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Case report

A giant atrial myxoma with fairly atypical features

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ABSTRACT

Atrial myxomas are the most common primary heart tumors. Because of nonspecific symptoms, early diagnosis may be a challenge. Most of these are attached to the inter-atrial septum and are mostly asymptomatic but eventually can cause symptoms depending on the size, mobility, and location of the tumor. We report a case of a large myxoma in the right atrium, which is an uncommon location for this type of tumor.

Case report

A 36-year-old man presented with atypical chest pain and palpitations for the last a month. On examination vital signs were as follows: blood pressure 110/70 mmHg, pulse 110 beats/min, and oxygen saturation 95% on room air, respiratory rate 18 breaths/min. The cardiovascular examinations revealed regular but tachycardic heartbeat with a diastolic murmur of grade 1–2/6 and also jugular venous congestion. The examination of other systems was normal. The ECG detected sinus tachycardia. A chest X-ray showed clear lung fields with cardiomegaly. Complete blood count and basic biochemical parameters were normal. Sedimentation rate was 11 mm/h, CRP was 3 mg/l. Transthoracic echocardiography revealed a mass almost completely fill the right atrium prolapsing through the tricuspid valve into right ventricle (10 × 5 cm diameter) (Fig. 1). The left ventricular size and function were normal in echocardiographic examination. Transesophageal echocardiography confirmed this well-circumscribed mass extending from the right atrium to the right ventricle which caused tricuspid regurgitation and stenosis. Transesophageal images could not be recorded due to technical reasons. The patient was referred to cardiovascular surgery. Right atriotomy was applied (Fig. 2). The tumor, originating from the base of the right atrium, was excised from sufficient depth completely (Fig. 3). Resected area was powered with pericardial patch after extraction process. The result of the pathological examination of tumor was compatible with a myxoma of dimensions 7.5 × 4.5 × 4.5 cm. The patient was discharged one week after the surgery without complaint.

Discussion

This clinical case is quite unusual, considering the massive dimensions of the myxoma, the uncommon localization in the right atrium, the rare clinical presentation as atypical chest pain and palpitation, the sex of the patient and the discrepancy between transthoracic echocardiogram findings and poor clinical presentation.

The clinical manifestations usually depend on the anatomic position and size of the mass. There are mainly 3 types of presentations: embolic, obstructive, and constitutional. Embolic manifestations include visceral infarctions, stroke, and myocardial infarction. In a related study evaluating the risk factors for embolism in cardiac myxoma, location of the myxoma and irregular tumor surface were independently associated with increased risk of embolic complications.[1]

Obstructive manifestations are usually mistaken for valvular stenosis. For instance, mitral valve obstruction due to left atrial myxoma presents with symptoms such as syncope and dyspnea, mimicking mitral valve stenosis. Although right-sided myxomas are rare, the signs and symptoms of RA myxomas are highly variable, to rely on the size, location, and mobility of the tumor.[2] Right atrial myxomas may remain asymptomatic or cause variable signs and symptoms, for example fever, weight loss, arthralgia, Raynaud's phenomenon, anemia, hypergammaglobulinaemia and elevated erythrocyte sedimentation rate.[2] The symptoms were gone after the tumor is removed. Our patient had fever, arthralgia, and anemia. The most prevent symptoms is dyspnea (in 80% of patients). Patients may present with right heart failure secondary to right ventricular outflow tract obstruction, or with syncope secondary to temporary complete obstruction of the tricuspid valve.[3]

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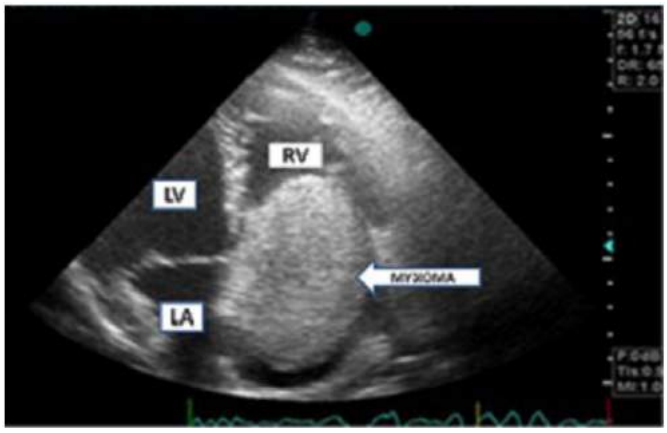

Fig. 1. Transthoracic echocardiography revealed a mass almost completely fill the right atrium prolapsing through the tricuspid valve into right ventricle.

