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Case Study

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SUCCESSFUL PREGNANCY OUTCOME IN A PATIENT WITH EBSTEIN'S ANOMALY AND WOLFF-PARKINSON-WHITE SYNDROME: A CASE REPORT

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ABSTRACT

Ebstein's anomaly is a rare congenital heart disease characterised by abnormalities of tricuspid valve and right ventricle. It has an extremely variable natural history depending upon a variety of pathological features. It occurs in 1% of congenital heart disease cases. Pregnancy is usually well tolerated unless cyanosis or arrhythmia appears. There is an increased risk of prematurity, foetal loss and congenital heart disease in the offspring. I report a case of 26 years $G_2P_{1+0}(L_0)$ unbooked patient, who presented for the first time in a tertiary care hospital at 24 weeks of gestation. On reviewing her history and previous records she was found to have ebstein's anomaly .On further investigations she was found to have wolff-parkinson-white (WPW) syndrome. Patient was admitted and regular fetomaternal monitoring was done. The patient went into spontaneous labor at 37 weeks of gestation and delivered a live female child with birth weight 2.2 kg. Her postnatal period was uneventful. This case report gives insight into the variable presenting features, complications and management strategies during pregnancy to achieve best possible maternal and foetal outcome in a patient with ebstein's anomaly.

KEYWORDS: ebstein's anomaly, congenital heart disease, pregnancy, wolff-parkison-white syndrome.

INTRODUCTION

Ebstein's anomaly is a rare congenital heart disease with a reported incidence of 1 in 200,000 live births.[1] The first case was reported by Welheim Ebstein in 1966 of a 19 year old man who died shortly after presenting with dyspnoea, palpitations, cyanosis and heart failure.[2] Ebstein's anomaly is characterised by apical displacement of septal and posterior tricuspid valve leaflets resulting in decrease in size of the right ventricle and atrialization of right ventricle which then behaves functionally as a part of right atrium.[3] These patients may have severe tricuspid regurgitation leading to enlargement of right atrium. The common cardiac anomalies associated with the condition are atrial septal defect (90%), pulmonary hypertension, ventricular and supraventricular tachycardia, ventricular septal defect, tricuspid atresia (30%), pulmonary stenosis and accessory pathway formation like WPW syndrome (20%). First degree heart block is seen in 50% of the patients.[4] The clinical presentation of the disease is varied, patient may remain asymptomatic throughout life or may require surgery in neonatal period. Pregnancy is usually well tolerated and the severity of complications is directly proportional to the degree of tricuspid regurgitation, right ventricular function and presence of cyanosis. Ebstein's anomaly in pregnancy is associated with increased risk of prematurity, foetal loss and congenital heart disease in the offspring.[5] Multidisciplinary approach involving obstetrician, cardiologist, anaesthesiologist and neonatologist is essential for prevention of maternal and foetal morbidity and mortality.

CASE REPORT

A 26 years $G_2P_{1+0}(L_0)$ unbooked patient, presented for the first time to the outpatient department of a tertiary care hospital at 24 weeks of gestation. In the previous pregnancy also she was unbooked and delivered a female child at home at around 32 weeks of gestation. The baby died 2 days after birth and the cause of death was not known. She conceived spontaneously 1 year after the last child birth. The patient gave history of some heart disease diagnosed in childhood but was not on any treatment or follow up for the same from the past 15 years. On checking her previous records it was found that she was a diagnosed case of ebstein's anomaly. The patient was then admitted for further investigations and monitoring. There was no history of shortness of breath, palpitation, fatigue, cyanosis and chest pain. She belonged to NYHA class I. At admission her blood pressure (BP) was 124/84 mm of Hg and pulse rate (PR) was 94/minute, regular, good volume with no radio-radial delay and no radio-femoral delay. Cardiovascular examination revealed systolic murmur in the pulmonary region with splitting of 2nd heart sound. Chest examination was normal with bilateral vesicular breath sounds and no added sounds. On per abdominal examination height of uterus (HOU) was corresponding

