

Submitral Aneurysm: A Rare Congenital Anomaly

Manish Ruhela*, Dinanath kumar, Chandra Bhan Meena, Rajeev Bagarhatta

Department of Cardiology, SMS Medical College and Hospital, Jaipur, Rajasthan, India

*Corresponding author: dr.manishruhela@gmail.com

Received May 22, 2014; Revised June 11, 2014; Accepted June 16, 2014

Abstract Submitral aneurysm is a congenital out pouching of the left ventricular wall invariably occurring adjacent to the posterior mitral leaflet. This rare congenital cardiac anomaly was initially described among the African population and is considered rare in the other parts of world. The clinical picture is dominated by congestive cardiac failure in the presence of mitral regurgitation. Echocardiography provides precise non-invasive diagnosis. The literature is reviewed to increase the awareness of clinicians, especially novice echocardiographers about this rare cardiac anomaly when coming across congestive cardiac failure with mitral regurgitation in young population.

Keywords: submitral aneurysm, congenital anomaly, mitral regurgitation

Cite This Article: Manish Ruhela, Dinanath kumar, Chandra Bhan Meena, and Rajeev Bagarhatta, "Submitral Aneurysm: A Rare Congenital Anomaly." *American Journal of Medical Case Reports*, vol. 2, no. 6 (2014): 114-116 doi: 10.12691/ajmcr-2-6-1.

1. Introduction

Submitral left ventricular aneurysm is a rare cardiac anomaly, recognized but with relatively sparse description in the literature. It was first reported from Nigeria and other African countries by Abrahams et. al where 12 patients with an unusual form of left ventricular aneurysm were described which they termed as "annular left ventricular aneurysm" [1]. Submitral aneurysm (SMA) is a congenital out-pouching of left ventricular wall, invariably occurring adjacent to the posterior leaflet of mitral valve [2]. Most of the cases of subvalvular left

ventricular aneurysms described in the literature are due to congenital weakness of the fibro-muscular annuli. Chesler and colleagues postulate that a dehiscence of the fibro-muscular union will result in aneurysm formation. [3] Cases have been described mainly from African countries but few case reports from India as well [4]. We report this case from the department of Cardiology, SMS Medical college, Jaipur, Rajasthan, India. This is the first case report of submitral aneurysm from the western part of India. Our case highlights the importance of considering SMA in the differential diagnosis of mitral regurgitation with left ventricular dysfunction and heart failure in young patients.

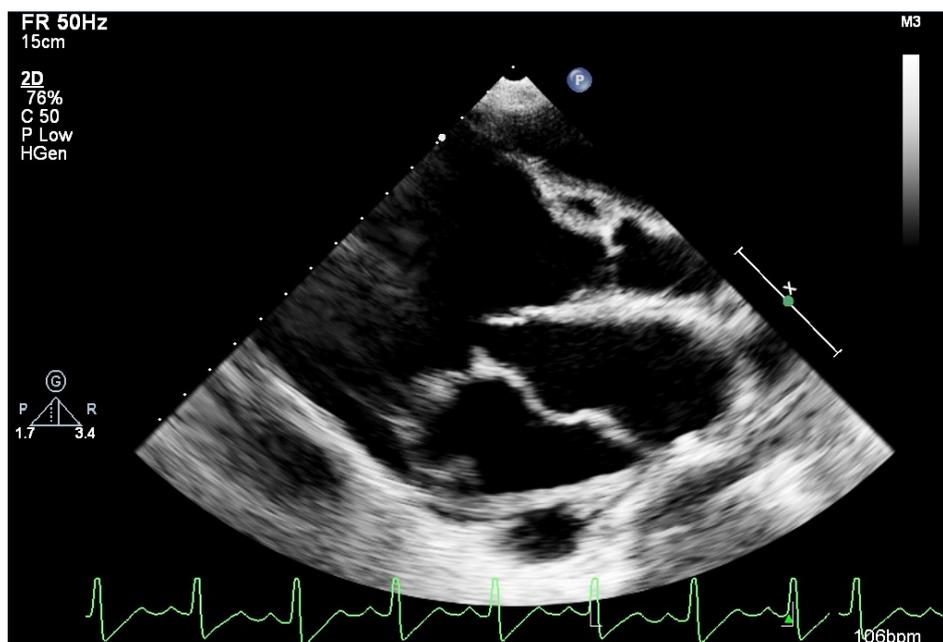


Figure 1. Transthoracic echocardiography (Parasternal long axis view) showing submitral aneurysm below posterior mitral leaflet

