

Ebstein's Anomaly - A Long Innings

L S Yashaswini¹
N Arun Kumar²,
S S Ramesh³,
M M Basavaraju⁴,
V Mohan Kumar⁵

¹Senior Resident, Rajarajeshwari Medical College and Research Institute, Bangalore, Karnataka, India, ²Senior Resident, ESIC MC & PGIMSR & MH, Bangalore, India, ³Assistant Professor, Mysore Medical College and Research Institute, Bangalore, Karnataka, India, ⁴Assistant Professor, Mysore Medical College and Research Institute, Bangalore, Karnataka, India, ⁵Consultant Physician, Columbia Asia Hospital, Mysore, Karnataka, India

Corresponding Author: Dr. N Arun Kumar, B-192, 10th Block, C.P.W.D Quarters, Domlur, Bangalore - 560071, Phone: 09986929708. E-mail: dreamfullofcream@gmail.com

Abstract

Ebstein's anomaly is a rare disorder with a reported incidence of 0.5% or less among patients with congenital heart disease. Ebstein's anomaly surviving for seven decades patients surviving after 50 years of age is < 5% but without symptoms is rare. Our patient presented with symptoms in the seventh decade only. Surviving till seventh decade is rare but surviving without symptoms is rarest of rare cases.

Keywords: Ebstein's anomaly, Seventh decade, Symptoms

INTRODUCTION

Ebstein's anomaly is a rare disorder with a reported incidence of 0.5% or less among patients with congenital heart disease. Ebstein's anomaly surviving for seven decades patients surviving after 50 years of age is < 5%. Anomaly of tricuspid valve very rare, accounting for <1% CHD. Incidence 1:2,00,000 live births. Apical displacement of septal and posterior tricuspid leaflets with dysplasia of valve. Inter atrial communication in 80-95% of patients. Cardinal symptoms being Cyanosis, right heart failure, arrhythmias, sudden cardiac death. Reports of patients surviving to adulthood with symptoms are very few! ECG-Himalayan p-waves, RBBB, Splintered QRS complexes, WPW Syndrome first degree heart block. Chest X Ray Vary from normal to typical globe shaped heart with narrow pedicle, cardiomegaly, normal or low pulmonary vascularity. 2D Echocardiography-Gold standard for diagnosis. Apical displacement of septal leaflet from insertion by atleast 8 mm. Dilation of right atrium, atrialized portion of right ventricle. Pulmonary artery pressure is typically low. Patients surviving after 50 years of age is <5%.¹⁻⁴

CASE HISTORY

A 65 year old male patient, previously asymptomatic, presenting with recent onset of breathlessness and swelling

of both lower limbs. Not a known hypertensive or diabetic. He was chronic smoker for 45 years.

GPE: Moderately built and nourished, conscious, oriented. Pulse - 72 beats/min, Regular, Normal Volume. BP - 120/80 mm Hg in upper limbs, 130/90 mm Hg in lower limbs.

Respiratory rate - 22 cycles/min. Grade 2 Clubbing-carpopedal (Figure 1), Jugular Venous Pressure - normal. Bilateral pitting pedal oedema present upto knee. Central Cyanosis present.

Post-axial polydactyly of right hand present (Figure 1). No Pallor, No Icterus, No Lymphadenopathy.

Height-167 cm, Weight-59 kg, BMI-21.14 kg/m².

Cardiovascular system: Apical impulse in left 5th intercostal space, 2.5 cm lateral to midclavicular line, hyperdynamic. Multiple heart sounds in mitral and tricuspid areas with split S1. Pansystolic murmur of grade 3/6 in left parasternal area. Split S2 at pulmonary area, P2 normal.

Respiratory system: Normal vesicular breath sounds heard.

Abdomen: Soft, bowel sounds + No Organomegaly.

Central Nervous System: No neurological deficits.

Routine Investigations

Hb - 14 g%, TLC - 6400 cells/cumm, DC - N⁷⁸, L²², ESR - 12 mm/hr.

Blood Urea - 39 mg/dl, Serum creatinine - 1.1 mg/dl.

Serum electrolytes - Na - 137, K - 4, Cl - 106.

Urine routine - normal.

Chest X ray PA View - Cardiomegaly with enlarged right atrium, normal vascularity of lung fields.