to the period of gestation (POG) with regular foetal heart sound~140/minute. Ultrasonography for foetal well being and to rule out any gross congenital anomaly (CMF) was done which revealed normal foetal growth parameters and no evidence of gross CMF. Her routine antenatal investigations were normal. ECG findings showed short PR interval with widening of QRS complex and presence of delta wave. Patient was advised maternal echocardiography and cardiology consultation. Echocardiography findings showed apical displacement of tricuspid valve (4.2 mm distal to Atrioventricular junction) with small functional right ventricle and enlarged right atrium along with moderate tricuspid regurgitation (TR). There was an atrial septal defect (ASD) and left to right shunt with satisfactory right/left ventricular systolic function. Left ventricular ejection fraction was 60%. On cardiologist consultation a diagnosis of ebstein's anomaly grade 2 with ASD with moderate TR and WPW syndrome was made. The patient was started on tablet metoprolol XL 50mg once daily. Foetal echocardiography revealed no obvious congenital heart anomaly. Regular fetomaternal monitoring was done. Patient remained asymptomatic throughout the antenatal period and went into spontaneous labor at 37weeks of gestation. Strict vital monitoring was done and labor was monitored partographically. To combat the pain and subsequent increased work load on the heart epidural analgesia was given to the patient. Prophylaxis against bacterial endocarditis was given. Outlet forceps were applied to cut short the second stage of labor and patient delivered a healthy female child with birth weight 2.2Kg. Postpartum period was uneventful. Patient was discharged on 5th postnatal day in good health and was advised regular follow up in the department of cardiology.

DISCUSSION

The exact etiology of the ebstein's anomaly is unknown. Environmental factors implicated in its aetiology include maternal ingestion of lithium or benzodiazepine and maternal history of miscarriage.[6] The embryological development of tricuspid valve leaflets and chordae involves undermining of the right ventricular free wall. This process continues to the level of the atrioventricular (AV) junction. In Ebstein's anomaly, this process of undermining is incomplete and falls short of reaching the level of the AV junction. In addition, the apical portions of the valve tissue, which normally undergo resorption, fail to resorb completely. This results in distortion and displacement of the tricuspid valve leaflets. Ebstein's anomaly may manifest clinically at any age. If it manifests in early neonatal period then surgical correction is feasible with good early and late survival and excellent functional status.[7] In adults the common symptoms include cyanosis, fatigue, dyspnoea, palpitations and decrease exercise tolerance.[8] In the present case report patient was asymptomatic with NYHA class I disease. In the presence of an interatrial defect risk of paradoxical embolization, brain abscess and sudden cardiac death increases. Pregnancy is a state of hyperdyanamic circulation because of an increase in cardiac output by 50% with an increase in both stroke volume and heart rate along with a decrease in systolic blood pressure due to decrease in peripheral vascular resistance and placental shunting of blood.[9] In ebstein's anomaly there is compromised right ventricular size and function therefore in such patients normal physiological changes of pregnancy may have an adverse haemodynamic consequence resulting in worsening of tricuspid incompetence and raised right atrial pressure.[10] The prognosis depends on the severity of anomaly, degree of tricuspid regurgitation, degree of heart failure and associated arrhythmias.[11] Patients with ebstein's anomaly and without significant cardiomegaly, cyanosis and arrhythmias are categorised into WHO risk class II. Pregnancy is usually well tolerated in these patients. In this case report also the patient had no significant symptoms and there were no complications in the antenatal, intrapartum and postnatal period. During the antenatal period the patient should be carefully monitored with proper counselling and adverse outcomes in the pregnancy as well as regular cardiology consultation and maternal echocardiography should be done.[12] Most of the patients go into spontaneous labor and the preferred mode of delivery is vaginal; caesarean section is reserved for obstetric indications only. During the intrapartum period to reduce maternal stress and further worsening of heart disease adequate pain relief in the form of epidural analgesia should be given. It is advisable to cut short the 2nd stage of labor by outlet forceps application to decrease the work load on the heart.[13] All these management strategies were also applied in the present case and she delivered a healthy female child vaginally without complications. Though endocarditis prophylaxis is not indicated in patients with ebstein's anomaly.[14]; however, it was given in our patient. Ebstein's anomaly is neither a contraindication for pregnancy nor an indication for termination of pregnancy. Pregnancy should be carefully monitored with close surveillance of maternal and foetal condition for early recognition and management of complications to achieve best possible outcome.

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