Case Report

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Right Ventricular Myxoma: Rare but a Known Entity Nowadays

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Abstract

Cardiac myxomas are the most common benign tumors of the heart. Ventricular myxomas are rarely described. We evaluated a young female who presented with syncope and dyspnea. 2D echo and CT pulmonary angiography were done which revealed a mass in RVOT. Surgical excision of the tumor was done via right atriotomy without any complications. Histopathology showed features of a myxoma.

Keywords: Myxoma; Right Ventricle; Excision.

Introduction

Myxomas constitute around 50% of the cardiac masses. Large percentages are located in the atrias, but intra-ventricular myxomas have been infrequently reported. Literature provides the percentage of distribution of myxomas in left atrium: 75-85%, right atrium: 15-20%, 5% occupying the right ventricular and <4% in left ventricle [1,2].

Clinical symptoms may be obstructive, embolic or constitutional. Differential diagnosis for an intra-cavitary cardiac mass includes thrombus, myxoma, lipoma and non-myxomatous neoplasms. Two dimensional real time echocardiography (2D-echo) can be used as a sole diagnostic modality. The advent of computed tomography (CT), has reduced the invasiveness and further enhancing the imaging quality. Surgery in the form of complete excision of the mass with adequate margins is the ideal mode of treatment.

Case Report

A 24 year old female was hospitalized with an episode of syncope upon exertion, with a 2 week history of sudden onset of dyspnea and the absence of other relevant cardiac symptoms. Family and personal history were unremarkable.

Physical examination revealed ejection systolic murmur in pulmonary area with no signs of cardiac failure in a moderately built Indian female (BMI of 22.2kg/m^2). Chest X-ray and electrocardiogram were unremarkable. Keeping in mind pulmonary thrombosis and right atrial mass, 2D-echo was done (Figure 1). Right ventricular mobile intra-cavitary pedunculated mass was observed at the inter-ventricular septum not causing inflow or outflow obstruction at rest. To reconfirm our suspicion a CT pulmonary angiogram was performed (Figure 2). A working diagnosis of myxoma was made and surgery was planned after discussing the pros and cons with the patient. Elective surgery was performed under cardiopulmonary bypass with aortic and bi-caval cannulation with cardiac arrest being achieved by means of ante-grade cold St Thomas II cardioplegia.

Right atrium was opened and Right ventricle was visualized through the tricuspid valve to reveal a 3*2cm mass attached to the outlet septum below the septal leaflet of the tricuspid valve with a stalk (Figure 3). Tumor was excised completely with a rim of 1cm endocardium. Rest of the RV was free of tumor. Thorough saline lavage was given. Tricuspid valve did not reveal any leak. Patient was weaned off bypass uneventfully in sinus rhythm. Post operative

recovery was smooth and the patient was discharged after 9 days. Follow up by 2D echo has showed no recurrences at the operative site or in other chambers.

Histopathological sections showed characteristics of a myxoma: central areas of homogenous eosinophilic material, surrounded by myxoid to fibrous stroma with loosely dispersed bland spindle cells. Areas of calcification and few hyalinised cells were seen. No mitosis or pleomorphism was observed (Figure 4).

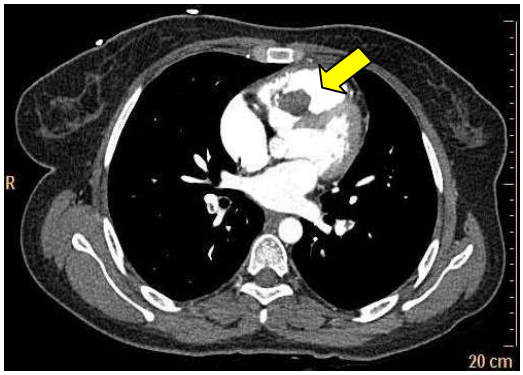

Fig. 2: CT-PA demonstrates clearly a hypodense mass attached to the inter-ventricular septum in the RV. (Dimensions: 27.0*20.0*28.0mm)

