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Right Atrial Myxoma with Critical Triple Vessel Disease: A Case Report

Narendra Kumar [*], **Dinesh Chandra, Harpreet Singh and Sanjay Kumar**

Department of Cardiothoracic vascular surgery, Max Superspeciality Hospital Shalimar Bagh, New Delhi – 110088

[*]**Corresponding Author:** Narendra Kumar, Department of Cardiothoracic vascular surgery, Max Superspeciality Hospital Shalimar Bagh, New Delhi – 110088, Tel: 917701967049, E-mail: dr.narendrasharma50@gmail.com

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Abstract

Cardiac myxoma are the most common primary heart tumor. It accounts for 40-50% of primary cardiac tumor. Approximately 75% occurs in left atrium and 15-20% in right atrium. Myxoma are usually polypoid, pedunculated lesion with smooth surface covered with thrombus. Most common site of attachment is at the border of fossa ovalis in left atrium but it can occur in any part of atrial wall. Symptoms are produced because of mechanical obstruction with cardiac blood flow, tumor embolization and constitutional symptoms due to increased expressions of IL-6. Echocardiography is the best diagnostic modality to localise the tumor. Surgical excision is the mainstay of treatment of myxoma present in any location.

In this case report, we presented a 62 year old male patient, known case of diabetes mellitus type II, hypertension and chronic obstructive pulmonary disease came to emergency on NIV and inotropic support with complain of chest pain and difficulty in breathing. He had history of 2-3 episode of ventricular arrhythmia needing CPR in previous hospitalization. CAG revealed critical triple vessel disease. Echocardiography revealed RA myxoma with severe left ventricular dysfunction (EF=20%). The case was challenging because of large size of tumor present near RA-IVC junction with critical TVD and presence of severe left ventricular dysfunction. The patient underwent coronary artery bypass with RA myxoma excision. The surgery was uneventful and patient was discharged on 6th post-operative day. Histopathology of mass confirmed myxoma.

Keywords: Arrhythmia; Myxoma; Critical triple vessel disease

List of Abbreviations

RA: Right atrium

EF: Ejection fraction

CAG: Coronary angiogram

CT: Computed tomography

CPR: Cardiopulmonary resuscitation

TVD: Triple vessel disease

IVC: Inferior vena cavae

LAD: Left anterior descending

LCx: Left circumflex

RCA: Right coronary artery

OM: Obtuse marginal

TTE: Transthoracic echocardiography

TEE: Transesophageal echocardiography

NIV: Noninvasive ventilation

Introduction

Cardiac myxoma are the most common primary heart tumor [1]. The incidence ranges between 0.0017% and 0.19% [2]. Approximately 75% of primary cardiac tumors are benign and 25% are malignant [3]. Myxoma is most common benign tumor. It accounts for 40-50% of primary tumors. Approx 75-80% occurs in left atrium and 15-20% in right atrium [4]. Myxomas are usually polypoid, round or oval. In 90% of cases are solitary and pedunculated lesions with a smooth surface that may be covered with thrombus.

The tumor ranges in size from 1 to 15cm. Myxoma generally arises from pleuripotent mesenchymal cells. Histologically, it consists of myxoid matrix of acid mucopolysaccharides [4]. The cells are usually polygonal or spindle-shaped [4].

Myxomas produces a variety of symptoms. Symptoms are caused due to mechanical interference with cardiac blood flow, tumor embolization and constitutional symptoms. These sequelae are related to the location, size and mobility of tumor.

Because of intravascular in nature, it produces embolic phenomenon depending on its location. Embolic phenomena are mainly systematic [5] but pulmonary embolism correlated with right atrial myxoma has also been reported [6,7]. A large myxoma may result in interference with valve function and may cause destruction of leaflet. Myxoma also produces constitutional symptoms like fever, malaise, rash, weight loss and myalgias because of increased expression of inflammatory cytokines, IL-6 [8].

The only treatment of cardiac myxoma is surgical excision. Here we report a case of a giant right atrial myxoma with critical triple vessel coronary artery disease, rarely reported in literature.

Case Presentation

A 62-year-old male patient was brought to the emergency of our hospital referred from other hospital with NIV and high inotropic support. The patient complained of chest pain and difficulty in breathing. Patient was a known case of diabetes mellitus Type II, hypertension, chronic obstructive pulmonary disease and was on regular medication. He had history of 2-3 episode of ventricular arrhythmia needing CPR and successfully cardioverted in previous hospitalization.

