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CASE REPORT

Anesthesia Management of a Patient with Sickle Hemoglobinopathy and Mitral Stenosis for Emergency Lower (Uterine) Segment Cesarean Section

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ABSTRACT

The pregnant patient with sickle hemoglobinopathy (SCD) and mitral stenosis (MS) presenting for emergency lower (uterine) segment cesarean section (LSCS) represents a challenge to the anesthesiologist. In such a case, the choice of anesthesia is dependent on the patient's clinical condition, urgency of surgery, and the reports of laboratory investigations. An understanding of physiological changes in pregnancy and the pathological impact of MS and SCD on pregnancy will help the administration of safe anesthesia for mother and baby. We have discussed the management of a pregnant patient with SCD and MS for emergency LSCS.

Keywords: Emergency lower (uterine) segment cesarean section, Mitral stenosis, Sickle hemoglobinopathy.

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INTRODUCTION

The pregnant patient with sickle hemoglobinopathy (SCD) presenting for emergency lower (uterine) segment cesarean section (LSCS) represents a challenge to the anesthesiologist. Most pregnant patients experience sickling complications during the peripartum period. Associated with comorbidities like rheumatic heart disease (RHD), mitral stenosis (MS) in these patients can become a nightmare for the anesthesiologist.

These patients are prone to hypoxia, hypotension, hypothermia, dehydration, acidosis, and congestive cardiac failure during the peripartum period, which can further lead to sickling complications.[1]

In such a case, the choice of anesthesia is dependent on the patient's clinical condition, type and indication of surgery, and laboratory investigations. We have discussed management of a pregnant patient with SCD and MS for emergency LSCS.

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CASE REPORT

A 29-year-old pregnant female, weighing 52 kg, 34 weeks of gestation, G6P1L0A4D1, and known case of sickle cell disease (SCD) and RHD with MS and tricuspid regurgitation (TR), was admitted for safe confinement due to severe anemia and precious baby. She had a bad obstetric history of four spontaneous abortions and fifth was intrauterine fetal death at 28 weeks of gestation. During the last pregnancy, she had dyspnea on exertion (DOE) grade II with palpitations and was diagnosed as RHD, MS with TR. Penidure prophylaxis was started. She had also been evaluated for persistent severe anemia; electrophoresis showed SCD. She had suffered from sickling crisis in her last pregnancy.

She presented to our hospital for 6th pregnancy with DOE grade II, palpitations, malaise, and pallor at 28 weeks of gestation. Routine investigations showed hemoglobin (Hb) as 7.7 gm/dL, with HbF as 9.7%, and HbS as 84.5%. Renal function test, liver function test, and coagulation profile were normal. The electrocardiogram showed sinus tachycardia, two-dimensional echocardiogram revealed a mitral valve area of 2.2 cm^2, mild MS and TR, no pulmonary hypertension (PH), and with an ejection fraction of 60%.

The arterial blood gas (ABG) was pH –7.409, pCO_2^- 30.6, pO_2^- 61.7, bone extracellular fluid –5.8, HCO_3^- 20.7, and SO_2^- 99.2%.

On examination, patient's general condition was moderate and poorly nourished. Pulse rate (PR) was 112/minute, blood pressure (BP) 90/60 mm Hg, pallor +, airway normal with buck teeth, respiratory system normal, and diastolic murmur of MS was present.

Patient was admitted for safe confinement. Blood transfusion was started for low Hb, but she went into labor and was taken up for emergency LSCS due to fetal distress. High-risk consent was taken for comorbid conditions and inadequate starvation. Inside the operating theater, monitors were attached, PR was 120/minute, BP 90/60 mm Hg, and SpO_2 was 94%. Due to scaly lesions

all over the body, venous access was difficult; one peripheral intravenous (IV) 22G and right external jugular vein was cannulated. Warm IV fluid and Ringer lactate (RL) solution was started 4 mL/kg/hour, to maintain central venous pressure 6 to 8 cm H_2O.

Defibrillator was kept on standby. Preoxygenation was started immediately with 3 number mask at the rate of 5 L/minute. For premedication, IV ranitidine, ondansetron, and hydrocortisone were given. Rapid sequence induction was done with cricoid pressure, using injection thiopentone 200 mg slowly in titrated manner and injection succinylcholine 75 mg. Patient was intubated with number 7 polyvinyl chloride-cuffed endotracheal tube. Injection atracurium 20 mg was given as long-acting muscle relaxant. Within 5 minutes, baby was delivered, cried immediately after birth, birth weight was 2.4 kg, and APGAR score was 9. After delivery, injection midazolam 0.5 + 0.5 mg and injection fentanyl 50 + 50 µg were given in titrated doses. Pitocin was started as slow-rate infusion and monitoring done for BP, PR, and saturation. Fentanyl patch was applied to forearm for 24 hours analgesia. Injection sodabicarb 40 cc was given prophylactically to prevent acidosis. Intraoperative hemodynamics was maintained to avoid hypotension and tachycardia. Intraoperative ABG showed pH at 7.409, pCO_2^- 30.6, pO_2^- 91.1, HCO_3^- 20.7, and saturation at 99.1%.

Blood loss was approximately 750 mL and urine output was 70 mL. Intraoperative IV fluid 1 pint of RL solution and 1 unit of blood transfusion was given. The patient was extubated after adequate reversal of neuromuscular blockade and return of adequate tone, power, and spontaneous respiration.

Postextubation, the patient was shifted to recovery ward for further monitoring, but she developed bilateral basal crepitations. These subsided after injection Furosemide 10 mg was given. Postoperative hemodynamics was well maintained. Oxygen was supplemented with Hudson mask 6 L/minute. Warm IV fluid RL 2 mL/kg/hour was given slowly with monitoring of pulse, BP, saturation, and urine output. Normotension and normothermia were maintained.

