Case Report

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Safe management of cesarean section in a patient of Eisenmenger syndrome

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ABSTRACT

We report our experience of a 29-year-old female with a complete atrio-ventricular septal defect leading to a single ventricle physiology and Eisenmenger syndrome. The patient successfully underwent spinal anesthesia for cesarean section in the 31st week of pregnancy. A multidisciplinary approach involving cardiologist, cardiac surgeon, obstetrician, and anesthesiologist was utilized to achieve a safe pregnancy and cesarean for the delivery of the baby. A close clinical assessment is required, especially during the third trimester when the risk of acute right ventricular dysfunction increases. The use of extracorporeal membrane oxygenation (ECMO) (as a bridge to recovery or bridge to salvage) was planned to support oxygenation and circulation in case of acute biventricular dysfunction. The delivery/cesarean section was performed in a cardiac surgery operating room, and to reduce the time-frame for ECMO institution the femoral vessels were exposed surgically before the cesarean section.

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INTRODUCTION

Despite the great interest of physicians in the management of congenital heart disease (CHD), the effects of pulmonary arterial hypertension (PH) during pregnancy are incompletely known.[1] The Eisenmenger syndrome consists of PH with a reversed or bidirectional shunt at the atrio-ventricular or aorto-pulmonary level[2] and the pathophysiological effects usually worsens during pregnancy and increases the risk of a poor maternal outcome.

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CASE REPORT

A 29-year-old female with a complete atrio-ventricular septal defect with functionally univentricular heart, presented with the complaints of effort dyspnea and cyanosis (SpO_2 about 70%); she also had history of recurrent episodes of bronchitis. The ECG revealed sinus bradycardia with a heart rate of about 50 beats per minute (bpm), requiring interruption of beta blocker therapy. The patient was discharged and recommended oxygen therapy through facemask at 4 l/min during night and in case of dyspnea. Cardiac catheterization study revealed an increase of pulmonary pressure up to irreversible PH and Eisenmenger syndrome, as confirmed by right heart catheterization (pulmonary artery systolic pressure 75 mmHg; and no reversibility to sodium nitroprusside infusion).

Although she was advised to avoid pregnancy an unplanned pregnancy occurred, and she refused medical termination of pregnancy. In the second trimester, she was admitted to the hospital for detailed clinical evaluation, revealing a right rotation of the heart with a 3/6 systolic murmur and an enlarged liver 2 cm below the costal margin. The SpO_2 was about 70% with significant cyanosis. Transthoracic echocardiography showed a complete atrio-ventricular septal defect resulting in functionally univentricular heart and moderate atrio-ventricular valve regurgitation. The left ventricle was mildly dilated with a

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preserved ejection fraction (60%); the right ventricle was hypertrophic and slightly dilated with an almost normal function—Tricuspid Annular Plane Systolic Excursion (TAPSE) 16 mm. The aortic valve was normal and pulmonary valve had trivial regurgitation while the pulmonary artery was dilated. Pulmonary artery systolic pressure (PASP) was severely increased to 80 mmHg and the inferior caval vein was mildly dilated without diameter reduction during inspiration. A stress test revealed increasing number of ventricular extra-systoles with the decrease of oxygen saturation to 55-60%.

In the 31st week of pregnancy, she was admitted to the hospital with complaints of hemoptysis followed by sudden respiratory arrest which resolved spontaneously. The patient was admitted to the intensive care unit (ICU); the arterial pressure, heart rate, and peripheral arterial saturation at admission were 150/90 mmHg, 76 bpm, SpO_2 68%, respectively. Nitric oxide and oxygen (the device to control nitric oxide was connected to the inlet of the face mask, administering 10 parts per million) was administered through a face mask with which the SpO_2 increased to 80%. Mechanical ventilation and other drugs were deliberately avoided. Blood investigations showed red blood cells count 6.36×10^6/ul, Hb 15.1 g/dl, hematocrit 49%, NTpro-BNP 670 pg/ml, fibrinogen 449 mg/dl, INR 1, and AT III 65%. Blood gases were pH 7.42, PaO_2 32 mmHg, $PaCO_2$ 58 mmHg, and HCO_3^- 31.2 mmol/l. The fetal cardiotocography revealed a normal heart rate; the ultrasound showed a normal structure of the fetus; however, the fetus weighed only about 1.6 kg and a breech presentation was detected. Dexamethasone 12 mg twice daily was administered to the patient for enhancing lung parenchymal maturity.

Because of low weight and Eisenmenger syndrome, an early cesarean section was decided, and a more aggressive approach of biventricular assistance was planned to support the hemodynamics if required. Thus, we decided to perform the cesarean section in the cardiac surgery operating room where facility for extracorporeal membrane oxygenation (ECMO) is readily available. Informed consent for cesarean section, possible ECMO institution, and surgical sterilization was obtained. Antibiotic prophylaxis with amoxicillin and gentamicin was administered to prevent infective complications. Preoperative thrombocytopenia (about 55×10^9 per liter) was treated by platelet infusion (1 pack, 300 ml) the day before delivery in order to obtain a platelet count higher than 75×10^9 per liter required for a safe spinal anesthesia.