Myxomas are the most common cardiac tumors. Myxomas arise in the left atrium in 75% to 80% of cases but they can also originate from other locations, such as the right atrium (RA) (18% of cases).[4] RA myxomas usually are located from the interatrial septum at the border of the fossa ovalis,[5] but in this case it was originated from the base of the right atrium.

Chest X-rays and electrocardiograms can be non-specific.[6] Transthoracic echocardiography shows an excellent sensitivity in detecting 95% of this unusually situation. On the other hand the sensitivity rises to 100% when transesophageal echocardiography does.[7] Thus echocardiography is the gold standard diagnostic technique. Computed tomography (CT) and magnetic resonance imaging may be useful to show in detail the point of fixation and associated complications. In our patient, an echocardiogram suggested the hypothesis of RA myxoma, which was confirmed by a histopathological exam.

Because of the risk of thromboembolic events, primary treatment is surgical removal of myxoma.[8] but complete resection of tumors is the most important factor. The survival rate after surgery is elevated. The recurrence rate of sporadic tumors is between 1% and 3%.[7] In our case, 6 months and one year after surgery echocardiographic examination showed no evidence of recurrence.

Conclusion

In summary, right atrial myxomas are rare and may have atypical presentation.

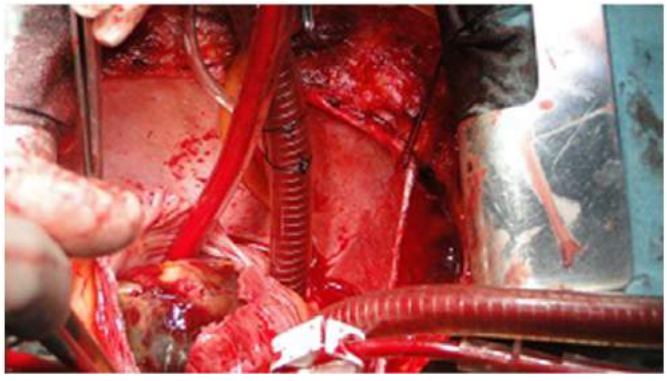

Fig. 2. Macroscopic appearance of the tumor after right atriotomy.

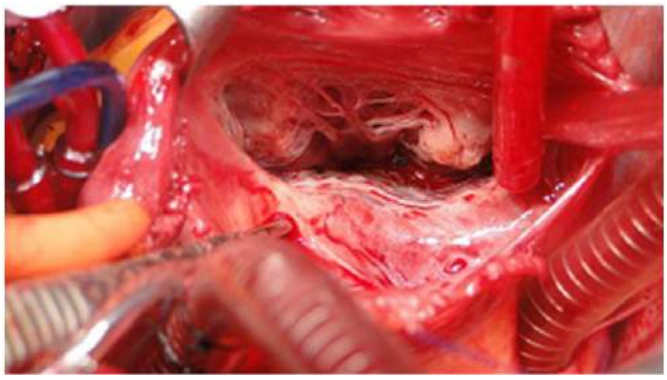

Fig. 3. The view of the right atrium and tricuspid valve after removing the tumor.

Heart tumors should be considered in differential diagnosis of atypical chest pain and palpitation and echocardiography should be performed without delay especially in adult patients with these symptoms. Echocardiography remains the best diagnostic method for locating and assessing the extent of myxomas and for detecting their recurrence, with a sensitivity of up to 100%.

Conflict of interest

The authors declare that they have no conflict of interest.

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