2D Echocardiography (Figure 2) - Congenital heart disease, Ebstein's anomaly of tricuspid valve, displaced STL by 5 cm, large sail like ATL. Dilated RA and RV with RV dysfunction, hypoplastic pulmonary artery, and severe low pressure TR (PASP-27 mm Hg). Normal left ventricular function.

ECG (Figure 3) - Sinus rhythm with QRS Axis of +60, normal p waves, prolonged PR interval, complete right bundle branch block, splintered QRS Complexes.³

Coronary angiogram: Normal.

DISCUSSION

Ebstein's anomaly of the tricuspid valve is an uncommon developmental abnormality with a reported incidence of less than 1% of all congenital cardiac malformations.¹ The natural history of this disease is variable, and it is believed that early death is often related to diagnostic procedures or thoracotomy.² Our case is a 65 year old male who was asymptomatic most of his life but presented with symptoms for the first time. ECG showing Sinus rhythm with QRS Axis of +60, normal p waves, prolonged PR interval, complete right bundle branch block, splintered QRS Complexes.³ Our case had Normal P waves indicate less symptoms in a patient with complete right bundle branch block without WPW syndrome.^{4,5}

Though the classical definition of Ebstein's anomaly emphasizes the downward displacement of a part or all of the tricuspid ring and valve, we have in addition, noted a wide range of abnormal features. We believe that dysplasia of valve leaflets is an inherent part of Ebstein's anomaly and this has been stressed by others as well.⁶

The indications for surgical treatment of Ebstein's malformation are not clearly defined and the ideal surgical mode of management remains controversial.^{7,8} The



Figure 1: Showing carpal clubbing

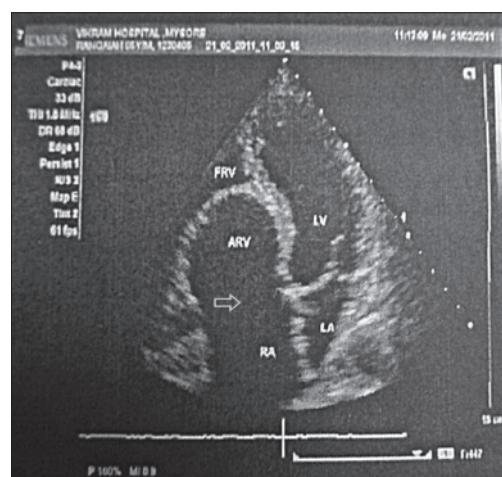


Figure 2: Trans thoracic 2D-Echocardiography showing dilated RA/RV

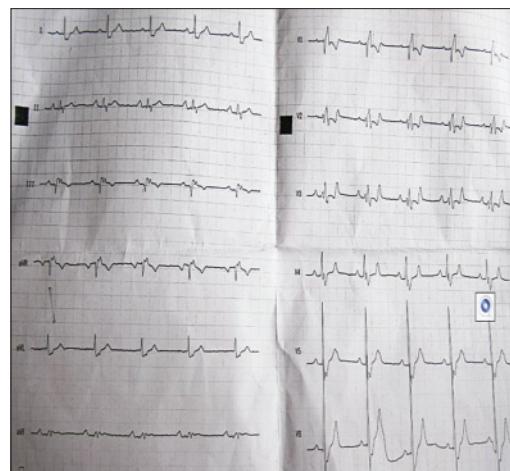


Figure 3: Electrocardiogram showing complete RBBB without himalayan P-waves

dysplastic leaflets and the dilated atrioventricular ring would both contribute significantly to the malfunctioning of the tricuspid valve and resultant cardiac failure. In

this setting, a valve replacement with plication of the thin walled atrialized right ventricle could possibly be the preferable mode of surgical treatment. However, because of the wide spectrum of anatomic variations in the tricuspid valve, the surgical approach to patients with Ebstein's anomaly needs to be individualised according to the specific morphology found at operation. Ebstein's malformation is often associated with other cardiac anomalies but in our case there is no other cardiac abnormality. This explains why our patient has survived without symptoms for this long.^{9,10}

CONCLUSION

A rare case of Ebstein's anomaly surviving for seven decades patients surviving after 50 years of age is <5% but without symptoms is rare. Our patient presented with symptoms in the seventh decade only. Surviving till seventh decade is rare but surviving without symptoms is rarest of rare cases.

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