In the operating room, a peripheral and a central venous catheter were placed for rapid fluid or drugs administration, if required; simultaneously, monitoring started with ECG, SpO2, central venous pressure, and invasive arterial pressure (radial artery). A 27-gauge spinal needle was inserted at L2-L3 intervertebral space while patient sitting. 11 mg of levo-bupivacaine and 10 μg of fentanyl were injected and the patient was made supine and slightly rotated to the left side in order to avoid caval vein compression. Additional, 20 mg ketamine and neo-synephrine (0.1 μg/kg) were administered intravenously to prevent vasodilation and hypotension. Modest preload augmentation with HES 500 ml (Hydroxyethyl starch) was performed after spinal anesthesia. After spinal anesthesia, the right femoral vessels were exposed and prepared with two polypropylene 5/0 purse strings, allowing for a safe cannulation of both artery and vein in case of hemodynamic decompensation. Echocardiography was not used intraoperatively for several reasons: trans-thoracic echocardiography was avoided to prevent the contamination of the surgical field whereas trans-esophageal echocardiography (TEE) was not considered because of the possibility of worsening of PH secondary to the TEE probe insertion in a high-risk awake patient. The information about the adequacy of the circulating blood volume was obtained by monitoring the central venous pressure.

The cesarean section was carried out without complication and surgical sterilization was obtained by removing both the fallopian tubes. No episode of hemodynamic instability occurred and the ECMO institution was not required. The infant (weight 1640 g, length 40 cm, cranial circumference 29 cm) showed Apgar score 4 and 7 at 1 and 5 minutes after delivery, and only oxygen was administered to the baby. The Doppler flow study of the maternal venous system of the legs was performed before discharge which did not show deep vein thrombosis or any other abnormality. The maternal postoperative course was uneventful, except for transfusion of two packs of platelets and one pack of plasma. At 16-month follow-up, the patient is alive and in stable clinical condition and the infant has attained a normal growth.

DISCUSSION

The common cause of PH in young women of childbearing age is related to a large intracardiac or aorto-pulmonary shunt due to CHD. The risk of maternal death in case of pregnancy for un-operated CHD with severe PH (Eisenmenger syndrome) ranges from 30% to 50%[2] and

it is closely related to the progressive increase in plasma volume, which reaches a peak value of more than 50% during the third trimester and burdens a compromised right ventricle. Normally, vasodilation in the pulmonary as well as systemic vascular bed allows adaptation to the volume overload of pregnancy. However, in presence of Eisenmenger syndrome, the irreversibly increased pulmonary vascular resistance does not allow for volume adaptation; moreover, the volume expansion in the presence of severe PH and decreased systemic vascular resistance increases the right to left shunt that worsens the hypoxia, which sets in a vicious cycle of further increases in the PH, RV strain and right to left shunt. The strained right ventricle is further strained due to decreased O_2 supply on account of worsening hypoxemia and decreased systemic arterial pressure. Arguably, the volume overload of pregnancy imposes a serious risk of right ventricular failure. At the time of labor and delivery severe hemodynamic compromise may occur due to labor-pain-associated increases in pulmonary vascular resistance and mobilization of blood volume during uterine contractions. The right ventricular strain can further increase due to acidosis commonly observed during labor.

Undoubtedly, the anesthetic goal in patients with Eisenmenger syndrome should be a stable cardiac output without worsening of the intra-cardiac shunt. However, both central neuraxial block and general anesthesia can potentially increase the right to left shunt: central neuraxial block by reducing the systemic vascular resistance whereas general anaesthesia by increasing the pulmonary vascular resistance due to catecholamine release particularly, during laryngoscopy, intubation, and surgical stimulation. Moreover, the techniques to prevent sympathetic stimulation invariably decrease systemic vascular resistance that too will increase the left to right shunt. Regional anesthesia has been reported as a safe method in patients with Eisenmenger syndrome.[3] We modified this approach and used a single shot of spinal anesthesia with levo-bupivacaine in place of bupivacaine and combined it with intravenous neo-synephrine, in order to achieve a more hemodynamic stability.

The use of ketamine in the presence of spinal anesthesia is not a common approach at our institution; however, we decided for this combination to increase the analgesia obtained by levo-bupivacaine[4] and for preventing hypotension. Peripheral vasoconstriction is a well-known effect of the ketamine. We used a low dose of ketamine and neo-synephrine to prevent vasodilation induced by a sympathetic blockade due to spinal anesthesia.

We considered epidural block for this patient as it provides a better control of hemodynamics than the spinal anesthesia. However, in the case of its failure, general anesthesia would be mandatory which might result in serious worsening of the hemodynamic status of the patient. The spinal anesthesia with ketamine and neo-synephrine led to a safe anesthesia without impairment of the hemodynamics.

CONCLUSION

In conclusion, a multidisciplinary approach (involving cardiologist, cardiac surgeon, obstetrician, and an anesthesiologist) achieved a safe pregnancy/delivery in a patient with Eisenmenger syndrome. A close clinical assessment is necessary especially during the third trimester when the risk of acute right ventricular dysfunction increases. The use of ECMO (as a bridge to recovery or bridge to salvage) was planned as a fast solution in the case of acute biventricular dysfunction. The cesarean section/delivery was performed in the cardiac surgery operating room because of the ready availability of ECMO and for reducing the time-frame for ECMO institution, the femoral vessels were surgically exposed before starting cesarean.

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