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Unusual Cause of Congestive Heart Failure with Severe Mitral Regurgitation

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Sub mitral aneurysm is a rare cardiac anomaly with varied clinical manifestations, usually due to congenital defect adjacent to posterior leaflet of mitral valve. We report 50-year-old male patient with submitral aneurysm who presented with features suggestive of congestive heart failure and severe mitral regurgitation. Echo cardiography and cardiac MRI aid in precise non-invasive diagnosis.

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INTRODUCTION

Sub mitral aneurysm is a rare cardiac anomaly characterized by outpouching of LV wall which arises in the fibrous ring usually due to congenital defect in the region of atrioventricular groove in the posterior portion of mitral annulus. It is a type of annular subvalvular LV aneurysm thought to be due to weakness of submembraneous curtain.(7) or internal cardiac dehiscence developing at junction between myocardium and fibrous skeleton heart. It was initially documented among Nigerians and the south African Bantu,

subsequently in other parts of world including India. Clinical features documented include mitral incompetence, systemic embolism, angina pectoris, myocardial infarction, tachyarrhythmias and cardiac failure (3). We report a 50-year-old male with submitral aneurysm presented as severe mitral regurgitation with congestive heart failure.

A 50-year-old male presented with the complaints of progressive breathlessness (NYHA -III) and bilateral leg swelling for the past two and half months along with history of paroxysmal nocturnal dyspnea.



Fig:1 submitral aneurysm with severe mitral regurgitation(Arrow) LV -Left Ventricle, RV- Right Ventricle

On examination, his blood pressure was 120/80mm Hg measured in right arm in sitting posture with pulse rate of 88/minute regular in rhythm, felt in all peripheral arteries. He had raised jugular venous pressure,6cm above the sternal angle with bilateral pedal edema. On palpation, he had hyper dynamic LV apex, left parasternal heave and apex was shifted to the left anterior axillary line in the sixth intercostal space. P2 was palpable. On auscultation, S1S2(+), S1 was soft and S2 close split, P2 loud and LV S3 were present. He had a 3/6 pansystolic murmur in the apex which conducted to the axilla along with a 2/6 mid systolic murmur in the pulmonary area. Abdominal examination revealed liver span of 12cms. ECG showed heart rate of 100/ min, sinus rhythm, PR interval 0.20sec, low voltage complex in the frontal leads, poor R wave progression from V1 to V5 with transition at V6 and left ventricular hypertrophy. His chest Xray -PA view showed cardio-thoracic ratio of 75% with evidence of pulmonary venous hypertension.



Fig:2 large submitral aneurysm (Arrow)

LV -Left Ventricle, RV- Right Ventricle, LA- Left Atrium

2D echocardiogram revealed large submitral aneurysm (pointed arrow) arising from posterior wall of left ventricle adjacent to posterior mitral leaflet with demonstrable color flow between LV and aneurysm,

measuring approximately in Plax view 5.1 cm X 3.7 cm with neck size of 2.2 cm and severe mitral regurgitation. There was mild aortic and tricuspid regurgitation with estimated pulmonary artery pressure of 56 mm Hg. In

addition, 17mm pericardial effusion was present posterior to left ventricle with preserved LV and RV function.

Cardiac MRI showed large submitral pseudoaneurysm of LV arising from posterior mitral annulus involving posterior mitral leaflet with layered small peripheral thrombus along the wall of the aneurysm, dilated left ventricle with good ventricular function (LVEF of 59%) and borderline high right ventricle volume with good ejection fraction (RVEF of 47 %) and severe mitral regurgitation. Coronary angiogram showed normal LAD, RCA and major OM, separate LAD and LCX origin with distal LCX occlusion. LCX was not dominant.

DISCUSSION

Submitral aneurysm is a rare cardiac defect dates back to 1812, first described by Corvisart with varied etiologies and clinical manifestations. Though etiology is thought to be congenital, there have been associations with conditions like Takayasu's arteritis and tuberculous pericarditis. It can be of several types namely having single neck or multiple necks or involving entire posterior mitral annulus (4). It can occur singly or coexist with aortic annular aneurysm.

Complications include congestive heart regurgitation, failure. severe mitral compression of LCX, arrhythmias, rupture, thrombus, calcifications and pericardial adhesions.

LVH, low voltage complexes, nonspecific ST segment and T wave changes, signs of myocardial ischemia or infarction, prolonged PR interval, left axis deviation are some of the ECG changes described(3).

Abnormal bulge in the left heart border and calcification are some of the documented radiological signs. Doppler echocardiography, MRI aid in noninvasive diagnosis.

Treatment of this condition is medical stabilization followed by surgical repair which poses technical difficulties due to fibrous

adhesions and proximity of left circumflex artery to ostium of aneurysm. In conclusion consideration of submitral aneurysm, in the diagnosis of young patients with congestive heart failure and severe mitral regurgitation is crucial.

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