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Case Report *Volume 6 Issue 1*

A Rare Congenital Heart Diseases - Ebstein Anomaly with Successful Pregnancy Outcome: A Case Report

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Abstract

Ebstein anomaly of tricuspid valve is a congenital disease of tricuspid valve associated with downward displacement and elongation of the septal and anterior cusp- resulting in severe valvular tricuspid regurgitation with grossly dilated right atrium, small size right ventricle (Atrialized right ventricle). Pregnancy complicated with uncorrected Ebstein Anomaly is uncommon and may pose a serious threat to maternal and fetal life in the clinical setting of altered hemodynamics of Pregnancy. Here we report a rare case of Ebstein Anomaly of tricuspid valve in a young primi patient -31 year old, whose oxygen saturation was remain between 70-75% in room air.

Keywords: Congenital Heart Disease; Ebstein Anomaly; Tricuspid Regurgitation; ASD; Pan Systolic Murmur; Pregnancy

Introduction

Ebstein anomaly (EA) is an uncommon complex congenital malformation of the heart with prevalence of 0.3 – 0.5% [1]. It accounting for <17 of all cases of congenital heart diseases [1,2].

It is characterized by dysplastic abnormalities of tricuspid valve which involves both basal and free attachments of the tricuspid valve leaflets, with downward displacement and elongation of the septal and anterior cusp resulting in tricuspid regurgitation [2], The proximal part of the right ventricle is "atrialized", becoming thin walled and poorly contractile, along with an enlarged right atrium [3].
Patients can have a highly variable clinical course related to the anatomic abnormalities of Ebstein anomaly and their hemodynamic effects or associated structural and conduction system disease [4] like Atrial Septal defect (ASD) 90%. Pulmonary hypertension, ventricular and supraventricular tachycardia, ventricular septal defect, tricuspid atresia (30%), pulmonic stenosis and Wolf – Parkinson – White syndrome (WPW Syndrome) (up to 20% of patients) [5].

With this anomaly, fertility is usually unaffected even in women with cyanosis [2] the average life expectancy at birth of Patients with Ebstein anomaly is 25 – 30 years. It is rare congenital cardiac abnormally associated with Cyanosis & arrhythmia, patient often reach to child bearing age and pregnant women pose a challenge to the treating physician [6]. Due to its rarify and short life expectancy at birth. There are not many cases and varied clinical presentation associated with Ebstein anomaly during pregnancy. Therefore, this case is presented to increase awareness about this entity among obstetricians [7].

Case Report

A 31 year old G_2P_1 at 37 weeks pregnancy with mild PIH with

severe oligohydramnios + IUGR was admitted at our hospital. In the present pregnancy she has regular antenatal checkups at another hospital. The antenatal period was uneventful except obstetric complication – H/O PIH + oligohydramnios with IUGR.

The past history narrated by her was that in childhood some doctor told her she has some valve problem in heart. No Echo or other reports are available with her. She didn't have any symptoms related to this heart condition, so she ignored it.

On admission the general condition of the patient was good. No pallor, icterus, cyanosis, mild edema present. On examination of the respiratory system no abnormality was detected. Her pulse rate was 90 beats per minute, regular, blood pressure was 130 / 90 mm of Hg. Her oxygen saturation was 75%. On Cardiovascular system examination pansystolic murmur was heard.

Echo

Grossly dilated RA, Small size RV (Arterialized RV) with fair RV function
Apical displacement of septal leaflet of tricuspid valve
Severe valvular tricuspid regurgitation
Mild pulmonary hypertension
Mitral valve structurally normal, No MS/MR
Aortic valve structurally normal, No AS/AR

- Congenital Heart Disease
- Ebstein's Anomaly
- Normal biventricular Function
- No RWMA at rest LVEF 55%(Visual)
- Severe Valvular Tricuspid Regurgitation
- Mild Pulmonary Hypertension

Other Investigations Reports were as Follows

HB: 12.1 gm/dl, TLC & DLC: WNL, Blood Group: O positive, FBS: 103 ms/dl. Coagulation profile: Normal, Liver function Test & Kidney function Test: Normal. Thyroid Profile: Normal. All viral marker were Normal.

USG Report

There is single live intrauterine fetus is vertex presentation. Gestational age based on measurements of BPD, FL and AC is about 33 weeks & 3 days.

EFW: 2132 + /-311gms. Severe Oligohydramnios with AFI 3.2
Placenta fundal anterior Gr. – II
The Doppler parameters are s/o Barcelona stage I.
Patient was advised elective LSCS for obstetric as well as cardiac condition.
Elective LSCS was done under epiduralanesthesia.

An IUGR male baby weighing 1.9 kg was delivered. Baby cried immediately after birth. No resuscitation was required. Baby had Tachypnea so shifted to NICU for further management. Echo of baby was done after 2 days of birth, which was normal. Baby was discharged after 4 days of birth from NICU and handed over to mother. After 5th post operative day patient with her baby was discharged with advice to come after 3 days and to see cardiology OPD after 1 week (Figure 1).

Figure 1: After 5th post operative day patient with her baby.

Discussion

Majority of patients with Ebstein's anomaly were among neonates and infants with cyanosis and congestive cardiac failure. Patients who survived to adulthood may be symptomatic with onset of arrhythmia or by pregnancy. The average life expectancy at birth of patients with Ebstein anomaly is 25 – 30 years [3].

In Ebstein's anomaly, there is compromised right ventricular size and function, further impaired by the increased blood volume and cardiac output during pregnancy. Increased right atrial pressure and volume both worsen tricuspid regurgitation. Raised catecholamine with maternal hypoxemia and stress level in pregnancy further predispose the cases to arrhythmia. The hemodynamic problems seen during pregnancy depend on the severity of TR and the functional capacity of the RV [8] Heart failure, stroke, arrhythmia, paradoxical embolism can occur even in asymptomatic patients [2].

Management of patients with Ebstein's anomaly during labour focuses on maintaining normal sinus rhythm, avoiding fluid overload and providing enough relief of pain to the patient by epidural analgesia [9].

The advantage of epidural anesthesia are minimal intravascular volume shift, decreased catecholamine levels, control of maternal hyperventilation and most importantly postoperative analgesia, Intrathecalanesthesia may complicate a right to left cardiac shunt due to sudden decrease in sympathetic vascular resistance [7].

Large doses of oxytocin have discernible vasodilating effects and should be administered cautiously. Methylergometrine and prostaglandins increase pulmonary vascular resistance and are generally avoided. Oxytocin (5-10 IU intramuscularly) is commonly followed in obstetric protocols [10]. We were lucky that our patient had no problem throughout the pregnancy, intrapartum and postpartum period).

Conclusion

Maternal and fetal prognosis is favorable in patients with Ebstein's anomaly and NYHA class I. But it can be complicated with major cardiac events like tachyarrhythmia or cardiac failure [11].

Due to varied clinical presentations associated with Ebstein anomaly during pregnancy, such women should undergo close surveillance with multidisciplinary approach during the antenatal period to be diagnosed in terms of complications and hence to be treated accordingly.

Maternal mortality due to Ebstein anomaly is considered to be less than 1% in asymptomatic patients but may be as high 5-15%, if aggravated by conditions like supraventricular arrhythmia or atrial fibrillation [12].

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