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A Giant Left Atrial Myxoma Inducing Mitral Stenosis

Mitral Stenozu İndükleyen Dev Sol Atrial Miksoma

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Öz

Miksomalar kalbin iyi huylu tümörleridir, sıklıkla sol ve sağ atriumdan sırasıyla %75 ve %18 oranında köken alır. Boyutlarına ve tümörün lokalizasyonuna göre semptomlar geniş bir spektrum şeklinde görülebilir. Atrium çıkım yolunu ciddi şekilde tıkayabilir. Olgu sunumumuzda 63 yaşında kadın hastada mitral stenoz semptomları oluşturan, literatürdeki ikinci dev sol atrial mixomayı sunmayı amaçlamaktayız.

Anahtar Kelimeler: Kalp Tümörü, Miksoma, Sol Atrium

Abstract

Myxomas are considered benign cardiac tumors, most commonly originating in the left atrium (75%) and less frequently in the right atrium (18%). The presenting symptoms can vary widely depending on the size and location of the tumor, and they may seriously occlude the atrial outflow. We herein report a case involving a giant left atrial myxoma in a 63-year-old woman presenting with symptoms of mitral stenosis.

Keywords: Cardiac Tumor, Myxoma, Left Atrium

Introduction

Cardiac myxoma is a benign neoplasm and the most common primary cardiac tumor in adults. Cardiac myxomas have a wide range of manifestations, mostly obstructive, but can also cause embolic events in severe cases. We herein present a case involving a 63-year-old woman who presented with dyspnea. After an echocardiographic evaluation, she was diagnosed with a giant left atrial myxoma. She underwent successful tumor resection with a favorable postoperative outcome.

Case

A 63-year-old woman was admitted to our hospital with dyspnea and symptoms of New York Heart Association class III heart failure. A large mass in the left atrium was detected by transthoracic echocardiography (Figure 1A). The mass was attached via a pedicle originating from the interatrial septum. It was mobile and measured 7.0 × 5.5 cm in size. In addition, the mass exhibited mobility that allowed it to prolapse into the mitral valve orifice. The mass had a dense, solid characteristic. The left ventricular ejection fraction was 55%. Preoperative transesophageal echocardiography was performed, and the findings were similar to the previous echocardiogram. Because of a technical problem involving the imaging device, however, we were unable to obtain a picture. The patient provided informed consent and permission for publication of this report.

The patient underwent an operation. After careful dissection around the heart, cardiopulmonary bypass was established. Exposure was obtained through a superior septal approach. The tumor was found to be filling most of the left atrium and had a close relationship with the superior aspect of the fossa ovalis. Optimal resection of the interatrial septum along with a limited superior part of left atrial wall was performed to prevent tumor recurrence. The tumor was 7.0 × 5.5 cm in size (Figure 1B, 1C). Patch closure for septal repair was not required. The residual interatrial septum was adequate in size; therefore, primary closure of the defect was performed. The patient had a favorable postoperative outcome and was discharged with no complications. Histological evaluation confirmed an atrial myxoma.

Discussion

Myxoma is the most common primary cardiac tumor in adults. It is generally found in the left atrium, and its prevalence ranges from 0.001% to 0.3% (1). Myxomas generally exhibit benign characteristics. However, they can cause constitutional symptoms or serious complications due to thromboembolic events of tumor fragmentation or a thrombus that forms on the tumor surface (2–4). Although rare, death may occur secondary to heart failure or mechanical obstruction of cardiac valves, mimicking the signs and symptoms of valve disease (4). In our case, a giant left atrial myxoma mimicked mitral stenosis.

Myxomas can be found as a component of Carney complex (5). Carney complex is a familial autosomal dominant disorder characterized by the development of atypical myxomas in different cardiac chambers, such as the ventricles, or multiple myxomas in approximately 45% of cases. It can occur sporadic

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manner in some of cases. Tumors related to Carney complex can be detected in patients of young ages, and the recurrence rate is 15% to 22%. It is also referred to multiple endocrine neoplasia syndrome with distinctive pigmented lesions of skin and mucosal membranes. A wide spectrum of endocrine abnormalities can occur in Carney complex. Additional tumors include myxomas affecting the skin, schwannoma, primary pigmented nodular adrenocortical disease (PPNAD), pituitary adenomas, thyroid tumors, testicular tumors, and ovarian lesions. Skin pigment abnormalities include small flat brown spots (multiple lentigines) and small, bluish-black spots (blue nevi). The most common symptoms and clinical manifestation of Carney complex can vary in population. In many cases, Carney complex is due to mutations of the PRKAR1A gene (5). The myxoma in our case was not related to Carney complex.

