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Case Report

Anesthetic Management and Transient Postoperative Hyperbilirubinemia in a 16-Year-Old Male with Hereditary Spherocytosis for Cholecystectomy

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ABSTRACT

Hereditary spherocytosis (HS) is a hemolytic disorder in which perioperative stress may exacerbate hemolysis. We report the anesthetic management of a 16-year-old male with HS undergoing cholecystectomy under general anesthesia with epidural analgesia. Postoperatively, there was a transient rise in bilirubin levels without a corresponding fall in hemoglobin or clinical evidence of hemolysis. The patient remained hemodynamically stable and improved with supportive management. This case highlights that postoperative hyperbilirubinemia in HS patients is not always indicative of hemolytic crisis and should be interpreted in the appropriate clinical context.

Introduction

Hereditary spherocytosis is a disorder of red blood cell membrane characterized by chronic hemolysis. It is one of the most common inherited hemolytic anemias, with an incidence of approximately 1 in 2000–5000 births [1-2]. Patients frequently develop jaundice and pigment gallstones, often necessitating a cholecystectomy.

Anesthetic management focuses on preventing factors that exacerbate hemolysis, including hypoxia, acidosis, hypothermia, and hemodynamic instability [2-3]. We present a case of postoperative hyperbilirubinemia without evidence of hemolytic crisis.

Case Report

A 16-year-old male, weighing 34 kg, with a known case of hereditary spherocytosis, presented with complaints of vomiting, fever, jaundice, and pallor.

Clinical Findings

On examination, the patient had icterus and pallor (Figure 1).

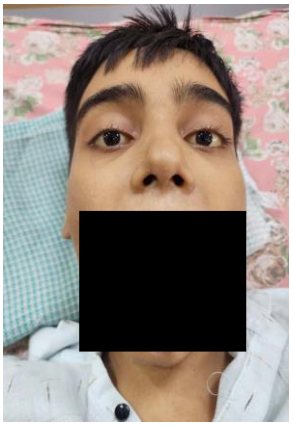

Figure 1- Patient with the features of Hereditary spherocytosis

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Timeline

- Preoperative: Hb 10.1 g/dL, total bilirubin 7.5 mg/dL
- Intraoperative: Uneventful with stable parameters
- Postoperative Day 1: Bilirubin increased to 13.4 mg/dL.
- Postoperative Day 2–4: Gradual clinical improvement and bilirubin decline

Diagnostic Assessment

Preoperative investigations showed:

- Hemoglobin: 10.1 g/dL
- Total bilirubin: 7.5 mg/dL
- Liver enzymes and renal function: Within normal limits

Postoperatively:

- Hemoglobin: 9.6 g/dL
- Total bilirubin: 13.4 mg/dL

There was no hemoglobinuria, no significant drop in hemoglobin, and no clinical evidence of hemolysis. Serum LDH, haptoglobin, and reticulocyte count were not available, which is a limitation.

Therapeutic Intervention

General anesthesia was administered with:

- Propofol 68 mg (2 mg/kg)
- Ketamine 68 mg (2 mg/kg)
- Fentanyl 34 mcg (2 mcg/kg), with titrated intraoperative doses
- Midazolam 0.68 mg (0.02 mg/kg)
- Glycopyrrolate 0.136 mg (0.004 mg/kg)

Endotracheal intubation was performed with a 6.5-mm cuffed tube. Anesthesia was maintained with sevoflurane in oxygen and air.

Muscle relaxation was achieved using vecuronium 3.4 mg (0.1 mg/kg), with additional doses as required.

A thoracic epidural catheter was placed at T9–T10, and ropivacaine infusion was started.

Intraoperatively:

- Normothermia maintained
- Adequate oxygenation ensured
- Hemodynamics stable (transient hypotension managed by adjusting epidural infusion).

Approximately 300 mL of packed red blood cells were transfused.

Neuromuscular blockade was reversed with:

- Neostigmine 1.7 mg (0.05 mg/kg)
- Glycopyrrolate 0.272 mg (0.008 mg/kg)

Patient was extubated uneventfully.

Follow-up and Outcomes

Postoperatively:

- Bilirubin increased transiently to 13.4 mg/dL.
- No clinical or laboratory evidence of hemolysis
- Epidural analgesia continued initially.

On postoperative day 2:

- The epidural stopped due to hypotension.
- Noradrenaline infusion started and was tapered by day 4.

Subsequently:

- Bilirubin decreased to 6.7 mg/dL.

Patient improved clinically and was shifted to the ward.

Discussion

Hereditary spherocytosis is characterized by chronic extravascular hemolysis due to defects in the red blood cell membrane [2-3]. During the perioperative period, various stressors such as hypoxia, acidosis, hypothermia, and hemodynamic instability can exacerbate hemolysis; therefore, careful anesthetic management is essential.

However, postoperative hyperbilirubinemia in such patients is not always indicative of a hemolytic crisis. Several non-hemolytic mechanisms may contribute to a rise in bilirubin levels, including increased bilirubin production due to surgical stress, transfusion-related bilirubin load, transient hepatic dysfunction, and the effects of anesthesia and perioperative hypotension [4]. These factors can result in elevated bilirubin levels even in the absence of active hemolysis.

Similar findings have been reported in the literature. Evans C et al [4] described postoperative jaundice in surgical patients without evidence of hemolysis, attributing it to multifactorial causes such as hepatic dysfunction and increased bilirubin load. In patients with hereditary spherocytosis, previous reports have also noted that bilirubin elevation may not necessarily correlate with hemolytic crisis, particularly when hemoglobin levels remain stable, and there are no clinical signs of hemolysis [2-3].

In the present case, the absence of a significant fall in hemoglobin, lack of hemoglobinuria, and overall clinical stability suggest that the postoperative rise in bilirubin was more likely due to non-hemolytic causes rather than an acute hemolytic episode.

Epidural analgesia played an important role in providing effective pain relief and attenuating the surgical stress response, which may help reduce triggers for hemolysis [5-6]. However, it was associated with transient hypotension in this case, requiring careful monitoring and timely intervention.

Limitations

This case report has certain limitations. Key laboratory parameters to definitively exclude hemolysis, such as serum lactate dehydrogenase, haptoglobin levels, and reticulocyte count, were not available.

Additionally, the study is a single case report with limited follow-up, which restricts the generalizability of the findings. Further studies are required to better

understand postoperative bilirubin changes in patients with hereditary spherocytosis.

Conclusion

Perioperative management of patients with hereditary spherocytosis requires careful optimization and avoidance of hemolysis triggers. Postoperative hyperbilirubinemia should be interpreted cautiously and not assumed to indicate hemolytic crisis in the absence of supporting clinical and laboratory findings.

Ethics approval and consent to participate

Institutional ethics committee approval was waived for this case report as per institutional policy. Consent was obtained from the patient and guardians.

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