2. Case Report

A 18 year old boy presented to cardiology OPD with progressively worsening dyspnea and palpitations for the last two year and history orthopnea for 7 days. There was no history suggestive of rheumatic fever. On examination he had a heart rate of 110/ minute, blood pressure of 108/60 mm Hg, raised jugular venous pressure (9 cms above the angle of Louis) and pedal edema. The apex was in the 6th left ICS, 2 cms outside the mid-clavicular line. On auscultation, S1 soft and LVS3 were present. A Grade 4/6 pan-systolic murmur radiating towards the axilla was heard. There was also a pan-systolic murmur in the left 3rd parasternal region which increased during inspiration. Chest auscultation revealed bilateral basal crepitations.

Abdominal examination revealed tender hepatomegaly (liver span 15 cms). ECG showed sinus tachycardia. Chest X- ray revealed cardiomegaly with evidence of pulmonary venous congestion. On transthoracic echocardiography, the significant finding was a submitral aneurysm (Figure 1, Figure 2) in the postero-lateral wall of the left ventricle. The patient also had severe mitral regurgitation with an eccentric jet with dilatation of the left atrium and left ventricle (Figure 3). He was also found to have severe tricuspid regurgitation (Figure 4). Left ventricular Ejection fraction was 48% and minimal pericardial effusion was also present. So, the diagnosis of submitral aneurysm was made and patient was stabilized with decongestive therapy and then refer to cardiothoracic department for further management.

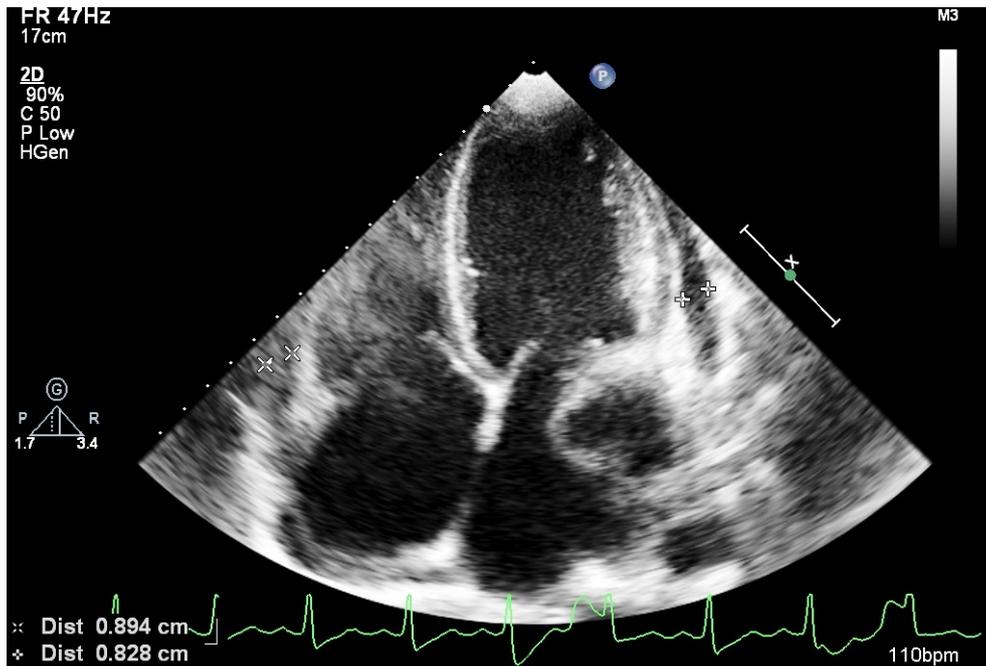


Figure 2. Transthoracic echocardiography (Apical 4 chamber view) showing submitral aneurysm and pericardial effusion.

