Diagnosis of a mild case of ebstein's anomaly by intracardiac ECG and pressure recording

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Abstract:
Ebstein's anomaly is a rare congenital heart disorder characterized by apical displacement of tricuspid valve. Clinical presentation depends on degree of tricuspid valve malformation, displacement, degree of tricuspid regurgitation and associated arrhythmia. Mild cases are often asymptomatic and diagnosis can be missed unless a high degree of suspicion is maintained. Mild cases can be diagnosed by Cardiac Magnetic Resonance (CMR) imaging or by simultaneous intracardiac ECG and pressure recording. Our case presented with recurrent episodes of palpitation and breathing difficulty. He was evaluated elsewhere, but the exact diagnosis was missed. Physical examination and chest X-ray was normal. ECG showed subtle notching in terminal QRS in lead II, III and aVf. Initial echocardiogram was reported normal. Later on with careful clinical evaluation, subtle ECG finding and detailed echocardiography ebsteins anomaly was suspected. Diagnosis was confirmed by recording ventricular EGM with RA pressure tracing from atrialised ventricle. With proper education and reassurance, he is asymptomatic at 1 year follow-up.

Keyword: 
"Ebstein's anomaly", "Intracardiac ECG","Atrialised ventricle"

Introduction:
Ebstein's anomaly is a rare congenital heart disease, characterized by atrialisation of the right ventricle owing to apical displacement of the septal and posterior tricuspid leaflets. The clinical presentation and hemodynamic consequences depend on degree of displacement and malformation of tricuspid leaflets and degree of resulting tricuspid regurgitation. Mild cases may be asymptomatic and frequently are undiagnosed. The signs and symptoms of the disease also depend on associated structural and conduction system disease.Chest radiographs may be entirely normal or show cardiomegaly with enlarged right atrial silhouette and narrow vascular pedicle.¹
Milder forms can be diagnosed by CMR imaging or simultaneous intracardiac pressure recording. Cardiac catheterisation can confirm diagnosis by simultaneous recording of right ventricular electrical activity with right atrial pressure waveform from atrialised ventricle. This case illustrates a mild form of ebstein’s anomaly. The diagnosis was missed on initial evaluation, but after careful clinical, ECG and echo correlation it was suspected and confirmed by intracardiac simultaneous ECG and pressure recordings.

Clinical History:
A 44 year old gentleman presented with exertional breathing difficulty for last six months. He also had recurrent episodes of palpitations. There was similar history six years back, which was evaluated and symptoms were attributed to iron deficiency anaemia due to bleeding duodenal ulcer. He was treated conservatively and symptoms subsided after correction of anaemia. He had an echocardiogram then, which reported structurally normal heart. He had recurrence of symptoms from last six months, which was again attributed to iron deficiency anaemia. However upper GI scopy showed healed duodenal ulcer. His heart rate and blood pressure were normal, and the physical exam was unremarkable. Electrocardiography showed sinus rhythm with subtle notching in terminal QRS in lead II,III and aVf (Fig. 1 & 2).

**Fig 1: 12 lead Electrocardiogram showing subtle changes in terminal QRS in lead II, III, aVf.**

**Fig 2: zoomed view of lead III, aVf showing notch in terminal part of QRS.**
His chest roentgenography was normal (Fig. 3).

**Fig.3 : Chest X-ray was normal.**
He underwent transthoracic echocardiography and on careful evaluation he was suspected to have an ebstein’s anomaly as tricuspid valve was 18 mm apically displaced (Fig. 4-A, B ). There was no evidence of right ventricular outflow obstruction, or any atrial or ventricular septal defects.
He had no history of cyanosis, swelling of feet, CVA, loss of consciousness or syncope in past. He was taken for catheterisation and EP study to confirm diagnosis of Ebstein’s anomaly and for evaluation of palpitation.

