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CASE REPORT

Pregnancy with Ebstein anomaly and early onset severe preeclampsia - a rare case

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ABSTRACT

Ebstein anomaly is a rare congenital cardiac abnormality, with a prevalence of 5.2 per 100,000 live births and accounts for 1% of congenital heart defects. The specific defect involves the abnormal attachment of tricuspid valve leaflets to the valve annulus, with displacement of septal and posterior leaflets, leading to downward displacement of effective orifice and division of right ventricle into two portions. The common cardiac lesions associated with Ebstein anomaly are ASD, PFO and WPW syndrome. We present a case of pregnancy with Ebstein anomaly (diagnosed at our hospital, with preeclampsia, resulting in preterm caesarean delivery. The patient had no complications and tolerated surgery, neonatal outcome was SGA (small for gestational age) birth. The pregnancy is safe in woman with Ebstein anomaly, who don't develop cyanosis or heart failure, and vaginal delivery is preferred. Preterm birth and SGA births are common in pregnancy with Ebstein anomaly. A close observation is required if mother develops cyanosis or arrhythmias.

Keywords: Ebstein anomaly, preeclampsia, echocardiography.

Ebstein anomaly is a rare congenital cardiac abnormality, characterized by specific structural deformity of the tricuspid valve. It accounts for approximately 1% of congenital heart defects, with a prevalence of 5.2 per 100,000 live births. It is associated with other cardiac anomalies in 38.3% and non-cardiac anomalies in 19.1% of cases respectively.[1] Imaging modalities such as echocardiography and cardiac magnetic resonance are used for diagnosis. The pregnancy is usually well tolerated as well as safe in women with Ebstein's anomaly, and woman can be advised to go ahead with pregnancy. The maternal complications that may develop include; arrhythmias, cyanosis, heart failure and paradoxical cerebral embolism. A close monitoring of clinical and haemodynamic changes is required during pregnancy for early detection of any maternal cardiac risk and to optimise the mode of delivery. [2,3]

Case

A 26 year old, G2A1 with period of gestation 32 weeks 3 days, came to the emergency with complaints of generalized edema for 5 days, high BP records for 3 days, with headache but no other signs/symptoms suggestive of impending eclampsia. She had a history of spontaneous complete abortion at 2.5 months gestation, 2 years back. The antenatal checkup was not adequate, with only two visits to local health worker. The patient did not give any past history of hypertension or history suggestive of heart disease (dyspnea, palpitation, chest pain, edema, syncopal attack). She was a non-smoker, nonalcoholic, and there was no history of teratogenic drug use. No histories of hypertension or heart diseases were present in her family.

On examination, pallor and pedal edema were present, no cyanosis was observed. Her pulse rate was 70 bpm, regular; BP was 140/100 mm Hg, and oxygen saturation at room air was 98%. The respiratory system examination was normal. On CVS examination, S1, S2 were heard with audible S3, a grade 2 pan systolic murmur was also heard in tricuspid

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area. On abdominal examination - uterus was 30weeks size, relaxed, presentation was cephalic and FHS was 130 bpm. The investigations were; Hb-11g%, 24 hour urine protein - 500mg, platelet count - 110,000/mL, serum albumin - 1.8 g/dl (rest of LFT normal), TLC, RFT, coagulation profile, and thyroid profile were normal. The antenatal ultrasound revealed a single live intrauterine fetus with 32 weeks gestation, cephalic presentation, an upper anterior placenta, oligohydramnios (AFI 3-4), mild uteroplacental insufficiency and EFWB of 1.8 kg. The fundus examination was normal.

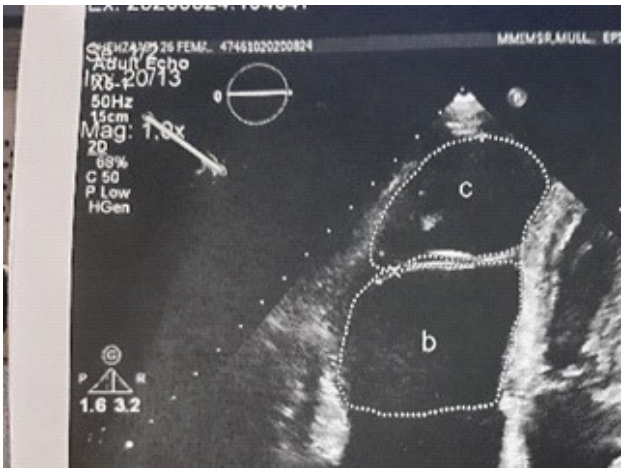

Figure 1: Figure shows a) Right atrium, b) Atrialized right ventricle, c) True right ventricle

The chest X - ray showed cardiomegaly and p-pulmonale was seen on ECG. After cardiology consultation 2D echo was performed. The echo findings were of Ebstein anomaly with severe tricuspid regurgitation (TR); apical displacement of septal tricuspid leaflet by 34mm, anterior tricuspid leaflet enlarged and sail like, dilated right atrium (RA area-16cm^2), true RV (right ventricle) area -15.2cm^2, atrialized RV area - 21.5 cm^2 and normal left ventricular systolic function (figure 1 and 2). The patient was admitted for preeclampsia with Ebstein anomaly and dexamethasone cover was given for fetal lung maturity.