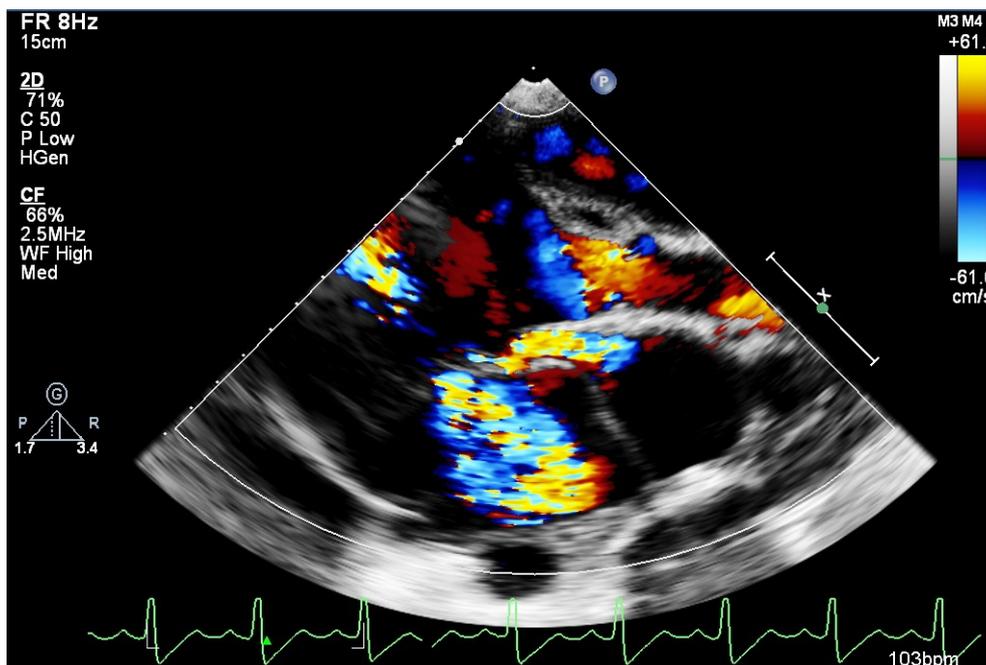


Figure 3. Transthoracic colour Doppler echo showing mitral regurgitation and flow into submitral aneurysm.

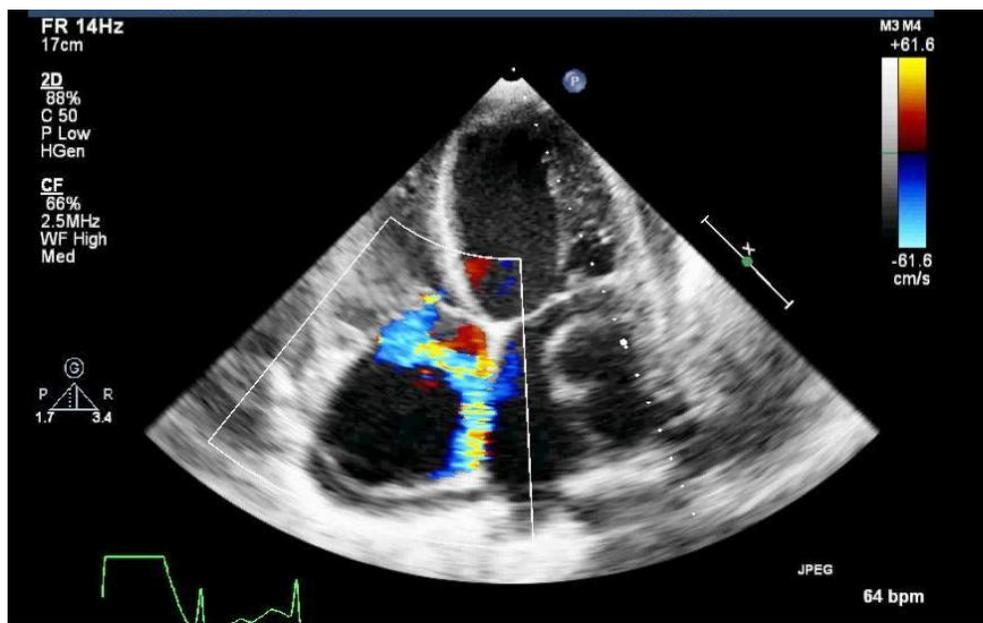


Figure 4. Transthoracic colour Doppler echo showing Tricuspid regurgitation and submitral aneurysm