He underwent systematic induction of arrhythmia as part of evaluation of palpitation, but it was negative for the same. However simultaneous recording of pressure as well as ECG and pull back from RV to RA confirmed the diagnosis. As shown in fig. 5 pull back from RV to RA depicts RV electrocardiogram and RV pressure wave form changing over to RV electrocardiogram and RA pressure waveform in atrialized RV. Contrast injection via the attain catheter confirmed the presence of atrialised ventricle (Fig. 6). Patient was educated about the disease and reassured about its benign nature. He is asymptomatic at 1 year of follow up.
Fig 6: showing catheter position in atrialised RV. Contrast injection confirms it to be proximal to tricuspid valve

Discussion:
Ebstein’s anomaly is a malformation of the tricuspid valve and right ventricle characterized by apical displacement of septal, posterior leaflets and less likely anterior leaflets of tricuspid valve. There is dilatation of the “atrialised” portion of the right ventricle and dilatation of the right atrioventricular junction resulting in tricuspid regurgitation. It accounts for < 1% of all cases of congenital heart disease and occurs in < 1 per 2,00,000 live births. This anomaly is most commonly associated with an ASD or PFO in approximately 80% to 94% of patients, whereas approximately 25% of the patients have an accessory conduction pathway. Additional anomalies may be ventricular septal defects and varying degrees of RVOT obstruction. The clinical presentation depends on severity of the malformation, hemodynamics, presenting age and degree of right to left shunting. Neonates and infants present with cyanosis or heart failure, where as adolescents and adults present with arrhythmias. They can present with decreasing exercise capacity, fatigue, signs of right-sided heart failure or progressive cyanosis. The splintered polyphasic terminal QRS pattern, characteristic of this anomaly result from fusion of a second QRS originating from atrialised right ventricle to the preceding normal QRS complex, but not always seen. The chest x ray may be completely normal or may show diagnostic configuration consisting of a right atrial enlargement with a narrow vascular pedicle. X-ray also prognosticates as greater the cardiomegaly worse is the prognosis. Diagnosis is usually by echocardiographic examination by showing apical displacement of tricuspid valve. But in mild cases diagnosis can be difficult because normally also, especially in association with ASD or tricuspid regurgitation there is some displacement of septal and posterior leaflets with considerable overlap. In Adults more than 20 mm or 8mm/m² displacement of the septal leaflet from the mitral annulus in apical 4 chamber view are considered diagnostic of ebstein’s anomaly. In subtle cases, diagnosis can be confirmed by simultaneous recording of right ventricular electrical activity and right atrial pressure from the atrialised ventricle, when the catheter is withdrawn from the right ventricle to the right atrium across the tricuspid valve. Alternatively cardiac MRI (magnetic resonance imaging) may be used to assess magnitude of septal displacement, ventricular size and function. Overall prognosis for Ebstein’s anomaly is poor, especially when they present during infancy or childhood but patients, who are diagnosed in adulthood, are having less associated cardiac defects and much lower morbidity and mortality. For asymptomatic patients only observation and follow up is required as arrhythmia and rapidly progressive heart failure can appear at any stage of life. Symptomatic patients may need surgical management, like tricuspid valve replacement, modified tricuspid annuloplasty or biventricular reconstructive surgery. This patient had completely normal chest X-ray and subtle changes in ECG and represented clinically mild form of ebstein’s anomaly. Initial echocardiographic exam (done twice) was reported normal. On careful evaluation there was suspicion of ebstein’s anomaly after detecting borderline apical displacement of tricuspid valve and tricuspid regurgitation. Diagnosis was confirmed by simultaneous intracardiac ECG and pressure recording. Hence if high level of suspicion is not exercised, then milder form
of ebstein's anomaly are clinically missed.

**Conclusion:**

Ebstein's anomaly is a complex congenital disorder with a variable anatomical severity and clinical presentation. Mild forms of disease can be missed and careful echocardiographic examination is required to indentify subtle cases. Diagnosis can be confirmed by intracardiac recording of ventricular EGM in atrialised RV or by CMR. Though periodic follow up and often masterly inactivity are exercised, yet patients can present with arrhythmias at any stage of life even with milder form.

**References:**