The patient's BP shot up and antihypertensive (labetalol) was started. Inspite of the antihypertensives her blood pressure continued to rise upto 200/120 mm of Hg and she complained of headache, vomiting with blurring of vision. The patient was catheterized, inj MgSo4 started according to Pritchard's regimen and induction of labor was done at 32 weeks 5 days gestation with PGE1 in view of impending eclampsia. The labor was later augmented with oxytocin. An emergency cesarean under GA (general anaesthesia) had to be performed for category 3 CTG (cardiotocograph), a live male baby of 1.4 kg delivered with APGAR-6 and 9 at 1 min and 5 min respectively. The patient tolerated surgery well and was transferred to cardiology department. The patient was put on diuretic, tab amlong 5mg OD and injection enoxaparin 40 mg OD. The post-operative period was uneventful and patient was discharged on 8th postoperative day on amlong, enoxaparin and diuretic (torsemide plus spironolactone), to be followed up in cardiology OPD.

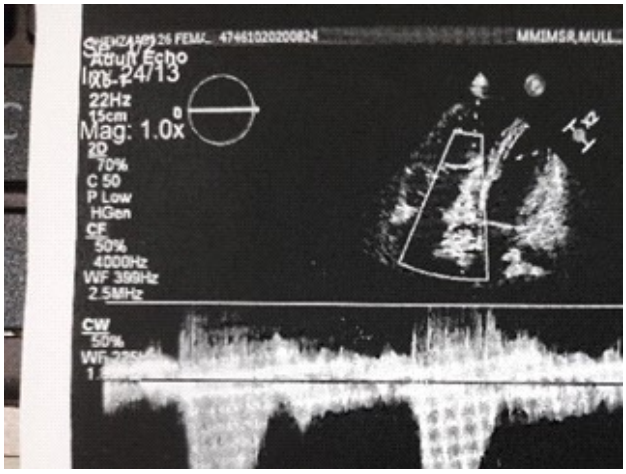

Figure 2: Figure shows tricuspid regurgitation

Discussion

Ebstein's anomaly is a rare congenital heart defect, and even rarer in pregnancy. A study reported 82 cases of Ebstein anomaly in a cohort of 7,850,381 woman delivered, with a prevalence of 1/100,000.[4] Morphologically in Ebstein's anomaly the tricuspid valve leaflets, septal and posterior leaflets, are displaced and the effective orifice is displaced downward into the right ventricular cavity dividing the right ventricle into two portions. The inlet portion (atrialized portion) is functionally confluent with the right atrium, while the other portion constitutes the functional right ventricle. Concomitant cardiac lesions are reported in about 38.3% of cases with Ebstein anomaly.[1] The common defects associated with Ebstein anomaly are atrial septal defect (ASD) and patent foramen ovale (PFO), the association being as high as 58.8%.[3] Arrhythmias are common in Ebstein anomaly and coexistent Wolf Parkinson White syndrome is observed in 16%-24% of cases.[2,3]

Pregnancy is accompanied by considerable hemodynamic changes, as a result of increase in circulating blood volume, and an increase in cardiac output. The uterine contractions, catecholamine release, rapid fluid-shift, and blood loss during labor further warrants the ability of woman with heart

disease to cope. The increased stroke volume may be poorly tolerated in Ebstein anomaly because of impaired RV size and function, it may lead to worsening of TR, increased RA pressures, and increased probability of right to left shunting.[2] A study monitoring hemodynamic changes during pregnancy in woman with Ebstein anomaly, showed that worsening of the CTR (cardiothoracic ratio), TRPG (tricuspid regurgitation peak gradient), and the TR severity are predictors of right heart failure during pregnancy.[3]

In the absence of cyanosis and heart failure, pregnancy is safe in woman with Ebstein anomaly (WHO risk class II). The vaginal delivery is well tolerated, and is preferred mode of delivery.[2] Some women may develop arrhythmia, cyanosis, heart failure and there is a risk of paradoxical cerebral embolus. A frequency of 18% was observed both for heart failure and arrhythmias by Kanoh M et al.[3] There is an increased frequency of preterm births (13.9%) and IUGR (11.11%) in infants born to Ebstein patients.[2] A study comparing Ebstein pregnancy with non Ebstein pregnancies found a significantly higher rate of major adverse cardiac event in Ebstein patients ($P < 0.001$), a higher frequency of preterm delivery, postpartum hemorrhage and caesarean delivery were also observed.[4]

The pregnancy and operative procedure were well tolerated in our patient, severe preeclampsia was the comorbidity observed, resulting in preterm induction and caesarean delivery for fetal distress. The newborn was small for gestational age but neonatal outcome was good. Not withstanding the safety of pregnancy in Ebstein patients, a close maternal and fetal observation is required in cases with arrhythmia or cyanosis .[2]

Conflict of interest: None. **Disclaimer:** Nil.

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