Case Report

DOI: http://dx.doi.org/10.18203/2349-2902.isj20171015

Atypical clinical presentation of right atrial myxoma: a case report

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Received: 28 February 2017 Accepted: 18 March 2017

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ABSTRACT

Primary intracardiac tumors are rare and approximately 50% are myxomas. Majority of myxomas are located in left atrium. Most of myxomas are attached to inter-atrial septum and have variable clinical presentation depending on size, location and mobility of the tumor. We report a case of large myxoma in the right atrium with atypical clinical presentation and location for this type of tumor.

Keywords: Cardiac tumor, Myxoma, Right atrium

INTRODUCTION

Cardiac tumors represent 0.2% of all tumors, secondary and metastatic form are 20 to 40 times more common than primary tumors. Among primary cardiac tumors, 75% are benign and 50% are myxoma with an incidence of 0.0017% in general population Most common site of myxoma is in left atrium (75%) followed by right atrium (18%) and more rarely in aorta, pulmonary artery, ventricles vena cava or even in other organ^{2,3,6} The differential diagnosis formed between thrombus or rhabdomyoma.

Sporadic myxomas are more common in adult female and they are mostly pedunculated and solitary, while familial form predominantly affects young male and they are multiple, sessile and have high rate of recurrence after surgical excision.^{5,7}

Signs and symptoms caused by myxomas depends on size, mobility and location of the tumor as well as physical activity and body position. Most of the patient present with the one or more symptoms of classical triad of intracardiac obstruction to blood flow, thromboembolic events and constitutional symptoms.^{3,5,7}

RA myxoma, in particular can obstruct tricuspid valve, causing sign and symptoms of right heart failure, peripheral edema, ascites, hepatic congestion and syncope.⁵

Transthoracic echocardiography alone is 95% sensitive in detecting myxomas and the sensitivity increases to 100% when followed by trans oesophageal echocardiography CT and MRI are useful to demonstrate the point of fixation and other complication.⁵

Surgical excision should be performed as soon as possible due to constant risk of thromboembolic event. Surgical treatment is definitive and recurrence is uncommon except in familial form.

CASE REPORT

A 52-year male presented with atypical chest pain, palpitation and syncope for last 3 month which became more frequent in last 1 month.

Cardiovascular examination revealed regular but tachycardiac heartbeat with splitting of S1 and no murmur and normal JVP. Examination of other system

was normal. ECG detected sinus tachycardia and chest X-ray shows clear lung field with cardiomegaly. ESR was 11mm/hr. and CRP was 3mg/L.

Transthoracic echocardiography revealed a mass (7x7x5cm) almost completely filling the right atrial cavity prolapsing through the tricuspid valve in to right ventricle and dilated IVC. Trans esophageal Echocardiography revealed well circumscribed mass extending from right atrium in to right ventricle causing tricuspid regurgitation and stenosis. MRI shows pedunculated and ovoid heterogenous mass confined to right atrium and bulging in to right ventricle during diastole (Figure 1 and 2).

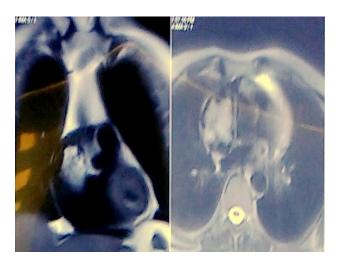


Figure 1: Cardiac MRI on T1 weighted imaging showing the well-defined mass of intermediate intensity in right atrium, pedicle attached to free wall of right atrium.

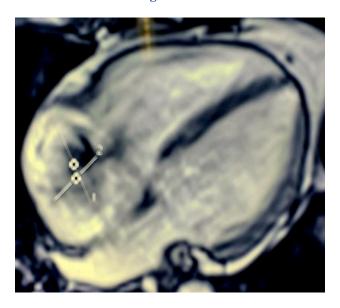


Figure 2: The cine-MR SSFP images in the fourchamber view, in diastole showing one well-outlined and polylobulated intracavitary mass in the right atrium prolapses through the triscuspid valve.

Right atriotomy was applied under cardiopulmonary bypass (Figure 3). The tumor was mobile pedunculated, lobulated and gelatinous mass attached to right atrial free wall and superior vena cava junction (Figure 3 and 4). Tumor was prolapsing in to right ventricle and inferior vena cava, tricuspid valve was normal.

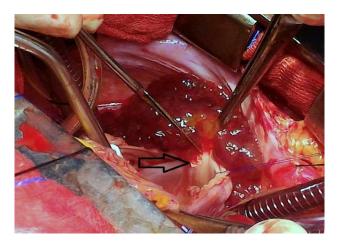


Figure 3: Operative picture showing arrested heart on cardiopulmonary bypass with right atrial cavity with gelatinous lobulated mass attached to right atrial free wall at SVC-RA junction. Arrow pointing to site of attachment.

On microscopy, there were abundant myxoid stroma with stellate cells. Necrosis, mitotic activity, atypia and pleomorphism were not detected.

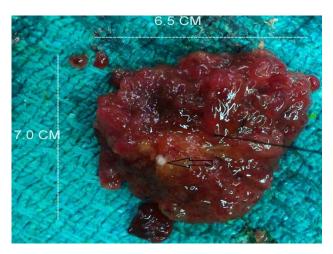


Figure 4: Excised specimen of 7cm X 6.5cm gelatinous, pedunculated mass with multiple lobe and focal area of haemorrhage. Arrow pointing to peduncle.

Post-operative course was unremarkable and he reported symptomatic improvement in follow up visit.

DISCUSSION

The right atrium is site of 15-20% of cases of myxoma, 70% of affected patients are women predominantly

between third and sixth decades of life. Common site of origin in right atrium is fossa oval is or base of interatrial septum. The signs and symptoms of RA myxoma are atypical and highly variable, depending on the size, location and mobility of the tumor. They may remain asymptomatic or may cause sudden death or eventually cause constitutional symptoms like fever, weight loss, arthralgias, anemia, Raynaud phenomenon and increased erythrocyte sedimentation rate. These symptoms disappear after tumor exicision. Most common manifestation is dyspnoea (80%) and right sided heart failure but patients may also present with atypical chest pain, palpitation pulmonary embolism, haemoptysis and syncope.

This clinical case is quite unusual considering, the rare clinical presentation as atypical chest pain, palpitation and syncope, in a male patient with no dyspnoea or constitutional symptoms. This is also unusual considering the uncommon location in right atrium at SVC-RA junction, the discrepancy between transthoracic echocardiogram finding and clinical finding. Echocardiography remains the best diagnostic method for location and extent of myxoma and for detecting the recurrence, however transthoracic echocardiogram may not identify tumor mass less than 5mm in diameter so transoesophageal echocardiogram is required in suspicion or in small size tumor.9 CT and MRI scan provide information regarding tissue characteristics, attachment to abnormal position an overview of cardiac and paracardiac morphology. Surgical excision of myxoma include large resection of their pedicle to prevent recurrence.^{1,8} In this case making an excision with large margin was potentially dangerous because of critical anatomical position at SVC-RA junction and consequent high risk of conduction disturbance.

CONCLUSION

Right atrial myxoma are rare and have atypical presentation, so they should be considered in differential diagnosis of atypical chest pain palpitation and syncope. Echo cardiography should be performed on urgent basis in adult patient with these symptoms. Early diagnosis and timely surgical intervention is mainstay of management to prevent possible fatal consequences such as sudden death.⁵

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

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Cite this article as: Rathor DKS, Jhajhria NS, Gupta VK. Atypical clinical presentation of right atrial myxoma: a case report. Int Surg J 2017;4:1444-6.