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PERIOPERATIVE ANAESTHESIA CARE IN A CHILD WITH HEREDITARY SPHEROCYTOSIS FOR LAPAROSCOPIC SPLENECTOMY

Dr. Deepak C. Koli [1], Dr. Tapas Mandal [2] and Dr. Hemant H. Mehta[3]

[1]Consultant Anaesthesiologist, Department of Anaesthesia and Pain Management, Sir H. N. Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India
[2]Consultant Anaesthesiologist, Department of Anaesthesia and Pain Management, Sir H. N. Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India
[3]Director and Head of the department, Department of Anaesthesia and Pain Management, Sir H. N. Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India

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ABSTRACT

Hereditary spherocytosis (HS) is a familial haemolyticdisorder characterizedby formation of abnormal red blood cells with fragile cell walls causing anaemia, jaundice, splenomegaly and gallstone formation. Most children have a mild disease and hencedo not require splenectomy, mainly reserved for a severe disease or in patients with symptomatic gall stone disease. The anaesthetic management of hereditary spherocytosis mainly depends upon the degree of haemolysis and severity of anaemia.We reportanaesthetic management in a case of HS with splenomegaly for elective laparoscopic SOS open splenectomy.

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INTRODUCTION

Hereditary spherocytosis is characterised by development of abnormal shaped red blood cells (RBCs) having fragile cell walls and transmitted as an autosomal dominant disorder with almost 25% of patients having no previous family history and predominantly constitute a new mutation [1.] Basic pathophysiology is due to protein defects in membrane of erythrocytes, causing cytoskeleton instability with changes in the erythrocyte shape in hereditary spherocytosis. Loss of erythrocyte surface area is mainly due to spectrin deficiency resulting in formation of spherical RBCs which are destroyed in spleen resulting in splenomegaly. The main abnormalities identified in red cell membrane are as follows spectrin deficiency alone or combined spectrin and ankyrin deficiency.

Corresponding author:** ***Dr. Deepak C. Koli,
Consultant Anaesthesiologist, Department of Anaesthesia and Pain Management, Sir H. N. Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India.

The other abnormalities being band 3 deficiency and protein 4.2 defect of which most common defect is spectrin deficiency [2]. The spherical shape of RBCs makes them susceptible to osmotic lysis which in turn results in anaemia, jaundice, cholelithiasis and splenomegaly along with altered liver function affecting metabolism of anaesthetic drugs in these patients [3]. The peri-operative management of hereditary spherocytosis is mainly divided into pre-splenectomy, splenectomy and post splenectomy care. The common perioperative management goals focus at avoiding hypoxia, hypothermia, acidosis, aggressive hydration and pre-emptive erythrocyte transfusion [(4, 5).]

CASE REPORT

A 9years old boywith hereditary spherocytosis, posted for laparoscopic sos open splenectomy. Pt. hadsevere pallor at the age of 20 days of life. On investigatingchild was diagnosed with hereditary spherocytosis as haemoglobin (Hb) was 5.2 gm%, reticulocyte count was 9.6%with increased osmotic

fragility. Since then, child has received 8-10 blood transfusions so far, once in every two to three years. Last transfusion received post laparoscopic cholecystectomydone 7 months back. Child had also received pneumococcal and meningococcal vaccination 3 months prior to surgery. On general examination, child was 125 cms tall and weighed 23.5kg. The child hadan evidenthaemolytic facies, he was afebrile with a pulse rate of 102beats/min, regularand a blood pressure of 100/60mmhg taken in right upper limb, respiratory rate 20 cycles/min, pallor ++, icterus + without any obvious lymphadenopathy.On systemic examination scars of previous surgery were visible, liver palpable 3 cm below costal margin, Spleen 5 cm below costal margin soft to firm,rest of the systemic examination findings were within normal limits. On airway examination mouth opening was adequate with mallampatti grade 1, however patient had a receding mandible hence difficult airway was anticipated.

Laboratory investigation showed HB 8.5 gm/dl, total leucocyte count 11600 and platelet count 2.26lacs, peripheral smear showed anisopoikilocytosis along with microcytes, elliptocytes and microspherocytes. Totalbilirubin 5.24mg/dl, direct bilirubin 0.53 mg/dl and indirect bilirubin 4.71mg/dl, pt. had an AB positive blood group with negative direct and indirect Coombs test. Ultrasonography showed Liver enlargement with normal parenchymal echogenicity. Liver span being14.2 cm. Spleen was enlarged in size and normal in echogenicity, span being 13 cm. Chest X ray and electrocardiogram done and werewithin normal limits.Considering a low Hb, patient was transfused with 1 packed cell a day prior to surgery and 2 packed cell were reserved for surgery. The patient was kept nil by mouth from midnight and maintenance fluidwas started in the ward.Our anaesthetic management goals were based on maintaining optimum hydration perioperatively, strict avoidance ofhypothermia and hypovolemia and strategies to prevent crisis perioperatively, transfuse blood or blood products post splenic vessel clamping or post splenectomy and postoperative pain management by IV PCA fentanyl and port site infiltration with 0.25% bupivacaine.

