A Case of Left Atrial Myxoma Associated with Atrial Septal Defect

Feridun Kosar, M.D.,* Nevzat Erdil, M.D.,† Hakan Gullu, M.D.,* Ibrahim Sahin, M.D.,† Vedat Nisanoglu, M.D.,† and Bektas Battaloglu M.D.‡

**Department of Cardiology; †Cardiovascular Surgery; and ‡Internal Medicine, Inonu University, Faculty of Medicine, Turgut Ozal Medical Center, Malatya, Turkey*

Abstract **Cardiac myxoma is the most frequent primary tumor of the heart. However, it is rarely associated with congenital cardiac anomalies such as atrial septal defect in the literature. We present a 72-year-old woman referred to the emergency department with loss of consciousness and finally diagnosed as a pedinculated mobile left atrial myxoma and concomitant occurrence of an ostium secundum type atrial septal defect. The mass was successfully excised, and atrial septal defect was safely repaired by primary suture. The patient is currently well after surgery. Atrial myxoma should be considered in the differential diagnosis when patients present with neurological consequences of systemic embolization.** doi: 10.1111/j.1540-8191.2005.200469.x *(J Card Surg 2005;20:475-477)*

INTRODUCTION

Myxoma is the most common type of primary tumors of the heart.[1,2] It is usually benign in nature, and commonly occurs between the third and the sixth decade of life. Approximately 75% of myxomas originate from the left atrium, and more than 90% are solitary.[1,2] More importantly, myxomas are quite infrequently associated with congenital cardiac anomalies such as atrial septal defect.[3–5] Cardiac myxoma can produce a wide spectrum of systemic findings including systemic and pulmonary embolism, and intracardiac obstruction.[6–9] The neurological consequences of systemic embolization include transient ischemic attacks, stroke, seizures, and syncope.[9] Mobile and/or pedinculated left atrial myxomas may prolapse to various degrees into the mitral valve orifice, infrequently resulting in obstruction of AV blood flow and mitral regurgitation. Therefore, the signs and symptoms of myxomas often mimic those of mitral valve disease.

In this report, we describe a patient who had a pedinculated mobile myxoma in the left atrium associated with an ostium secundum type atrial septal defect.

CASE REPORT

A 72-year-old woman was evaluated in the emergency department for loss of consciousness. This was her first episode, and her detailed medical history revealed dyspnea on exercise and episodes of palpitation. On her physical examination, there was a grade 2-3/6 systolic ejection murmur that was most prominent at the left second intercostal space. An ECG recording showed normal sinusal rhythm and nonspecific ST-T wave changes in precordial derivations. Chest X-ray showed normal cardiothoracic ratio. Laboratory tests were nonspecific and nondiagnostic. Also, her cranial computed tomography revealed an ischemic necrotic zone in the brain. Since the patient complained of dyspnea and palpitation, and had a sign of cerebrovascular event, transthoracic echocardiography was immediately performed after hospitalization. Two-dimensional transthoracic echocardiogram showed that there was a pedinculated mobile mass in the left atrium (Fig. 1). The mass was lobulated and pedinculated in shape and it had been connected to the posterior wall of the left atrium thorough a pedinculum, and it was prolapsing into the mitral valve orifice reaching at the tips of the mitral valves without obstructing mitral valve orifice. Her clinical and echocardiography findings pointed out atrial myxoma. Transesophageal echocardiography clearly demonstrated a lobulated and pedinculated mobile nonhomogenous mass with a broad base of 15 mm attached to the posterior wall of the left atrium (Fig. 2A), and also revealed a left to right shunt thorough the atrial septal defect demonstrating an ostium secundum type atrial septal defect which was measured 12 mm in diameter (Fig. 2B). To exclude any underlying occult coronary artery disease, and to demonstrate the vascular supply to tumor, coronary angiography was performed prior to cardiac surgery. Coronary angiograms revealed that there was no atherosclerotic lesion including wall irregularities and no collateral vessels.

