Ebstein's Anomaly - A Rare Finding in Congenital Rubella Syndrome (CRS)

Bhavesh Talaviya[1], Rohan Parikh, Amey Beedkar, Mahendra Maske
Sunil Washimkar, Pradeep Deshmukh, Mukund Deshpande

ABSTRACT

Ebstein's anomaly is very rare (0.5 %) cardiac anomaly seen in patient of congenital rubella syndrome. Epidemiology and clinical data are lacking to guide management strategy. Congenital rubella syndrome is manifestation of rubella infection in foetus during period of embryogenesis. A verity of structural heart defect are known to occur with CRS but Ebstein's anomaly has not been described in association with CRS, but we are reporting case of CRS with EA. The case describe here is 32 year old woman presented with complaint of bilateral pedal oedema was evaluated and diagnosed as having EA. ECG and 2D Echo show right atrial enlargement, septal leaflet detachment and at rialization of right ventricle. Risk of death is increased by having other extra cardiac defects.

Introduction :

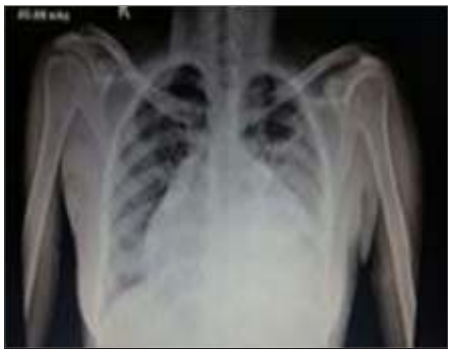
First describe by Wilhelm Ebstein in 1866, Ebstein's anomaly accounts for < 1% of all congenital heart defects, and < 0.5 % in congenital rubella syndrome[1]. In CRS Cardiac defects usually occurs in more than 50% of infants if infected before 8th month of gestation. These include PDA, peripheral pulmonary artery stenosis, pulmonary and aortic valve stenosis, septal defect, TGA, TOF, TA and stenosis of other major vessels[2].

The case described here is a 32 year old woman presented with bilateral pedal oedema. She was diagnosed to have EA. After detailed examination and investigations it was found to be part of congenital rubella syndrme. Because of rarity of such cases, it is unlikely to build complete scientific data from single centre. Hence reporting of even isolated case is important and will help medical science over rest of the world to solve the problem.

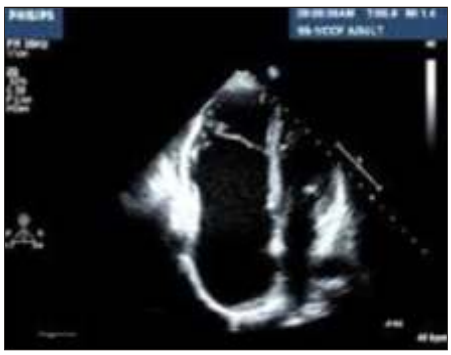
Case Report :

A 32 years old woman presented with bilateral pedal oedema since 1 month. There was no history of breathlessness, chest pain, fatigue or palpitation. On general examination apart from mild pallor and bilateral pitting oedema and raised JVP, there were no other remarkable findings. Her CVS examination revealed palpable thrill in tricuspid area with single loud second heart sound and pan-systolic murmur along lower left sternal border.

Chest X-Ray s/o cardiomegaly with a predominance of right heart enlargement and reduction in bronchovascular markings due to reduced pulmonary blood flow.



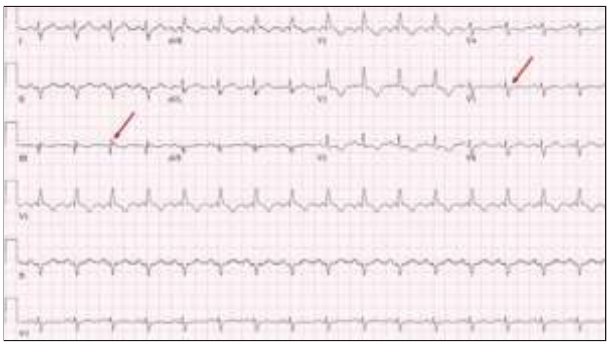
2D-echo revealed apical displacement of tricuspid valve, TR, Small RV cavity, large RA cavity.



