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Case Report

Successful pregnancy outcome in a case of Eisenmenger syndrome

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ABSTRACT

Eisenmenger syndrome is a very rare heart disease in pregnant women. Eisenmenger syndrome is defined as pulmonary hypertension resulting from an uncorrected left to right shunt of a VSD, ASD or patent ductus arteriosus (PDA), with subsequent shunt reversal and cyanosis. Pulmonary hypertension is the prognosis index. The high mortality risk (25-30%) is associated with pregnancy and the peripartum period due to cardiovascular collapse. Pregnancy should be discouraged and reliable contraception, preferably permanent sterilization, advised because of the extreme risk associated with pregnancy. Depo-Provera or progesterone implants are nonsurgical alternatives. Even first-trimester termination is associated with a maternal mortality rate of 5-10%. However, despite all the risks, a few patients deliver successfully with a good maternal and neonatal outcome. We presented a 27-year-old booked G1P0+0 admitted at 34 weeks gestation with Eisenmenger syndrome. She was treated medically during pregnancy, underwent elective caesarean section at 34 weeks of gestation delivered a baby and was subsequently discharged on the 15th postoperative day without any serious complications.

Keywords: Eisenmenger syndrome, Pregnancy, Pulmonary hypertension

INTRODUCTION

Eisenmenger's syndrome (ES) was first described in 1897. ES is rare in pregnant women with an incidence of about 3%.[1] Pregnancy with ES has a high risk due to hypoxia, thrombosis, cyanosis, arrhythmia and cardiac failure, which can appear as common complications as early as the second trimester. Sudden cardiac arrest can occur during caesarean section or soon after birth.[2] The objective of this study was to highlight the risk factors for pregnancy complicated with ES.

CASE REPORT

27 years old primigravida women came to antenatal OPD R. G. Kar Medical College at 10 weeks of gestation. She had h/o exertional dyspnoea since childhood. She was diagnosed with congenital heart disease (ventricular septal defect) at 12 years of age. No corrective surgery was performed at that time. Despite the life-threatening risk to the mother, she opted to continue the pregnancy.

She was followed up in OPD on regular basis after that. She was a known case of type 2 DM and was on tablet metformin (500 mg) TID, during pregnancy insulin lispro was added in the early 2nd trimester. She was taking metoprolol (12.5) and tablet furosemide (40) and tablet sildenafil (20). A cardiology opinion was sought. Baseline echocardiography and ECG were done and she was kept on medical treatment with sildenafil (20 mg) and bosentan (62.5 mg). Throughout the follow-up she was normotensive and her insulin dose was titrated according to the increasing gestational age requirement. Fetal growth was evaluated using serial ultrasound. Due to polycythaemia, phlebotomy was performed at the 25th week of gestation. The pregnancy progressed uneventfully until the 26th week when the patient had a bronchial

infection and was treated with amoxycillin, which improved her condition. She was now admitted at 34 weeks gestation with mild dyspnoea.

Figure 1: Peripheral cyanosis.

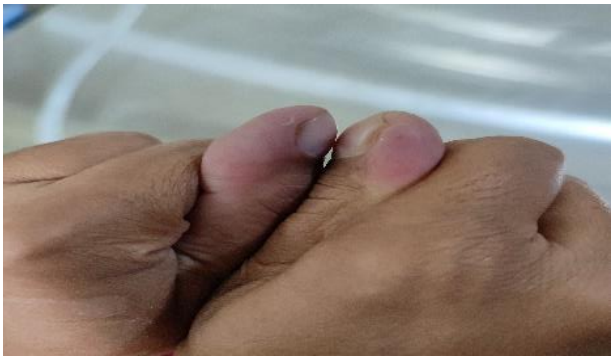

Figure 2: Clubbing with cyanosis.

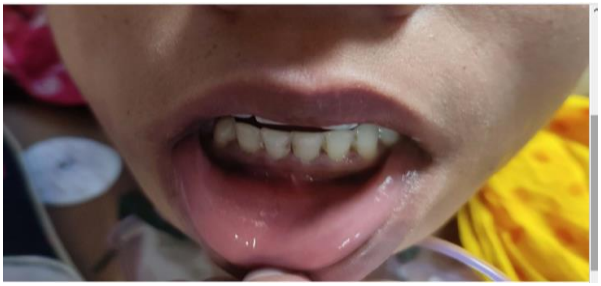

Figure 3: No central cyanosis.

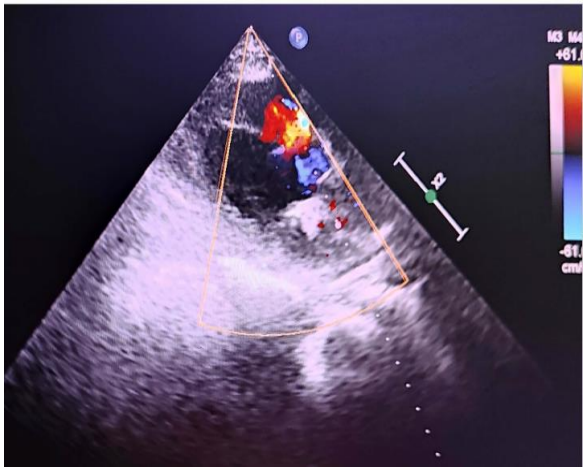

Figure 4: Echo with Doppler revealed bidirectional flow.

