Case Report:

Congenital sub mitral left ventricle aneurysm with normal mitral valve - A rare presentation

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Abstract

Sub mitral Left Ventricular Aneurysm (SMLVA) is an unusual, non-ischemic Left Ventricular (LV) Aneurysm. SMLVA is rarely reported in Indian subcontinent. It is most commonly found in Black population, among the natives of south and West Africa. SMLVA is basically a congenital out-pouching of the LV wall occurring adjacent to the posterior leaflet of Mitral Valve\textsuperscript{2}. It is typically diagnosed in young adults, presenting mostly with features of Mitral regurgitation, congestive heart failure, systemic embolization, arrhythmia and sometimes sudden death. But in our case mitral valve was almost normal and the patient presented with progressive worsening dyspnoea, palpitation and occasional chest pain secondary to gradual decline in ejection fraction. His coronary angiogram was normal. Differential diagnosis of SMLVA must be kept in mind when a young individual present with features of progressive LV dysfunction though it is rare.

Key words: Sub mitral left ventricular aneurysm, LV dysfunction, congenital.

Introduction

Sub Mitral Left Ventricular aneurysm is an uncommon cardiac lesion with very little description in the Indian literature. In 1962 Abrahams et al\textsuperscript{1} reported 12 patients with an unusual form of Left Ventricular aneurysm from Nigeria and other African countries and they termed as “Annular Subvalvular left ventricle aneurysm”. The anatomy of mitral valve annulus is such that its 2/3 portion is related to posterior leaflet which is attached to LV myocardium through annular ring\textsuperscript{3}. A weakness or disruption of this muscular–fibrous union results in occurrence of sub mitral aneurysm below mitral leaflet. Cases of SMLVA has been reported from all parts of the world but very few from Indian subcontinent\textsuperscript{4,5}.

Few case of SMA has been reported in Caucasians in which all presented with significant Mitral regurgitation.

Our patient is a rare presentation of a large SMA without involvement of Mitral Valve Apparatus. He presented with progressive dyspnoea, occasional chest pain and palpitation. Our case emphasise on considering a differential diagnosis of SMA in young adult presenting with features of low ejection fraction or LV dysfunction.

The typical location of the aneurysm with normal coronaries on angiography confirms the diagnosis of SMLVA. Surgical correction is mandatory to treat the cardiac failure secondary to increasing size of the aneurysm and to prevent the potential cardiovascular events.
Case Report
A 23 years young male presented to our department with progressive worsening dyspnoea for last 1 ½ years associated with occasional chest pain and palpitation. There was no past history of rheumatic fever, any episode of syncopal attack, asthma or Tuberculosis. No family history of any cardiovascular disease. On examination he had heart rate of 110 per minute, regular rate and rhythm, Blood pressure of 90/60mmHg, JVP was raised and had pedal oedema. The apex was in the 6th left intercostal space, 2 cm outside the mid clavicular line. On auscultation, soft S1 and Left ventricular S3 were present. Chest auscultation revealed bilateral basal crepitation. All peripheral pulses were bilaterally and equally palpable. ECG showed evidence of sinus tachycardia. Chest X-ray revealed cardiomegaly with evidence of pulmonary venous congestion. Echocardiography finding revealed normalsize Right and left atrium. Right ventricular outflow tract was also normal in size. Left ventricle was dilated (60mm) with a large aneurysm, present in the inferior wall in submitral position. Mitral valve, Tricuspid valve and Aortic valve were normal with significant reduction of ejection fraction (<30%), Interventricular septum was normal and moderate pericardial effusion was present.
Cardiac MRI was done which shows normal LA and LV was relatively small. LV shows a large saccular defect in the inferior wall, in submitral region. The saccular lesion measures 6.1x4.5x4.0 cm with a neck measuring 4.4cm in diameter. The angiogram is suggestive of normal coronaries.

Fig 1: Showing relatively small LV with a large aneurysmal sac (AN)

Fig.2: Showing normal angiogram.

