Case Report

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Successful Pregnancy Outcome in A Case of Eisenmenger Syndrome: A Rare Case Report

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ABSTRACT

Pregnancy complicated with Eisenmenger syndrome is associated with high risk to the fetus as well as the mother. There is approximately 50% risk of sudden maternal death, frequently occuring a few days postpartum and the overall fetal wastage is reported to be up to 75%. Patients with Eisenmenger syndrome are advised to refrain from pregnancy or to terminate pregnancy by the end of first trimester itself. Management of these patients requires a co-ordinated multi-specialist care when such pregnancies reach a stage where safe termination is not advisable. However, inspite of all the risks, a few patients deliver successfully with a good maternal and neonatal outcome. We present a 27-year-old unbooked $G_3P_1A_1L_0$ admitted at 34 wk gestation with Eisenmenger syndrome. She was treated medically during pregnancy, underwent elective Caesarean section at 37 wk of gestation delivered a healthy baby and was subsequently discharged on the 10th postoperative day without any serious complications.

Keywords: Pulmonary hypertension, Pregnancy, Sildenafil

CASE REPORT

A 27-year-old G3P1A1L0 came to our OPD at 34 wk 4 d gestational age with chief complaint of pain abdomen for 2 d which was not increasing in frequency or duration nor associated with bleeding or leaking per vaginum. She also had pedal oedema of one month duration and history of breathlessness on exertion for past three years. During her previous delivery at home two years back, she had an attack of breathlessness with cyanosis after expulsion of a pre-term dead female fetus at six months gestational age weighing 1 kg. She was admitted to hospital and diagnosed with heart disease. Our patient was a known case of Ventricular Septal Defect (VSD) with Eisenmenger syndrome on medication (Sildenafil) for the last two years, which was discontinued four months ago on her own. She had a spontaneous abortion one year back at three months gestational age for which no dilatation and curettage was required. Her antenatal visits were not regular. Family history revealed nothing significant with respect to heart disease.

On examination she was short statured (height 136 cm) and poorly built weighing 47kg. General examination revealed oedema, clubbing and central cyanosis. Her vitals were stable with pulse rate of 100 beats/minute and blood pressure 130/100 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 with P2 loud. 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 uterus to be 32 wk size, relaxed with cephalic presentation and fetal heart 132 beats/minute. Parietal wall oedema and vulval oedema were present. On per vaginal examination cervix was uneffaced, firm, posterior and internal os closed.

On investigation, hemoglobin was 11 g/dl and hematocrit was 60%. Complete blood count and urine examinations were found to be normal. ECG showed features of biventricular hypertrophy and echocardiography reported large perimembranous VSD with severe Pulmonary Artery Hypertension, Tricuspid Regurgitation and bidirectional shunt with ejection fraction 40%. Her ultrasound report at the time of admission showed a single live intrauterine fetus of 33 wk gestational age with adequate liquor. Placenta was located anteriorly, fundal, grade III and the expected baby weight was 1.1 kg. She was managed conservatively; cardiology consultation was obtained after which was she was put on Sidenafil, furosemide, spironolactone and nifedepine.

She was advised complete bed rest, injection ampicillin and gentamycin, betamethasone for fetal lung maturity, iron, calcium tablets and anxiolytics. She underwent elective lower segment caesarean section at 37 wk under epidural anaesthesia in the presence of cardiologists and after high risk consent. A term female baby weighing 1.6 kg was delivered by vertex with application of forceps. Liquor was clear and adequate and the baby cried soon after birth with APGAR score 8 at 1' and 9 at 5'. Intraoperative period was uneventful and fluid input was monitored throughout strictly by central venous pressure monitoring.

On the first postoperative day, she developed fever which did not subside on paracetamol infusion, for which she was shifted to intensive care unit (ICU). Her vitals were stable but respiratory system examination revealed bilateral rhonchi. In ICU she was given oxygen inhalation via nasal cannula, higher antibiotics, nebulisation with β_2 agonists and steroids and kept for observation. Uterus was well contracted with no active bleeding per vaginum. Urine examination showed plenty of pus cells and she was treated for urinary tract infection. Oxygen saturation was maintained at 88% on room air.

Her condition improved and she was shifted to ward on seventh postoperative day. She was discharged on tenth postoperative day with advice of strict use of barrier contraception and to attend cardiology outpatient department (OPD) for follow up. At six weeks follow up, she was well with healthy baby, exclusively breast feeding, advised for barrier contraceptive and regular cardiac check up. At three month follow up she was asymptomatic with healthy baby and was advised for permanent sterilization.

DISCUSSION

Eisenmenger's syndrome was first described in 1897. The incidence of Eisenmenger's syndrome is approximately 3% of patients with congenital heart defects. Maternal mortality in this situation is high but that associated with VSD (60%) is higher when compared with ASD or PDA [1]. A high incidence of maternal death was associated with hypovolemia, thromboembolic phenomena and preeclampsia [2]. Cartago et al., in their study also showed two cases of

Eisenmenger syndrome treated with Sildenafil as monotherapy caused stabilization of the maternal condition and good clinical outcome [3].

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, may be the best approach to anesthetic management of these difficult patients [4]. There is no evidence to support the choice of either vaginal or cesarean delivery for cardiac reasons: vaginal delivery is associated not only with a lower average blood loss but also increased maternal effort. Gleicher et al., reported a 34% mortality associated with vaginal delivery and a 75% mortality associated with cesarean section [5].

Third trimester fetal surveillance with ultrasound and antepartum testing is important because at least 30% of the fetuses will be growth retarded. Maternal arterial partial pressure of oxygen should be maintained at a level of 70 mmHg or above [6]. This is explained by a high incidence of spontaneous abortions, a 30-50% risk of premature delivery and low birth weights as maternal hypoxemia disturbs fetal growth [5]. Fetal outcome correlates well with maternal haematocrit and successful pregnancy is unlikely with a haematocrit >65% [7].

Kansaria reported a case of Eisenmenger Syndrome in pregnancy where patient died three weeks after delivery [7]. There is no survival case of Eisenmenger syndrome complicated by severe preeclampsia reported in literature. Phupong et al., reported the fatal outcome of a pregnant woman with Eisenmenger syndrome associated with VSD who underwent caesarean section because of severe preeclampsia and an unfavourable cervix, and died from pulmonary embolism in the postpartum period [1]. In a review of seven patients with this syndrome who received prophylactic anticoagulation, Pitts et al., implicated secondary hemorrhage as the cause of death in five [8].

The unique feature in our case is that despite having an Eisenmenger syndrome with associated VSD our patient conceived, continued pregnancy without much major symptoms and antenatal visits; delivered a healthy baby and survived the surgery well.

CONCLUSION

In conclusion, although pregnancy should be discouraged in women with Eisenmenger's syndrome, it can be successful as seen in our case wherein co-ordinated multi-specialist care and proper monitoring helped in avoiding any adverse maternal or fetal outcome. This case may help in giving ray of hope to all such pregnancies complicated with Eisenmenger syndrome at an advanced gestational age where termination is not possible.

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