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Case Report

An Atypical Giant Right Atrial Myxoma Presented with Minimal Symptoms

Abstract

Myxomas are the most common primary cardiac tumors, rarely found in the right atrium. Myxomas usually seen in women, typically between the ages of 40 and 60. We present a case of giant right atrial myxoma that prolapsing through the tricuspid valve into the right ventricle, in a 37-year-old man with constitutional symptoms. There was a discrepancy between atrial mass and intensity of symptoms. Our patient underwent surgical treatment with excision.

Introduction

Primary tumours of the heart are not common and the prevalence of cardiac tumors ranges from 0,001% to 0,3% at autopsy [1]. Over 70% of primary cardiac tumors are benign and the most common form of these primary tumours are myxomas. Most of the myxomas are located in the left atrium (75–80), arising from the interatrial septum at the border of the fossa ovalis [2]. Larger tumors are more likely to be associated with cardiovascular symptoms [3]. Commonly observed symptoms and signs are dyspnea, pulmonary edema, cough, peripheral edema and fatigue. Constitutional symptoms (fever, weight loss) are seen in around 30% of patients. Laboratory abnormalities (anemia and elevations in the erythrocyte sedimentation rate, C-reactive protein) are present in 35 % of patients [4].

This case report show us discrepancy between giant right atrial myxoma and atypical symptom of patient. Once a cardiac myxoma is diagnosed, surgical excision should be performed without delays because of the risk of thromboembolic events [5-7], syncope and sudden cardiac death. Generally, surgical treatment is definitive and recurrence is uncommon.

Case Presentation

We report a 37-year old male who came to hospital with cough, malaise, fewer and fatigue. There was no history of chest pain, palpitations, syncope, hypertension, diabetes mellitus, alcohol intake, cigarette smoking. Vital signs were stable with a blood pressure of 115/75 mmHg, pulse rate of 98 bpm and temperature of 37.0 °C. On the cardiovascular physical exam, there was a grade 2/6 holosystolic murmur at the left parasternal region. Pulses were normal in lower extremities with no edema. On the laboratory findings; the haemoglobin was 9,3 g/dl, hct 28 %. An elevated CRP level (24 mg/l) and erythrocyte sedimentation rate (ESR: 53) were reported in his blood test; otherwise, blood tests were normal. An electrocardiogram showed sinus rhythm. He had normal cardiac size with clear lung fields on chest X-ray. Transthoracic echocardiography (TTE) showed

mild-moderate tricuspid regurgitation and a large echo-dense mass in the right atrium, attached to lateral free wall, filling the whole atrial chamber with prolapsing through the tricuspid valve into the right ventricle during diastole (Figure 1 and Movie Clip).

Our patient underwent median sternotomy. Cardiopulmonary bypass (CPB) was established between two venous cannula (superior and inferior vena cava) and aortic cannula. The right atrium was opened through an incision (Figure 2). The tumor was excised en bloc with safe margin of normal atrial wall at its attachment. Macroscopically, the tumor presented as a 7,4x4,7 cm, soft and lobulated with capsule (Figure 3). The pathologic report of the tumor was consistent with myxoma. On the histologic specimen, myxoma cells exist as elongate, stellate in a myxoid stroma with vacuolated cytoplasm and colorless matrix (Figure 4). Post-operatively, TTE showed no intracardiac mass and minimal tricuspid regurgitation.

Discussion

Myxoma is the most prevalent primary cardiac tumor and the right atrium is an unusual location. Myxomas can present at any age group but occurs more often between the 4th and 6th decades of life. The signs and symptoms of the right atrial myxoma are atypical

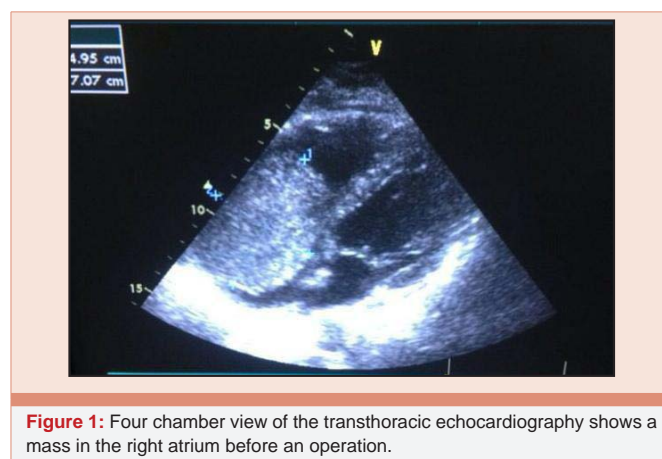
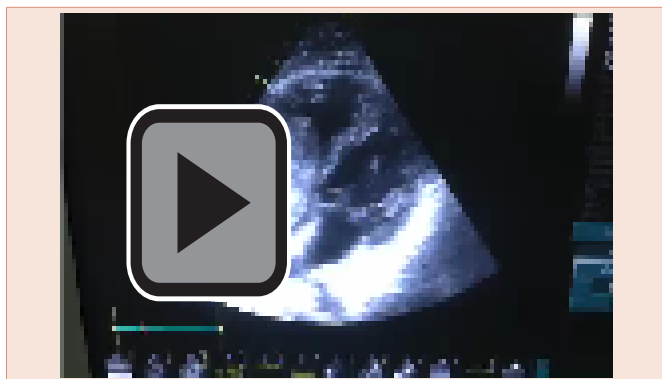


