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

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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 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 pansystolic 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.

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 syndrome¹. 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 vessels².

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 syndrome. 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 pansystolic 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.

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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 atrialization of right ventricle. Risk of death is increased by having other extra cardiac defects.
As she had congenital cardiac defect in the form of EA and associated ocular and auditory findings she was evaluated for rubella infection. Her Rubella IgG was found to be positive and finally she was labelled congenital rubella syndrome. 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 atrialization of right ventricle occurs in association with varying degree of tricuspid stenosis and regurgitation due to abnormally placed tricuspid valve leaflet. 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. 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:**