Ebstein’s Anomaly with Various Unexplained Arrhythmias in a 57-Year Old Man

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Abstract
There are very few congenital cardiac disease which may remain asymptomatic up to adulthood, Ebstein’s anomaly is one of them. With rapid advancement in the field of echocardiography many new cases identified in day-to-day practice. Timely surgical intervention in these cases may distinct survival advantage. We present a case of Ebstein’s anomaly of adult age group which is seen with typical manifestation and seen in less than 5% cases.

Introduction
Presentation of arrhythmias in adult patients is generally suggestive of ischemic heart disease, valvular heart disease or cardiomyopathy but presentation of multiple types of arrhythmias in elderly patients may sometimes give clue of congenital heart disease. Ebstein’s anomaly is otherwise a rare disease after 50 years of age, only 5% of patients live beyond fifth decade. With advancing age, the enlarged right atrium predisposes to atrial fibrillation or atrial flutter, which with reentrant supraventricular tachycardia occurs in approximately one-third patients.

Case Report
A 57-year old previously healthy man presented with complaint of palpitations which lasted up to 30 minutes and associated with dyspnea. History of similar two episodes were present around 18 and 2 years back. On his cardiovascular examination a grade 2/6 holosystolic tricuspid regurgitation murmur (best heard at the left sternal border with inspiratory accentuation) and wide splitting of second heart sound in pulmonary area, and other systems normal.

His ECG on different days showed various types of rhythm and arrhythmia (Figure 1 - 4) and chest x-ray showed cardiomegaly with left ventricular type apex and pulmonary oligemia (globular box-shaped heart) (Figure 5).

Echocardiography finding showed, apical displacement 15 mm/m of septal leaflet of tricuspid valve with right atrial enlargement with presence of low pressure tricuspid regurgitation. These finding were suggestive of Ebstein’s anomaly (Figures 6 - 8).

Discussion
The tricuspid valve anomaly described by Ebstein in 1864 consists of apical displacement of the septal and posterior tricuspid leaflets, which results in an enlarged right atrium functionally integrated with the inlet region of the right ventricle (“atrialized” right ventricle). The outlet and trabecular portions of the right ventricle constitute an often hypoplastic, “functional” ventricle. Ebstein’s anomaly occurs in 1 per 20,000 live-births, accounting, for 0.3% to 0.7% of all cases of congenital heart disease while only 5% of the patients survive beyond the fifth decade.

With advancing age, the enlarged right atrium predisposes to atrial fibrillation or atrial flutter, predisposing...
to re-entrant supraventricular tachycardia in about one-third of patients. Risk factors believed to be associated with the condition are a family history of Ebstein’s anomaly or other congenital heart disease, northern European ancestry and maternal exposure to benzodiazepines or lithium. More than 30% of patients with Ebstein’s anomaly have associated cardiac defects.

The clinical manifestations of Ebstein’s anomaly depend on the degree of tricuspid valve malformation and consequent regurgitation, and any associated cardiac defects. Many patients first experience symptoms as adults, but the onset can occur after birth or in infancy or childhood. In adults, the anomaly commonly presents with arrhythmia.

Reentrant supraventricular tachycardia, atrial fibrillation, atrial flutter and junctional tachycardia occur in 25% to 30% of patients and preexcitation caused by right-sided accessory pathways occurs in 5% to 25% of ECG.

As our case Day 1 ECG (Figure 1) showed regular narrow complex tachycardia with no visible P wave with ventricular rate 280/min suggestive of AVNRT and day 3 (Figure 3) ECG show regular narrow complex tachycardia with no visible P wave with ventricular rate 240/min suggestive of AVNRT and Day 2 (Figure 2) ECG showed irregular narrow complex tachycardia with no visible P wave with ventricular rate 120/min suggestive of atrial fibrillation. In a study by Yu et al, PSVT is present in 10.4% and atrial tachyarrhythmia is present in 13.8% of the adult patient with Ebstein’s anomaly. In another study by Fause et al showed SVT present in 48.6% and AF/AFL present in 20.8% of adult patients with Ebstein’s anomaly.

In 75% to 95% of cases, the QRS complex is characterized by a right conduction defect of right bundle branch type.

As in our case, Day 1 to Day 4 ECG (Figures 1-4) show RBBB (R’R pattern in V1-V4). In study by Yu et al, RBBB was present in 65.3% patients. In another study by Fause et al, RBBB
was present in 68.1% patients. WPW preexcitation occurs in 5% to 25% of the electrocardiograms in Ebstein’s anomaly.

Atrial arrhythmias without evidence of pre-excitation can be treated pharmacologically, whereas percutaneous radiofrequency ablation is indicated in the presence of an accessory pathway. In general, surgical intervention with tricuspid valve repair or replacement is restricted to patients with severe heart failure, cyanosis, intractable arrhythmias or paradoxical embolization (passage of thrombi from the venous circulation into the arterial circulation through a right-to-left shunt at the atrial level). Patients with Ebstein’s anomaly should be assessed regularly for signs of deterioration in functional capacity, increasing cyanosis or presence of arrhythmia. Prophylaxis against infective endocarditis is warranted in all cases.

In conclusion, there is increase in survival of adult congenital heart disease due to vast progress in congenital cardiac surgery, so this type of cases may be seen more and more in routine clinical practice. Presentation with various types of arrhythmias in adult patients with minimal symptom and physical findings needs further evaluation by echocardiography because these findings may be manifestation of a congenital heart disease in adult as in our case.

References