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Mitral Annular Disjunction with bileaflet mitral valve prolapse

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ABSTRACT

Mitral annular disjunction (MAD) is a structural abnormality of the mitral valve whereby there is a distinct separation of the mitral valve annulus-left atrial wall continuum and the basal portion of the posterolateral ventricular myocardium, a region which would normally be attached. It is usually associated with myxomatous mitral valve disease. MAD has been reported in various studies since the last four decades as constituting around 42-90% of patients with myxomatous mitral valve disease and mitral valve prolapse. It is often associated with arrhythmias ranging from benign ventricular ectopics to malignant ventricular fibrillations causing sudden cardiac deaths in the young. This condition seems to be more common in women. Last few decades have witnessed many case reports and prospective studies about this entity enhancing our understanding about its pathophysiology. We report a case of a 34 year old male with mitral valve prolapse and MAD resulting in severe mitral regurgitation who underwent mitral valve replacement.

Key words: Mitral annular disjunction, Myxomatous mitral leaflet prolapse, Mitral regurgitation

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INTRODUCTION:

Mitral annular disjunction (MAD) is characterized by a discernible separation between the left atrial wall-mitral valve junction and the proximal, basal left ventricular wall. The term mitral annular disjunction was first used by Bharati et al [1] in 1981 and described by Hutchins et al [2] in 1986. Hutchins examined 900 hearts after post-mortem arteriography from autopsies of adults at the Johns Hopkins Hospital. He showed that floppy mitral valve was significantly associated with disjunction of the mitral annulus. Since then, there has been a lot of research about the pathophysiology and mechanisms of MAD. There is enough evidence implying that MAD is associated with ventricular arrhythmias and sudden cardiac deaths. Thus, the significance of early detection and subsequent management of MAD cannot be overemphasized.

CASE PRESENTATION:

A 34-year-old male patient presented with complaints of palpitations and fatigue since one month. His electrocardiogram showed T wave inversions in inferior leads, ventricular premature complexes(VPCs) and ventricular bigeminy. Echocardiography revealed myxomatous mitral valve with mitral valve prolapse (MVP) with severe MR (mitral regurgitation). Both the mitral leaflets were thickened and prolapsing. Anterior and posterior mitral leaflets were displaced 8 mm and 12 mm respectively, away from the annulus towards the left atrium suggesting mitral annular disjunction. Left ventricle was significantly dilated. 24-hour Holter monitoring was done to characterize the ventricular arrhythmias. VPC burden was less than 1%, though VPCs were of multiple morphologies. Cardiac magnetic resonance imaging reported prolapse of both leaflets, severely dilated left ventricle with normal systolic function, bileaflet mitral valve prolapse with MAD, severe MR and partial fibrosis of posteromedial papillary muscle. Patient was taken up for surgery. Under general anesthesia, cardiopulmonary bypass was instituted with standard aortic and bicaval cannulation and aorta was cross clamped. Myocardial protection was by antegrade cold blood St.Thomas

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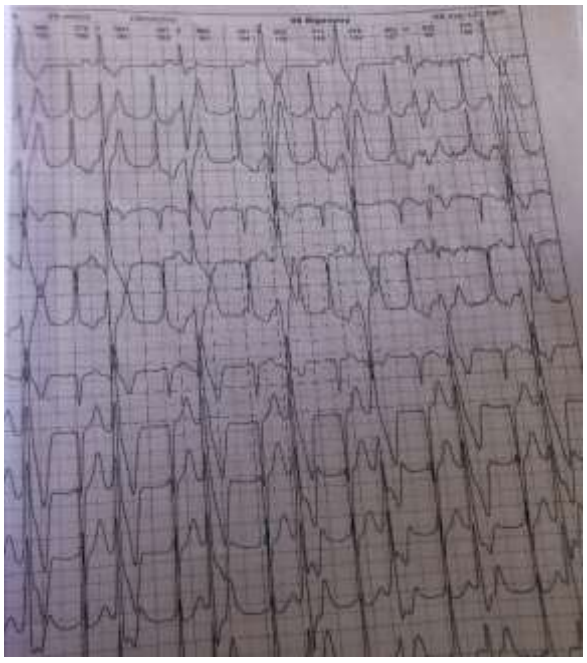
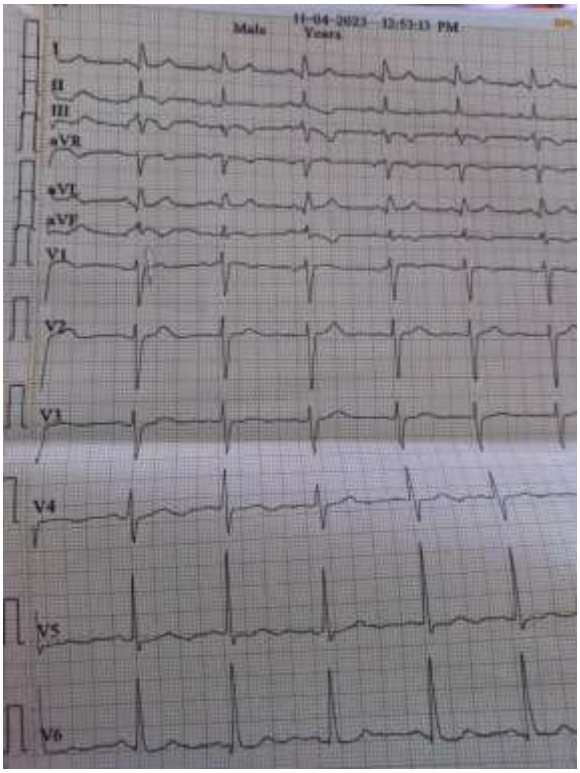
cardioplegia solution. The mitral valve was approached through the right atrium trans-septally. Anterior mitral leaflet and posterior mitral leaflets showed myxomatous changes with elongated chordae, dilated annulus with disjunction. Both mitral leaflets were excised. Mitral annulus was sized to be 29mm with St. Jude Medical valve sizer and 29mm St. Jude Medical mechanical valve seated and secured with 2-0 polyester pledgeted sutures. The tricuspid valve was assessed and found to be competent on saline insufflation test. Interatrial septum and right atrium were closed in a continuous manner with prolene sutures in two layers. Patient was gradually weaned off cardiopulmonary bypass and routine chest closure was done. Patient was shifted to intensive care unit in a hemodynamically stable condition in normal sinus rhythm with minimal inotropic support.





