

# STORMY COURSE OF A HUGE SUBMITRAL ANEURYSM CAUSING LOW CARDIAC OUTPUT STATE

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Submitral aneurysm is a rare structural abnormality of congenital or acquired aetiology. Most reported cases are from Africa. Unless promptly treated surgically this condition is invariably fatal. We report a case of a young Indian male who presented with dyspnea of recent onset, diagnosed to have a massive submitral aneurysm causing low cardiac output and compression of cardiac structures.

**KEY WORDS:** Submitral aneurysm · Congenital · Huge.

## INTRODUCTION

Submitral aneurysm is an exceedingly rare cardiovascular structural abnormality of congenital or acquired aetiology characterized by outpouching of the left ventricular wall, classically occurring adjacent to the posterior mitral leaflet. Although most of the reported cases are from Africa,<sup>1,2)</sup> cases have been reported from other countries as well, including India.<sup>3)</sup> The aetiopathogenesis of this condition still remains enigmatic to the medical community. Here we describe the case of a young Indian male (hailing from the Rajasthan state in Western India) who presented with this condition. The condition was provisionally diagnosed with two dimensional transthoracic echocardiography and confirmed by cardiac catheterization. He had a stormy course which started with acute onset dyspnea culminating in death within a span of 26 days. We report this case to highlight the atypical presentation, and to reemphasize the lethality of this condition unless promptly corrected surgically.

## CASE

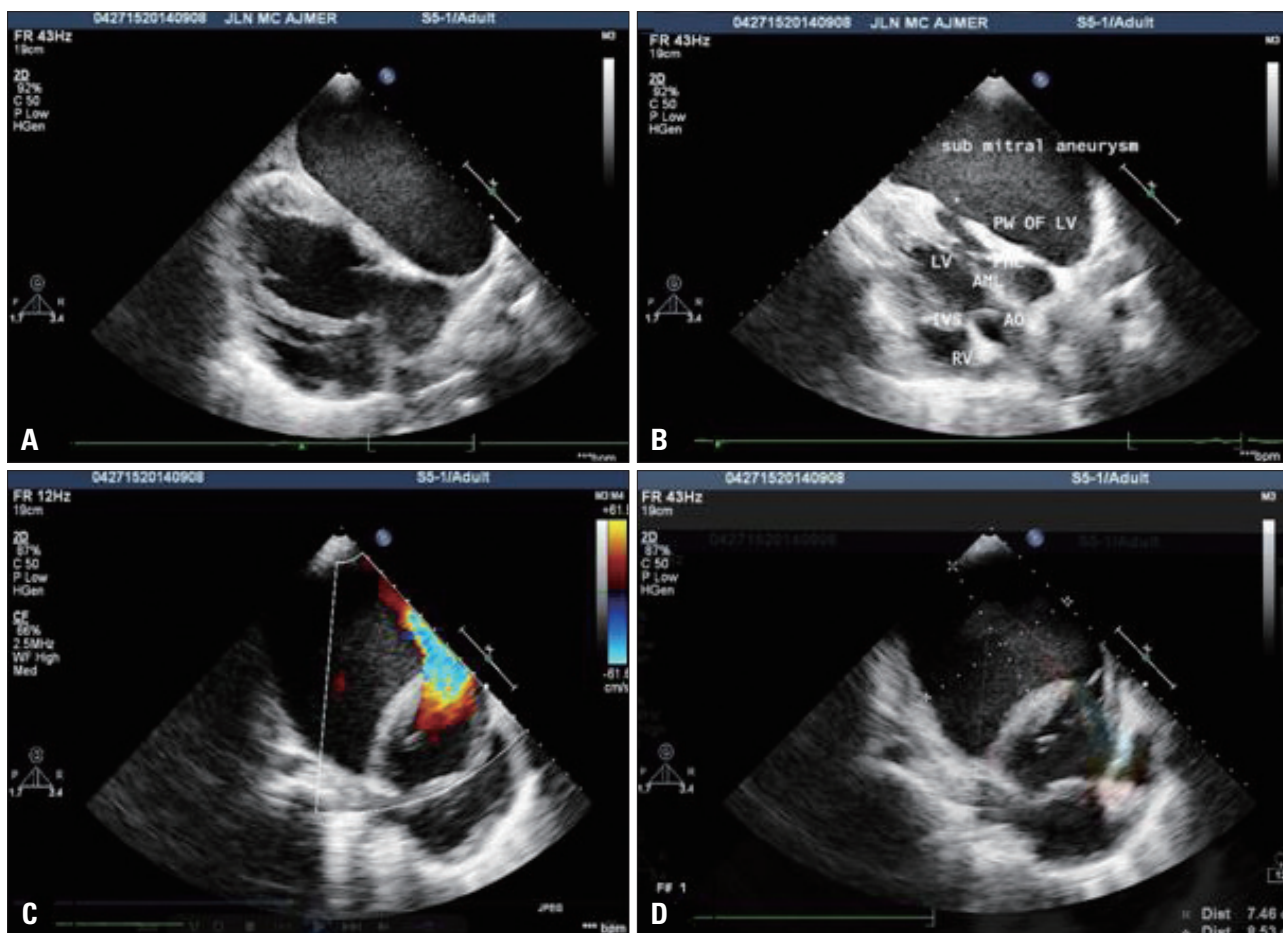
A 24-year-old male, presented with history of sudden onset shortness of breath of 20 days duration, which had worsened from New York Heart Association (NYHA) class I to NYHA class IV over a course of 3 days along with, orthopnea and ex-