The mass was successfully excised from the left atrium, and atrial septal defect was closed with primary suture. Histologic examination of the mass confirmed a benign myxoma including a lax connective tissue with myxomatous cells together with endothelial cells, macrophages, and muscle cells. The patient made an uneventful recovery in the postoperative period, and he is currently well after surgery.

Address for correspondence: Nevzat Erdil, M.D., Inonu University, Faculty of Medicine, Department of Cardiovascular Surgery, 44315 Malatya, Turkey. Fax: +90 422 3410728; e-mail: nerdil@inonu.edu.tr

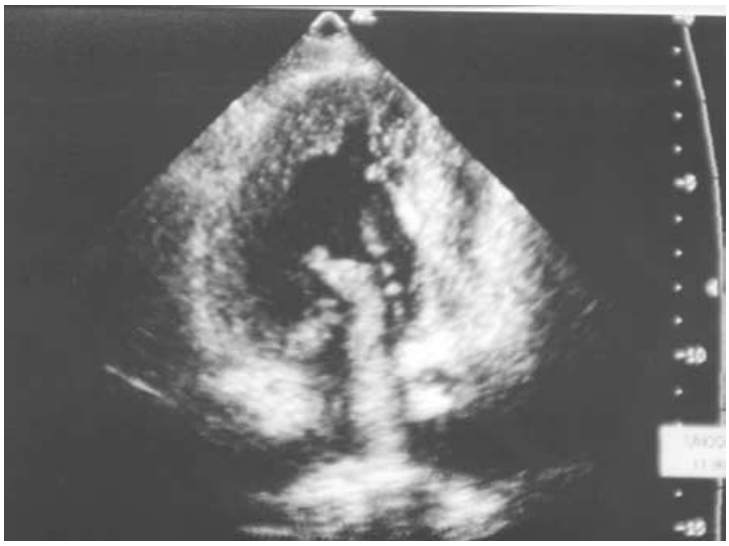

Figure 1. *Transthoracic echocardiography showing a mobile left atrial myxoma protruding towards the left ventricle.*

