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Case Report

Sole Epidural Anaesthesia for Spleenectomy in patient of Hereditary Spherocytosis with Massive Spleenomegaly

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Abstract
Sole Epidural anaesthesia is suitable for massive Spleenomegaly as it has definite advantages over General anaesthesia. The segmental sole Epidural block denotes the use of a segmental blocked involved in field of surgery and also avoids haemodynamic changes in comorbids conditions.
Keywords: *Hereditary Spherocytosis, Segmental Epidural Anaesthesia, Spleenomegaly, General Anaesthesia.*

Introduction

Hereditary spherocytosis (HS) is an extremely rare autosomal dominant disorder. HS is a familial hemolytic disorder with marked heterogeneity of clinical features ranging from asymptomatic condition to a fulminant hemolytic anemia.[1]

It occurs due to an intrinsic defect in the red cell membrane as a result of which cells have a spherocytic shape[2]. The estimated prevalence in the Caucasian population ranges from 1:2000 to 1:5000[3]. It is characterized by a deficiency of ankyrin or spectrin (transmembrane proteins) that links the bilayer of red cells to the membrane skeleton. The spherocytes are susceptible to osmotic lysis.

In 80% instances, the inheritance of HS is autosomal dominant and in others autosomal recessive and in the remaining as sporadic mutations or recessive genes[4]. HS is diagnosed by strong family history of anemia, jaundice, splenomegaly and cholelithiasis. Altered liver function and metabolism of anesthetic agents in the liver in these patients can be very challenging. Epidural anaesthesia[5,6] is the choice because patient will be heamodynamically stable[7,8] and administration of General Anaesthesia agents can be avoided.

Aim

To avoid the effects of General Anaesthesia as patient had cardiovascular and respiratory compromise.

Case Report

A 60 yrs female presented with high grade fever, vomiting since 2 days.

Pain over left side of abdomen since 2 years insidious in onset gradual in progression, intermitent, aching type relieved with medication

H/O High grade Fever present, intermitent in onset relieved with medication

H/O Vomiting present 2 episodes, bilious, non projectile relieved with medication

Known case of asthma since 20 years and on medication

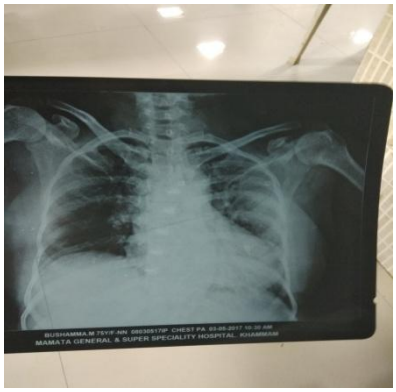
Past history of blood transfusion 3 years back with 4 units of packed RBC. No similar complaints in family members

On Examination patient was febrile, pale, icterus present, but no lymphade nopathy. On abdominal examination patient liver enlargement -½ inch below costal margin with grade VI spleen extended in the pelvis. Rest systemic examination was normal

Laboratory investigations: Revealed hemoglobin 7.8 gm%, total bilirubin: 2.3 mg/dL direct bilirubin: 0.4 mg/dL indirect bilirubin: 1.9mg/dL, enzymes - alanine transaminase: 122 IU/L, aspartate transaminase: 102 IU/L, alkaline phosphatase: 128 IU/L, Reticulocye count 3%, Malarial parasite test was negative .Coagulation profile was normal.

Peripheral blood smear: showed spherocytosis and abnormally shaped poikilocytes. Osmotic fragility of incubated blood cells was positive. Direct and indirect Coombs test were negative hemoglobin electrophoresis was normal.MCV 81fL, MCH 33.4 pg ,MCHC 49%

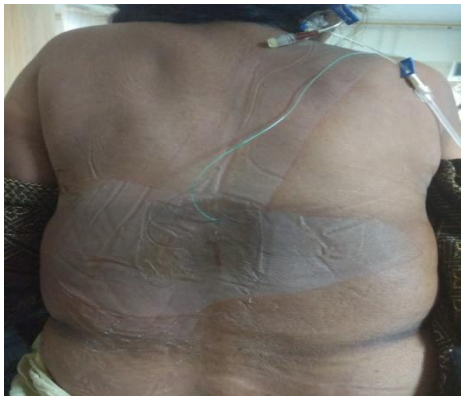
Ultrasonography: Mild hepatomegaly, massive splenomegaly was present. chest X-ray findings cardiomegaly present. PFT showed severe lung obstruction.



2D Echo findings: Mild dilated RA, RV, Severe TR, Moderate PAH, Mild MR, Mild PR, Sclerotic Aortic valve with Mild AR , Normal LV Systolic function, Grade I Diastolic Dysfunction

The patient received 1 units of packed red blood cells (PRBCs), injection Vitamin K 10 mg IM and Vaccination against Pneumococci, Haemophilus influenzae and menningococal given 14 days prior to surgery. Patient was counselled and risk consent was taken.

