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Case Report

Undiagnosed cyanotic heart disease - A challenge to emergency medical professionals

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ABSTRACT

Cyanotic heart disease characterized by pulmonary hypertension with reversal or bidirectional shunt associated with atrioventricular septal defects or patent ductus arteriosus is known as Eisenmenger syndrome. Pregnancy-associated decreased systemic vascular resistance increases the degree of right to left shunting and hypoxemia, thereby increasing substantially both the maternal mortality and fetal wastage which is reported to be as high as 70%. Ideally, this is a contraindication for conceiving but if there is strong desire of pregnancy, it should be a supervised antenatally by multidisciplinary team. We present a case of a young primigravida who had cesarean delivery under subarachnoid block at 37 weeks of gestation and was referred after fall in oxygen saturation. It can be a challenge to manage these cases at a peripheral unit if the condition is undiagnosed.

Keywords: Eisenmenger syndrome, Cyanotic heart disease, Hypoxemia, Postpartum

INTRODUCTION

Eisenmenger syndrome is a rare cyanotic heart disease characterized by pulmonary hypertension with shunt reversal or bidirectional shunt associated with atrioventricular septal defects or at aortopulmonary level.[1] Pregnancy can worsen Eisenmenger syndrome due to fall in systemic vascular resistance resulting in higher fraction of right to left shunt and hypoxemia that is associated with high maternal mortality and adverse fetal outcome ranging from 30 to 50% and even up to 70%.[1,2] ES can be seen in 3% of the pregnant females.[3] As ES is associated with high maternal mortality and fetal demise, the diagnosis and anesthetic management is crucial for successful outcome. After obtaining consent, we present a case of 25 years primigravida who presented to a peripheral center for the management of labor and underwent emergency cesarean delivery due to fetal distress and was referred to our center after desaturation in the post-operative period.

CASE REPORT

A 25-year-old primigravida and no antenatal visit, with 37 weeks of gestation, presented to a peripheral hospital with labor pains and fetal distress, as this was an emergency she was taken for emergency cesarean surgery under single-shot spinal anesthesia. She gave birth to a dead fetus. After the surgery, she had a fall in oxygen saturation (SpO2-45%), so she was referred to

our center. When received in emergency room, her pulse was 130/min, blood pressure 100/70 mmHg, and oxygen saturation 60% with oxygen supplementation. She was cyanosed. On auscultation, there was a systolic murmur at the pulmonary area, bilateral rhonchi in the lungs. Immediately, trachea was secured with an endotracheal tube and was ventilated with 100% oxygen. She was treated on as peripartum cardiomyopathy. She was given antibiotics, diuretics, steroids, nebulization with beta agonist, and other supportive treatment. Electrocardiogram revealed right ventricular hypertrophy. Echocardiography was done on urgent basis which revealed large muscular 12 mm ventricular septal defect with right to left shunt, severe pulmonary hypertension (PASP – 110 mmHg), severe TR, and LVEF – 45%. Her hemoglobin was 14.2g % and hematocrit of 42. Chest radiograph revealed prominent pulmonary conus and pulmonary vascular congestion. Her arterial blood gases showed pH – 7.30, PaO2 – 54, PaCo2 – 49, HCO3 – 18, and BE – (-8). All other routine investigations were within normal limit. She was ventilated for 24 h. The very next day the trachea was extubated as she was conscious, oriented and her hemodynamics were stable, her oxygen saturation remained between 80 and 86% with oxygen supplementation. She was started on oral sildenafil. On the 5th day, she was discharged from the hospital with advice to follow up in the OPD.

DISCUSSION

Eisenmenger syndrome is ideally a contraindication for pregnancy, if pregnancy is continued, early birth of the child without any prolongation of labor is desired by meticulous planning by multidisciplinary team of obstetrician, anesthesiologist, cardiologist, and neonatal physicians. In this patient, differential diagnosis was acute respiratory distress syndrome, peripartum cardiomyopathy, and pulmonary thromboembolism and was treated on these lines but exact diagnosis was known after the echocardiography findings. Hypovolemia can lead to increased hematocrit and increased PVR thus right to left shunt so fluid management is important.

