Windsock deformity of submitral left ventricular aneurysm communicating into left atrium – role of transesophageal echocardiography

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ABSTRACT
Submitral left ventricular aneurysm (SMLA) is a rare condition. We report here a 38-year-old male patient, presented with mitral regurgitation and features of congestive cardiac failure (CCF) with New York Heart Association (NYHA) function class III, diagnosed to have SMLA. We discuss here the etiology, types, clinical presentation, and management of SMLA and also the role of transesophageal echocardiography in diagnosis.

Keywords: Submitral left ventricular aneurysm, transesophageal echocardiography, three-dimensional echocardiography

INTRODUCTION
Submitral left ventricular aneurysm (SMLA) is characterized by outpouching of the left ventricular wall in the posterior portion of the mitral annulus. Echocardiography plays an important role in the diagnosis of SMLA. Real-time three-dimensional (3D) echocardiography, nuclear magnetic resonance, and multidetector cardiac tomography (CT) also have been used recently in the diagnosis of SMLA.

CASE REPORT
A 38-year-old male, known case of rheumatic heart disease with mitral regurgitation presented with progressive dyspnea on exertion for the past 7 months and orthopnea for 1 week. He was on penicillin prophylaxis from his 18 years of age. His preoperative blood investigations revealed deranged liver function tests and serum creatinine. An electrocardiogram showed normal sinus rhythm with heart rate 110/min. Chest-X-ray revealed cardiomegaly. Transthoracic echocardiography (TTE) revealed submitral aneurysm below lateral annulus with dilated left atrium (LA), moderate mitral regurgitation (MR), and moderate tricuspid regurgitation (TR) with moderate pulmonary artery hypertension and good biventricular function. His coronary angiogram showed normal coronaries with no external compression of the coronary artery. The patient presented with symptoms suggestive of CCF with features of pulmonary edema. He was initially treated with diuretics and inotropes, two days after which he recovered. He was posted for mitral valve repair/replacement. Patient was administered anesthesia as per institutional protocol, no major hemodynamic instability occurred during...
induction, and transesophageal echocardiography (TEE) probe (IE33 Philips with X7-2t) was inserted. TEE revealed SMLA below the posterior mitral leaflet of size 6.4 × 3.3 cm [Figures 1 and 2], bulging into LA creating a windsock deformity and SMLA was seen communicating with LA through a small slit in its roof along with moderate MR [Figure 3 and Videos 1, 2] and moderate TR. Two distinct jets contributed to MR one through the valve leaflets and other through SMLA into LA. TEE findings were confirmed intraoperatively. Intraoperatively a 4 × 3 cm swelling was seen externally at atrioventricular groove just below the left atrial appendage. Under moderate hypothermic cardiopulmonary bypass (CPB) with cold blood cardioplegic arrest, interatrial septum (IAS) was opened along patent foramen ovale and walls of the SMLA inside LA were excised. The neck of the aneurysm was seen opening just below the PML annulus and the aneurysm was seen extending as a pouch downward and outward from left ventricle, which was seen externally. The neck of the aneurysm from left ventricle (LV) closed with interrupted 4-0 pledgeted sutures. The excess edges of the aneurysm inside LA were plicated. Mitral valve ring annuloplasty was done with 28 mm Medtronic ring. During ring annuloplasty, to reinforce the aneurysm closure, the annuloplasty ring sutures were passed through both the PML annulus and the closed aneurysm neck. Three interrupted pledgeted sutures were used to plicate tricuspid valve annulus from posterior to anterior leaflet. Patient was gradually weaned from CPB with Inj Milrinone 0.35 µg/kg/min and Inj Adrenaline 0.05 µg/kg/min. Post CPB, TEE showed an echo-free space on lateral aspect of PML annulus, but not communicating with LV [Video 3]. There was mild MR, mild TR, and no flow across IAS was seen. Postoperative period was uneventful and followup of the patient for 6 months showed favorable prognosis.

**DISCUSSION**

SMLA was first described in 1812 by Corvisart and first case series was reported by Abrahams et al. in 1962 from Nigeria. Even though, it was more commonly documented in African blacks, over the last three decades it was also reported in Indian population.

SMLA is most commonly seen below the PML annulus. Du Toit et al. classified SMLA into three types based on the extension of the aneurysm as Type I: single localized neck, Type II: multiple necks (separate distinct openings), and Type III: involvement of the entire mitral annulus.

The etiology can be due to congenital defect in the posterior mitral valve ring or due to infective and inflammatory causes like Takayasu’s arteritis and tubercular pericarditis. The clinical presentation of SMLA may vary from an incidental finding in an asymptomatic patient to more common forms of presentations like significant mitral regurgitation, congestive heart failure, thromboembolism, ventricular arrhythmias, and myocardial ischemia due to compression of the coronary artery and sudden death.

TTE plays an important role in the diagnosis of SMLA, a subpericardial echo-free space is noted below the...
posterior mitral leaflet and communicating with LV.\(^\text{[10]}\) However, TEE plays a key role in identifying the rupture of the aneurysm into LA,\(^\text{[11]}\) which should be suspected in TTE whenever paravalvular jet of MR is seen. Real-time 3D TEE also help in delineating the aneurysm and better visualization of its extension and also its relationship with surrounding structures.\(^\text{[12]}\) Management of SMLA includes initial stabilization with diuretics and afterload reducing drugs. Definitive treatment is surgical management, which includes closure of the neck of aneurysm and mitral valve repair using ring annuloplasty or mitral valve replacement in case of mitral leaflets are distorted.

**CONCLUSION**

Even though SMLA is a rare condition, it should be considered as differential diagnosis in young population presenting with CCF with severe MR in decompensated state. TEE both 2D and 3D plays an important role in the diagnosis and understanding the relation of the aneurysm with other cardiac structures.