treme fatigue. The patient had been completely asymptomatic prior to these 20 days. There was no history of specific triggering events or undue exertion prior to symptom onset. General physical examination was unremarkable. The patient was profoundly dyspneic at rest. His pulse rate was 110/min, feeble in volume and of no special character. Blood pressure was 82/58 mm Hg in the left upper limb in sitting position. Jugular venous pulse was elevated to 12 cm with prominent v waves. He had a left parasternal heave and on auscultation the cardiac apex was laterally displaced 3 cm lateral to the left mid clavicular line. The first heart sound was soft, pulmonary component of second heart sound was loud, left ventricular third heart sound was present and a mitral regurgitation murmur of grade 4/6 was noted. Haematological and biochemical investigations were within normal limits. Chest X-ray revealed cardiomegaly with straightening of the left heart border, electrocardiogram showed sinus tachycardia and left atrial abnormality. On two dimensional echocardiography, modified apical 4 chamber view revealed a grossly dilated (8.53 × 7.46 cm) submitral aneurysm<sup>4,5)</sup> arising from the wall of the left ventricle adjacent to the posterior mitral leaflet. It is one of the largest aneurysms ever reported.<sup>4,5)</sup> The aneurysm was compressing the left ventricle practically preventing any reasonable cardiac output from the left ventricle (Fig. 1, Supplementary movie 1). The

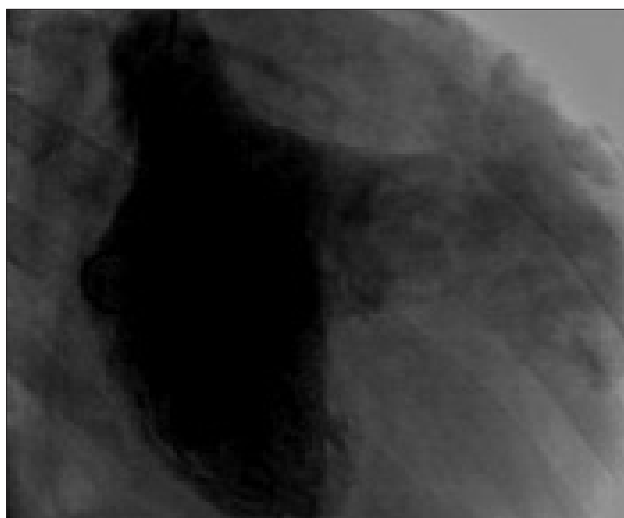
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**Fig. 1.** Two dimensional echocardiographic images acquired using Philips IE33/S5-1 probe. A: Apical 4 chamber view showing a large cystic mass of size comparable to both the ventricles put together. B: Same view with plane shifted anteriorly to visualize the communication (\*) between the cyst and the left ventricle (just beneath the posterior mitral leaflet). C and D: Parasternal short axis view at the basal level (with and without color Doppler, respectively), showing the giant aneurysm compressing left ventricle, with bidirectional flow between the two, largest dimension of the aneurysm being 7.46 × 8.53 cm. AML: anterior mitral leaflet, PML: posterior mitral leaflet, PW: posterior wall of LV, IVS: inter ventricular septum, Ao: aorta, LV: left ventricle, RV: right ventricle.