The child was premedicated with IV glycopyrollate 0.1mg, IV midazolam 0.5mg and IV ketamine 20mg in preoperative room with vital monitoring. Once child was wheeled in the operative room standard routine ASA monitors wereattached.Core temperature and end tidal CO2 monitoring was also done throughout intraoperative period post induction. Considering anticipated difficult intubation, all necessary difficult airway equipments were kept ready during induction. Induction of general anaesthesia was done with IV fentanyl 40mcg, IV propofol 60mg, and IV atracurium 15mg given for muscle relaxation after confirming bag and mask ventilation. On laryngoscopy with Macintosh blade 3, patient was found to have Cormac lehane grade 3 hence bougie guided endotracheal intubation was done with no. 5 cuffed endotracheal tube. Maintenance of anaesthesia was done with O2:N2O 50%:50% and sevoflurane to MAC of 1.0 - 1.2. Post induction a large bore 20G IV was taken and patient positioning was done with all due care. Injection clonidine 25 mcg and injection fentanyl 20mcg given just before incision, laterintermittent boluses were given intraoperatively as per requirement.IV fluids were given judiciously throughout the intraoperative period. Vitals were stable throughout the surgery. However, considering a large sizedspleen, left sided subcostal incision was taken for splenectomy. Ultrasonography guided left sided paravertebral block with a total of 12ml of 0.2ml ropivacaine along with 2mg dexamethasone and 30mcg buprenorphine was administered from T6 TO T9 level. Intraoperative blood loss was around 70 to 80ml and total urine output was100 ml. The intra-operative temperature was maintained between 37°C-35.5°C using warm IV fluids and mattress warmer. After surgery patient made supine and then reversed with injection glycopyrolate 0.2mg and injection neostigmine 1.2mg once fully awake, with good muscle tone and regular breathing attempts. Post extubation child was shifted to post anaesthesia care unit (PACU) for observation. In PACU child was comfortable and pain free with stable vitals.Rest of the postoperative course was uneventful.

DISCUSSION

Hereditary spherocytosis occurs due to an intrinsic genetic defect of RBC membrane proteins, which are major components of the cytoskeleton for maintaining shape of RBC. The resultant Qualitative or quantitative abnormalities in these proteins results in spheroidal shaped red blood cells, these abnormal shaped cells are then targeted in spleen during their passage into sinusoids due to spherical shape and gets phagocytosed resulting in extravascular haemolysis.(6) In chronic cases or other concomitant illness it causes haemolytic crisis leading to anaemia, hypoxia, splenomegaly.(6) Direct / indirect bilirubin accumulates in gall bladder to form gallstones, altering liver function and causing hepatomegaly. Splenomegaly is usually mild to moderate, however the size of spleen per se is not an indication for splenectomy. Laboratory diagnosis mainly involves peripheral blood smear, MCV, MCHC & reticulocyte count. The presence of spherocytes is confirmed by an osmotic fragility test the sensitivity of which varies from 48 to 95% although this may be increased to 99% by glycerol lysis test.(7)

Patients with severe haemolysis requires medical management based on folate therapy and surgical management for such cases requires a splenectomy which would result in cessation of haemolysis and restoration of haemoglobin to normal levels along with improvement in jaundice. In order to prevent risk of gall stone formation and haemolytic crisis surgical management is indicated in all patients except in asymptomatic and well compensated patients.[8] Patients posted for splenectomy should be vaccinated with Pneumococcal and Haemophilus influenzavaccines preoperatively to prevent severe infections in postoperative period. Intraoperative anaesthetic management goals include adequate hydration in order to prevent stasis, avoid hepatotoxic drugs, hypoxia and acidosis, prevent hypothermia by using warm fluids intravenously and maintaining optimal room temperature along with use of warming blankets/ mattress warmers as hypothermia can lead to vasoconstriction and circulatory stasis. Replacing blood losses with packed cell PRBC and if required Fresh frozen plasma FFP transfusionshould be done on priority basis. Supplemental oxygen, proper antibiotics and fluid administration along with effective pain management are postoperative goals in these cases. [9, 10] Volatile agents are preferred for maintenance of general anaesthesia and isoflurane causes minimum reduction in hepatic blood flow hence it is preferred volatile agent in these cases. [9, 10] The risk of life threatening infections and sepsis due to encapsulated organisms is highest post splenectomy in post-operative period, hence these patients require a continuous monitoring with repeat complete blood count and renal function test and

chest X ray. Postoperative pain management is also crucial in suchpatients, in our case we gave ultrasonography (USG) guided paravertebral block for post-operative pain relief. USG guided paravertebral block is good alternative to thoracic epidural for thoracic and breast surgery but lately it has also been used for abdominal surgeries, especially in hepatic and renal procedures. [(11)] The recent review of various randomized controlled trials which compare thoracic paravertebral block with other anaesthetic techniques shows the evidence that it can be used in minor abdominal surgeries like herniorraphy or ventral hernia repair in which mainly somatic analgesia is needed.Thoracic paravertebral block provides satisfactory anaesthesia during the entire surgery [(12).] The main complications of paravertebral block include epidural spread of local anaesthetic drug, vascular puncture, pneumothorax and failed block but with the use of ultrasound the incidence of these complications has significantly reduced making paravertebral block convenient and safe to administer for wider anaesthesia community [(13)] To conclude, our patient had good pain relief in immediate postoperative period because of paravertebral block along with intravenous paracetamol and patient remain hemodynamically stable and pain free throughout the perioperative period.

CONCLUSION

Perioperative vigilant care with hemodynamic and temperature monitoring along with proper measures to avoid hypotension, hypoxia and acidosis along with good post-operative pain relief will definitely improve the surgical outcome and prognosis.

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