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Different Surgical Approaches, Similar Outcomes: A Two-Case Report of Spleenectomy in Hereditary Spherocytosis

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Abstract:

- ***Background***

Hereditary spherocytosis is a congenital disorder predominantly inherited in autosomal dominant pattern. It is characterised by haemolytic anemia which presents as Anemia , Jaundice ,Spleenomegaly and gall stones (pigmented gall stones) . Splenectomy is definitive treatment with laparoscopic approach increasingly preferred but open splenectomy also favoured in selected cases

- ***Case Presentation***

We present two cases of hereditary spherocytosis managed with two different surgical approaches. A 33-year-old male patient with symptomatic splenomegaly and cholelithiasis underwent combined laparoscopic splenectomy with cholecystectomy, resulting in rapid postoperative recovery. A 47-year-old patient presented with severe anemia, jaundice, and massive splenomegaly who underwent open splenectomy after preoperative optimization. Both patients showed significant clinical and hematological improvement postoperatively.

- ***Conclusion***

Both approaches laparoscopic and open splenectomy are almost equally effective in managing HS , minimally invasive surgery has advantage of faster recovery, while open splenectomy remains a safe and effective option in cases of massive splenomegaly or resource limitations . Individualised surgical planning is essential for best outcomes.

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I. INTRODUCTION

Hereditary spherocytosis is an inherited hemolytic anemia caused by mutation in various genes, most commonly ANK1 gene followed by SPTB,SLC4A1,SPTA1 and EPB42. These mutations result in defects in erythrocyte membrane proteins such as spectrin, ankyrin, and band 3. These abnormal proteins result in spherical and osmotically fragile red blood cells that are mechanically trapped and hemolyzed in the spleen, leading to chronic extravascular hemolysis.

Clinically, HS presents with the classical triad of anemia, jaundice, and splenomegaly which presents with symptoms of fatigue, weakness pallor , icterus ,abdominal pain and discomfort.

Chronic hemolysis predisposes patients to pigmented gallstone formation. Splenectomy remains the definitive treatment for moderate to severe disease, as it removes the primary site of red cell destruction and significantly improves clinical symptoms and laboratory parameters .

With advances in minimally invasive surgery, laparoscopic splenectomy has become the preferred approach. However, open splenectomy also performed in selected cases, like in patients with massive splenomegaly or other technical considerations.

We present two adult cases of HS managed with two different surgical approaches to highlight outcomes and surgical decision-making.

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II. CASE 1

A 33-year-old male, a known case of hereditary spherocytosis, presented in opd with intermittent abdominal pain, fever, and jaundice. He had a history of multiple similar episodes in past and prior endoscopic intervention for CBD stones .

On general physical examination, pallor and icterus were present, on per abdomen palpation gross splenomegaly found extending up to the umbilicus. Laboratory investigations revealed hemoglobin of 9.9 g/dL , total bilirubin 10.98mg/dL out of which 10.11mg/dL was indirect bilirubin.. Peripheral smear showed spherocytes, and ultrasonography demonstrated gross splenomegaly with cholelithiasis.

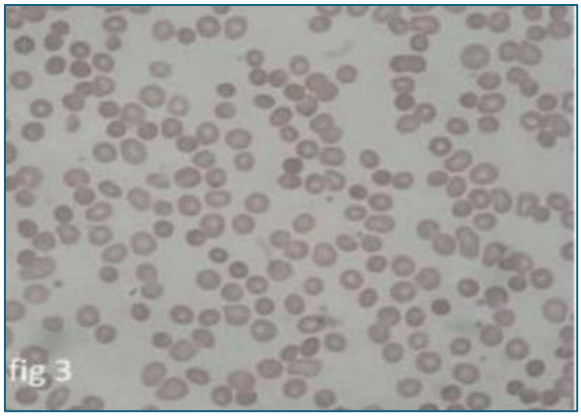

Fig 1 - Spherocytes

After proper preoperative vaccination, the patient underwent combined laparoscopic splenectomy with cholecystectomy. Intraoperatively, the spleen was markedly enlarged, intraop period was uneventful.

The postoperative course was uneventful. The patient resumed oral intake on postoperative day 1, and jaundice improved rapidly and indirect bilirubin fall to 5.02mg/dL. He was discharged on postoperative day 10 with normal physiological limit bilirubin levels.. At follow-up, there was complete resolution of symptoms with normalization of hemoglobin and bilirubin levels.


Fig 2 -Specimen

III. CASE 2

A 47-year-old female patient presented in surgery opd with chief complaints of abdominal pain and jaundice for 2 months which was gradually progressive , patient also complaints for generalized weakness and fatigue since 2 years.