Figure 1: Four chamber view of the transthoracic echocardiography shows a mass in the right atrium before an operation.

and highly variable, depending on the position, size, mobility of the tumor, physical activity and body position of the patient. The most common symptoms of right atrial myxoma have been reported to be those of congestive heart failure, while other symptoms constitutional, thromboembolic and obstructive [8]. Impaired left ventricular function without obstructive coronary artery disease rarely reported in literature [9]. Our patient presented with constitutional symptoms of malaise, fatigue and rising of CRP, ESR levels. Constitutional symptoms in patients with cardiac myxomas have been attributed to releasing of the cytokin, especially interleukin-6 which causes inflammatory and autoimmune manifestations. The symptoms disappear after the tumor is removed. Right atrial myxomas usually



Movie Clip:

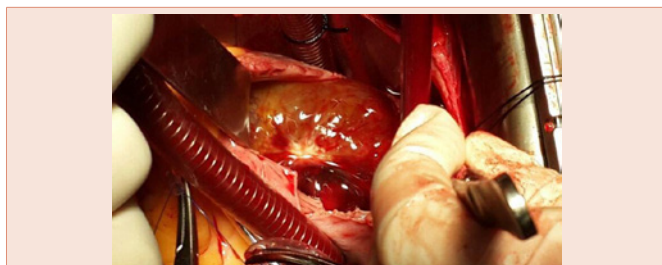


Figure 2: Intra-operative photograph of the tumor showing the right atrium with lobulated mass.

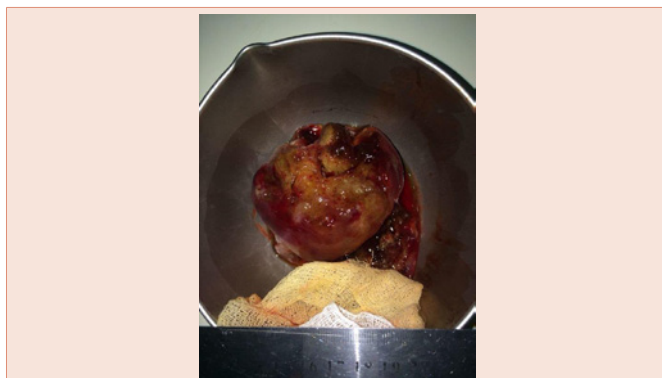


Figure 3: A lobulated mass with an irregular surface and with capsule.

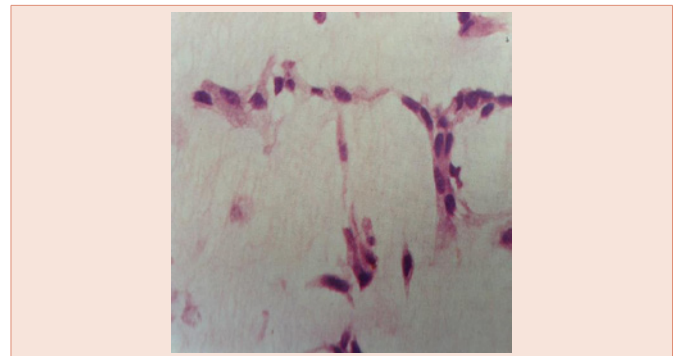


Figure 4: Microscopic section of the mass showing the myxoid stroma, elongate cells, vacuolated cytoplasm with colorless matrix (hematoxylin and eosin, X 300).

originate in the fossa ovalis or base of the interatrial septum. But in our case, mass was originate from lateral free wall of the right atrium.

Diagnosis of all patients was done by left heart catheterization before 1980. Nowadays, transthoracic echocardiography is the first diagnostic method, particularly transesophageal echocardiography became the preferred methods for the accurate and reliable diagnosis with 100 % sensitivity [1,10]. Computerized tomography and magnetic resonance image are the other diagnostic methods [11,12]. With this diagnostic modalities, sectional views of thoracic and pulmonary structures can be obtained and the tumor size, localization, attachment point can be visible accurately [12]. Also MRI and CT can detect small tumors (<1cm) and additional information about right atrial myxoma that extend into vena caval or tricuspid valve orifice.

During surgical treatment, venous cannulation problems can occur for right atrial myxomas. While superior and inferior venae cavae were cannulated, before aortic cross-clamp, the heart should be manipulated minimally, in order to prevent embolization. Superior cava cannulation can be done close to the innominate vein, for avoiding right atrial manipulation and embolization.

The treatment choice for myxomas is surgical removal. When the atrial myxoma diagnosis is confirmed, urgent surgery is necessary to prevent mortal complications such as embolic complications and sudden death. Even, mortality can be as high as 8% in asymptomatic patients in some series [13]. Complete resection of the tumor and its implantation base with a good safety margin is essential to cure the disease. The survival rate after surgery is elevated [14-16].

Conclusion

In this case report, we emphasize the rarity of giant right atrial myxoma in young patient and the discrepancy between myxoma size, trans thoracic echocardiogram findings and symptoms. And also cannulation can be done with carefully for avoiding embolization complications.

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