Cardiovascular examination revealed that the heart rhythm was regular with plop sound at 3rd intercostal space of the left parasternal region. Respiratory examinations showed wheeze in bilateral lung field. Prominent bilateral IJV was noted in neck. Other systemic examinations were unremarkable.

Electrocardiogram showed normal sinus rhythm with inverted T wave in V3-V5 and Q wave in lead II, III, aVF. A transthoracic echocardiography showed following findings: Hypokinetic anterior septum, basal septum, anterior wall, antero-lateral wall, inferior, infero-lateral wall. Thickness of myocardium preserved indicating viable myocardium. A hyperechoic, non-calcific mobile mass of size approx. 3.8 X 4 cm present in right atrium and possibly attached to lateral wall of right atrium prolapsing through tricuspid valve causing obstruction in valve function (Figure.1). Fossa ovalis intact. Low-moderate mitral regurgitation with ejection fraction of 20%. IVC was dilated with < 50% respiratory variation.

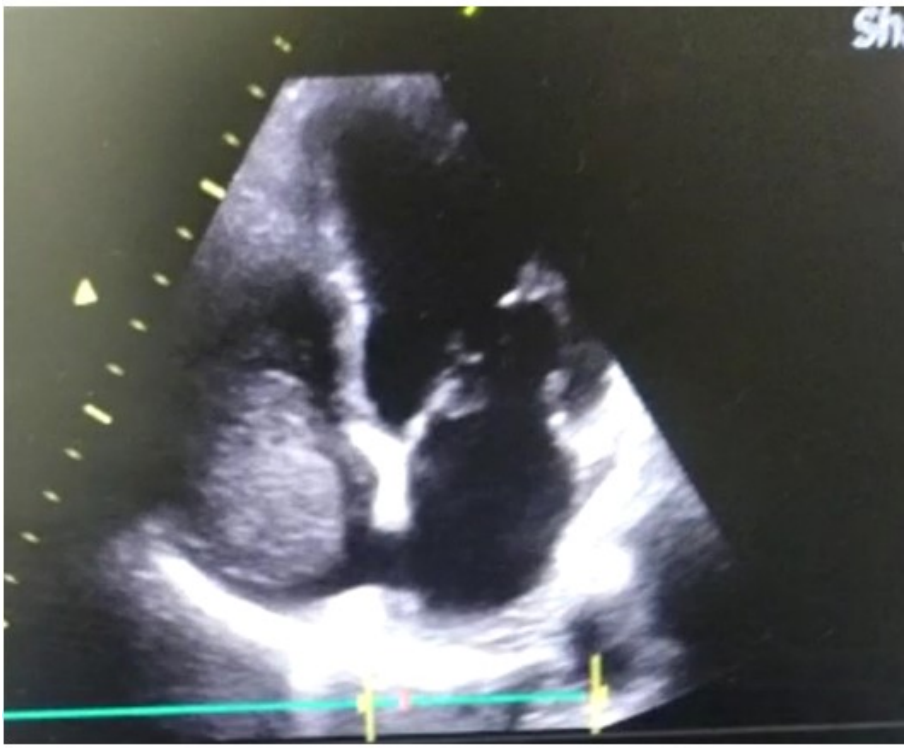

Figure 1: Echocardiogram showing tumor prolapsing through the tricuspid valve

Liver function test was abnormal with an aspartate aminotransferase (AST) level of 760U/L and alanine aminotransferase (ALT) level of 628U/L. Renal function test was normal with creatinine level of 1.03 mg/dl. Coronary Angiogram revealed: LM distal plaquing. LAD proximal 100% stenosis, distal segment filling retrogradely via collaterals from OM1. LCx distal diffuse disease, OM 1 normal. OM2 ostio-proximal 90% stenosis, RCA dominant vessel proximal 100% stenosis, distal segment filling retrogradely via collaterals from left system.

The diagnosis of triple vessel coronary artery disease, right atrial myxoma with congestive heart failure was confirmed and urgent surgical resection with coronary artery bypass was scheduled a day after diagnosis. On the day of surgery just before shifting the patient to operation theatre he developed one episode of ventricular arrhythmia which was successfully cardioverted to sinus rhythm by giving 150 joule of DC shock.

