Longest Surviving Patient with Ebstein's Anomaly in Indian Subcontinent

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Abstract

Ebstein's anomaly accounts for 0.3% to 0.7% of all cases of congenital heart disease. The condition is characterized by abnormal tricuspid valve and right ventricle with apical displacement of tricuspid valve leading to atrialization of right ventricle. There have been case reports of patients surviving up to ninth decade. It is unusual for these patients to be asymptomatic in adulthood for long duration. We describe a patient with Ebstein's anomaly in the ninth decade with coronary artery disease.

Introduction

Ebstein's anomaly is a rare congenital heart disease and accounts for 0.3% to 0.7% of all cases of congenital heart disease[1] and occurs in approximately 1 in 20,000 lives births.[3] It is characterized by apical displacement of tricuspid valve into right ventricle due to abnormal attachments of posterior and septal leaflets of tricuspid valve to the ventricular wall leading to atrialization of the right ventricle. Very few reports are there of patients surviving to adulthood, even less so for patients in their ninth decade of life.[2] The patients present mainly with arrhythmias in adulthood.

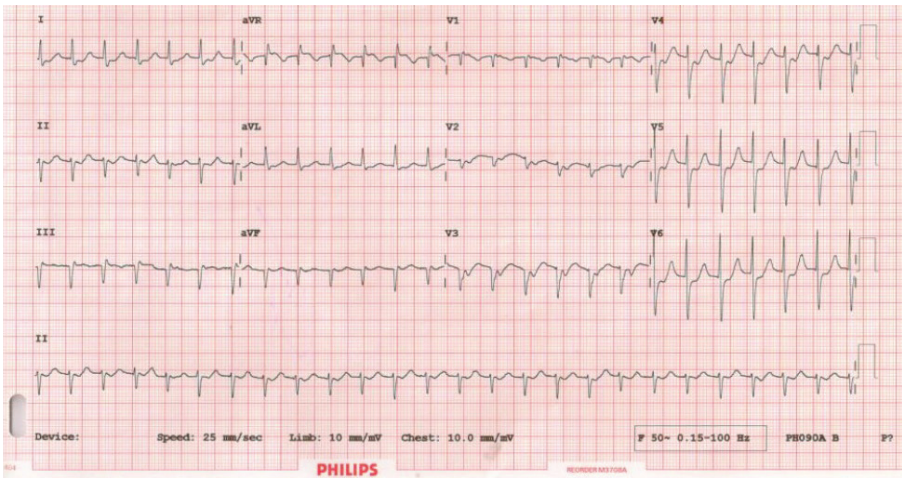

Fig. 1: ECG showing supraventricular tachycardia

Case Report

An 84-year-old male patient, presented to our hospital with complaint of chest pain for 1 day. He also had associated Grade II dyspnea with palpitations. He was a diagnosed case of systemic hypertension. He was a non-smoker.

His physical examination was unremarkable with the exception of BP which was 180/80. His ECG showed a regular narrow QRS tachycardia (Figure 1). His ECHO revealed dilated RA (46 mm), atrialised RV (39 mm), shift of septal tricuspid leaflet towards RV apex (19 mm), Moderate tricuspid regurgitation, normal RV and LV function, LVEF – 69% (Figure 2). He was taken up for coronary angiogram which showed double vessel disease with 80% stenosis in proximal Left Anterior Descending Artery and 90% stenosis of Left Circumflex Artery which was non-dominant. Right coronary artery was normal (Figure 3). He was advised EP study followed by myocardial revascularisation. He however opted to continue on medical management and has been on follow up.

Discussion

Ebstein's anomaly is characterized by apical displacement of tricuspid leaflet into right ventricle due to abnormal attachments of posterior and septal leaflets. This displacement leads to atrialization of right ventricle. This condition has a wide spectrum of presentation ranging from cyanosis and heart failure during infancy to minimal symptoms and signs in adulthood.

In general, symptoms are related to the degree of anatomic abnormality. Patients with mild apical displacement and mild dysfunction of the tricuspid valve may remain asymptomatic or present in adulthood with arrhythmia or paradoxical embolic event. Approximately 20 to 30 percent of cases present with atrial tachyarrythmias across age groups with greater frequency in adolescents and adults. Up to 20 percent of patients present with arrhythmias which may be due to accessory conduction pathway. The symptoms which do occur during adulthood are due to arrhythmias which are usually supraventricular. This is usually attributed to a right atrioventricular accessory pathway.[4]

The patients can sometimes present with polymorphic Ventricular tachycardia and sudden cardiac death. Tachyarrhythmic sudden death is responsible for the decline in survival rate in the fifth decade.[5] Wolff-Parkinson-White syndrome in otherwise healthy individual carries an estimated sudden cardiac death risk of 0.02%, but in Ebstein's anomaly, atrial flutter or fibrillation with accelerated conduction is accompanied by a major increase in the risk of sudden death.

Following the repair of adult congenital heart disease atherosclerotic

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coronary artery disease may be seen and concomitant CABG may be required at the time of correction of ACHD.[6] Oldest recorded patient with Ebstein's anomaly lived to 85 years and was devoid of cardiac symptoms until age 79 years.[2] Our patient is the oldest

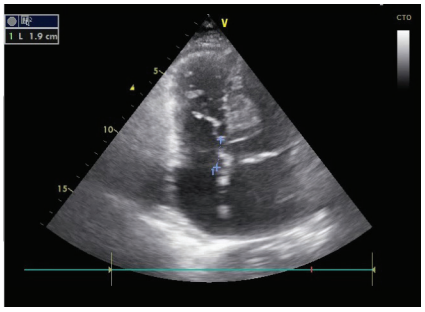

Fig. 2: Apical 4 chamber view – showing apical displacement of tricuspid valve

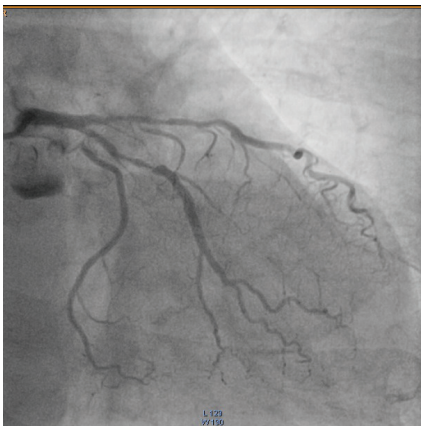

Fig. 3: Right Anterior oblique view – Left coronary angiogram showing significant stenosis in left circumflex and left anterior descending

with Ebstein's anomaly reported in the Indian subcontinent with underlying coronary artery disease. Since our patient had not been willing for any intervention he has been on medical management since.

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