Large mobile hemangioma arising from interatrial septum which mimicks mitral stenosis

Mitral stenozu taklit eden interatriyal septumdan kaynaklanan büyük mobil hemanjiyom

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Cardiac hemangioma is extremely rare and accounts for 2.8% of all detected benign heart neoplasms. In this article, we described the large hemangioma originating from interatrial septum and mimicking mitral stenosis. Opposite of myxoma, cardiac hemangioma mimicking mitral stenosis has not been previously reported in the English literature. The mass which was pathologically diagnosed as cavernous hemangioma was removed surgically. The postoperative course was uneventful. No recurrence of the tumor was noted at six months after operation.

Key words: Cardiac hemangioma; interatrial septum; mitral stenosis.

Cardiac hemangiomas are rare, benign cardiac tumors that occur at a rate of 2.8% when compared with other primary tumors of this type.^[1] Their origin is uncertain, but they are thought to be either true neoplasms or hamartomas. Cardiac hemangiomas are localized in the atria, the ventricles, the ventricular septum, and the pericardium. They are also found on very rare occasions in the mitral valve. Although most hemangiomas are asymptomatic, they can present with arrhythmias, conduction disturbances, pericardial effusion, coronary insufficiency, outflow tract obstruction, or congestive heart failure. Sudden cardiac death as a result of conduction disturbances or rupture and tamponade has been reported.^[2]

Diagnosis is usually made by echocardiography, computed tomography (CT), or magnetic resonance imaging (MRI). In adults, in contrast to the neonatal

Kardiyak hemanjiyom son derece nadir olup, tespit edilen tüm iyi huylu kalp tümörlerinin %2.8'ini oluşturur. Bu yazıda, interatriyal septumdan kaynaklanan ve mitral darlığı taklit eden büyük bir hemanjiyom olgusu sunuldu. Miksomanın aksine, mitral darlığı taklit eden kardiyak hemanjiyom daha önceden İngilizce literatürde bildirilmemiştir. Patolojik olarak kavernöz hemanjiyom tanısı konulan kitle cerrahi olarak çıkartıldı. Ameliyat sonrası seyir olaysızdı. Ameliyat sonrası altı aylık takipte tümör nüksü görülmedi.

Anahtar sözcükler: Kardiyak hemanjiom; interatrial septum; mitral darlık.

period and infancy, no alternative curative treatment to surgery has been reported for hemangiomas.^[3] Histologically, these can be classified as cavernous, capillary, and capillary-cavernous types.^[4]

To our knowledge, this is first case of a hemangioma originating from the interatrial septum which caused mitral valve stenosis. In this article, we report the case of a female with an intracavitary left atrial hemangioma mimicking mitral stenosis that was detected via transthoracic echocardiography (TTE) and confirmed by surgery and histology.

CASE REPORT

A 56-year-old female was hospitalized due to shortness of breath with palpitations on exertion New York Heart Association (NYHA) class III experienced over a one-month period. The patient's past medical



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history revealed no chronic illnesses, and her general appearance was normal. Auscultation of the heart revealed 2/4 diastolic rulman at the apex. The patient's blood pressure was 105/73 mmHg, and the pulse was 90/min and regular. No apparent abnormalities were found via electrocardiography or chest X-ray, and the laboratory test results were within the normal range. Transthoracic echocardiography showed a large (5.27x3.8 cm), mobile, spherical, well-defined, encapsulated dense mass with a homogenous echo arising from the interatrial septum, which was later revealed to be a smooth oval nodule with a pedicle that was attached to the interatrial septum. This mass protruded into the left ventricle and caused both incompetence and relative narrowing in the mitral valve during left ventricular diastole (Figure 1a). A 30 mmHg peak gradient along with a 15 mmHg mean gradient were present over the mitral valve. No mass was detected in any other cardiac chambers. Cardiac catheterization showed normal coronary and feeding blood vessels extending from the right coronary artery (RCA) toward the tumor, which had the characteristic sign of tumor blush that is common in hemangiomas.

The patient was taken to the operating room to remove the cardiac mass. The surgery was performed via a median sternotomy, and cardiopulmonary bypass (CPB) was established after placing an arterial cannula into the ascending aorta and bicaval venous cannulations. The ascending aorta was then clamped, and the heart was arrested with cold blood cardioplegia. The patient was cooled down to 30 °C, and the left and right atriums were then opened. An soft, smooth, elastic, reddish brown mass was found adhering to the interatrial septum from its pedicle which occupied more than half of the left atrium with a dimension of 6x5x4 cm (Figure 1b). The mass was resected from the rim of the septum, and the iatrogenic atrial septal defect was then repaired with a primary suture. The left atriotomy was then directly sutured. The postoperative clinical course was satisfactory, and the patient was discharged on the fifth postoperative day.

Macroscopically, the tumor was capsulated and consisted of a uniformly spongy structure filled with blood. A histopathological examination of the excised mass demonstrated high vascularity and vascular channels filled with red blood cells. There were also many large cystic spaces lined with endothelial cells, which was compatible with a cavernous hemangioma (Figure 1c). The tumor tested negative for malignancy.

DISCUSSION

Hemangiomas can present in any age group with a mild predominance in females. In a previous review of 56 cases of cardiac hemangiomas, 36% were found in the right ventricle, 34% in the left ventricle, 23% in the right atrium, and 7% on the interatrial septum and in the left atrium.^[5] Echocardiography is a frequently available imaging modality for detecting cardiac hemangiomas which is accurate and noninvasive.^[6] However, coronary angiography, CT, or MRI is recommended for selected patients with symptomatic coronary artery disease or in cases of unsatisfied echocardiography. In our patients, TTE was satisfactory to determine the characteristics of the mass and any hemodynamic changes, so no further evaluation was needed. At first, we thought the mass might be a myxoma because it originated from the interatrial septum, had a large volume, and was obstructing the mitral valve. We performed coronary angiography to assess whether there was possible concomitant coronary artery disease and to determine the vascular structure of the mass. A tumor blush on the coronary angiography suggested the mass was a hemangioma rather than a myxoma. The mass was removed at surgery and was identified as a cavernous hemangioma.



Figure 1. (a) Apical four-chamber view on echocardiography with the left atrial mass protruding into the left ventricle through the mitral valve. (b) Gross specimen of the tumor. (c) Microscopic view of the tumor demonstrating the proliferation of enlarged vessels filled with blood (H-E x 40). A myxoid background that is diagnostic of a cavernous hemangioma can be seen.

The symptomatology of tumors depends on their anatomic location and extension. Most cardiac hemangiomas are discovered incidentally, and they may cause dyspnea, palpitation, atypical chest pain, and arrhythmia. Shortness of breath and palpitation on mild exertion were present in our patient due to obstruction of left ventricular inflow due to mitral stenosis.^[2,7]

Our patient underwent surgery due to deterioration in hemodynamic status. When diagnosed, hemangiomas should be removed because of the possibility of rupture, tamponade, and sudden cardiac death. However, spontaneous tumor resolution during a two-year follow-up also has been reported. ^[8] In cases involving the removal of a hemangioma, follow-up is recommended to identify any recurrence of the tumor. The prognosis is usually satisfactory following simple resection as long as multiple lesions do not recur.^[3,7]

In conclusion, hemangiomas should be kept in mind as a possible diagnosis in rare cases in which a left atrial intracavitary mass originating from the interatrial septum obstructs the mitral valve.

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