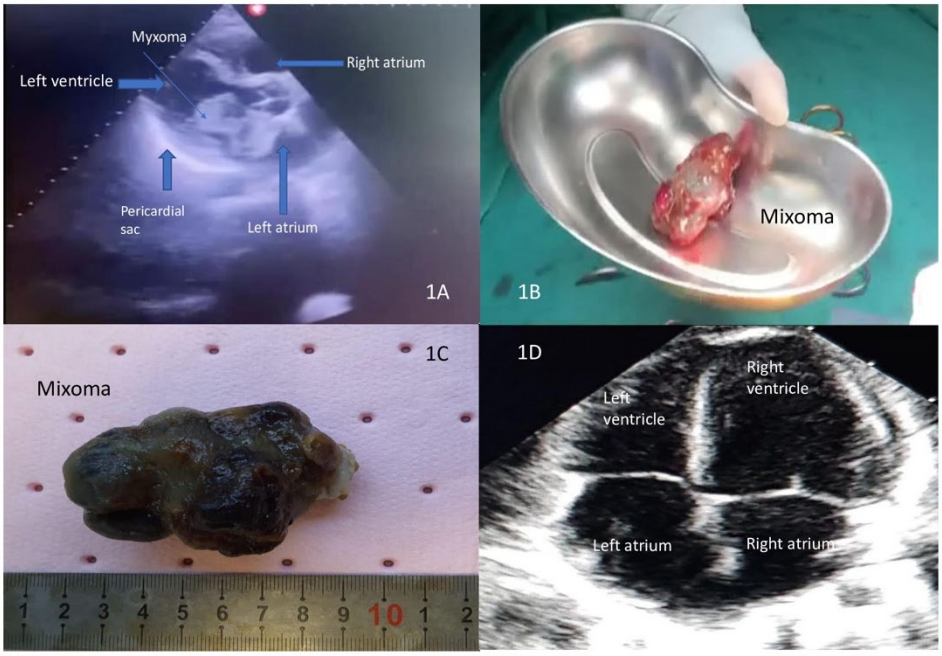

Figure 1. A. Preoperative view of giant left atrial myxoma in echocardiogram. **B.** Intraoperative view of excised mixoma. **C.** Macroscopic view and the size of giant myxoma. **D.** Postoperative view in echocardiogram.

Myxomas should be excised surgically. To obtain adequate exposure for large or multifocal tumors, a biatrial or superior septal incision is sometimes required (6,7). In addition to tumor resection, adjacent valve repair may be needed due to annular enlargement, a valve defect after tumor resection, or septal defect patch closure. No valvular structures required repair in the present case.

A myxoma may recur if the resection was not complete or the pedicle was not totally excised. Another important point is that before the aortic cross clamp is applied, manipulation of the heart should be avoided to prevent distal embolism due to tumor fragmentation or thrombus formation on the surface of the tumor (8). In our case, we also resected septal tissue around the pedicle along with part of the left atrial wall due to its adhesion to the tumor.

Many studies have recommended periodic echocardiography to detect any recurrence (8) (Figure 1D).

Conclusions

Our patient had developed only dyspnea and symptoms of mitral stenosis. Echocardiography revealed a giant left atrial myxoma mimicking mitral stenosis, and surgical resection was successfully performed.

Resection with adequate margins and careful intraoperative manipulation are crucial to prevent recurrence and reduce the risk of emboli.

Even when adequate septal resection has been performed, echocardiography is crucial for detecting recurrence during long-term postoperative follow-up.

We believe this case report represents one of the largest left atrial myxomas described in the literature to date.

Conflict of interest statement

The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

Written consent: Written informed consent was provided by the patient (07.06.2022).

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