Large Submitral Aneurysm Compressing Left Main Coronary Artery: Rare Presentation of a Rare Disease

Parveen Kumar1, Suryakant Jana2, Kumar Kenchappa3, Geetesh Manik4

Abstract
Submitral left ventricular aneurysm is a rare cardiac anomaly that was first reported from African countries and initially termed as “annular left ventricular aneurysm”. Submitral aneurysm (SMA) causes out-pouching of the left ventricular wall, adjacent to the posterior leaflet of the mitral valve. Generally, SMA opens into left ventricle (LV) with a wide mouth and not into left atrium (LA). We report a case of Submitral Aneurysm with two openings: one into LV and the other into LA. This case also highlights the compression of coronary arteries by the submitral aneurysm. Large SMA can cause compression of left main coronary artery rarely. Having a knowledge of this point can help the clinician. SMA generally have an opening in LV but in this case SMA has two openings (One in LV and another in LA). This knowledge can help in proper surgical management.

Introduction
Submitral left ventricular aneurysm is a rare cardiac anomaly that was first reported from African countries initially termed as “annular left ventricular aneurysm”.1 The etiology is debatable; although the current consensus is that they are most likely due to a congenital weakness of the fibro-muscular annuli.2,3 Submitral aneurysm (SMA) causes out-pouching of the left ventricular wall, adjacent to the posterior leaflet of the mitral valve.4 Generally SMA opens into left ventricle (LV) with a wide mouth and not into left atrium (LA).

Case Report
A 19 years old girl presented to cardiology clinic with gradually progressive worsening shortness of breath on exertion associated with exertional palpitations and atypical chest pain for the past 2 years. There was no history of orthopnoea and paroxysmal nocturnal dyspnea. There was no history suggestive of rheumatic fever in past. On clinical examination, she was found to have mitral regurgitation. Chest X-ray revealed cardiomegaly with evidence of pulmonary venous congestion. On two-dimensional transthoracic echocardiogram (TTE), the significant finding was a submitral aneurysm of the LV with the mouth of the aneurysm just below the posterior mitral annulus opening into the LV and into the LA (Figure 1). Three dimensional TTE showed the same findings (Figure 2). LV angiogram showed significant mitral regurgitation (Figure 3). Coronary angiogram done showed systolic narrowing of the lumen of the distal left main, proximal left anterior descending artery & left circumflex artery due to extrinsic compression by a sub mitral aneurysm (Figure 4). This patient underwent successful surgical repair of submitral aneurysm and mitral valve replacement.

Discussion
Submitral aneurysm, although uncommon, should always be considered in differential diagnosis in young patients presenting with mitral insufficiency or signs and symptoms of heart failure. The main clinical manifestations of SMA are mitral regurgitation, thromboembolism, arrhythmias, sudden death and heart failure.5-7 In some cases, it presents as myocardial ischemia due to compression of the coronary arteries by the aneurysm or as cardiogenic shock.8-9 The mechanisms of mitral regurgitation are multifactorial and...
The definitive diagnosis is made by transthoracic echocardiography in the presence of an aneurismal dilatation in submital location behind the posterior mitral leaflet that communicates with the left ventricular cavity through one or more necks. The spatial extent of these aneurysms can be challenging to delineate, thus resulting in surgical failure, which is often attributable to either failure to identify additional aneurysm necks (50% of failures) or inadequate closure of the aneurysm. Preoperative assessment has previously been done using transthoracic echocardiography, 2-dimensional TEE, and invasive angiography. Real-time three-dimensional echocardiography is not necessary for the diagnosis of SMA. However it is very useful in the evaluation of the relationship of the aneurysm with the other cardiac structures. In the evaluation of the anatomical characteristics of the aneurysm allowing the identification of one or more apertures through which aneurysm communicates with the left ventricle, providing additional data to two-dimensional echocardiography, and improving the plan for surgery.

References