DISCUSSION

Sickle cell disease is hereditary hemolytic anemia, characterized by formation of abnormal HbS on deoxygenation and leading to sickling of red blood cells. Conditions, such as hypoxemia, acidosis, hypothermia, anemia, hypertonicity, and increased 2,3 diphosphoglyceric acid levels result in sickling and then vaso-occlusion of micro- and macrocirculation. Thrombosis may occur in spleen, brain, lungs, liver, and heart.[1]

While the general care of patients with SCD has improved since the last two decades, pregnant patients with SCD still present an ongoing challenge to the anesthetist. Sickle-related events (painful crisis, acute chest syndrome, and stroke) can occur in 50% women with SCD in the postoperative period. Among these, the incidence of postnatal sickling and vaso-occlusion crisis is 7.7 to 31%. So, when such pregnant patients with SCD present for emergency LSCS in view of fetal distress, we have to evaluate whether she is in labor or having a sickling crisis. Detailed history, physical examination, and lab investigations will help in decision making, but the impact of anesthetic technique on occurrence of sickling complications has been a source of controversy. But, whatever the choice of the anesthetic technique, it is important to avoid hypothermia, hypoxemia, hypovolemia, acidosis, hypotension, vasoconstriction, and stasis.

Camous et al[2] experienced sickling complications in 25% patients after delivery in cases of general anesthesia (GA). The GA was a risk factor even when the severity of illness was taken into consideration.

The choice of anesthesia for the obstetric patient with SCD depends on general condition and preference of the anesthetist.

Regional anesthesia could be beneficial for SCD by inducing vasodilatation and enhanced blood flow in anesthetic area as well as optimum pain control. Epidural analgesia is preferred to treat vaso-occlusive crisis; however, in our case, we opted for GA because of fetal bradycardia.

Goodwin[3] found that nonsickle cell-related complications, such as fever and infection were higher in the regional anesthesia group, mainly in obstetric patients receiving epidural anesthesia.

Some authors suggest that compensatory vasoconstriction in nonblocked area, lack of control of ventilation, and potential for stasis during regional anesthesia create an environment in which sickling occurs. But, in one series, epidural anesthesia was successfully used to treat vaso-occlusive crisis.[3]

Goals of anesthesia management include avoidance of circulatory stasis, maintaining arterial oxygenation, and avoiding acidosis due to hypoventilation as these events trigger sickling in patients with SCD. Also, maintenance of normal body temperature is desirable to minimize vasoconstriction and associated circulatory stasis.[4] Preoperative need for exchange transfusion depends on the general condition of the patient and the type of surgical procedure. Exchange transfusion is generally recommended before major surgical interventions in order to minimize sickling and reduce the circulating HbS concentration below 30%.[4] Prophylactic transfusion significantly reduced the incidence of painful crises of SCD ($p < 0.01$) and substantially reduced the cumulative incidence of other complications of this disorder.[5] But, posttransfusion, our patient went into labor and developed fetal distress.

Rheumatic MS complicating pregnancy is still a frequent cause of maternal mortality. It is a fixed cardiac output state. Women with severe MS often do not tolerate the cardiovascular demands of pregnancy. Increased volume load and tachycardia limit the time available for left ventricular filling, resulting in enlargement of left atrium and pulmonary congestion due to back pressure changes. Enlarged left atrium, causing atrial fibrillation, worsens the scenario.[6]

The goals of anesthetic management of patients with MS are[6]

- Avoiding tachycardia and immediate treatment of atrial fibrillation;
- Maintenance of venous return and adequate systemic vascular resistance;
- Avoidance of aortocaval compression;
- Prevention of pain, hypoxemia, hypercarbia, and acidosis.

The risk of maternal death due to cardiac decompensation is greatest during labor and immediate postpartum period due to uterine autotransfusion, oxytocic drugs, and labor pain.[7]

There are no controlled studies examining the best type of anesthetic technique in these patients, and standard guidelines are lacking. Therefore, individualizing the anesthetic management according to parturient's condition and experience of existing treatment options is the key to success in these patients.[8]

Regional anesthesia has proved to be a safe technique for cesarean section in cardiac patient with mild-to-moderate MS. Epidural anesthesia causes segmental blockade, gradual onset, and less hemodynamic variations. However, neuraxial blockade in anticoagulated patients has a high risk of epidural hematoma.[9]

General anesthesia has the disadvantages of increased pulmonary artery pressure, tachycardia during laryngoscopy and intubation, difficult airway, and pulmonary aspiration. Modified rapid sequence induction using etomidate, remifentanyl, and scoline is an ideal choice in severe MS with PH and in case of fetal distress. High-dose opioid induction may cause fur ther respiratory depression in neonates. Due to nonavailability of above drugs, we used drugs like thiopentone, scoline, midazolam, and fentanyl, but in a titrated manner.[8]

In our case scenario, we had to deal with SCD, MS with PH, and fetal distress. We administered general anesthesia with rapid sequence induction maintaining cricoid pressure.

During perioperative management, we planned meticulously for fluid infusion, stress response alleviation, hemodynamic stability, temperature monitoring, maintenance, and pain management.

CONCLUSION

A team approach is mandatory for the management of such a type of patient with RHD and SCD for emergency LSCS. The choice of anesthesia is also dictated by the patient's clinical condition, investigations, and urgency of procedure. Finally, the understanding of physiological changes in pregnancy and pathological impact of MS and SCD on pregnancy will help us to administer safe anesthesia for mother and baby. It will reduce morbidity and mortality.

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