After taking informed written consent, we performed primary median sternotomy under general anesthesia. Intra-operatively tumor mass size and location was re-assessed with the help of transesophageal echocardiography (Figure 2). A full study was performed with TEE and no mass was found in other chamber of heart. Cardiopulmonary bypass was established with conventional hypothermia at 32 degrees Celsius. The aorta was cross-clamped and myocardial protection was achieved by giving cold blood cardioplegia antegrade through aortic root. The right atrium was opened in oblique fashion. The tumor was found to be mobile, pedunculated, fragile gelatinous mass covered with thrombus attached by stalk to the posterior wall of the right atrium near RA-IVC junction. The tumor was excised in toto along with stalk (Figure 3). RA was closed in layers. Three distal anastomosis performed. Rewarming initiated. During re-warming three proximal anastomosis performed. The cardiopulmonary bypass was terminated after rewarming and thorough de-airing. The tumor was sent for histopathological examination. Post-operatively patient had 2-3 episode of short run non-sustained ventricular arrhythmia (reverted spontaneously) on 3rd day. The patient recovery was smooth and discharge on 6th day post-op in clinically stable condition and further follow-up advice. The patient came for follow-up as advised. The stitches was removed and he was recommended for further medications and regular follow-up.

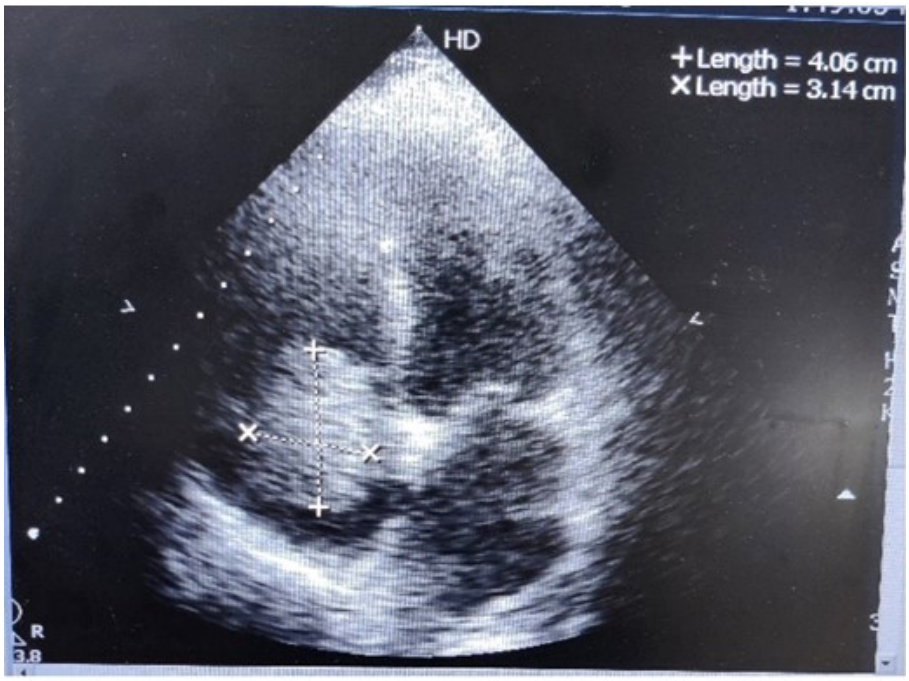

Figure 2: Intra-operative TEE showing mobile, pedunculated tumor of size 4.06 X 3.14 cm