[1]Senior Resident
The Department of Cardiology,
Govt. Medical College & Super Speciality Hospital, Nagpur.

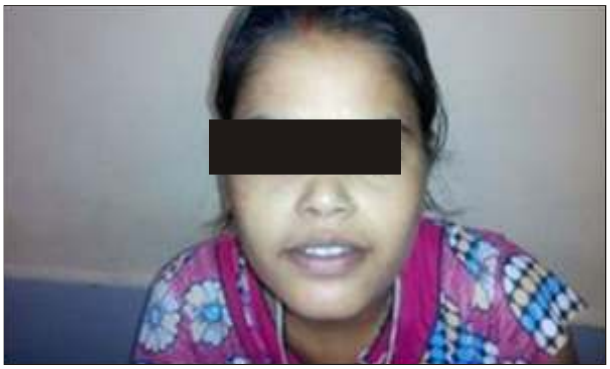
Address for Correspondence -
Dr. Bhavesh Vithhalbhai Talaviya
E-mail : bvtalaviya@gmail.com

ECG was s/o RBBB and Himalayan "P" wave in lead II s/o RAH. ECG s/o sinus rhythm, right bundle block, right ventricular hypertrophy and right atrial enlargement. Red arrows demonstrate the fragmented QRS complex noted in Ebstein anomaly. Othere findings in a twelve-lead electrocardiogram (ECG) may demonstrate accessory pathways, "Himalayan" p waves referring to giant p waves. Atrial arrhythmias including atrial fibrillation, atrial flutter or atrial tachycardia may be present. Fragmented QRS complex on 12-lead ECG, a marker of myocardial scar, has been associated with larger atrialized RV area and an increased risk of arrhythmic events in adult patients with EA.



Cardiac MRI revealed grossly dilated RA measuring 8cm with mild dilatation of RV associated with atrialisation of right ventricle. There was also apical displacement of the septal and posterior leaflets of the tricuspid valves with tricuspid incompetence. These findings were consistent with the diagnosis of Ebstein's anamoly.

Other finding : Patient had cataract with lens induced glaucoma and early RAPD in right eye for last 10 years. ENT evaluation s/o moderate to severe bilateral sensory neural hearing loss.



- As she had congenital cardiac defect in the form of EA and associated occular and auditory findings she was evaluated for rubella infection. Her Rubella IgG was found to be positive and finally she was lebelled congenital rubella syndrme. She was treated with Angiotensin-converting enzyme (ACE) inhibitors, diuretics, and digoxin for heart failure and advised about endocarditis prophylaxis.

Discussion :

Ebstein's anomaly is rare form (0.5%) of cyanotic congenital heart disease where is atrialization of right ventricle occurs in association with varying degree of tricuspid stenosis and regurgitation due to abnormally placed tricuspid valve leaflet[3]. High index of suspicion is required to diagnose CRS. Any patient with congenital cardiac defect with associated visual, hearing or neuropsychiatric defects should alert this rare condition[4]. The diagnosis of CRS in our case was made on basis of suggestive clinical picture and positive serology. Clinical picture, X-Ray findings and ECG were suggestive of EA, which was confirmed by echocardiography and cardiac MRI. Treatment of Ebstein anomaly is complex and dictated mainly by the severity of the disease itself and the effect of accompanying congenital structural and electrical abnormalities. Treatment options include medical therapy, radiofrequency ablation, and surgical therapy.

References :

1. Oster ME, Riehle-Colarusso T, Correa A (January 2010). "An update on cardiovascular malformation in congenital rubella syndrome. "Clin Mol Teratol.88 (1)1-8.
2. Sudharshan S, Ganesh SK, Biswas J (2010). " Current approach in diagnosis and management of posterior uveitis." Indian j ophthalmol. 58 (1): 29-43.
3. Cooper LZ aiford CA Jr. Rubella. In: Infectious diseases of the foetus and new-born, 5th edn. Eds. Remington JS, Klein JO, Philadelphia. W.B Saunders, 2001; pp. 347-388.
4. Gupta V, Verma S, Ramkrishnan S etal. Congenital rubella syndrome presenting in adulthood with fatigue. Circulation. 2013;128:2542-2545.