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Case Report

Successful Outcome of a Post-dated Pregnancy in a Patient with Uncorrected Tetralogy of Fallot with Pulmonary Atresia

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Abstract

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease, with an overall incidence of around 10% of all congenital heart diseases. TOF with pulmonary atresia is at the extreme end of the anatomic spectrum which is difficult to correct surgically. We have discussed here a rare case of a post-dated pregnancy with uncorrected TOF with pulmonary atresia and its successful pregnancy outcome in a 24-year-old woman at a tertiary care centre with a multidisciplinary approach.

Key words: Congenital heart disease, Pulmonary atresia, Tetralogy of fallot

INTRODUCTION

Tetralogy of Fallot (TOF) is characterized by pulmonary stenosis, right ventricular hypertrophy, overriding aorta, and non-restrictive ventricular septal defect (VSD). TOF is the most common form of cyanotic congenital heart disease occurring in approximately 1 in 3600 live births. It accounts for 3.5% of all infants born with congenital heart diseases. To reach adulthood, most patients end up requiring correctional surgery for TOF. Only few patients survive up to adulthood without correction of TOF.[1,2] Survival into the fourth decade without surgery is extremely rare (only about 3%).[3] The principle danger for a pregnant woman with TOF is cardiac decompensation due to inability to meet the additional demands imposed by the physiological changes of pregnancy and parturition. Maternal complications in these women include worsening cyanosis and dyspnea, right ventricular failure, thromboembolism, pulmonary hypertension, arrhythmias, and maternal mortality. Fetal complications include abortions, intrauterine growth restriction, prematurity, and intrauterine demise. Obstetrical events include spontaneous abortions, premature rupture of membranes, and preeclampsia. The risks of maternal cardiac complications depend on the severity of residual lesions at the time of conception. Maternal death is rare. Pregnancy is generally well tolerated with no long-term sequelae.

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Patients with TOF often present with cyanosis in the 1st year of life. Survival of these patients largely depends on the degree of pulmonary obstruction and the pulmonary blood supply. Limitation of blood to the lungs combined with VSD results in supply of oxygen-poor blood to the body, causing cyanosis. Pink TOF is characterized by mild pulmonary stenosis, and the VSD is in balance resulting in no significant right to left shunt. The degree of right ventricular outflow is minimal and no cyanosis develops in these patients. These patients have mild symptoms and diagnosis may be delayed, and these patients are often asymptomatic up to adulthood.

Heart diseases complicate 0.2–3% of pregnancies and congenital heart lesions now constitute at least half of all these cases. TOF with pregnancy is even rarer. The incidence of TOF with pulmonary atresia is 0.007/1000 live births and accounts for one-fifth of all cases of TOF.[4] Uncorrected TOF with pulmonary atresia in pregnancy is a rare entity with limited literature. Pregnancy in uncorrected TOF entails serious risks including increased maternal morbidity and mortality up to 15% and poor perinatal

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outcome.[5] Most women with repaired TOF can have successful pregnancies. In agreement with the WHO classification, the repaired TOF is ranked in Class II category and the cyanotic unrepaired heart diseases in Class III category.

CASE REPORT

A 24-year-old Primigravida residing from Golaghat, Assam, was referred from Jorhat Medical College and Hospital. She was referred as a case of Primigravida at term pregnancy not in labor with subaortic VSD with bidirectional shunt and valvular pulmonary stenosis. The patient had presented to Jorhat Medical College with occasional complaint of cough and easy fatigability, and she was classified as NYHA Class II. The patient had an uneventful childhood. During the third trimester (8th month), she had occasional cough, easy fatigability, and shortness of breath which was not relieved on over the counter drugs for which she consulted her obstetrician. On investigation, a cardiac lesion was detected which on further investigation was found to be acyanotic TOF. She was hence referred to Assam Medical College and Hospital for further management on July 22, 2017 and was admitted in labor room the same day.

On general examination, she was fully conscious and oriented with a pulse rate of 92/min regular and normovolemic, blood pressure of 102/82 mmHg, respiratory rate of 22/min, afebrile on touch, no evidence of central cyanosis, clubbing, or peripheral edema. On cardiovascular and respiratory examination, she had normal vesicular breath sounds and bilaterally symmetric chest movements. S1 was normal and S2 was single and loud with a pansystolic murmur. Obstetric per abdominal examination showed a term sized uterus, longitudinal lie, cephalic presentation, fetal heart sounds localized on the right spinoumblical line which was 144/min in rate and regular in nature. The patient was shifted to antenatal ward and was investigated further. Investigations showed hemoglobin of 12.0 g%, white blood cell 7200/cc, and platelet count 1.9 lakh/cc. The hematocrit was mildly raised (49%). O_2 saturation at room air was 97%. Coagulation profile, blood chemistry, urine analysis, and serum electrolytes were within normal limit. Ultrasonography revealed a single live fetus in cephalic presentation with a gestational age of 35 weeks and 5 days, BPP of 8/8 and estimated fetal weight of 2768 g. No gross fetal congenital anomaly was noted. Doppler was normal.

