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Case Report

Innovative Management of Congenital Hereditary Spherocytosis: Combined Laparoscopic Cholecystectomy and Splenectomy in A Paediatric Patient

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Abstract:

Congenital Hereditary Spherocytosis (CHS) is an inherited dysfunction or a deficiency in one of the erythrocyte membrane proteins. It is an inherited form of haemolytic anaemia which is noted by the presence of spherical erythrocytes. This leads to increased fragility and haemolysis. This case report is based on an 8-year old girl with CHS. She presented with acute abdominal symptoms and was subsequently managed with a combined cholecystectomy and splenectomy. This strategy seeks to address both the frequent haemolytic crises and the presence of gallstones, ultimately providing an extensive management strategy for paediatric patients with CHS.

Keywords: combined; cholecystectomy; splenectomy; pherocytosis

Introduction

CHS is the most common form of haemolytic anaemia for which a splenectomy is indicated. It is inherited in an autosomal dominant fashion with the prevalence in Western populations being 1 in 5000.[1]

It is characterised by dysfunction or a deficiency in one of the erythrocyte membrane proteins which results in spherical erythrocytes prone to haemolysis. Patients commonly present with the following; mild jaundice, splenomegaly, anaemia and gallstones.

A splenectomy and cholecystectomy both are pillars of treatment to alleviate haemolysis and prevent cholecystitis due to gallstones.[2]

Risks and benefits should carefully be assessed before either procedures are performed in patients with CHS. The main indication is symptomatic haemolytic anaemia. If gallstones coexist with spherocytosis, gallbladder removal should ensue. Drastic clinical improvement, despite persistent haemolysis, occur often after a splenectomy in patients with severe disease.[3]

This case report presents a case of an 8-year old paediatric patient who underwent a combined laparoscopic approach to address both splenomegaly and cholecystitis.

Case Presentation:

An 8-year old girl was admitted to the Clinical Centre of the University of Sarajevo (KCUS) with complaints of abdominal pain which has been last five days, followed by vomiting and loose stools for the past two days. The patient presented with fever which was reaching up to 38.9°C.

Her past medical history was significant for CHS. She required multiple blood transfusions to manage her anaemia.

Upon physical examination, the patient presented with pallor and jaundice as well as notable splenomegaly.

Laboratory investigations revealed an elevated C-reactive protein (CRP) of 286.6 mg/L, a white blood cell count of 28 x 10^9/L, hemoglobin of 74 g/L, hematocrit of 0.22, red blood cell count of 3.05 x 10^12/L, and platelet count of 478 x 10^9/L. Her liver function tests showed an AST of 20 U/L, ALT of 11 U/L and LDH of 281 U/L. Her total bilirubin was elevated at 115 µmol/L, with unconjugated bilirubin at 83 µmol/L.

An abdominal ultrasound revealed a distended gallbladder with an echogenic and thickened wall up to 4 mm. The intraluminal contents indicated calculous cholecystitis. Her spleen was significantly enlarged and measured approximately 177 mm in length. She also demonstrated signs of diffuse peritonitis, especially in the ileocecal region, with a

reactive appendix that suggested acute enterocolitis. There was a moderate amount of free fluid peri-splenically and inter-intestinally.

Initial Management:

Based on the severity of the patient's symptoms as well as ultrasound findings, she was originally treated conservatively and scheduled for an appendectomy.

The treatment of the complicated appendicitis was a three-port laparoscopy mixed technique. This included; intra-corporeal ligation of the appendix vermiformis artery, extracorporeal trans-umbilical ligation of the appendix and the appendectomy.

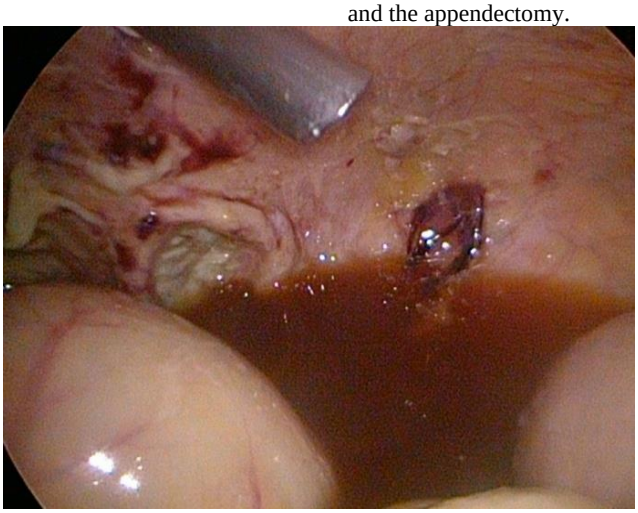

Figure 1: Perforated appendicitis.

