

Case Report:

Anaesthetic considerations for patient of Hereditary Spherocytosis for splenectomy and cholecystectomy: A case report

¹Dr Anuradha Malliwal, ²Dr Sonal Mehta, ³Dr Rochana Bakhshi, ⁴Dr Bibekananda Mahapatra

[¹ Associate Professor , ² PG Student 3rd year , ³ Professor , ⁴ Associate Professor]

Department of Anaesthesiology, DY Patil Medical College, Navi Mumbai , Maharashtra , India

Corresponding author: Dr Sonal Mehta (drmehtasonal@gmail.com)

Date of submission: 12 November 2014 ; Date of Publication: 15 December 2014

Abstract:

Hereditary spherocytosis (HS) is an inherited haemolytic disorder characterised by anaemia, jaundice and splenomegaly. The primary lesion is reduced deformability of erythrocytes which are trapped and destroyed in the spleen resulting in haemolysis and anaemia. Common complications include cholelithiasis, haemolytic episodes, and aplastic crisis. Splenectomy is curative. Here we report the case of a 19-year old female HS patient who underwent splenectomy and cholecystectomy under general anaesthesia. The chief considerations in perioperative management include pre-emptive erythrocyte transfusion, proper hydration and avoidance of hypoxia, hypercarbia and acidosis.

Key Words: Hereditary Spherocytosis, splenectomy ,cholecystectomy, perioperative management.

Background:

Hereditary spherocytosis (HS) is an inherited haemolytic disorder with clinical features ranging from an asymptomatic condition to a fulminant haemolytic anaemia requiring erythrocyte transfusion.. Incidence is 1:5000. The primary lesion is reduced deformability of erythrocytes due to defects in the membrane proteins ankyrin, spectrin, or protein 4.2. Abnormal spherocytes are trapped and destroyed in the spleen resulting in haemolysis. Characteristic features include anaemia, jaundice and splenomegaly. The patient was posted for surgery under general anaesthesia after correction of anaemia with blood transfusion and vaccination against encapsulated organisms.^(1,2)

Case Report:

We report a case of a 19-year old female HS patient, weighing 35kg scheduled for splenectomy and cholecystectomy. She presented with anaemia, repeated episodes of jaundice, loss of appetite, easy fatigability, pain in both hypochondria and progressive dyspnoea on exertion since two years.

On examination, she had pallor, icterus and splenomegaly.

Breath-holding time was 20 seconds. HS was diagnosed from spherocytosis on peripheral blood smear and increased erythrocyte osmotic fragility. Haemoglobin (Hb) -9.4gm%, vitamin B₁₂ levels were decreased (84pg/ml).

Thalassemia studies, direct Coomb's test, serum iron studies and tests for other haemolytic anaemias were normal. Ultrasonography abdomen showed splenomegaly (splenic span-18cms) and echogenic sludge in the gallbladder. Parents and siblings tested negative for the same.

The patient was posted for surgery under general anaesthesia after correction of anaemia with blood transfusion and vaccination against encapsulated organisms. Post-transfusion investigations were: Hb-11.5gm%, platelets 245K/ml, prothrombin time 14.5secs, INR 1.06, total bilirubin 5.28mg% (with indirect bilirubin 4.65mg%), serum aminotransferases mildly elevated and LDH 590 U/L. Other routine investigations were

unremarkable. Folic acid supplementation was started.

In the operation theatre, two wide-bore cannulae were secured. She was premedicated with inj. glycopyrrolate and cautious, intermittent doses of inj. midazolam and inj. fentanyl. After induction with inj. pentothal sodium and intubation facilitated by inj. atracurium, anaesthesia was maintained with N₂O, O₂ and isoflurane. Intraoperatively, care was taken to avoid hypoxia, hypothermia and acidosis. ECG, NIBP, oxygen saturation (SpO₂), end tidal carbon dioxide (EtCO₂) and urine output were monitored throughout surgery.

Blood loss was approx. 1000ml, which was replaced with one colloid and one unit PCV (after clamping of splenic vessels). Surgery lasted 3hrs and was uneventful. Multimodal analgesia was provided with inj. tramadol, inj. paracetamol and local infiltration of the surgical site with inj. bupivacaine 0.25%. Patient was extubated after surgery and shifted to ICU for observation.

Postoperatively, she received two units PCV and four fresh frozen plasma. Chest physiotherapy, nebulisations and incentive spirometry were started. On postoperative day 2, patient had a febrile episode with diminished air entry in bilateral lung bases. Day 3: she had fever spike, tachycardia, tachypnoea, basal creptations. Complete blood count (CBC), peripheral smear (PS) for malarial parasites (MP), liver function test (LFTs), serum creatinine, serum electrolytes and blood for culture/sensitivity was sent, which showed neutrophilic leukocytosis on CBC, PS for MP negative, others WNL. Higher antibiotics were started.

