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Case Report
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Management of Challenging Ebstein's Anomaly Case in a Teenager Patient

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Background

Ebstein's anomaly is a relatively rare form of congenital malformation of the tricuspid valve and right ventricular failure [1] with prevalence of 1/20,000 live births and accounts for 1% of all cases of congenital heart disease [2-4]. It was first described by Wilhelm Ebstein in 1866 [5]. Ebstein's anomaly has a broad clinical spectrum depending on the severity of the tricuspid regurgitation; age of clinical presentation, hemodynamic variations and degree of right-to-left interatrial shunt, right ventricular functions and associated abnormalities. The cardinal symptoms in Ebstein's anomaly are progressive cyanosis, right ventricular myopathy, arrhythmias, and sudden cardiac death. Many cases of Ebstein anomaly can be missed on routine Echocardiography and remain undiagnosed for longer time period which can delay definitive treatment in a significant symptomatic patient, by the time of diagnosis many patients develop advance heart failure having unstable hemodynamics specially in cases coexistence with arrhythmias leads to increase morbidity and mortality and many cardiac surgeons refuse their surgical intervention due to unstable hemodynamics. In such cases newly advanced specific cardiac modalities should be used 1st line for early diagnosis and prompt treatment where available.

Case Presentation

An 18-year-old female from rural part of Afghanistan, presented hemodynamically unstable to the emergency room having sign and symptoms of advanced heart failure. ECG shows atrial flutter with FVR and patient urgently managed according to the GDMT. She has past medical history of progressive exertional dyspnea NYHA Class II and palpitations of 6 years duration, and had episodes of syncopal attacks at home with no history of cyanosis, chest pain, or history of sore throat, joint pains in childhood. Initially she was managed by cardiologist in Afghanistan on "line of heart failure with unknown cause" and patient responded well to medical therapy but from last 06 months despite good compliance to medication her symptoms gradually progressed to NYHA Class III and she was referred to our hospital for further workup and management. On general physical examination normal looking girl with irregular pulse rate of 150/min, the blood pressure was 90/55 mmHg, Prominent jugular venous pulse noted with "V" wave and peripheral edema, a sustained left parasternal heave was palpable and on auscultation, the second heart sound was widely split and fixed, right ventricular third heart sound was heard with a pansystolic murmur of tricuspid regurgitation. Chest examination showed bilateral basal crepitations and hepatosplenomegaly were absent.

Investigations

Patient ECG showed narrow complex tachycardia 2:1 Atrial flutter with heart rate 150/min, loss of isoelectric baseline and saw tooth waves in inferior lead, chest X-ray revealed cardiomegaly with a CTR of >65%, right atrial enlargement and increased vascularity.

A two-dimensional echocardiogram in the apical 4 chamber view showed, apical leaflet point is >10 mm and tricuspid valve coaptation is also apical displaced. Part of RV is an "atrialized" (Figure 1). A moderate size atrial septal defect of about 20 mm × 23 mm × 30 mm was found in the middle of the atrial septum with right-to-left shunt *via* ASD.

Patient subjected to CMR which shows moderate size ASD secundum with dimensions sagittal 20 × 23 × 30 (4x) × (2x) hugely dilated RA. RA area 59 cm^2 and dimensions are 82 mm × 78 mm. RA volume in ventricular systolic phase 278 ml and 265 ml in ventricular diastole. Some part of RV apparently atrialized. Septal leaflet of tricuspid valve is apically displaced and separation of TV-MV

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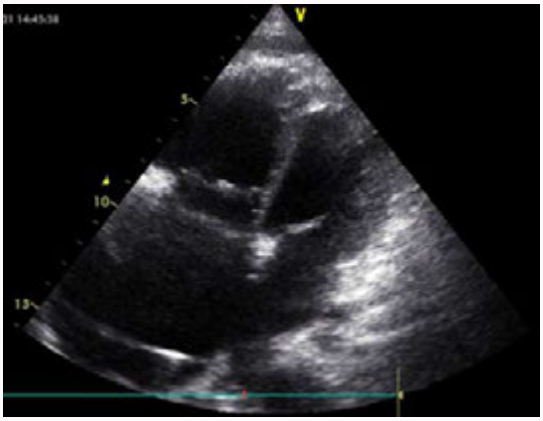

Figure 1: Two-dimensional echocardiogram.

