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Case Report

A 9-year Old Boy with Ebstein's Anomaly

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Abstract

Ebstein anomaly is the apical displacement of the septal and posterior leaflets of the tricuspid valve, which results in an enlarged right atrium, functionally integrated with the inlet region of the right ventricle. Its clinical manifestations depend on the degree of tricuspid valve regurgitation and any associated cardiac defects. We are reporting a case of 9 years old, apparently healthy boy, hailing from Salop,Ullapara, Sirajganj, came to cardiology out patient department (OPD) of North Bengal Medical College Hospital, Sirajganj, with 6 weeks history of palpitation on exertion. Clinical examination revealed, pansystolic murmur in tricuspid area with end inspiratory accentuation. Transthoracic echocardiography showed, apical displacement of septal and posterior leaflets of tricuspid valve with atrialised right ventricle, which is termed as Ebstein anomaly.

Key words: *Atrialization, Apical displacement, Posterior leaflet*

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Introduction

The tricuspid valve anomaly was 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 anomaly occurs in 5 per 100 000 live births, accounting for 0.5% of all cases of congenital heart disease.[1] 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.[1,2,3] More than 30% of patients with Ebstein's anomaly have associated cardiac defects.[1] In this study, we reported a rare case of Ebstein's anomaly.

Case Report

A 9 years old, previously healthy school boy, hailing from Salop, Ullapara, Sirajgonj, came to the cardiology OPD of North Bengal Medical College Hospital, Sirajganj, with a 6-week history of palpitations. The symptoms occurred during exertion, 1-2 times per week, lasted up to 20 minutes at a time and were associated with dyspnoea. On physical examination, there was pansystolic murmur of grade 2/6, in the left sternal edge, which was best heard after breath holding on inspiration. ECG showed no abnormality, but cardiac shadow was enlarged in postero-anterior view of chest X-ray. Transthoracic echocardiography demonstrated the presence of Ebstein's anomaly of the tricuspid valve, with apical displacement of the valve and formation of an "atrialized" right ventricle (a functional unit between the right atrium and the inlet [inflow] portion of the right ventricle) (Figure 2). The anterior tricuspid valve leaflet was elongated, whereas the septal leaflet was rudimentary. Ethical clearance had been taken form institutional ethical committee and the informed consent of the subject had been taken from patient's guardian for study.

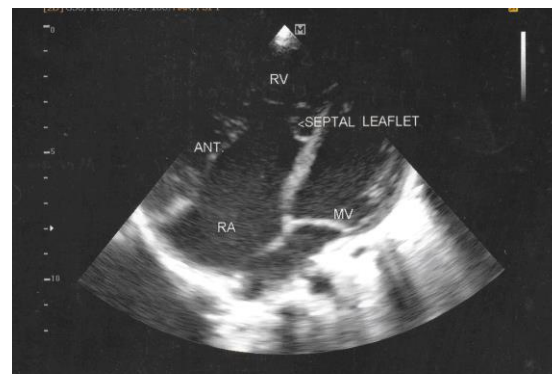

Figure 2: Echocardiogram showing Ebstein's anomaly (apical displacement) of the tricuspid valve with long anterior leaflet and rudimentary septal leaflet (Arrowed) and formation of an "Atrialized" Right Ventricle (ARV)

Discussion

The clinical manifestations of Ebstein's anomaly depend on the degree of tricuspid valve malformation and consequent regurgitation, and any associated cardiac defects.[2,3] Many patients first experience symptoms as adults, but the onset can occur after birth or in infancy or childhood. In newborns, the anomaly often presents as cyanosis and, in the absence of surgical repair, is associated with a 20% mortality in the first year of life. In infants, it may present as congestive heart failure and in children as an incidental murmur. In adults, the anomaly commonly presents with arrhythmias. Factors associated with a worse outcome are young age at diagnosis, male sex, cardiothoracic ratio of more than 0.65 and the presence of cyanosis.[2]

The treatment of Ebstein's anomaly has to be tailored to the individual patient. Patients with heart failure and little impairment in functional capacity can be managed medically. 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.

References

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