Patient recovered and was shifted to ward on day 6 and discharged on day 11 with advice for penicillin prophylaxis life-long. (Tab pentid 400mg bd).

Discussion:

HS is the commonest cause of inherited haemolytic anaemia in North America (1:5000 births), but comparatively rare in Asians. Most cases are inherited as autosomal dominant; approximately 25% are sporadic and may be due to spontaneous mutations. Diagnosis is generally made in childhood or young adult life.^(3,4,5)

HS results in the formation of abnormal red blood cells with fragile cell walls leading to anaemia, jaundice, splenomegaly and ultimately, gall stone formation. Intrinsic membrane defect results in increased fragility of RBCs predisposing to hemolysis.

Four abnormalities in red cell membrane proteins have been identified-

- (1) Spectrin deficiency alone (**most common defect**)
- (2) Combined spectrin and ankyrin deficiency,
- (3) Band 3 deficiency, and
- (4) Protein 4.2 defects.

These defects result in uncoupling in the vertical interactions in the lipid bilayer skeleton and loss of membrane microvesicles. This leads to loss of membrane surface area without a proportional loss of cell volume and causes sphering of the RBCs.

Diagnosis of HS is based on increased osmotic fragility, spherocytes on peripheral blood smear and increased MCHC.

Complications include (1) Cholelithiasis, a consequence of chronic hemolysis (2) aplastic crisis- most commonly after Parvovirus B19 infection (3) hemolytic crisis during intercurrent infection and

(4) megaloblastic crisis in presence of folic acid deficiency. Acute chest syndrome --onset of new lobar infiltration on chest x-ray (excluding atelectasis) associated with fever, respiratory distress or chest pain has been reported.

Clinical management varies with the degree of the disease. Surgical treatment involves splenectomy, which is indicated in all patients to prevent the risk of gallstone formation and hemolytic crises, except in those who are well-compensated and asymptomatic. It results in cessation of haemolysis, return of the haemoglobin levels to normal and clearance of jaundice. Splenectomy should be performed after the age of 6 years and with appropriate counselling about the risk of infection. If gallstones are present, cholecystectomy may be performed simultaneously, or at a later date. Immunisation with pneumococcal and haemophilus influenza vaccines should precede splenectomy because of risk of life-threatening sepsis due to encapsulated organism. Anaesthetic management of HS involves pre-splenectomy, splenectomy and post-splenectomy care. Preemptive erythrocyte transfusion, aggressive hydration and avoidance of hypoxia, hypothermia and acidosis form the basis of preoperative management. Intraoperatively, avoidance of hypoxaemia is the key. Sedatives and opioid analgesics should be

used with extreme caution to prevent respiratory depression, hypoxia and sickling. Blood loss should be replaced whenever necessary. Normothermia should be maintained to minimize vasoconstriction and associated circulatory stasis.

Post-splenectomy, risk of life threatening sepsis due to encapsulated organisms necessitates life-long penicillin prophylaxis. Increase in average haemoglobin levels, platelet levels and cholesterol concentration are associated with an increased risk of atherosclerotic events, such as stroke and myocardial infarction.⁽⁵⁾

Conclusion:

Peri-operative management of HS largely depends on the severity of anaemia and the degree of haemolysis. Anaesthetic goals include avoidance of hypoxia, acidosis, and hypothermia. Vaccination before splenectomy is a must to prevent post-operative infections. Patients of HS are at increased risk for developing perioperative complications like aplastic crisis and haemolytic episodes for which awareness and vigilance are of vital importance.

References:

1. Bolton-Maggs PH, Stevens RF, Dodd NJ, Lamont G, Tittensor P, King MJ, *et al.* Guidelines for the diagnosis and management of hereditary spherocytosis. *Br J Haematol* 2004;126:455-74.
2. Agre P, Asimos A, Casella JF, McMilan C: Inheritance pattern and clinical response to splenectomy as a reflection of erythrocyte spectrin deficiency in hereditary spherocytosis. *N Eng J Med* 1986 Dec 18; 315 (25): 1579-83.
3. Steinberg MH. Management of sickle cell disease *N Eng J. Med* 1999; 340: 1021-30.
4. Shapiro ND, Poe MF. Sickle cell disease, an anaesthesiological problem, *Anaesthesiology* 1995; 16: 771-80.
5. Hassan A, Babadoko AA, Isa AH, Abunimye P. Hereditary Spherocytosis in a 27-year old female: case report. *Annals of African Medicine* vol 8, No1;2009:61-63 .
6. Mariani M, Barcellini W, Vercellati C, Marcello AP, Fermo E, Pedotti P, *et al.* Clinical and hematologic features of 300 patients affected by hereditary spherocytosis grouped according to the type of the membrane protein defect. *Haematologica* 2008;93:1310-7.