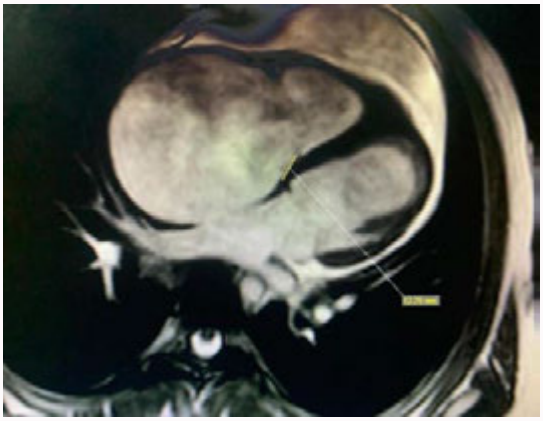

Figure 2: Cardiac magnetic resonance.

AML hinge point is >10 mm, (Figure 2) mild pericardial effusion is also observed and RV has been analyzed for microaneurysms, thin out segments, fat infiltration and regional wall motion abnormality. No stigmata of ARVC in pre-contrast studies.

The patient underwent an electrophysiologic study (Figure 3), which confirmed the 2:1 atrial flutter followed by successful radiofrequency ablation.

Case Specific Management

This patient was deemed inoperable by many Cardiac Surgeons in our country. In our case for better outcomes patient underwent radiofrequency ablation for typical atrial flutter initially and then surgical correction of ASD II closure with autologous pericardial patch and tricuspid valve repair using modified cone repair with autologous fixed pericardial band and Prolene stitches. Additionally, LAA was plicated through ASD using Prolene stitches and RA reduced in size by plication at closure time. At one year's follow up, the patient is having good quality of life, on optimum medical therapy including Digoxin, Diuretics and ARB; with Mild to Moderate Tricuspid Valve Regurgitation; and she has got married.

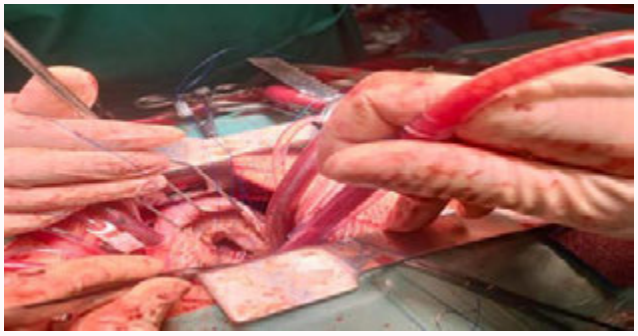

Figure 4: Intraoperative image of Ebstein's Anomaly correction surgery.

Discussion

Ebstein's anomaly is an uncommon congenital malformation characterized by a shift of the septal and posterior tricuspid valve towards the right ventricular wall of the apex with different presentations due to its heterogenic nature [1]. These patients can remain asymptomatic for years, but the common symptoms are abnormal heart rhythms; shortness of breath; fatigue and bluish discoloration of the skin [6]. In the era of cardiovascular intervention, the survival of Ebstein's anomaly has increased but still <50% of patients survive beyond 10 years [7]. A significant issue in patients with Ebstein's anomaly is determining the appropriate time for therapeutic intervention and timely diagnosis. Another cardiac anomaly known to be associated with Ebstein's anomaly is ASD with the prevalence of 80% to 94% and arrhythmias more common in adults with atrial flutter or fibrillation in 25% cases [8,9]. Although echocardiography could be the first line investigation but it is not as sensitive and specific as CMR because few cases of Ebstein's anomaly remain undiagnosed with echocardiography. Therefore, CMR is the best modality to make accurate diagnosis of Ebstein's anomaly and

Figure 3: An electrophysiological study shows a typical atrial flutter with 2:1.

right ventricular assessment, where available.

With the ongoing medical and surgical advancement, several treatment options are described by several authors. Surgical modalities depend upon the severity and type of defect in Ebstein's anomaly. Therefore, each case should be individualized. Factors associated with a worse outcome are young age at diagnosis, male sex, cardiothoracic ratio of more than 0.65 (>65%) and the presence of cyanosis [10]. Indications for surgery in Ebstein's anomaly is not clearly defined and the ideal mode of surgical management remains controversial [11,12]. However, in cases where patient is having hemodynamically significant arrhythmias making surgical intervention difficult for cardiac surgeons, radiofrequency catheter ablation can be applied to arrhythmias before surgical intervention making patient amenable to surgical intervention and preventing sudden cardiac death.

Conclusion

In a Nutshell, in Ebstein's Anomaly cases having clinically significant symptomatic arrhythmias, Patient's heart failure decompensates and progresses more quickly therefore arrhythmias ablation before surgical correction is the best approach followed by early surgical intervention to prevent morbidity and mortality. Additionally, CMR should be used 1st line cardiac modality where diagnosis of Ebstein's anomaly is uncertain, and when adequacy, of Right Ventricular functions, is in question.

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