Original Research Paper

Volume - 13 | Issue - 02 | February - 2023 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Cardiology

A RARE CASE OF MITRAL VALVE WITH SINGLE LEAFLET

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ABSTRACT A forty-year-old male is diagnosed on echocardiography as an isolated uni-cuspid mitral valve deformity leading to MS, MR, TR, and PAH. Almost all cases of uni-cuspid mitral valve are associated with absent/hypoplastic posterior valve leaflet; which can be either be associated with other cardiac malformations or isolated entity; as in our case. None of the cases described as yet had absent/hypoplastic anterior valve leaflet in available literature.

KEYWORDS :

INDORDUCTION:

Isolated uni-cusp mitral valve is a rarest condition, has seldom been described in literature. However, it has been described in association with other conditions. Few cases of uni-cuspid Mitral valve have been described in association with other cardiac malformations, in one such case it was associated with ASD (ostium secundum) with MR and PAH (Espinola-Zavaleta et al, 2019). In most of the cases including our study the posterior mitral leaflet was absent or hypoplastic as described in limited available academic literature.

Case Study:

A 38-year old 50 Kg male, 174 cm tall, teetotaller and non-smoker, presented with the complaints of gradually increasing dysponea on exertion for last 10 years. Four years ago he had undergone cardiac evaluation at other center by 2D- ECHO, cardiac catheterisation, ECG and was diagnosed a case of CHD- moderate MR, severe TR, and Severe PAH. Despite been receiving medication since then, he has benefitted slightly, however, exertional dysponea has been persisted and increased gradually. In Addition, he also had weakness and weight loss. Moreover his past history revealed that he had contracted measles (he was not immunized against measles) at the age of around 10 years and approximately 10 more years later he had pulmonary TB for which he had received 9 months' anti-tubercular treatment and felt he that he has developed persistent cough since then. Then from the age of around 28 years he gradually developed progressive dysponea on exertion and presented to us now.

On examination of the patient the AP diameter of the chest was increased (Barrel shaped) although lungs were clear. Apex was hyper dynamic and apical impulse felt just lateral to mid axillary line at 5-6th intercostal space. The parasternal heave over left sternal edge (LSE) was present and on auscultation grade 3/6 pan-systolic murmur in the apical region was present and was radiating towards axilla. Furthermore, over LSE the first heart sound was loud and preceded by mid diastolic murmur with pre-systolic accentuation which was loudest in the tricuspid region. Current ECG showed P pulmonale, with right axis deviation and incomplete RBBB (Figure 1); his ECG 4 years back did not showed any RBBB (Figure 2).

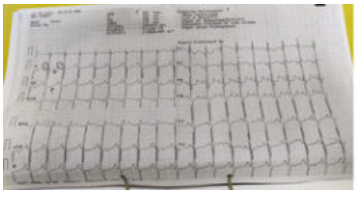

Figure 1: current ECG

Figure 2: Previous ECG

Current 2D-ECHO revealed both atria and ventricles were dilated (Figure 3 a & b). The mitral valve showed uni-cusp in the form of anterior mitral leaflet with normal valve apparatus and posterior leaflet was absent although the other three valves were normal (Figures 4, 5)

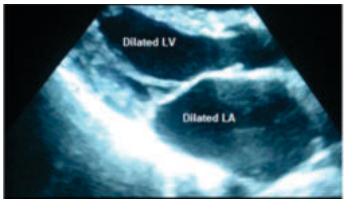

Figure 3 a: ECHO shows dilated LV & LA

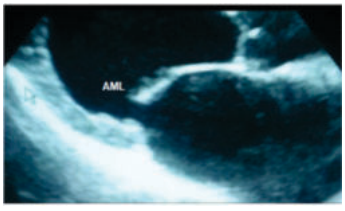

Figure 3 b: ECHO LV & LA with uni-cusp

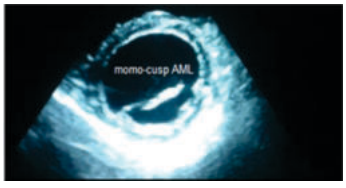

Figure 4: ECHO short axis view

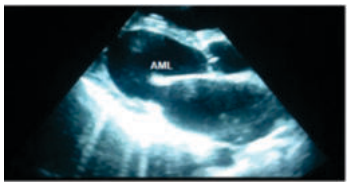

Figure 5: ECHO no posterior cusp (PML)

The colour Doppler study revealed mild MS (MG/PG-5/15 mm HG), severe MR (Figure 6, 7) with posterior jet, mild to moderate TR with PAH, RVSP 86 mmHg. Earlier ECHO (4 years back) was similar although uni leaflet of mitral valve was not reported then.

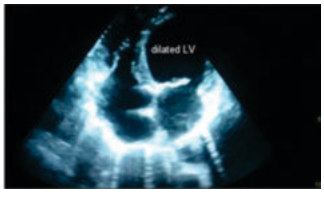

Figure 6: Apical four chamber

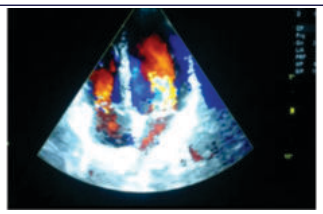

Figure 7: apical four chamber with colour flow study

In an earlier study done elsewhere, described a case of uni-cuspid mitral valve in a known case of Marfan's syndrome also associated with ostium secundum ASD with mitral regurgitation and pulmonary arterial hypertension (Espinola-Zavaleta et al, 2020). Similarly, another case of an isolated uni-leaflet mitral valve, the patient was in a 28-year-old asymptomatic female with minimal mitral regurgitation (Vincelj, J et al 2013). Likewise In a case of 40-year-old female in which post leaflet of mitral valve was hypoplastic with mitral regurgitation, simillarly, there was an another case of uni cuspid mitral valve but without mitral regurgitation and symptoms (Espinola-Zavaleta et al, 2002).

DISCUSSION:

All the cases of uni-cuspid MV including this one; the posterior leaflet was absent or hypoplastic, some of them were association with OS ASD, Marfan's etc. whereas others were isolated. Since this patient has developed severe pulmonary arterial hypertension hence advised regular cardiology follow ups and medical/surgical management. This case was an isolated uni-cuspid valve and was not associated with any other congenital conditions.

Conflict of interest: none

Funding: self

Purpose: Academic

Consent:patient's consent has taken for the article (attached: appendix)

Ethical Approval: Not Applicable

Availability of data and materials: Not Applicable

Abbreviations Used:

MS Mitral Stenosis
MR Mitral Regurgitation
TR, Tricuspid Regurgitation
PAH pulmonary Arterial Hypertension
OS ASD Ostium Secundum atrial Septal Defect
RBBB right Bundle Branch block
RVSP Right ventricular Systolic Pressure

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