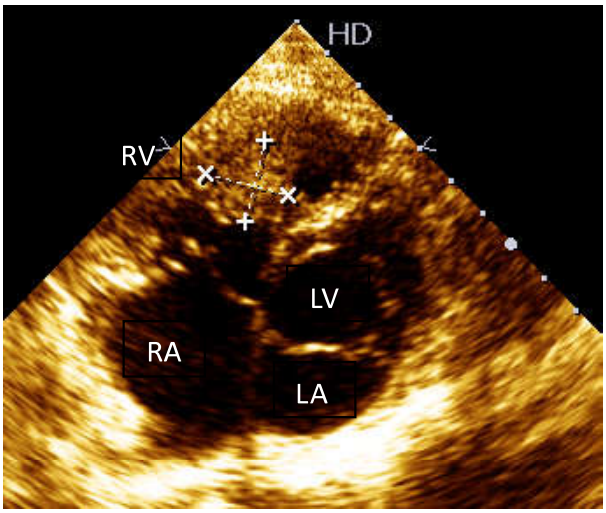

Fig. 1: Mass in the right ventricle attached to the inter-ventricle septum measuring 2.0*1.7cm

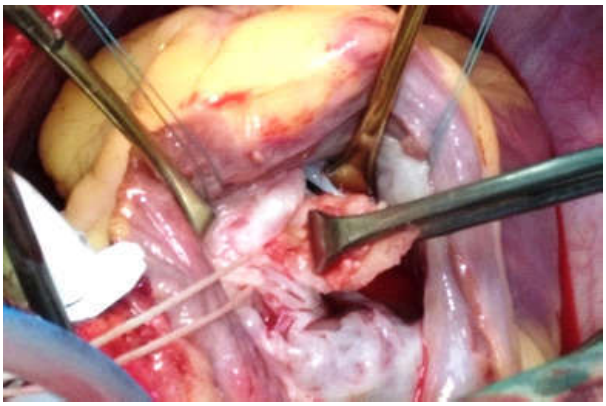

Fig. 3: Lobulated non calicfic, pale mass. Septal leaflet being retracted to expose the mass in RV. (Photo taken from head end)

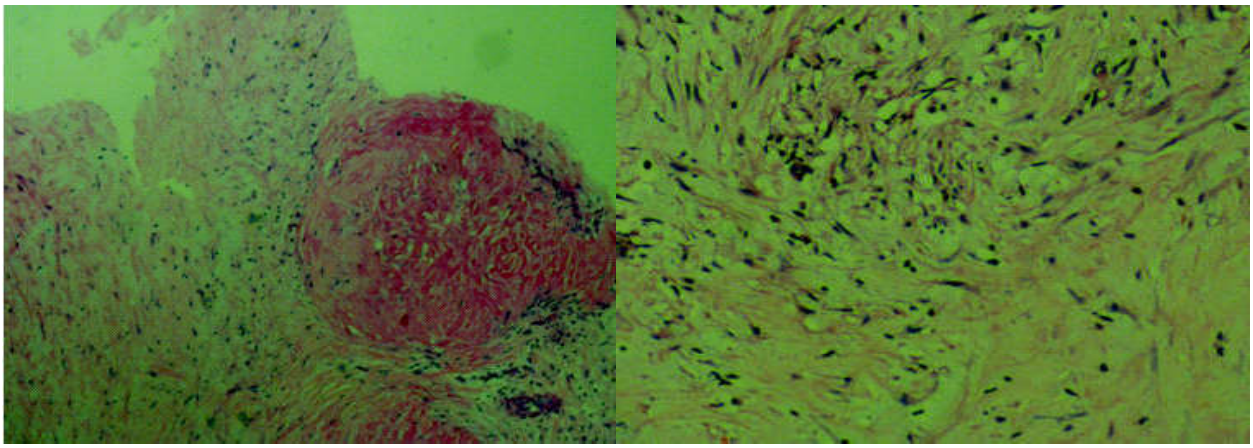

Fig. 4: Hemotoxylin & Eosinophil stain: spindle cell arrangement, low power and high power

Discussion

Benign tumors constitute 80% of primary cardiac tumors, myxomas being most common [3]. Previously, sarcomas were thought to be more common in ventricles [4]. But this can be questioned with new evidence of increasing reporting of ventricular myxomas (>100 cases have been reported). More than 90% of myxomas are sporadic, solitary and more commonly seen in young females [5]. Right ventricular outflow tract tumors can produce inflow and outflow tract obstruction depending on the size of the mass. Sometimes they can also present with arrhythmias, pulmonary embolism, RV dysfunction or sudden death [6].

