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Case Report

ANAESTHETIC MANAGEMENT OF A CASE OF HERIDITARY SPHEROCYTOSIS FOR SPLENECTOMY AND CHOLECYSTECTOMY

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ABSTRACT:

Objectives: *Hereditary spherocytosis is a heterogeneous group of disorder that results in the formation of abnormal red blood cells with fragile cell walls causing anaemia, jaundice, splenomegaly and ultimately gall stone formation. Most children have mild disease do not require splenectomy. Splenectomy is reserved for those with severe disease or who develop symptomatic gall stone. Individuals with symptomatic gallstones usually have a cholecystectomy and at the same time splenectomy if indicated.*

Case Report: *A 20 years old female diagnosed as hereditary spherocytosis since age of one year. After 19 years she was diagnosed as splenomegaly with cholelithiasis. After proper investigations and vaccination patient was posted for surgery splenectomy and cholecystectomy at the same time which is challenging from the anaesthetic point of view because the sickling oriented anaesthetic approach. Commonly recommended perioperatoive management includes preemptive erythrocyte transfusion, aggressive hydration and avoidance of hypoxia, aplastic crisis, hypothermia and acidosis.*

Conclusion: *Patients with Hereditary spherocytosis, as they are more prone to develop infection, were meticulously controlled through out the perioperative period. Removing the spleen does not cure the disease, but it does allow the red blood cells to live longer so that a child no longer became anaemic during periods of stress or infection. It is very important that all these patients should receive all of the normal childhood immunizations and a few special immunizations (pneumococcal and meningococcal) to prevent infection.*

Key Words*: Hereditary Spherocytosis, Splenectomy, cholecystectomy, aplastic crisis.*

INTRODUCTION:

Hereditary spherocytosis is an inherited disease that results in the formation of abnormal red blood cells with fragile cell walls which is usually transmitted as an autosomal dominant disorder. 25% of patients with hereditary spherocytosis have no previous family history and mostly represent as new mutation[1]. In hereditary spherocytosis erythrocytes shape changes are caused by membrane protein defects resulting in cytoskeleton instability. Spectrin deficiency leads to loss of erythrocyte surface area, which produces spherical RBCs. Spherocytic RBCs are culled rapidly from the circulation by the spleen ultimately develop splenomegaly. Four abnormalities in red cell membrane proteins have been identified and include-(a) spectrin deficiency alone, (b) combined spectrin and ankyrin deficiency, (c) Band 3 deficiency and (d) protein 4.2 defect. Spectrin deficiency is most common defect[2].

Normal RBC is shaped like disc.Spherocytes is round and fragile and does not change shape to pass through certain organs as easily as normal RBC. Because spherocyts cannot change their shape easily, they stay in the spleen longer than normal RBC and the membrane surrounding the cell becomes damaged. After circulating through the spleen many times, the cell eventually becomes so damaged that it is destroyed by the spleen which leads to anaemia and jaundice.Frequently patients with hemolytic anamia develop gallstones as a complication of the increased red cell hemolysis.

For practical purpose, the treatment of hereditary spherocytosis involves presplenectomy care, splenectomy and post splenectomy care. The presplenectomy care was taken before planed for surgery. The new pathophysiological model has a number of implications for anaesthgetic management. Commonly recommended perioperative management includes preemptive erythrocyte transfusion, aggressive hydration, avoidance of hypoxia, hypothermia and acidosis[3,4].

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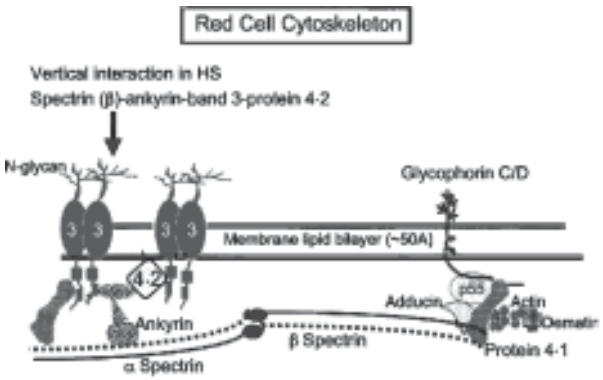

Fig.-1: *Schematic presentation of structural organization of red cell cytoskeleton(Bolton-Maggs,2004). â Spectrin is the key component in that it pairs with á spectrin to form a heterodimer, and it binding sites for ankyrin and protein 4.1.The common protein defects are associated with spectrin (á and/or â), ankyrin, band 3 protein and protein 4.2.*

Avoidance of hypoxaemia is the key goal in sickling-based management. Premadication and opioid based analgesia has been traditionally been used with extreme caution because of concern about respiratory depression, hypoxia and sickling.[5.] There is also a high level of analgesic tolerance in these type of patient due to recurrent episodes of sever pain. Hypothermia has also been suggested as a perioperative trigger of complications. Acute chest syndrome (ACS) is another important complication in which the onset of new lobar infiltration on chest x-ray, excluding atelectasis, accompanied by fever, respiratory distress or chest pain. After concerning all these complications anaesthetist should therefore concentrate on the basic standards generally accepted anaesthetic practice.

CASE REPORT :

A 20 year old female with known hereditary spherocytisis, presented with right upper quadrant pain and nausea. Physical examination was unremarkable except for right upper quadrant pain and moderately icteric sclera. There was also moderate splenomegaly. Her past medical history was typical of a patient with hereditary spherocytisis with frequent admission usually for fever and blood transfusion, but no history of major complications such as acute chest syndrome, Stroke etc. According to her clinical presentation, cholelithiasis was confirmed by abdominal ultrasound, which revealed a distended gallbladder with containing echogenic structures within it with normal wall thickness. Liver is enlarged in size and spleen is measuring 19.41 cm in its long axis. Subsequently patient was admitted under general surgery department for both splencetomy & cholecystectomy at the same sitting.

