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Case Report

Recurrent left atrial myxoma: A case report

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Abstract. Cardiac myxomas are the most common primary cardiac tumors. A 69-year-old female patient with a history of operated left atrial myxoma 3 years ago readmitted to the hospital for evaluation of progressive dyspnea and atypical chest pain. The echocardiographic study revealed a new tumorous mass at the same localization. The tumor was resected with the underlying heart tissue by transseptal approach. Histologic analysis confirmed a recurrent myxoma. Postoperative echocardiographic examination revealed no abnormalities. We present this case because of its recurrence and accompany with idiopathic thrombocytopenic purpura.

Key words: Left atrium, myxoma, recurrence

1. Introduction

Atrial myxoma is the most common benign tumor of the heart. They are most commonly found in the left atrium, attached to interatrial septum by a stalk. Patients who have atrial myxoma usually present with cardiac obstruction, arrhythmia, or peripheral embolization. In this case, we report an adult female patient, who presented with a recurrent left atrial myxoma.

2. Case report

A 69-year-old woman was admitted to our cardiology clinic with a history of dyspnea, backache and nonproductive cough for 3 years. Her functional capacity was II according to the New York Heart Association classification. She had a 15-year history of hypertension and a positive family history for coronary artery disease. Physical examination was unremarkable except for the irregular heart rate at 82 beats per minute. Multiple hyperpigmented skin spots (lentigo) were also noted (Figure 1). Electrocardiogram revealed atrial fibrillation with normal ventricular rate. The patient's hemoglobin level was 12.9 g/dL (normal range 12-14 gr/dL in women), her total white blood cell (WBC) count was $15x10^3/mm^3$ (normal range 4-$10.8x10^3/mm^3$), and her platelet count was $65x10^3/mm^3$(normal range: 150-$450x10^3/mm^3$). All other coagulation and biochemical analyses were normal. Chest radiography revealed moderate cardiomegaly. The computed tomography of thorax performed on admission has revealed a mass in the left atrium. Two dimensional echocardiogram demonstrated moderate mitral regurgitation, mild left ventricular concentric hypertrophy with normal systolic function (ejection fraction: 64%) and a big left atrial tumor attached to the left lateral atrial free wall (2.0x3.0x2.0 cm). Transesophageal echocardiography confirmed a large pedunculated mass in the left atrium. There was no history of peripheral or pulmonary embolization. Selective coronary angiography was performed before the operation and it revealed minimal coronary artery disease. On the operation, the mass was found to originate from

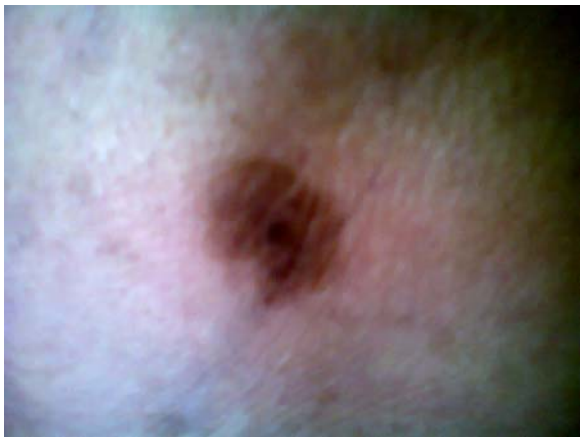

Fig. 1. The patient has brown, irregularly shaped lentigos on her face.

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the left lateral atrial free wall. The patient recovered well in the immediate postoperative period and discharged after one week. She was readmitted to our hospital three years after the initial operation with effort dyspnea and atypical chest pain. A complete blood count showed a low hemoglobin level of 11.7 gr/dl and hematocrit value of 33.6%. Likewise the platelet count was low ($38x10^3/mm^3$), WBC count was normal ($6.3x10^3/mm^3$). Serum cross-reacting protein and erythrocyte sedimentation rate were 5.2 mg/L and 10 mm/hr, respectively (normal ranges: 0.01-5 mg/L; 0.01-30 mm/hr, respectively). The echocardiographic study demonstrated a new tumorous mass at the same place (Figure 2).

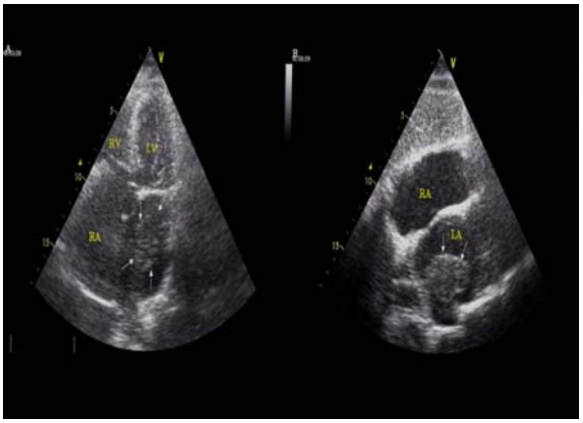

Fig. 2. Two dimensional echocardiography images revealed a left atrial myxoma attached to the left lateral atrial free wall (marked by arrow, 1-A, apical four chamber view; 1-B, subcostal view) LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