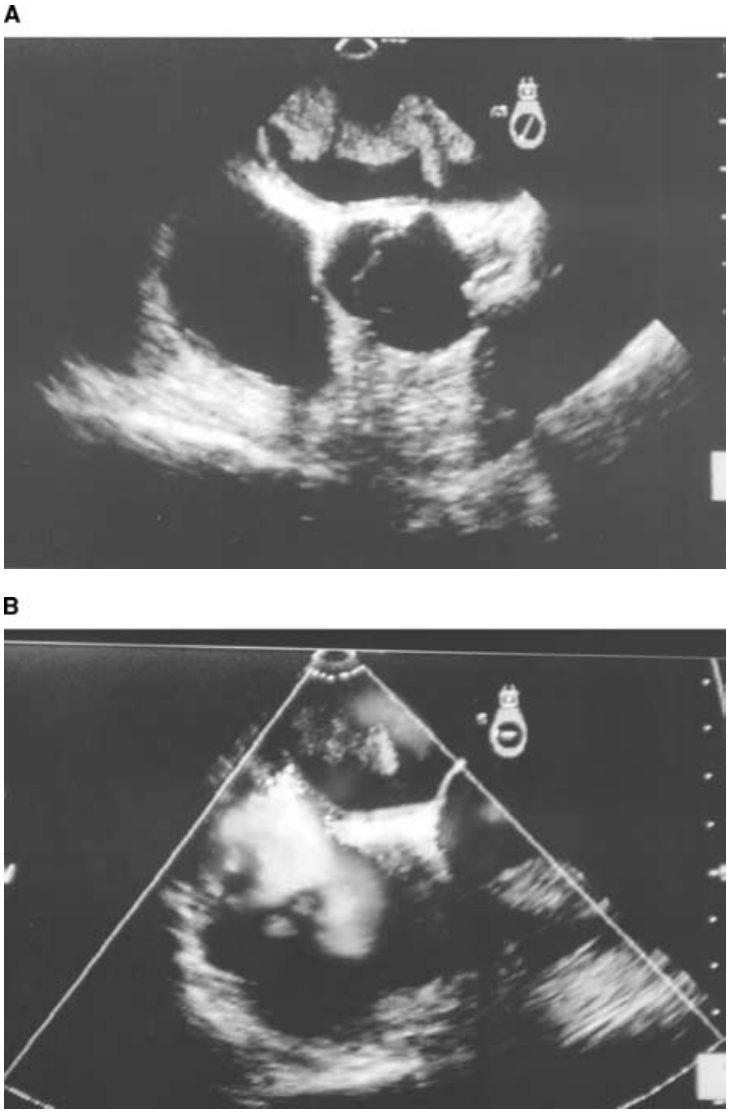

Figure 2. *Transesophageal echocardiography showing a pedinculated mobile nonhomogenous mass attached to the posterior wall of the left atrium (A) and a left to right shunt thorough ostium secundum atrial septal defect (B).*

DISCUSSION

Primary tumors of the heart are rare. Of these, myxomas are the most common one.[1,2] The high portion of myxomas occurs in the left atrium, usually begins interatrial septum (fossa ovalis) and grows into the atrium.[1,2] But, they can also originate from the posterior atrial wall, the anterior atrial wall or the atrial appendage. The mass may protrude through the mitral valve into the left ventricle and obstruct the left ventricular inflow in diastole. Our patient had a left atrial myxoma attached to the posterior wall of the left atrium, protruding towards the left ventricle without obstructing mitral valve orifice in diastole. Also, myxomas are very infrequently associated with congenital cardiac anomalies. Only in a few reports, myxoma has been reported with atrial septal defect.[3-5] We reported rare case of atrial myxoma with an secundum type atrial septal defect.

The majority of cardiac myxoma may produce the symptoms and signs of heart diseases. The clinical manifestations depend on their size and location; dyspnea is the most common manifestation in up to 80% of patients. The patients can also present with other cardiac symptoms such as chest pain, palpitation, and syncope.[6-9]

Cardiac myxoma may be a source of emboli to the central nervous system and elsewhere in the vascular tree. Embolization is one of the critical complications of myxoma, and occurs in about 30% to 40% of the patients. The site of embolism is dependent upon the location and the presence of an intracardiac shunt. In most cases, cardiac tumors have thrombi on their surfaces, and both thrombi and necrotic tumoral particles may cause systemic and pulmonary embolization. The neurological signs or symptoms may occasionally be the first or only clinical manifestation of a cardiac tumor as in our case.[9] An embolic stroke without evidence of cerebrovascular disease, particularly in the presence of sinus rhythm, should raise the suspicion of cardiac myxoma. Also, there appears a conflict regarding therapeutic approaches of embolic stroke; if thrombus causes embolization, thrombolysis may be useful, on the contrary, if tumoral particles lead to embolization, the patient would not benefit from thrombolytic therapy, and even would be vulnerable to the side effects of thrombolysis.

Two-dimensional transthoracic echocardiography is usually adequate for diagnosing cardiac masses and/or myxomas, and if necessary, transesophageal echocardiography can be used to determine the location, size, shape, point of attachment, and mobility characteristics of a myxoma. Additionally, other noninvasive diagnostic imaging methods including computed tomography and magnetic resonance imaging may be helpful in such cases. Coronary angiography is indicated to demonstrate the vascular supply to tumor and to exclude any underlying occult coronary artery disease.

Surgery for atrial myxoma is the treatment of choice, and helps in avoiding the occurrence of complications. After surgical removal of myxomas, patients usually remain symptom-free and have a normal lifespan.[10]

In conclusion, atrial myxoma should be considered the differential diagnosis of patients presenting with neurologic symptoms or with signs of embolization. And concomitant occurrence of atrial septal defect must be considered in patients with atrial myxoma.

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