On admission blood work performed include a complete blood count, coagulation profile, Hb-Electrophoresis, S. Bilirubin, Liver function tests(LFTs), osmotic fragility tests, S. creatinine, S.urea, urine analysis & anaemia type & blood cross matching. The coagulation profile was normal. The complete blood count was remarkable for anaemia (Hb-7.5gm/dl). The Hb electrophoresis was in normal pattern. The presence of spherocytes on the peripheral smear associated with increased osmotic fragility test which is the usual pattern for spherocytosis reveals the diagnosis of hereditary spherocytosis. After conformation with ultrasonology our patient was decided to do the splenectomy and cholecystectomy at the same sitting. However before surgery she was given vaccine against pneumococci, H-influenza & hepatitis B. With all aseptic precaution, induction of anaesthesia was done with propofol, fentanyl, and rocuronium and was maintained with 50%N_2O in O_2 and vecuronoum and isoflurane.

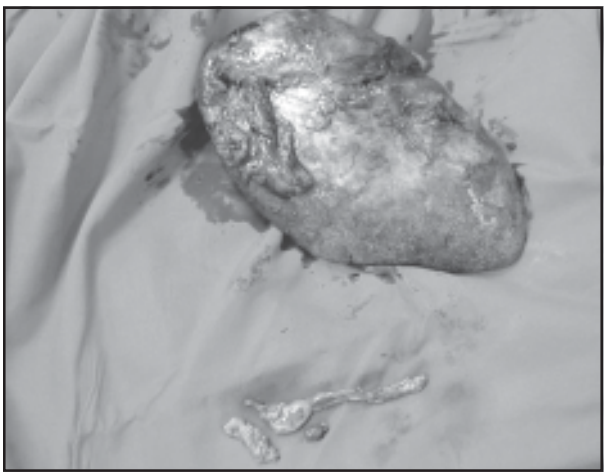

Fig.-2: *The dissected Spleen(19.41 cm) with spleenecules.*

Post operative analgesia was maintained with conventional intramuscular injection of Pethidine. Per operative 4 units of packed cell was given after

clamping of splenic vessels. 24hr after operation haemoglobin becomes 15gm/dl but 72 hr after Hb becomes 10gm/dl but the platelet count becomes > 7 lakhs and injection Clexane subcutaneously, 12 hourly was started as thrombo-prophylaxis. From the 1st POD prophylaxis injection Penicillin was also started in addition to other medication.

DISCUSSION:

Cholelithiasis is a known complication of hemolytic anaemia & is frequently associated with hereditary spherocytosis. Splenectomy is very effective in reducing haemolysis, leading to a significant prolongation of the red cell life span, although not necessarily to normal. The clinical manifestations & complications (anaemia & gallstones) are much reduce in sever hereditary spherocytisis, but at the price of one increased risk of life threatening sepsis from encapsulated organisms, particularly streptococcus pneumonia[6].

Our patient was also selected for splenectomy on the basis of her clinical symptoms & presence of complication such as gallstones, not simply on the basis of the diagnosis alone.

An analysis of decision making for mild hereditary spherocytosis, based on the available lecture & computer modeling, suggests that spleenectomy is of no benefit in the absence of gallstones (Marchetti, et. all, 1998), children or young adults with mild hereditary spherocytosis, who also have gallstones are likely to benefit from combined splenectomy & cholecystectomy in terms of life expectancy[6] .

Moreover many patients with such sickling disorder have impairment to oxygen delivery secondary to pulmonary damage, widespread macro and microvasculopathy, increased blood viscosity, anaemia, impaired vascular regulation and disturbed nitric oxide signaling[7].They therefore may have limited reserve to cope with further reduction in oxygen delivery. Thus the avoidance of hypoxaemia is the foundation of anaesthetic management of this patient.

The risk of postoperative sepsis is not completely eliminated by the current recommended preoperative vaccinations & post splenectomy antibiotic prophylaxis (BCSH, 1996, Davis et .all, 2002). The small but definite remaining risk of sepsis is also clearly explained to the parents of our patient & the indication for splenectomy must be clear.

For pre splenectomy vaccination & post splenectomy follow up., our pt was preoperatively vaccinated and also advised to repeat pneumococcal vaccination at 5 years intervals, although there is no clear evidence in the literature. Penicillin prophylaxis is recommended for life (reid 1994). Although preventive measures are very successful, it is clear that they do not completely eliminate risk. The risk of infection remains highest in the youngest patient but was reported to be reduced by 47% & the mortality by 88% in one study (Jugenberg et al 1999).

The risk of late past spleenectomy thrombosis[8] is also sought out for our patient, and standard thrombo prophylaxis that is subcutaneous heparin was also started.

CONCLUSION:

After family studied, this case may be a new genetic mutation case. As our patient. was diagnosed from the age of 1yr 9m[n], the complication of gallstone is obvious. Splenectomy is reserved for those with sever disease or who develop symptomatic gall stone, when cholecystectomy should be performed at the same time. After concerning all probable complications like aplastic or megaloblastic crisis, hemolytic crisis, acute chest syndrome, stroke etc anaesthetist should have better understanding of silent but insidious end –organ damage in the brain, kidney and lungs. This allows for more accurate preoperative assessment, with better grasp of the potential for perioperative organ dysfunction in the individual patient. It also points to the development of potentially effective ways of perioperative sickle cell complications.

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