A preoperative hematology consultation was requested. The peripheral blood smear showed that the morphology of red blood cells and leukocytes was normal. Also, morphology of platelets was typically normal, with varying numbers of large platelets. It was determined that there were no blood abnormalities rather than a low platelet count. Abdominal ultrasonography revealed that normal liver and spleen size. The patient was diagnosed with chronic idiopathic thrombocytopenic purpura (ITP) and was recommended to keep the platelet count over $70x10^3/mm^3$ for a safe operation. Intravenous immunoglobulin (400 mg/kg/day) was started. We also performed a coronary angiography which revealed minimal atherosclerotic plaque progression without any significant stenosis. The tumor was resected with the underlying heart tissue by the transseptal approach and measured 6.0x4.0x3.0 cm, was slightly pedunculated, glistening, pinkish red, and of a firm, gelatinous consistency (Figure 3). The operative and postoperative courses were uneventful. The histology confirmed an identical myxoma as the primary mass. Postoperative echocardiographic examination showed no abnormalities. She was discharged without any complications. The patient is currently on follow up with medical treatment.

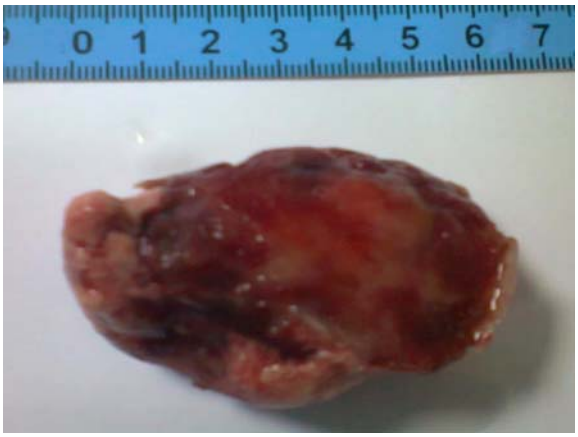

Fig. 3. Postoperative finding of the resected myxoma.

3. Discussion

Myxomas are mesenchymal tumors, which can occur at any age; however, they mainly exist between the third and sixth decade of life. They are histologically benign tumors but they may be lethal because of their strategic position. This disease is approximately two to three times more prevalent in women than in men (1-3). They most frequently present with signs and symptoms of mitral valve disease or with embolic phenomena (1). Recurrent myxoma is reported in sporadic (4%-7%) and familial cases (10%-21%) with the interval between the formation of the new tumor of more than 4 years (4).

Myxoma with thrombocytopenia has been rarely described in the literature (1). In accordance with the literature, the patient in this case had thrombocytopenia. The mechanism by which intracardiac tumor leads to thrombocytopenia remains unclear, although it has been postulated that abnormal mechanical shear stress, caused by tumor-induced flow obstruction, may be responsible for the increase in the breakdown of platelets. To clarify this issue, genetic and hematological studies are needed.

The Carney complex is a rare, familial autosomal dominant syndrome characterized by spotty pigmentation of the skin, endocrine

dysfunction, myxomas, schwannomas, pituitary adenomas, thyroid tumors, testis tumors, ovarian tumors, and breast tumors. Not all patients with Carney complex develop cardiac myxomas, but affected individuals usually have at least two components of the complex (5). The patient in this case has recurrent cardiac myxoma and presented with multiple lentigos on her face. The diagnosis of carney complex usually relies on clinical diagnostic criteria. Mutations are usually detected in the PRKAR1A gene. This coding region, available on a clinical basis, has a mutation detection rate of approximately 60% (6). The genetic analysis was suggested but the patient refused this procedure due to socio-economic reasons. Therefore, genetic and endocrinological examinations were planned after the discharge.

Early diagnosis and surgical treatment can lead to rapid resolution of symptoms with avoidance of probable complications like peripheral embolism and valve dysfunction. Excision of atrial myxomas using cardiopulmonary bypass has been established with generally good clinical results. However, one must use an individualized approach based on the location of tumors to control possible embolization. Transseptal approach was preferred in the present case since it allows total resection of the left atrial myxoma (7). Multifocal growth of a benign myxoma or malignant transformation, inadequate resection, intraoperative implantation or embolization play an important role in development of recurrent myxoma. Familial disposition may also play a role in recurrent development. The abnormal DNA ploidy pattern of myxoma patients showed a high recurrence (8).

Recurrence of the myxoma in this case could be due to inadequate resection. Although recurrence of myxoma is rare, in our case it points to an invasive nature of the tumor. We feel, therefore, that excision of the underlying atrial septum or wall is justified in every case. Such a procedure will be more accurate in keeping with the surgical principles of tumor surgery without adding greatly to the technical difficulties of the operation. The defect created in the septum or the atrial wall can be easily closed primarily or with a Dacron patch to restore normal anatomical and physiological function. In follow-up examinations as well, two-dimensional echocardiography is essential. As a result, clinicians should be aware of the clinical symptoms of the myxomas and its complications. We think that a long-term follow-up by echocardiography in all patients after myxoma resection should be advised for an early detection of any recurrence.

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