3. Discussion

Submitral aneurysm is rare and most cases have been described in black and Negroid people. Sub-mitral aneurysm was first described in 1812 by Corvisart and, since then, only 100-120 cases of these aneurysms have been reported worldwide. [5] Sporadic case reports in the last two decades have documented their existence in the Indian population also. Even though the etiology of the condition is thought to be a congenital, co-existence of the condition with Takayasu's arteritis [6] and tuberculous pericarditis [7] has been reported. Although it can present as life threatening complications such as ventricular tachycardia due to compression of the left main coronary artery, the most common presentation is severe mitral regurgitation [8]. Doppler echocardiography has proved to be a useful technique for the noninvasive diagnosis of submitral aneurysms in the clinical setting. Management of submitral aneurysm involves initial medical stabilization with diuretics and afterload reducing agents. Surgical repair is the definitive treatment and includes pericardial patch repair, valvuloplasty through a transmitral approach [9], transatrial repairs with sutures [10], or mitral valve replacement [11]. Mortality is high in un-operated cases and operative repair under cardiopulmonary bypass is the most appropriate management. Post-operative outcome is directly related to size of the aneurysm and severity of LV dysfunction.

In conclusion, submitral aneurysm, although uncommon, should always be considered in the list of differential diagnosis in young patients presenting with a murmur suggestive of mitral insufficiency or signs and symptoms of heart failure. The definitive diagnosis is made by transthoracic echocardiography in the presence of an aneurysmal dilatation in submitral location behind the posterior mitral leaflet that communicated with the left

ventricular cavity through one or more necks. Novice echocardiographers should be aware about this rare cardiac anomaly when coming across congestive cardiac failure with mitral regurgitation in young population.

Statement of Competing Interests

Authors have no competing interests.

References

- [1] Abrahams DG, Barton CJ, Cockshott WP, et al. Annular subvalvular left ventricular aneurysms. *Quart J Med* 1962; 31: 345-360.
- [2] Antunes MJ. Submitral left ventricular aneurysms. Correction by a new transatrial approach. *J Thorac Cardiovasc Surg* 1987; 94: 241-245.
- [3] Chesler, E., Mitha, A.S., and Edwards, J.E. Congenital aneurysms adjacent to the annuli of the aortic and/or mitral valves. *Chest*. 1982; 82: 334-337.
- [4] Chockalingam A, Gnanavelu G, Alagesan R, Subramaniam T. Congenital submitral aneurysm. *Echocardiography* 2004; 21: 325-328.
- [5] Deshpande AV, Vaidya SV, Kumar A. Submitral aneurysm. *Heart* 2004; 90: 988.
- [6] Rose AG, Folb J, Sinclair Smith CC, Schneider JW. Idiopathic annular submitral aneurysm associated with Takayasu's arteritis. *Arch Pathol Lab Med* 1995; 119: 831-5.
- [7] Sharma S, Daxini BV, Loya YS. Profile of Submitral Left Ventricular Aneurysm in Indian Patients. *IJH* 1990; 42: 153-6.
- [8] Ruiz D, Domigo P, Canalis E, Mata J. Annular submitral left ventricular aneurysm. *Eur Heart J* 1992; 13: 424-425.
- [9] Janeira LF, Talit U, Parker R, Hughes CE, Tuna IC. Surgical management of ventricular tachycardia in sub annular left ventricular aneurysm. *Ann Thorac Surg* 1995; 60: 438-40.
- [10] Sutorius DJ, Helmsworth JA, Majeski JA, Miller SF. Repair of a subvalvular left ventricular aneurysm following mitral valve replacement. *Ann Thorac Surg* 1981; 32: 92-6.
- [11] Almeida - Filho OC, Schmidt A, Sgarbieri R, Marin- Neto JA., Maciel BC. Large submitral left ventricular aneurysm associated with mitral valve aneurysm. *Echocardiography* 2002; 19:391-3.