



Diagnostic Perplexity: Right Atrial Mass

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Introduction

The differential diagnoses of intracardiac masses include vegetation, thrombus or tumors. Presenting clinical features along with its characteristics; size, shape, location, mobility and attachment of the mass, help differentiate etiology. Echocardiography is the gold standard test for the diagnosis of intracardiac masses and transesophageal echocardiography (TEE) has further improved its diagnostic accuracy.

The prevalence of primary cardiac tumors is approximately 0.02%. About 75% of primary tumors are benign, and 50% of benign tumors are myxomas. Approximately 75% of sporadic myxomas occur in females. In a retrospective review of 171 patients from India, the mean age of presentation was 37.1 years.^[1]

Most of the cardiac myxoma occurs on the left side of heart in particular in the left atrium. Although myxomas are the most frequent primary tumors, right atrial is an atypical location leading to a diagnostic dilemma^[2-4]. Differential diagnosis with malignant tumors, metastasis and vegetations must be taken into account.

Case Presentation

We present a case of 42-year old woman who presented to us with history of dyspnoea on

exertion which had worsened gradually from NYHA II to grade III over a period of two years. During the last two months, she also had five episodes of unprovoked syncopal attacks not associated with any seizures. She reported history of paroxysmal palpitations since last two month and increasing difficulty in breathing on lying down. She reported a gradual loss of appetite and generalized weakness. She has three children and gave no history of oral contraceptive use. Prior to this, she had never needed hospitalization or blood transfusions. Family history was unremarkable.

Chest examination revealed a normal first heart sound and a widely-split second heart sound with a soft systolic ejection murmur at the upper right sternal border. Normal breath sounds were heard. Abdominal examination showed no hepatomegaly. The rest of the physical examination was unremarkable. The 12-lead electrocardiogram demonstrated sinus rhythm of 92 bpm with a right axis deviation.

Transthoracic echocardiography revealed a large (65 x 42 mm), soft, hyperechoic mass present in an enlarged right atrium, collapsing in the right ventricle through the tricuspid valve on four-chambered view. Right atrial and right ventricular enlargement with dilated IVC was noted on the subcostal view. She underwent cardiac

catheterization and coronary artery disease was ruled out.

Given the patient's significant symptoms and the large size of the mass, with possible risk of stroke, surgical resection was planned. Intra-operatively, transesophageal echocardiography confirmed the findings. Midesophageal bicaval view showed a large (67x48 mm) right atrial mass with irregular borders and heterogenous echogenicity; it did not appear to have any clear attachment to the right atrial wall. The IVC was clear and non-collapsing. The patient underwent median sternotomy under general anesthesia. Cardiopulmonary bypass (CPB) was instituted with conventional mild hypothermia (34.0°C). Cardiomegaly due to right atrial and right ventricular enlargement was observed. Under a single aortic cross-clamp for 15 minutes, the mass was completely excised through a right longitudinal atriotomy. The mass was mobile, lobulated, and necrotic appearing with attachment to the right atrial roof near the commissure of the anterior and septal leaflet of the tricuspid valve via a one cm stalk. There was no further invasion into any other structures. No tricuspid regurgitation was noted after removal. The resected mass was sent for histological assessment. CPB was discontinued without any difficulty and she was transferred to the intensive care unit (ICU) in good hemodynamic condition.

The histopathological report was consistent with the sections showing paucicellular myxoid material containing polygonal and spindle cells with eosinophilic myxoid cytoplasm, bland nuclei, inconspicuous nucleoli and focal multinucleation (myxoma cells). There was no nuclear atypia. The features were consistent with myxoma.

She showed full clinical recovery during this period and was discharged in good clinical condition. Repeat TTE after one month has not revealed any right atrial mass.

Discussion

Intracardiac primary tumors are rare and highly variable in their clinical expression. They occur most often between the 3rd and 6th decades of

life^[9] Presentations may range from incidental diagnosis, as in our case, to sudden death.

Cardiac tumors, especially myxomas tend to become quite large before the onset of symptoms which explains the indolent worsening of the patient's symptoms over a period of two years. She presented to us with rapid deterioration of symptoms within the last two months, with orthopnea as the major symptom. She did not have any neurological deficit, although she gave history of syncopal attacks.

Although myxomas are the most frequent primary tumors, right atrial is an atypical location leading to a diagnostic dilemma^[5-7]. A far more common cause of right atrial tumors are metastatic tumors that arise in the abdomen and pelvis, especially hepatoma and renal cell carcinoma, which directly invade the inferior vena cava and extend into the right heart^[10,11]. Secondary infiltrative tumors of the heart usually extend into the right atrium from the IVC (and less commonly from the right atrial free wall). Tumor extension along the IVC and into the right atrium is the mechanism of intracardiac tumor spread, most frequently described in renal cell carcinoma, Wilm's tumor, hepatoma and uterine leiomyoma^[12]. Contrary to this, our patient did not have a history of carcinoma with lymph node metastasis, and there was no evidence suggesting metastatic involvement of the liver or kidneys.

The histopathological report gave us the diagnosis of myxoma. Unlike in this case, a majority of right atrial myxomas are found to be attached to the interatrial septum, usually at the border of the fossa ovalis^[8]. In this case the myxoma was found to be attached by a thin stalk to the annulus of the tricuspid valve which is an unusual site.

Intraoperatively the mass was found to filling whole of the right atrium, we postulated that the syncopal attacks were caused by the incomplete filling of the right heart resulting in decreased forward flow. The thin stalk of attachment allowed free mobility of the tumor into the right ventricle through the tricuspid valve with diastole. Unlikely the mass was found to be attached to the

septal leaflet portion. The usual attachment is in the interatrial septum, as discussed before. Surprisingly, there was no evidence of damage to the tricuspid valve or no tricuspid regurgitation was noted after removal of the mass. This has to be kept in mind, since no events of paroxysmal atrial fibrillation, unusual sites of thrombosis, tricuspid valve stenosis/ regurgitation or family history were known, making the presence of a primary mass in the right atrium atypical and uncharacteristic.

Conclusion

Though a rare location, a high index of suspicion of a myxoma is needed in a right atrial mass presenting with a variety of unexplained symptoms. The patient should be counselled regarding possible recurrence once diagnosis is confirmed.

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