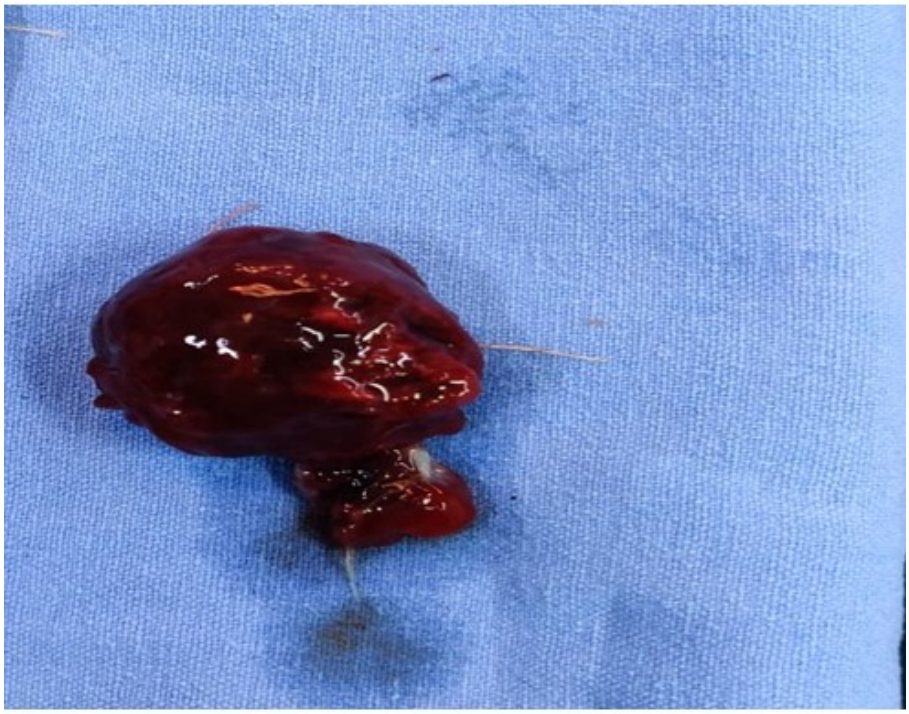

Figure 3: Excised tumor mass covered with thrombus

On histopathological examination, there was atypical cells in small cords, nests and as isolated cells against abundant myxoid stroma admixed with cells comprising of oval nuclei with abundant cytoplasm. Fair number of hemosiderin laden macrophage and inflammatory cells noted, all confirming the diagnosis of myxoma.

Discussion

Myxomas are most common primary cardiac tumor. They are usually benign and have been reported in both sexes but most often occur in women. Most common site of attachment is at the border of fossa ovalis in left atrium but it can occur in any part of atrial wall. In our case tumor was found to be attached in posterior wall of right atrium near RA-IVC junction. Morbidity is mainly related to symptoms produced by tumor embolism, heart failure, mechanical valvular obstruction and various constitutional symptoms. Case of pulmonary embolism in RA myxoma is reported in few series. Cemalettin et al. [6] reported a case of simultaneous pulmonary embolism detected on chest computed tomography as filling defects in branches of pulmonary artery with a giant right atrial villous myxoma. Aref Obagi et al. [7] also reported a case of bilateral lower lobe pulmonary emboli with right atrial myxoma detected on CT scan.

RA myxoma shows atypical and highly variable symptoms depending on size, position and mobility of tumor. They are usually asymptomatic and rarely cause constitutional signs and symptoms including fever, weight loss, arthralgias, Raynaud's phenomenon and increased erythrocyte sedimentation rate. In our case, patient presented with chest pain, dyspnea. Hepatic congestion, prominent IJV noted on examination indicating tumor obstructing valvular function. In addition, there was presence of critical triple vessel coronary artery disease.

Echocardiography is the best diagnostic modality to locate extent, size and mobility of tumor. Transesophageal echocardiography can be used to delineate the tumor presenting in far-field like right atrium and the posterior wall of left atrium (sensitivity TTE-95% Vs TEE-100%)[9]. In this case, myxoma was present in posterior wall near RA-IVC junction.

Surgical excision is the mainstay of treatment. Complete resection of tumor with base is essential to cure the disease and to prevent recurrence. Our case was challenging as large size of myxoma located near RA-IVC junction need careful IVC cannulation to prevent tumor mass fragmentation causing embolization. In addition, presence of critical triple vessel disease, the patient old age with multiple co-morbidities, severe left ventricular dysfunction carries a high risk to proceed for surgical intervention. In spite of that right atrial myxoma excision with coronary artery bypass was successfully performed without any intraoperative complication.

In our case, we applied cross-clamp over both aorta and pulmonary artery to avoid embolization of fragments while doing excision of myxoma. Femoral cannulation may be another option if there is suspicion of intrusion of mass in IVC.

The prognosis of surgical excision of myxoma is good with low risk of morbidity and mortality (0-3%) [10] while in case of familial myxoma it ranges from 12-20% [11]. So, regular echocardiographic follow-up is essential in familial myxoma group.

Conclusion

Surgical excision is the mainstay of treatment in case of myxoma present in any location. Transthoracic echocardiography plays a major role in diagnosis of myxoma. TEE is helpful in delineating the better characteristics of myxoma. Presence of RA myxoma with critical TVD provide a challenging task to surgeons so surgical intervention should be done with great experience and expertise.

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