, Sidhu P, Aherne T, et al. 1993. Atrial myxoma: national incidence, diagnosis and surgical management. *Ir J Med Sci* 162:223-6.
- Sugeng L, Sahoo S, Lang RM. 2004. Atypical cardiac myxomas. *Echocardiography* 21:43-7.
- Terada Y, Wanibuchi Y, Noguchi M, Mitsui T. 2000. Metastatic atrial myxoma to the skin at 15 years after surgical resection. *Ann Thorac Surg* 69:283-4.