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ANAESTHETIC MANAGEMENT OF A 26 YEARS OLD PREGNANT WOMEN WITH A KNOWN CASE OF SICKLE CELL DISEASE PLANNED FOR EMERGENCY CESAREAN SECTION

Anaesthesiology

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ABSTRACT

A 26 years old pregnant women who is a known case sickle cell disease (ss) presented with respiratory distress and severe anemia planned for emergency cesarean section . After preoperative optimisation with 4 units of blood transfusion and oxygenation via facemask operative intervention was carried out under combine spinal epidural anaesthesia . A sensory block level of T6 is achieved with intrathecal injection 10 mg of hyperbaric bupivacaine with 25 microgram of injection fentanyl .Intraoperatively 1 units of blood transfusion was done under regular ABG monitoring to prevent acid -base inbalance, hypoxia, acidosis , hypothermia and hypercarbia . Postoperative analgesia was provided through the epidural catheter with 6 ml of (inj 0.125% of bupivacaine + 25 microgram of inj fentanyl) every 12 hourly for 3 days. She was discharged from the postnatal ward on 5 th day following surgery with a healthy male baby . Pregnancy associated with sickle cell disease carries a high risk of maternal and neonatal mortality and morbidity. It provide a great challenge to anaesthesiologist during perioperative management. The uneventful course of anaesthesia in this case is managed with systemic evaluation and careful anaesthetic strategy.

KEYWORDS

INTRODUCTION

Sickle cell disease is an autosomal recessive disease with wide spectrum of severity and manifestation. SCD arises as a result of single gene mutation (GAG-GTG and CTC-CAC) in chromosome 11, causing substitution of the nonpolar amino acid valine in place of the negatively charged glutamate in the sixth position of the β-chain of the haemoglobin molecule. Due to this, the normal adult 'haemoglobin A' is replaced with 'haemoglobin S' {HbS (α2/β2)}.[1] Erythrocytes in SCD undergoes rapid but reversible alternation in shape on deoxygenation (PaO2 <50 mmHg) and intracellular polymerization of abnormal HbS molecule which deforms the cell by stretching the normal flexible biconcave shape into elongated rigid form and thus imparting a 'sickle' shape to the cell.[2] However, the polymers will disaggregate with oxygenation and restore normal shape. But repeated sickling cycles and extensive polymerization of HbS will produce erythrocyte metabolic abnormalities and membrane damages leading to irreversible sickling regardless of oxygen concentration and intravascular haemolysis of red cells. The sickle cells are rapidly cleared from the circulation by the reticuloendothelial system. Therefore, the life span of sickle erythrocytes is reduced to approximately 12 days. As a result of damaged red cell membrane, there is increased adherence to peripheral blood mononuclear cells, platelets and the vascular endothelium via P-selectin ligands present on the endothelial surface leading to vaso-occlusion, ischaemic-perfusion injury and end-organ damage. Lysis of erythrocytes will release of free haeme and depletion of nitric oxide and arginine will worsen the vascular endothelial injury leading to vasculopathy and its complications such as stroke, pulmonary hypertension, vaso-occlusion, acute painful crisis and end-organ damage.[3] Pregnancy with SCD has always posed as a challenge to anaesthesiologists as the physiological changes that occur during pregnancy can over burden various organs that have been already affected due to SCD which will subsequently increase in maternal and foetal complications. Although many developments have come up in the management of SCD ,pregnancy complicated by this disease remains associated with adverse maternal and perinatal outcomes.[1] This case report highlights and reviews an anaesthesiologist perspective on the management of labour pain, caesarean delivery, and postoperative analgesia.

CASE REPORT

A 26 year old pregnant women (GRAVIDA 3 , PARA0+2) presented to our hospital with respiratory distress and severe anemia . She was a known case of sickle cell disease for 11 years and had a history of cholecystectomy 5 years back under general anaesthesia. She had registered for antenatal care at 10 weeks of gestation with a history of multiple blood transfusion for her persistent low haemoglobin count . She had a history of two spontaneous abortion in the past for which multiple blood transfusion were done due to low haemoglobin count . Her present pregnancy is diagnosed as gravida 3 para (0+2) at 36 week of gestation with severe anemia with breathing difficulty with sickle cell disease .