On examination, she was of average height (152 cm) and 53 kg weight compared to 1st antenatal visit which was 46 kg. No history suggestive of orthopnoea, dizziness or palpitations was present. General examination revealed oedema, clubbing and peripheral cyanosis (Figure 1 and 2). Her vitals were stable with a pulse rate of 102 beats/minute and blood pressure of 120/80 mm of Hg in the right upper limb. Her respiratory rate was 24 /minute and JVP was raised. Thyroid and breast examination revealed no abnormality. On CVS examination, she had thrill over the apex, pansystolic murmur (G2) with P2 loud, but no features of right heart failure. Her saturation was 88-90% in room air. The apex beat was shifted 1 cm lateral to the midclavicular line and left parasternal heave was present. Respiratory system examination revealed bilateral vesicular breath sounds in all lung fields. Abdominal examination revealed the uterus to be 34 weeks in size, relaxed with cephalic presentation and fetal heart at 132 beats/minute. On per vaginal examination, cervix was uneffaced, firm, posterior and internal os was closed.

On investigation, haemoglobin was 14.1 g/dl and haematocrit were 42%. Complete blood count and urine examinations were found to be normal. ECG showed features of right axis deviation and echocardiography reported large perimembranous VSD (17 mm) with severe pulmonary artery hypertension (34 mmHg), tricuspid regurgitation and bidirectional shunt with ejection fraction 62% (Figure 4). She was managed conservatively; a cardiology consultation was obtained after which she was put on complete bed rest, restricted physical activity, oxygen therapy and anxiolytics.

Her ultrasound report at the time of admission showed a single live intrauterine fetus of 33 weeks gestational age with adequate liquor. Placenta was located anteriorly; grade III and the expected baby weight was 1.4 kg. Antenatal corticosteroid was administered for fetal lung maturation.

Vaginal delivery under epidural analgesia was 1st thought of and dinoprostone gel (0.5 mg) was given for cervical ripening, but after 24 hours no improvement in Bishop's score was found. She underwent elective caesarean section under epidural analgesia in presence of a cardiologist and after high-risk consent. A term female baby weighing 1.6 kg was delivered by vertex. Liquor was clear and adequate and the baby cried soon after birth with an APGAR score of 7 at 1' and 8 at 5'. The intraoperative period was uneventful and fluid input was monitored strictly by central venous pressure monitoring.

She was shifted to ICU immediate postoperative period. Her vitals were stable but respiratory system examination revealed bilateral rhonchi. In ICU she was given oxygen inhalation via nasal cannula, higher antibiotics and diuretics and kept for observation. The uterus was well contracted with no active bleeding per vagina. She stayed in ICU for 3 days. Vasodilator agents, sildenafil tablet 20 mg two times a day and Bosentan 62.5 mg were added

on day 4; injection LMWH (40IU) was started and the stitch was removed on day 10.

Her baby was admitted to the neonatal ICU for 11 days given low birth weight. The baby was discharged on day 12, with a weight of 1985 g. Both the mother and the baby were healthy on discharge. She was advised to barrier contraceptives and to follow up in the cardiology outpatient department.

DISCUSSION

ES is associated with a risk of sudden death. The increase in blood volume and decrease in systemic vascular resistance in pregnancy can lead to right ventricular failure, with a decrease in cardiac output and sudden death. Maternal mortality with ES is as high as 40% in pregnancies that continue past the first trimester.[3] In contrast, the 15-year survival rate is more than 75% in non-pregnant patients.[4] Postoperative fluid shifts associated with caesarean delivery pose an even greater risk, with mortality rates approaching 70%.

Moreover, straining during labour may result in an increased right ventricular pressure, which may cause fatal arrhythmia and even sudden fetal death.[5] Pregnancy should be avoided in women with ES due to high morbidity and mortality.[6]

The preferred mode of delivery is vaginal with adequate pain relief using intravenous analgesics or low-dose epidural anaesthesia. Care must be taken during the conduct of anaesthesia as hypotension can be life-threatening in these patients.[7] Benefits of vaginal delivery include less blood loss, less risk of infection and lesser risk of clot formation; but disadvantages include increased pain and stress and increased sympathetic flow, thus increasing the load on the ventricles. A caesarean section should be reserved for conditions such as severe intrauterine growth retardation and obstetrics indications.[8] Vaginal delivery remains an acceptable option and still accounts for the majority of births in some series.[9] 65% Maternal mortality in ES is reported with caesarean section.[10]

Anaesthesia for patients with pulmonary hypertension is controversial. The use of epidural or intrathecal morphine sulphate, a technique devoid of effect on systemic blood pressure, maybe the best approach to the anaesthetic management of these difficult patients.[11]

FGR is a common fetal complication. If the maternal oxygen saturation is <85%, babies usually die in utero before reaching a viable gestation and early miscarriage is very common. Preterm delivery is also frequent reaching up to 85% of pregnancies.[3] Fetal outcome correlates well with maternal haematocrit and successful pregnancy is very unlikely with a haematocrit >65%.[12] Despite the maternal and fetal complications, the neonatal survival rate of babies alive at birth approaches 90%.[13] Congenital heart defects are seen in approximately 5% of offspring.[13]

CONCLUSION

Thus, to conclude, pregnancy should be avoided in a woman with ES because of a high maternal mortality rate. If a patient with congenital heart disease and ES comes with pregnancy, she should be managed in a tertiary care centre with a multidisciplinary approach, a team of obstetricians, cardiologists, skilled anaesthetists and paediatricians.

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