He was planned for surgical correction. Conventional median sternotomy was done. Cardiopulmonary bypass was achieved by aortic and bicaval cannulation. The aorta was cross clamped under moderate hypothermia and myocardial protection was provided with cold blood antegrade root cardioplegia. We found dense adhesion between the sac and pericardium. We approached through left atrium. Mitral valve was assessed after opening left atrium which was normal. The aneurysm was approached from outside (external approach). Aneurysm was opened, the clots and debris was removed, and excess wall of the aneurysm was excised keeping healthy LV margins. The ‘neck’ of the aneurysm was closed with interrupted pledgetted 3–0 Polypropylene suture.
horizontal mattress sutures in such a way that, the sutures goes through the neck of the aneurysm and through the posterior mitral annulus. The defect in the left ventricle was narrowed down with 2–0 Polypropylene purse string suture and the defect was closed with PTFE Felt using 2–0 Polypropylene interrupted pledgetted sutures. Mitral valve was tested and was found to be competent. Left atrium was closed directly.

Intraoperative pictures:

Fig.3- Showing dense adhesion of the sac with the pericardium.

Fig.4- The aneurysmal sac is separated with a patch of pericardium which could not be separated.

Postoperative echocardiography was done which shows normal mitral valve with no gradient across
the valve. Patient was discharged in stable condition after 7 days. He was followed up after 3 months with echo which suggest no mitral regurgitation and ejection fraction of 51%.

Discussion
LV aneurysm is more common occurrence in post myocardial infarction patients. Submitral Left Ventricular Aneurysm (SMLVA) is a rare lesion that occurs most often in the blacks\textsuperscript{1} and still rarest in Indian population\textsuperscript{4,5}. The prevalence of this lesion among blacks appears to indicate a congenital predisposition\textsuperscript{6}.

SMLVA is basically a congenital outpouching of the LV wall occurring adjacent to the posterior leaflet of Mitral Valve\textsuperscript{2}. It is typically diagnosed in young adults, presenting mostly with features of Mitral regurgitation, congestive heart failure, systemic embolization, arrhythmia and sometimes sudden death. Submitral left ventricular aneurysm seems to be caused by a junctional defect between the cardiac muscle and the fibrous structure of the heart\textsuperscript{3}. The anatomy of mitral valve annulus is such that its 2/3 portion is related to posterior leaflet which is attached to LV myocardium through annular ring. A weakness or disruption of this muscular–fibrous union results in occurrence of submitial aneurysm below mitral leaflet\textsuperscript{3}. Clinical symptoms arise as a result of valvular regurgitation or occasionally from compression of cardiac structures by the aneurysmal sac\textsuperscript{2} or patient can be asymptomatic for many years\textsuperscript{5}. In our patient mitral valve was almost normal and the patient presented with progressive worsening dyspnoea, palpitation and occasional chest pain secondary to gradual decline in ejection fraction. Diagnosis by chest x ray is easy if calcification is present in the aneurysm wall\textsuperscript{7}. Now days, the transthoracic echocardiography is the most accurate diagnostic tool\textsuperscript{8}. The typical location of the aneurysm and the absence of coronary artery disease on angiography confirm the diagnosis of SMLVA. Surgical treatment is mandatory as soon as the diagnosis of SLVA is made, in order to treat the cardiac failure that arises from expansion of the aneurysm and to avoid other potential cardiovascular events. The inherent difficulties associated with the extracardiac approach included inadequate exposure of the mitral annulus, residual mitral regurgitation, and technical difficulties in approaching the aneurysm due to adhesions. Antunes described the transatrial approach in 1987\textsuperscript{2}. We approached through both extra and intra cardiac approach. Surgical failure can be related to the failure to identify additional aneurysm necks or inadequate closure of the aneurysm and lack of support to the mitral annulus leading to recurrent aneurysm formation. Successful repair is dependent on the appropriate understanding of the relationship between the aneurysm, the mitral valve and its annulus. If mitral regurgitation is associated with aneurysm then it is advisable to do mitral ring annuloplasty than direct suturing to prevent recurrence of regurgitation. Mitral Valve Replacement (MVR) is one more option in cases where mitral valve leaflets are distorted or damaged and when it is non-repairable. Also the mitral valve replacement may be an option in cases of postoperative severe mitral regurgitation.

Conclusion
SMLVA is very rare cardiac lesion in Indian subcontinent. The patient may not always present with symptoms of mitral regurgitation but with a normal mitral valve, as in our case. Differential diagnosis of SMLVA must be kept in mind when a young individual present with features of progressive LV dysfunction though it is rare. Echocardiography is sufficient to make the diagnosis. Surgery is the treatment of choice.
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References