On admission her HB- 5.5 gm%, HBS-75.0%,HBF -16.1% with anisocytosis , poikilocytosis , few sickle cell , polychromatic red cells and fragmented red cells on peripheral blood smear .USG obstetric shows a live intrauterine pregnancy of 36 week with fetal heart rate 140 BPM and active limb movement with an estimated body weight of 2.9 kgs in cephalic presentation . Her biochemical investigations are SERUM Na – 134mmol /l , K – 4.8 mmol/l ,TLC-24600 ,PLT COUNT -1.2 L, PROTEIN -5.44GM/DL, ALBUMIN -2.91GM/DL, GLOBULIN -2.53GM/DL , TOTAL BILIRUBIN -3.36MG/DL,AST -75U/L, ALT -148U/L, ALP -138U/L , PT – 16.2 , INR -1.6 . On general examination BP – 105/78 mmhg , PR -124 bpm , chest – bilateral air entry with basal crepitation ,CVS –s1s2 Heard ,GCS– E4V5M6,with dehydration , pallor, and icterus .Her hemodynamic status was stabilised with 3 units of blood transfusion through out the day of admission.

On the following day the patient went into spontaneous labour and was planned for emergency caesarean section under ASA grade 3 after a complete preanesthetic examination and informed consent. The monitors were all connected including electrocardiogram , non invasive blood pressure,pulse oximeter and an arterial line for ABG monitoring .The patient's coagulation profile was normal so ,a combine spinal epidural anaesthesia was planned . Two peripheral lines are secured with 18 G cannula in both the arms . Her preoperative vitals are - BP – 118/74 mmhg , PR – 99 BPM , SPO2 – 95 % in room air . Premedication done with inj ranitidine 50mg, inj ondansetron 4 mg, inj metaclopromide 10 mg . Under all asepsis and antiseptic measures ,epidural anaesthesia is given via 18G Tuohy's epidural needle in L2-L3 space in sitting posture. At 4.5 cm, the epidural space was reached, and the catheter was secured at 10 cm followed by which spinal anaesthesia is given via L3-L4 spine using 25 G quincke spinal needle.10 mg of hyperbaric bupivacaine with 25 microgram of injection fentanyl was given intrathecally after confirmation of free

and clear csf flow . Patient was then positioned in 15 degree left lateral tilt and sensory level of spinal anaesthesia is achieved uptoT6 level of dermatome.Oxygen was administered via facemask @5l /min . Intraoperative hypotension was managed with crystalloid and vasopressor .Care was taken to avoid hypoxia , hypercarbia ,acidosis and hypothermia .Continuous ABG analysis was done to avoid any acid base inbalance and to prevent hypothermia warm intravenous fluid and warmer were applied . A healthy male baby was born with good APGAR score following which 10 units of inj oxytocin was given in 500 ml DNS and 5 IU IM in buttock . As intraoperative blood loss was around 600 ml,she was given 1 units of whole blood intraoperatively .Her intraoperative period was uneventful with controlled vitals and she was maintaining saturation of 99-100% via facemask.

She was shifted to postoperative anaesthesia care unit after surgery and her vitals were BP -122/88 , PR -89 BPM , SPO2 – 97 % in room air , chest was clear bilaterally . Oxygenation was done via nasal prong @ 3 L /MIN in PACU. Post operative analgesia was given with epidural top with INJ (0.125 % bupivacaine +25 microgram of fentanyl) 6 ml every 12 hourly for 3 days. Post operatively her ABG was normal , haemoglobin of 9.8 gm% and normal coagulation profile .She was shifted to ward after 24 hrs .She was discharged from postnatal ward on 5 th day following surgery with a healthy male baby .

DISCUSSION

In this case we can clearly understand the importance of preoperative assessment and meticulous anaesthetic planning before caesarean section to prevent the dreaded complications of sickle cell anaemia in the perioperative as well as postoperative period. Hypothermia throughout the procedure must be avoided as it will lead to shivering and peripheral stasis which in turn will lead to hypoxia and increased sickling. Epidural analgesia with regular top-up has been given to prevent vaso occlusive crisis. Vaso occlusion leads to recurrent painful episodes and a variety of serious complications which leads to serious organ system complications and death. Hypotension and hypoperfusion from epidural analgesia was prevented with pre-loading of IV fluids and patient maintained good oxygenation and normocarbia throughout the procedure.[4] Acute chest syndrome is one of the most feared complication of SCD which is difficult to differentiate from respiratory distress caused by bacterial infection. Therefore it is adviced to start broad spectrum antibiotics to patient in perioperative period. Incentive spirometry was started to prevent pulmonary complications. Efficient anaesthetic management in form of avoiding acidosis, hypoxia, hypothermia, hypovolemia, good postoperative pain relief with epidural top-ups, postoperative oxygen therapy, postoperative thromboprophylaxis, chest physiotherapy, nebulization, incentive spirometry with early ambulation and regular ABG monitoring proved to drastically improve patient outcome.[4]

CONCLUSION

Our case report demonstrates that cesarean section can be safely performed in pregnant women with sickle cell disease under regional anaesthesia with proper planning and optimal perioperative preparation.

Conflict Of Interest:-There is no conflict of interest.

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