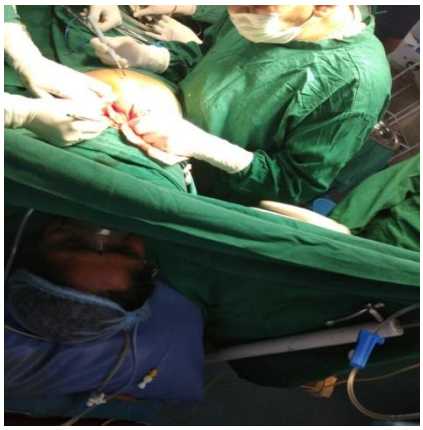
After 6 hrs of NBM, patient shifted to OT. Monitors such as 5 lead ECG, pulse oximeter and temperature probe kept. And 18 G peripheral line, 7Fr central line and arterial line was secured. Preoperative Vitals HR 96/min, BP 140 /90 mmHg and SpO_2 100%.



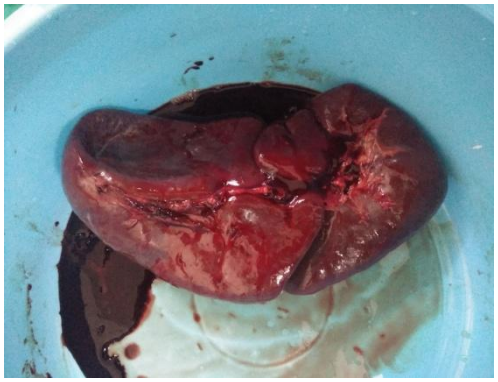
In sitting position and the back was cleaned, painted and draped. Under all aseptic precaution. With 18 G epidural needle, epidural space[9] was identified with loss of resistance technique and hanging drop method, at T10-T11 level and epidural catheter was introduced with tip of catheter at T 7 level. Test dose with 3 mL of 2% plain lignocaine was given and placement of catheter was confirmed. Inj midazolam 1mg iv, inj Fentanyl 30 mcg iv, inj ranitidine 50 mg iv was

given.O2 supplementation given through Hudson mask.

An initial dose of 5 mL 0.5% bupivacaine[10], 5 ml of 2% xylocaine and fentanyl 50 mcg was given 5 min after test dose. After confirming level of block from T4 to T12, surgery was commenced. Ryle's tube was inserted. Effect of first dose lasted for 45 min. Epidural top ups of 5 mL of 0.5 % Bupivacaine were given at an interval of 30 min each for 6 times, total dose was180 mg . Later ropivacaine 0.2% 6ml every 30 mins was given and total dose was 24 mg .Surgical relaxation was excellent.



Continuous monitoring of Pulse Rate, Arterial Blood pressure, ECG, SPO2 recorderd every 3 mins. Duration of sugery was 4½hrs .Intraoperatively vitals were stable throughout operative procedure. ABG measurement to monitor acid-base status[11] was normal.



Patient did not have complication like bradycardia, hypotension, respiratory depression, shivering, nausea and vomiting. Patient shifted to ICU for monitoring.

Postoperatively Epidural top up with 6ml/ hr of 0.2 % Ropivacaine was given as continous infusion for 8 hrs. The patient received 3 units FFP and 1 unit of PRBCs after clamping the spleenic vessels and 1500 ml of crystolloids given totally. Total blood loss was 500 mL and urine output 400 mL. The intra-operative temperature was 35°C-35.5°.

Discussion

HS is an autosomal dominant disorder. It occurs due to a defect in the genes coding for proteins ankyrin or β spectrin.

Commonly recommended peri-operative management includes erythrocyte transfusion, avoidance of hypoxia, hypotention, hypothermia, and acidosis. Perioperative goals by providing adequate analgesia, to avoid hepatotoxic drugs, to maintain hepatic blood flow.

Laboratory diagnosis involves a peripheral blood smear, in which spherocytes are seen. Acantho-cytes or speculated red cells may also be seen.

MCHC is increased. The reticulocyte count is increased. Osmotic fragility test is often positive.

Epidural anesthesia was chosen in our patient primarily because she had Bronchial Asthma, cardiac disease and jaundice. General Anaesthesia can potentiate deterioration in liver function and liver blood flow.

Regional anesthesia probably preserves liver blood flow as long as normotension is maintained. Epidural anesthesia is desired as the stress associated with general anesthesia can lead to the release of catecholamines, which can decrease liver blood flow.

Conclusion

The management of HS rare disorders is largely dependent on the severity of anemia and degree of hemolysis. Anemia should be corrected preoperatively before a major surgery. Epidural Anaesthesia was chosen because patient had severe airway obstruction, elevated LFT and cardiac disease. Segmental epidural is the choice in such situations as it causes minimal physiological alterations[12].

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