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Ebstein's Anomaly With Mitral Valve Prolapse Surviving Till 52 Years Without Treatment : A Rare Case Report

KEYWORDS: Anasarca,Cardiac cirrhosis, Mitral valve prolapse.

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ABSTRACT *Ebstein's anomaly is a congenital heart defect in which the septal and posterior leaflets of the tricuspid valve are displaced towards the apex of the right ventricle of the heart.Here we are reporting a rare case of 52years male who presented with generalised anasarca ,cardiac cirrhosis and mitral valve prolapse without any previous medical illness.*

SUMMARY:
A 52year old male patient with complaints of anasarca,shortness of breath with pansystolic murmur in tricuspid area mid systiloc click with late systolic murmur in mitral area ,routine blood investigations which showed normalcy.chestxray showing gross cardiomegaly.ultrasound showing cirrhosis of liver,gross ascitis.2D echo showedebstein's anomaly with mitral valve prolapse,which is confimed with cardiac catheterization

CASE REPORT:
- A 52 year old male,farmer by occupation presented with complaints of on&off episodes of pedal edema since 10years ,present episode lasted since 2months insidious in onset,gradually progressive initially present upto the ankle now extended till knee
- Patient also complaining of abdominal distension since 2years insidious in onset,gradually progressive aggravated since 2months
- Facial puffiness since 1month
- SOB since 15days grade 4 NYHA associated with dry cough
- H/O Orthopnea and Paroxysmal nocturnal dyspnea present
- Episodes of palpitations and syncopal attacks during exertion present
- No H/O chest pain
- No H/O cyanotic spells
- No H/O recurrent respiratory tract infections
- Past H/O No history of similar complaints in the past
- No H/O diabetes mellitus,hypertension
- Not a known alchoholic,known smoker since 30 yrs 3 to 4 cigarettes per day
- General physical examination - moderately built and poorly nourished,bilateral pitting type of pedal edema upto knee · Vitals -pulse:88bpm regular, highvolume,BP 100/60 mm hg,in sitting position in right arm

CARDIOVASCULAR SYSTEM EXAMINATION:
Apical impulse is at 5th and 6th intercostal spaces diffuse and hyperdynamic

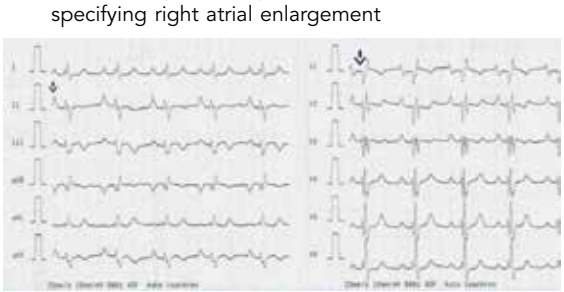
JVP raised 8cms above the sternal angle ,with prominat " a" waves and "cv " complexes ,giant ''v'' waves

Cardiac borders -enlarged

Auscultation:
Mitral area:mid systolic click followed by high pitched ,late systolic murmur of grade 3 radiating towards axilla better heard with diaphragm of stethoscope during inspiration

Tricuspid area:high pitched soft and blowing pansystolic murmur of grade3 radiating towards epigastric area better heard with diaphragm of stethoscope accentuated during inspiration(carvallo s sign) in supine position

LAB INVESTIGATIONS
- Heamoglobin:12.9gm/dl
- ESR 50mm/1hr
- Complete urine examination normal
- HbsAg,HCV are non reactive
- Serum creatinine 1.1 mg/dl
- USG abdomen:cirrhosis of liver,grossascitis,bilateral minimal pleural effusion
- ECG showing sinus rhythm with tall P waves in lead II specifying right atrial enlargement



2D ECHO showing ebstein's anomaly plus MVP,grossly dilated right atrium,tricuspid valve apically displaced,anterior mitral leaflet prolapsed,paradoxicalIVS,minimal pericardial effusion

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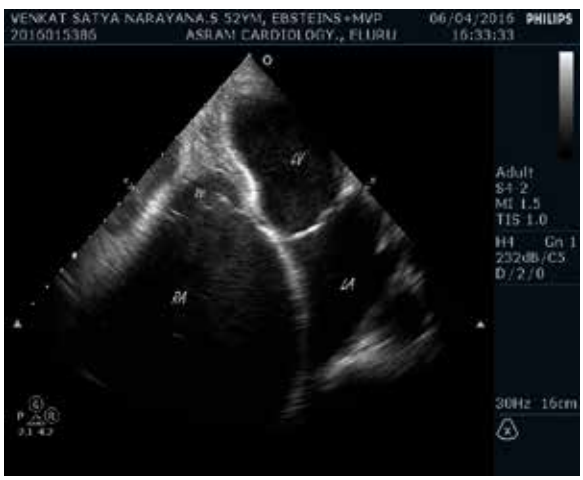
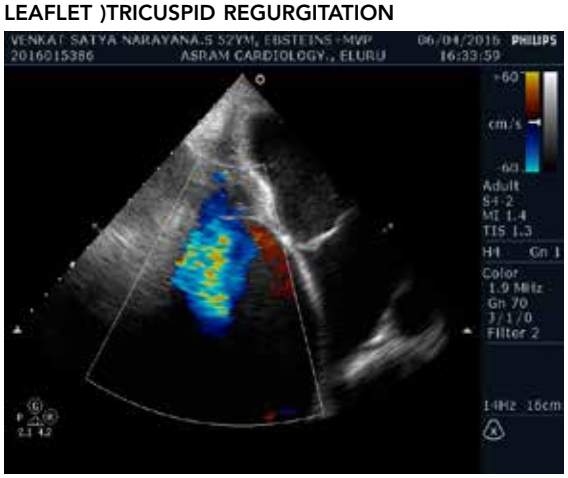
Tricuspid valve : Tricuspid regurgitation
jet velocity –2.6 m/sec ,severe low pressure TR
Mitral valve : mitral valve prolapse > 2 mm ,anterior mitral leaflet prolapse