Electrocardiogram showed sinus rhythm with right axis deviation and right ventricular hypertrophy. Echocardiogram (ECHO) showed situs solitus with AV and VA concordance with normal biventricular function, non-restrictive sub-aortic VSD with bidirectional shunt with overriding aorta, right ventricular hypertrophy with mild pulmonary stenosis, and small pulmonary atresia. No evidence of any vegetation or effusion. No coarctation of the aorta and no collaterals were seen from descending aorta. Ejection fraction was 67%. Cardiologists advised infective endocarditis prophylaxis before delivery of ceftriaxone 2 g and gentamycin 120 mg, and the patient was considered as moderate risk for delivery. The patient and party were counseled about the situation of patient and induction was considered. PGE2 gel was instilled intracervically on July 28, 2017 in morning. Strict maternal and fetal monitoring continued. The patient delivered a healthy 2.830 kg male baby in morning of July 29, 2017 by outlet forceps application. Active management of 3rd stage of labor was done. Post-delivery immediately 40 mg furosemide was administered intravenously, uterotonics were used and fluids were restricted. After delivery, her general condition was fair along with pulse rate of 92/min, regular and BP was 104/82 mm of Hg. Oxygen inhalation was continued throughout delivery and after delivery as well. Post-delivery vital monitoring of patient continued. Uterus was well contracted. No post-delivery complication was noted. The patient was shifted to cardiac intensive care unit the same day for intensive monitoring. The patient was stable and shifted back to obstetric HDU on July 31, 2017. The patient was discharged on August 1, 2017 (4th postnatal day). Fetal ECHO was advised to be done for the baby. Risks of next pregnancy were well explained to the patient and her husband, and the husband was advised to use barrier method for contraception. The patient came for follow-up 12-week postpartum and reviewed in cardiology department as well. Both the mother and the baby enjoyed good health. Regular follow-up was advised from cardiology point of view. Fetal ECHO was not done by the patient and was again reminded to do so. A surgical correction was offered for the same [Figures 1 and 2].

DISCUSSION

Uncorrected TOF in adulthood is rare and TOF with pregnancy is even rarer. Although survival into adulthood without correction is rarely reported, the development of congenital systemic-pulmonary collaterals may enable patients with pulmonary atresia to reach adult age as in our patient. It is classified as moderate risk for pregnancy with heart disease with mortality rate reaches 5–15%. Management of these patients in pregnancy is challenging due to associated hemodynamic alterations. To reduce these problems, sufficient rest and oxygen therapy have been suggested. Careful postpartum monitoring is essential in these patients as sudden cardiovascular alterations in the postpartum period may be detrimental in these patients.

Figure 1: The patient with pink tetralogy of Fallot at 12-week follow-up

Figure 2: The infant born to mother with pink tetralogy of Fallot at 12-week follow-up

Active management of 3rd stage of labor and prevention of postpartum hemorrhage is mandatory as even lesser blood loss can lead to decompensation in these women.

During pregnancy, the additional observations needed include blood gas analysis (especially pO2 and saturation of O2) and hematocrit/hemoglobin. Fetal growth and well-being need to be monitored carefully. Vaginal birth is the preferred mode of delivery in women with TOF as in our patient. Cesarean section is indicated only for obstetrical indications. Infective endocarditis prophylaxis is recommended, which was given in our patient.

Genetic counseling and transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5–50%, compared to a background risk of 1% of having a baby with congenital heart disease. Incidence of cardiac defects in infants of TOF patients ranges from 3 to 17%.[6] The risk of transmission is dependent on associated genetic syndrome. A discussion about contraceptive methods is appropriate in all women with repaired or unrepaired TOF. Combined hormonal contraceptives are contraindicated in women with unrepaired TOF. Regular follow-up should be done from cardiology point of view.

A multidisciplinary teamwork is essential for the management of patients with uncorrected TOF and pregnancy. Without optimum intensive obstetrical and medical management, prognosis of these patients remains poor with high morbidity and mortality rates.

CONCLUSION

Pregnancy in patients with uncorrected TOF is a life-threatening condition for both mother and fetus. Various maternal, fetal, and obstetrical complications are associated in pregnant patients with uncorrected TOF. A multidisciplinary approach is required for the management of patients with uncorrected TOF and pregnancy.

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