Myxomas are a neoplasm of uncertain histogenesis that occurs only on the endocardial

surface. These originate from the reserve totipotential sub-endocardial cells capable of forming vascular structures that have endothelial and neural markers [7]. Grossly one thirds of myxomas are gelantinous and friable and thus prone to fragment and embolize [8].

Echocardiography is an excellent method to diagnose myxomas by non invasive means [9]. CT was done in our case to rule our pulmonary embolism. Gadolinium enhanced cardiovascular magnetic resonance imaging is used primarily to differentiate myxoma and thrombus based on their enhacement [10].

Based on the location of the tumor and size, varying approaches have been described. In the early periods of bypass evolution, right ventriculotomy was used as an approach which provided greater visibility, completeness in excision of large tumors but caused more post-operative arrhythmias and dysfunction. Right atrial approach is ideal for all right sided intra-cavitary tumors as it provides good exposure and the added advantage of inspection of the fossa ovalis [11]. Rarely bicameral approach (combined pulmonary artery and right atrial incision has been described to excise large tumors in toto [12].

Complete Excision of the right ventricular myxomas can be obtained limiting to the endocardium and myocardium surrounding the pedicle. Full thickness excision is not recommended [11]. Minimal manipulation of the tumor and thorough saline lavage after excision is helpful in preventing implantation of tumor cells. Reports of recurrences have been highlighted in few case reports. The cause for recurrence may be due to embolisation, incomplete excision, growth from a second focus [13]. Hence regular biannual 2D echo follow up needs to be highlighted [14]. RV myxomas are infrequently found in general population. Thus we are providing increasing insight into the rarely found tumor.

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References

1. Mittle S, Makaryus AN, Boutis L, Hartman A, Rosman D, Kort S. Right sided myxomas. J Am Soc Echocardiogr 2005;18:695.
2. Hirota J, Akiyama K, Taniyasu N, Maisawa K, Kobayashi Y, Sakamoto N, Komatsu N. Injury to the tricuspid valve and membranous atrio-ventricular septum caused by huge calcified right ventricular myxoma: report of a case. Circ J;2004; 68:799-801.
3. Perchinsky MJ, Lichtenstein SV et al. primary cardiac tumors; forty years experience with 71 patients. Cancer 1997;79:1809-15.
4. Allister HA Jr: primary tumor and cysts of the heart and pericardium. Curr Probl Cardiol. 1979;4:1-51.
5. Reynen K. Cardiac myxomas. N Engl J Med 1995; 333:1610-17.
6. Rym Gribaa, Mehdi Slim et al. Right ventricular myxoma obstructing the right ventricular outflow tract: A case report. J of Medical Case reports 2014,8: 435.
7. Pucci, A, Gagliardotto P, Zanini C, Pansini S, Di Summa M and Mollo F. Histopathologic and Clinical Characterization of Cardiac Myxoma-Review of 53 Cases from a Single Institution. American Heart Journal. 2000;140:134-8.
8. Pinede L, Duhaut P and Loire R. Clinical Presentation of Left Atrial Cardiac Myxomas: A Series of 11 Consecutive Cases. Medicine. 2001;80: 159-72.
9. Vander Heusen FJ, Stratmann G and Russell IA. Right Ventricular Myxoma with Partial Right Ventricular Outflow Tract Obstruction. Anaesthesia and Analgesia, 2006;103:305-6.
10. Satitthummanid S, Tumkosit M, Benjacholamas V, Chattranukulchai P, Boonyaratavej S and Puwanant S. Right Ventricular Myxoma. Journal of American College of Cardiology 2011;57:630.
11. Bortolotti U, Mazzucco A, Valfré C, Valente M, Pennelli N, Gallucci V. Right ventricular myxoma: review of the literature and report of two patients. Ann Thorac Surg. 1982;33:277-8.
12. Trushar P. Gajjar, Gaurang B. Shah, Neelam B. Giant ventricular myxoma obstructing right ventricular outflow tract. Rev Bras Cir Cardiovasc 2011;26: 663-6.
13. Colin R Dang, Edward J Hurley. Contra-lateral recurrent myxoma of the heart. The Ann of Thorac Surg 1976;21:59-62.
14. Andres Diaz, Carmelo Di Salvo, David Lawrence, Martin Hayward. Left atrial and right ventricular myxoma: an uncommon presentation of a rare tumour. Interactive CardioVascular and Thoracic Surgery 2011;12:622–5.