**Fig. 2.** Left ventricular angiogram in right anterior oblique view with 30° angulation showing the giant aneurysm in free communication with the left ventricle at the basal level, its huge dimensions completely displacing the left ventricle from its original site. Pig tail catheter *in situ* seen.

compressive effect of the aneurysm was also hindering venous blood from entering the collapsed right ventricle. Pulse wave Doppler revealed bidirectional flow between the left ventricle and the aneurysm. On cardiac catheterization, left ventricular angiogram in right anterior oblique 30 degree view further confirmed the diagnosis (Fig. 2, Supplementary movie 2). His coronary angiogram was normal. The patient and his relatives were informed about the lethality and poor prognosis of the condition unless surgically corrected and was referred to the surgical unit on an urgent basis. Despite the expedient management and our best efforts for faster intervention, the patient succumbed to the condition before corrective surgery could be attempted.

## DISCUSSION

The very first case of submitral aneurysm to be described dates back to 1812.<sup>4)</sup> Submitral aneurysm as an entity became clearly established after an eloquent series of 12 cases reported in 1962.<sup>2)</sup> The first case reported from Asia was from India in

1978.<sup>3)</sup> While the focal clustering in the sub-Saharan African population suggested an acquired aetiology of infectious nature,<sup>1)</sup> the focused concentration in a particular ethnic group hinted at a genetic basis. The attributed causes have been varying what with syphilis, tuberculosis, Chagas disease and endocarditis of the mitral valve dominating the picture in the 1970s to vasculitis and valve replacement surgery which were implicated as probable causes later on.<sup>1)5)</sup> Although most of the initial records were exclusively from Africa, non-African patients too have been reported subsequently.<sup>4)</sup> Possibility of it being a congenital defect surfaced with reports illustrating co-existent aneurysms.<sup>1)6)</sup> Antenatal diagnosis by fetal echocardiography allows anticipatory management although most of the cases diagnosed in fetal life have remained asymptomatic till early childhood.<sup>7)8)</sup>

The underlying pathology is believed to be faulty fusion between the left ventricular musculature and the fibrous skeleton of the heart at the left atrium-mitral valve region. The consequence of this is electrical isolation of this segment creating an arrhythmogenic nidus potentially capable of triggering complex and fatal arrhythmias, which are the most common cause of death in this condition. Enlargement of this segment and displacement of the posterior mitral annulus leads to failure of the supporting apparatus and failure of leaflet coaptation with secondary mitral regurgitation fuelling further enlargement and volume overload of the left ventricle.<sup>6-8)</sup> Although congenital, this condition usually manifests by the middle of the third decade.<sup>5)</sup>

There have been few reports and series on surgically corrected submitral aneurysms.<sup>5)9)</sup> While results at early follow up have been promising,<sup>9)</sup> data on long term follow up and outcomes are lacking. The condition is uniformly fatal unless surgically corrected and patients have succumbed to the condition even after surgical correction.

Our case was remarkable for the massiveness of the aneurysm (among the cases reported till date, few have had bigger dimensions),<sup>4)5)10)</sup> and the puzzling symptoms caused by the altered haemodynamics owing to the compression of the right and left ventricles by the enormity of the aneurysm which was causing a low cardiac output state (responsible for the patient's extreme fatigability) and congestive features attributable to the impaired venous return.

In conclusion, although rare, in cases presenting with acute onset congestive cardiac failure, especially in a young adult, the possibility of a submitral aneurysm should always be kept in mind. Echocardiography is diagnostic in most cases, and re-

veals an aneurysmal dilatation beneath the mitral valve and behind the posterior mitral leaflet, which has communication with the left ventricle through one or more necks. Once diagnosed emergent surgical intervention is of utmost importance and shall be the decisive factor in determining the outcome as a stormy course as in the present case can prove rapidly fatal unless operated in time.

#### SUPPLEMENTARY MOVIE LEGENDS

Movie 1. Two dimensional echocardiography obtained using Philips iE33/S5-1 probe (Philips Medical Systems, Bothell, WA, USA). Modified apical 4 chamber view showing a huge sub mitral aneurysm in free communication with the left ventricle (just beneath the posterior mitral leaflet) compressing and displacing both the ventricles.

Movie 2. Left ventricular angiogram in right anterior oblique view with 30° angulation, showing a giant aneurysm in free communication with the left ventricle at basal level, its huge dimension, completely displacing the left ventricle from its original site. Pig tail catheter *in situ* seen.

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