Since the fall in systemic vascular resistance seen in pregnancy can worsen the right to left shunt with a very high rate of mortality reaching 30–50%.[1] The single-shot spinal is further associated with decrease in systemic vascular resistance and hence right to left shunt. There have been few case reports with better maternal and fetal outcome under combined spinal and epidural anesthesia. Maternal mortality is higher when associated with VSD in such patients as compared to atrial septal defects or patent ductus arteriosus.[3] The outcome can be improved by antenatal diagnosis and meticulous planning. Sildenafil therapy produces better maternal outcome.[4] Oxygen supplementation can improve the pulmonary hypertension and thus decrease right to left shunt.

Meticulous deairing of all the intravenous lines is of utmost importance to prevent paradoxical air embolism. Infective endocarditis prophylaxis has been advised in all patients of cyanotic heart diseases.[5]

Intermittent positive pressure ventilation during general anesthesia can increase the PVR due to increase in intrathoracic pressure and resultant fall in venous return. Titrated epidural anesthesia prevents hypotension. However, only epidural anesthesia produces segmental action and may provide inadequate anesthesia with need to convert into general anesthesia. In this scenario, combination of intrathecal opioid and titrated epidural local anesthetics helps to increase the safety and reliability of the neuraxial block. Furthermore, post-operative epidural analgesia helps to reduce pulmonary hypertension.[6] The choice of anesthesia depends on the prevailing hemodynamic and pathophysologic condition. Early post-operative mobilization also helps to reduce other complications. Delivery by a pregnant woman with Eisenmenger syndrome represents an increased risk of pulmonary thromboembolism and sudden death, often occurring within the first few days of postpartum.[7] e degree of maternal hypoxemia is the most important predictor of fetal outcome as it has been noted that pre-pregnant arterial oxygen saturation is in a direct proportion to live births. ere are high chances of premature delivery and intrauterine growth retardation. Poor prognostic signs in maternal congenital heart disease include maternal hematocrit >60%, arterial oxygen saturation <80%, right ventricular hypertension, and syncopal attacks. A fixed PAH not responsive to oxygen also carries a grave prognosis and may be an absolute indication to terminate the pregnancy.[8]

The diagnosis in this patient was missed as there was no antenatal clinic visit and there was no history suggestive of any cardiac disease. There was some degree of polycythemia which can be thought of retrogradely. There are limited resources at the periphery and diagnosis of these diseases is difficult and if any complication occurs, the management becomes difficult. Sometimes in the emergency situation, the proper history and examination can be missed.

As this condition has no shunt murmur at later stage, the diagnosis by clinical examination is difficult.

CONCLUSION

Cyanotic heart disease is ideally a contraindication for pregnancy, if pregnancy is continued prenatal diagnosis and patient counseling is of utmost importance for fruitful maternal and fetal outcome. Careful history taking, examination, and hemodynamic parameters even in emergency situation are desired.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Cole PJ, Cross MH, Dresner M. Incremental spinal anaesthesia for elective caesarean section in a patient with Eisenmenger's syndrome. Br J Anaesth 2001;86:723-6.
2. Gurumurthy T, Hegde R, Mohandas B. Anaesthesia for a patient with Eisenmenger's syndrome undergoing caesarean section. Indian J Anaesth 2012;56:291-4.
3. Rathod S, Samal S. Successful pregnancy outcome in a case of Eisenmenger syndrome: A rare case report. J Clin Diagn Res 2014;8:OD08-9.
4. Cartago RS, Alan PA, Benedicto J. Pregnancy outcomes in patients with severe pulmonary hypertension and Eisenmenger syndrome treated with sildenafil monotherapy. Chest 2012;142:999.
5. Wilson W, Taubert KA, Gewitz M, Lockhart PB, Baddour LM, Levison M, *et al*. Prevention of infective endocarditis: Guidelines from the American Heart Association: A guideline from the American Heart Association rheumatic fever, endocarditis, and Kawasaki disease committee, council on cardiovascular disease in the young, and the council on clinical cardiology, council on cardiovascular surgery and Anesthesia, and the quality of care and outcomes research interdisciplinary working group. Circulation 2007;116:1736-54.
6. Abboud TK, Raya J, Noueihed R, Daniel J. Intrathecal morphine for relief of labor pain in a parturient with severe pulmonary hypertension. Anesthesiology 1983;59:477-9.
7. Kahn ML. Eisenmenger's syndrome in pregnancy. N Engl J Med 1993;329:887.
8. Kandasamy R, Koh KF, Tham SL, Reddy S. Anaesthesia for caesarean section in a patient with Eisenmenger's syndrome. Singapore Med J 2000;41:356-8.

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