Picture 1 Picture 2

ECG : Picture 1 -before surgery. Picture 2- two years after surgery

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DISCUSSION:

Mitral annular disjunction was initially described over 30 years ago in an autopsy report of 900 hearts. At that time, MAD was not attributed to clinically adverse outcomes and as such it received little attention. However, over the decades, studies have deduced that MAD may be of more clinical significance than previously thought [1,2]. In MAD, junction of mitral leaflet with the atrial wall is separated from the left ventricular wall; it is functionally decoupled with a paradoxical annular dynamics in which the mitral annulus moves in concordance with the left atrium during the cardiac cycle instead of with the left ventricle, described as systolic curling [3]. This leads to an expansion and flattening of mitral valve annulus in systole increasing the stress on mitral leaflets' chordae tendinae. Abnormal mechanical stretch causes inflammation, local ischemia and replacement fibrosis becoming the initiating area for arrhythmogenesis [4]. 33% of patients with MAD present with frequent ventricular ectopics and 10% with severe arrhythmic events [3,4]. Life threatening arrhythmias are more often seen in female patients and those with higher burden of ventricular ectopy (VE), bileaflet myxomatous mitral valve degeneration, MR severity, and presence of flail leaflets [5].

Diagnostic role of various cardiac imaging modalities like transthoracic with transesophageal echocardiography and cardiac magnetic resonance imaging cannot be overemphasized [4,5]. The best view to assess MAD on echocardiography is the parasternal long axis view. Transesophageal echocardiography may be used to quantify the lesions better. A distance of more than 5mm between posterior mitral leaflet insertion and left ventricular myocardium on transesophageal echocardiography is considered diagnostic. Cardiac magnetic resonance has the additional advantage of characterizing associated myocardial and papillary muscle fibrosis apart from depicting circumferential extent of MAD and systolic curling on T mapping /late gadolinium enhancement. The separation distance has been described to vary between 1 and 15mm [3,4,5]. Dejgaard et al found that MAD most frequently involves the

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left ventricular anterior wall (28%) followed by inferior wall (26%) and posterolateral wall (9%). MAD involves the mitral annulus circumferentially with a range of 30^0 to 240^0 more exclusively along the posterior mitral leaflet [6].

Treatment of severe mitral regurgitation in these cases warrants early surgical intervention in the form of valve repair or replacement. Surgical intervention resolves the altered annular dynamics, reduces arrhythmia burden and hence minimizes the chances of sudden cardiac death. Catheter ablation can be attempted in symptomatic, drug refractory ventricular arrhythmias especially in those who have a well defined ventricular ectopic foci on electrophysiological mapping [7]

Conclusion :

Mitral annular disjunction must be looked for in cases of mitral regurgitation; more so, in myxomatous mitral valve disease. Early recognition and prompt surgical interventions can be life saving from malignant ventricular arrhythmias that can result in sudden cardiac death. Since this entity is gaining attention, more prospective studies may help understand the altered mitral annular dynamics and chart appropriate interventions curtailing the disease progress.

REFERENCES:

1. Bharati S,Granston AS, Liebson PR, Loeb HS, Rosen KM, Lev M. The conduction system in mitral valve prolapse syndrome with sudden death. Am Heart J. 1981; 101:667–70.

2. G M Hutchins, G W Moore, D K Skoog: The association between floppy mitral valve with disjunction of the mitral annulus fibrosus New England Journal of Medicine 1986;314(9):535-40.

3. Chakrabarti AK, Bogun F, Liang JJ. Arrhythmic Mitral Valve Prolapse and Mitral Annular Disjunction: Clinical Features, Pathophysiology, Risk Stratification, and Management. J. Cardiovasc. Dev. Dis. 2022; 9: 61

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 07, 2024

4. Miller MA, Adams DH, Pandis D, Robson PM, Pawale A, Pyzik R and et al. Hybrid positron emission tomography/magnetic resonance imaging in arrhythmic mitral valve prolapse. JAMA Cardiol. 2020; 5:1000–5.

5. Sriram CS, Syed FF, Ferguson ME, Johnson JN, Enriquez-Sarano M, Cetta F, et al. Malignant bileaflet mitral valve prolapse syndrome in patients with otherwise idiopathic out-of-hospital cardiac arrest. J Am Coll Cardiol. 2013; 62:222–30

6. Dejgaard LA, Skjølsvik ET, Lie ØH. The mitral annulus disjunction arrhythmic syndrome. J Am Coll Cardiol 2018;72(14):1600–9.

7. Wu S, Siegel RJ. Mitral annular disjunction: A case series and review of the literature. Front. Cardiovasc. Med. 2022;9:9760-66.