PAH : Pulmonary capillary wedge pressure -22 mm of Hg
Mild PAH present

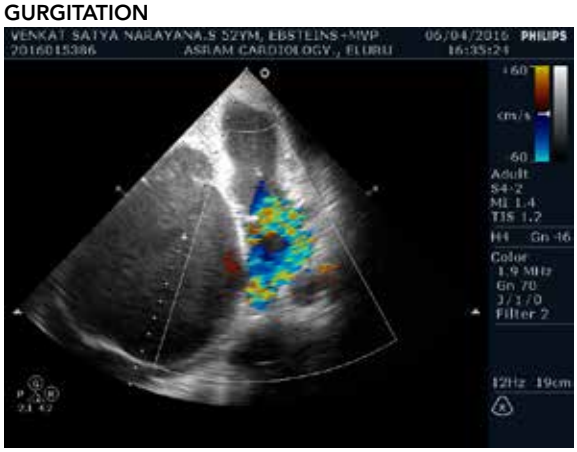
1.GROSSLY ENLARGED RIGHT ATRIUM EBSTEINS ANOMALY



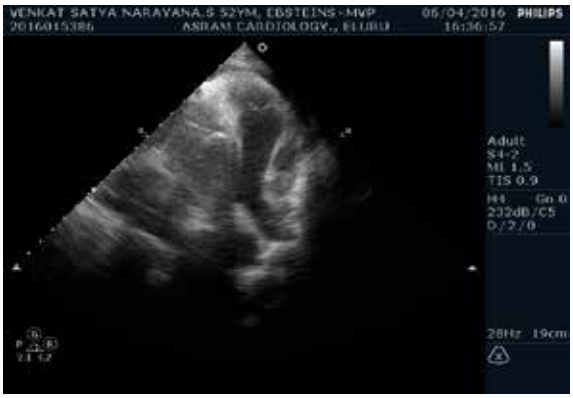
2.PICTURE SHOWING SEVERE LOW PRESSURE (SEPTAL LEAFLET)TRICUSPID REGURGITATION



3. PICTURE SHOWING MITRAL VALVE(ANTERIOR MITRAL LEAFLET) PROLAPSE WITH SEVERE MITRAL REGURGITATION



4.NORMAL PULMONARY ARTERY



CARDIAC CATHETERISATION :
right atrial presuure : systolic/diastolic 15/10 mm of Hg,

Right ventricular pressure :systolic/diastolic 20/10 mm of Hg
left atrial pressure : systolic /diastolic -20/15 mm of hg

DISCUSSION
Ebstein anomaly is characterised by apical displacement of the septal (and often the posterior) leaflet of the tricuspid valve into the RV CAVITY .the RV is therefore divided into a proximal " atrialized " portion and a distal " functional " portion .the effective volume of the functional RV is often small[1]

An ASD or patent foramen ovale is present in more than 1/3 rd of cases . other associations lesions far less common are pulmonary stenosis, VSD, & PDA .ebsteins anomaly is commonly found in patients witrh CCTGA·Patients presenting in infancy represent the worst end of the spectrum with severe tricuspid regurgitation and a high incidence of associated abnormalities such as pulmonary stenosis or atresia[2]

In adults ,palpitations secondary to atrial arrythmias is the most common clinical presentation. upto 20% of unoperated patients may die from HEART FAILURE &approximately 5 % may die suddenly ,presumbly from atrial or ventricular arrythmias.heart failure results from RV dysfunction & tricuspid regurgitation & may be exacerbated by LV fibrosis and dysfunction[3]

Dysrhythmias were the mostfrequent cause of death and often persisted after surgical correction[4]

When the leafletdysplasia is particularly severe and associated with right ventricular myocardium hypoplasia, the clinical manifestation occurs very early, even in fetal or infant life, with an early death due to congestive heartfailure[5]

There are numerous variations on the surgical management of ebstein anomaly. [6]

Valve repair may not be feasible in most severe forms of ebstein anomaly in which the long anterior leaflet of the tricuspid valve is adherent to the RV endocardium and there is almost complete atrialization of the RV valve replacement is good option in these patients[6,7]

The results of the surgery in patients with poor RV function can be improverd considerably if the RV is unloaded by a concomitant CAVOPULMONARY(GLENN) SHUNT also

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known as ONE AND HALF VENTRICLE REPAIR[6,7]

In most severe cases with near complete absence of a functioning RV auniventricular repair leading to a Fontan palliation may be necessary[8,9]

Intra operative radiofrequency or cryoablation (modified MAZE) and division of accesory pathways that are usually located in the posteroseptal or RV free wall is recommended at the time of valve repair or replacement [10]

Overall surgical mortality was 13% in a multicenter analysis with young age at the time of operation as the only multivariate risk factor [6]

The long time survival and functional capacity of operated patients is very good[11]

CONCLUSION:

survival of cases of ebstein's anomaly has been noted till adulthood hence careful evaluation of all cases is necessary to improve the outcomes.

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