On evaluation, blood investigations revealed severe anemia with hemoglobin of 6.6 g/dL, reduced RBC count . elevated total bilirubin of 15.51 mg/dL (predominantly indirect) Physical examination showed pallor and icterus, per abdomen palpation suggested spleenomegaly. On osmotic fragility test, increased osmotic fragility of red blood cells was present and Ultrasonography showed gross splenomegaly measuring approximately 18 cm.

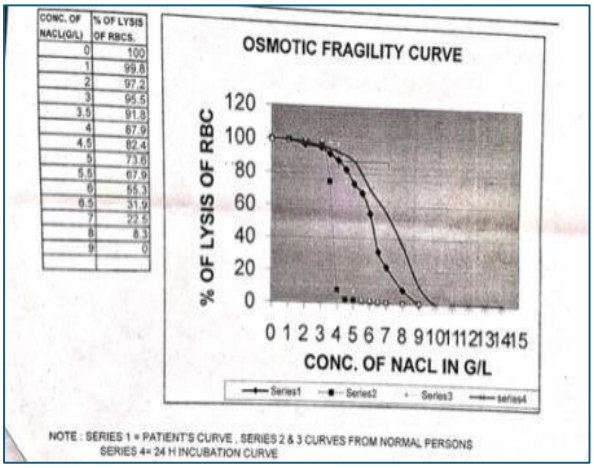

Fig 3- Increased Osmotic Fragility

The patient was improved preoperatively with transfusion of two units of packed red blood cells. Preoperative vaccination against pneumococcal and meningococcal infections was administered. Repeat investigations showed improvement in hemoglobin to 10.5 g/dL.

The patient underwent open splenectomy. Intraoperatively, a massively enlarged spleen measuring approximately 22 × 10 × 7 cm was removed. One unit of packed red blood cells was transfused intraoperatively.

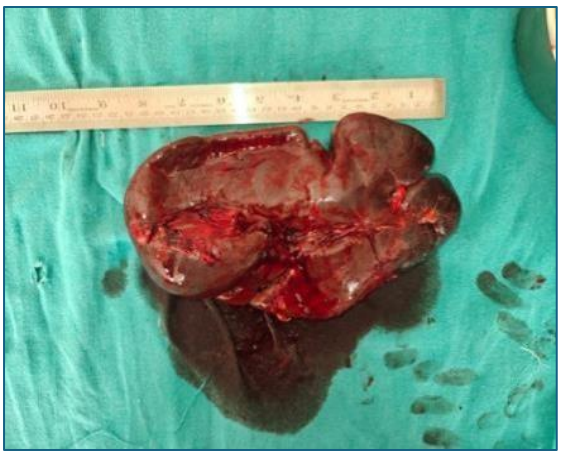
Fig 4– Specimen

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The postoperative period was uneventful, and no further transfusions were required, on postoperative day 2 jaundice improved significantly with indirect bilirubin level 3.93mg/dL ,By postoperative day 5, the patient showed marked clinical improvement with resolution of abdominal pain and jaundice. Laboratory investigations showed haemoglobin of 10.8 g/dL and normalisation of bilirubin levels.

At 30-day follow-up, the patient was asymptomatic with haemoglobin levels above 12 g/dL and no residual complaints.

IV. DISCUSSION

Hereditary spherocytosis results in chronic extravascular hemolysis due to destruction of spherocytes in the spleen. So splenectomy remains treatment of choice in chronic symptomatic cases as it effectively eliminates the hemolysis , leading to correction of anemia and rapid reduction in bilirubin levels, as we observed in both cases.

The first case shows the advantages of a minimally invasive approach. Laparoscopic splenectomy, combined with cholecystectomy, which allowed simultaneous management of splenomegaly and gallstone disease resulting in minimal postoperative pain, early recovery, and shorter hospital stay.

The second case proves that open splenectomy is still a valuable surgical choice, particularly in patients with massive splenomegaly or when laparoscopic facilities and resources are limited. Despite being more invasive and morbid to patient it provided equally effective hematological correction and clinical symptoms improvement.

Both cases showed:
- Postoperative rise in hemoglobin
- Dramatic fall in bilirubin levels(>50% fall in 48 hrs and >90%fall in 5 days.
- Resolution of clinical symptoms

These findings tells us that the primary determinant of outcome is removal of the spleen, rather than the surgical approach itself.

However, laparoscopic splenectomy offers clear advantages:
- Reduced postoperative pain
- Faster recovery
- Better cosmetics

Open splenectomy remains preferable in:
- Massive splenomegaly
- Hemodynamic instability
- Limited laparoscopic resources

Thus, surgical approach should be finalised based on patient factors and available resources.

V. CONCLUSION

Splenectomy is the definitive treatment for hereditary spherocytosis, resulting in excellent clinical and hematological outcomes. While laparoscopic splenectomy is the preferred approach due to its minimally invasive nature and open splenectomy continues to be a safe and effective alternative in selected cases. Tailoring the surgical approach to individual patient characteristics ensures optimal outcomes.

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