LEFT ATRIAL MYXOMA WITH ATYPICAL CLINICAL PRESENTATION

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ABSTRACT

Myxoma is the most commonly seen primary cardiac tumour, with an incidence of 80-90% in the left atrium, followed by the right atrium and ventricle. Patients with atrial myxomas present with intracardiac obstruction, embolization to the pulmonary and systemic circulation, or constitutional symptoms. Hepatomegaly and increase of the liver transaminases levels' involvement in myxomas are rare, has been described to cause hepatic insufficiency.

We describe an interesting case of a 47-year-old patient with systemic findings and symptoms, which was primary symptomatically treated for several months in psychiatry, neurology and chest diseases clinics in other centres but never diagnosed, and undergoing emergency surgery in our clinic following determination of left atrial myxoma at tests performed by our cardiology department. The patient had no symptoms at examinations performed 6 months and 1 year postoperatively, and no pathological findings were determined.

Key words: Left atrial myxoma, emergency surgery, cardiac tumors.

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Introduction

Myxoma is the most commonly seen primary cardiac tumour in adults; 80-90% appear in the left atrium, followed, in order, by the right atrium and ventricle. Right atrial myxomas are more solid compared to left ones and tend to have a wider base. Transthoracic echocardiography is approximately 95% sensitive for the detection of cardiac myxomas, and transesophageal one approaches 100% sensitivity. Though the majority of atrial myxomas are sporadic, it is imperative that first-degree relatives of patients with documented myxomas undergo screening for occult myxomas. Myxoma frequently produces symptoms associated with cardiac obstruction and thromboembolism. Unless they are not familial, myxomas generally consist of a single lesion. They are more common in women than men, and are often seen in women aged between 30 and 601–3.

Case

A 47-year-old woman presented to our hospital with respiratory difficulty, palpitation and confusion persisting for 3 months and much worsened in the previous 10 days. Symptomatic therapy (sertraline, betahistine dihydrochloride, ipratropium bromide and salbutamol) had been started in psychiatry, neurology and chest diseases clinics in other centres, but no myxoma diagnosis had been established at that time, due to myxoma’s enlargement, respiratory difficulty, palpitation and confusion get more and more symptomatic. Myxoma was subsequently diagnosed by our cardiology department, and the patient was assessed by our clinic for emergency surgery when her confusion worsened. At physical examination, the heart was rhythmic and pretibial oedema was present. Myxoma filling the entire left atrium was determined at echocardiography (Figures 1-3), (Table 1).
No previously determined coronary disorder was present in her history. For 5 years, she had been hospitalized and treated with a diagnosis of depression in various centres. There was no known disorder in the family history, apart from chronic obstructive pulmonary disease. Coronary angiography revealed normal coronary arteries. Transesophageal echocardiography was not performed due to non-compliance. Hepatomegaly was observed at abdominal ultrasonography. No pathology was detected at other system examinations. At laboratory examinations, ALT was 714 U/L (N:<32) and AST 567 U/L (N:<33). A median sternotomy approach was adopted at surgery, and an easily friable 4x3x2 cm mass with a myxoid appearance was excised from the left atrium (Figure 4).

<table>
<thead>
<tr>
<th>Echocardiographic Data</th>
<th>Patient</th>
<th>Normal Values</th>
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<tbody>
<tr>
<td>Left ventricle end diastolic diameter</td>
<td>50 mm</td>
<td>37-55mm</td>
</tr>
<tr>
<td>Left ventricle end systolic diameter</td>
<td>40 mm</td>
<td>20-40mm</td>
</tr>
<tr>
<td>Left atrium systolic diameter</td>
<td>38 mm</td>
<td>20-40mm</td>
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<tr>
<td>Right ventricle diastolic diameter</td>
<td>58 mm</td>
<td>18-25 mm</td>
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<tr>
<td>Right atrium diastolic diameter</td>
<td>52 mm</td>
<td>20-32 mm</td>
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<tr>
<td>Ejection Fraction %</td>
<td>50</td>
<td>&gt;50-55</td>
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<tr>
<td>Pulmonary artery pressure</td>
<td>90mmHg</td>
<td>15-30 mmHg</td>
</tr>
</tbody>
</table>

Table 1: Echocardiographic data of the patient compared with normal values.

No problems arose with cardiopulmonary bypass. At laboratory examinations on the 2nd day postoperatively, ALT was 77 U/L and AST 37 U/L (N:<33). Laboratory values were within normal limits on the 5th day postoperatively. The preoperative symptoms had resolved on the 6th day postoperatively, and the patient was discharged. Benign myxoma with polygonal fusiform and star-shaped cells on a dense myxoid base forming papillary structures was determined from the material sent for pathology analysis. Immune staining was observed at immunohistochemical examination with vimentin, SMA, CD 34 and desmin, while no immune staining was reported with S 100.
The patient had no symptoms at follow-up after 6 months and 1 year postoperatively, and no pathological findings were determined.

Discussion

Size of myxoma is associated with the symptoms it causes. Myxomas of small diameter have irregular structures, and embolization-related findings are more common. Neurological symptoms in patients with low general condition who could not be operated have been reported to decrease with thrombolytic therapy. The first imaging technique in diagnosis of myxoma is often transthoracic echocardiography, although transesophageal one is of greater value in the analysis of intracardiac structures, identifying thrombus, assessment of intratral septum and determining mass character.

Our patient presented with respiratory difficulty, palpitation and confusion. Anamnesis revealed admissions to various psychiatry and neurology clinics over the previous 3 months. Hepatomegaly was detected at abdominal ultrasound, elevated pre-operative ALT and AST values were determined, and diagnosis was made with transthoracic echocardiography. Although there are generally normal and non-specific ECG findings in myxoma cases, atrial fibrillation, conduction disorder and branch block can be seen. Cardiomegaly leading to obstruction can be proved radiologically. There were no pathological ECG or radiological findings in our case.

Surgical removal of the myxoma is the treatment of choice and usually curative; however, myxoma recurrence does occur and is most frequently associated with a familial syndrome. The myxoma patients can get symptomatic treatment followed till the patients go surgery. Treatment of myxoma consists of surgical excision of the entire mass together with surrounding healthy tissue. Heart manipulation must be performed with care to avoid tumour embolization, and the tumour must be removed without fragmentation. All heart chambers and valves must be carefully inspected. Myxomas resolve with complete healing in the absence of traumatization and inoculation. Recurrence is not observed in these patients at long-term follow-up.

In our case, extracorporeal circulation was applied after sternotomy, the tumour was excised from the left atrium, the chambers were checked and irrigation was performed, with primary closure of the defect. The patient’s confusion resolved after the operation and ALT and AST values returned to normal 5 days after surgery.

Postoperative recurrence in myxomas is known to be seen at the 6th month at the earliest and at the 11th year at the latest, with a mean figure of 30 months. Follow-up with echocardiography should be performed at 6-monthly intervals. No recurrence was determined in our case at echocardiographic examination at follow-ups over 14 months. Recurrence is common in cases with distribution in several chambers, early distant metastasis, atypical origin and a familial history, and follow-up after surgery is of vital importance.

In conclusion, atrial myxomas may exhibit a wide range of clinical findings, and symptomatic therapy without diagnosis may be tried in different clinics. Diagnosis of myxoma must not be overlooked in such patients. Symptomatic therapy may be given in situations in which surgical mortality is high. Extraction of the tumour and pedicle together with surrounding tissue without loss of time once myxoma has been diagnosed increases surgical success.

References


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