The patient was discharged on the seventh postoperative day. A multidisciplinary team which included haematologists, based on her recurrent need for blood transfusions and the presence of multiple gallstones, agreed on a combined elective splenectomy and cholecystectomy.

Pre-operative Preparation:

The patient received a Pneumovax vaccine 15 days before the scheduled surgery in order to mitigate the risk of postoperative infections.

Surgical Procedure:

Three months post-appendectomy the patient had undergone a combined laparoscopic cholecystectomy. This procedure was performed using five ports: a 10-mm umbilical port for the laparoscope, two 5-mm ports in the left and right upper quadrants, and two additional 5-mm ports for instrumentation.

Laparoscopic Cholecystectomy:

Trocar placement:

A 10-mm trocar was inserted under the umbilicus for the laparoscope. A 10-mm trocar was placed in the epigastrium. Two 5-mm trocars were inserted in the right upper quadrant (Figure. 2).

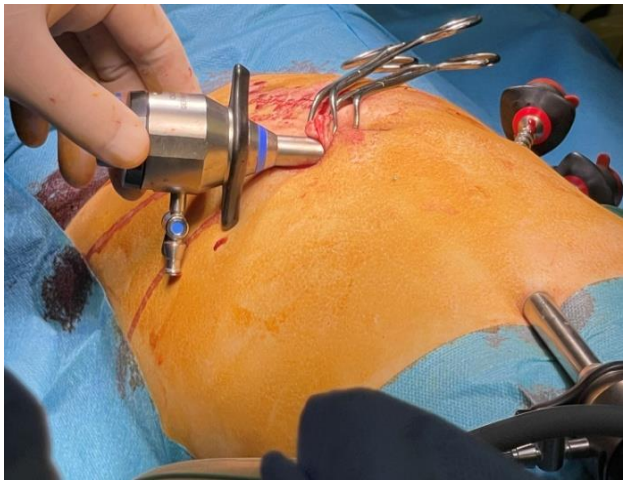

Figure 2: Trocar placement for the cholecystectomy.

Procedure:

After careful identification of the cystic duct and artery, both these structures were clipped (Figure.4). There after the gallbladder was dissected from the liver bed using electrocautery (Figure. 5). The gallbladder was removed through the umbilical port.

Figure 3: Distended gallbladder prior to removal.

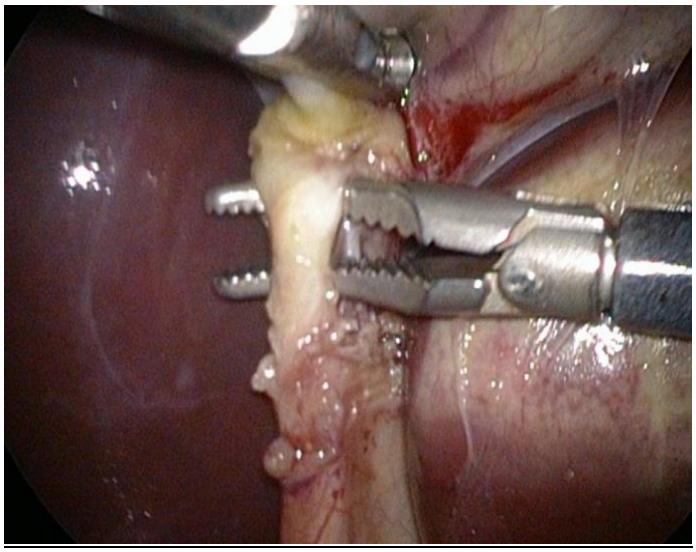

Figure 4: Identification of cystic duct and artery

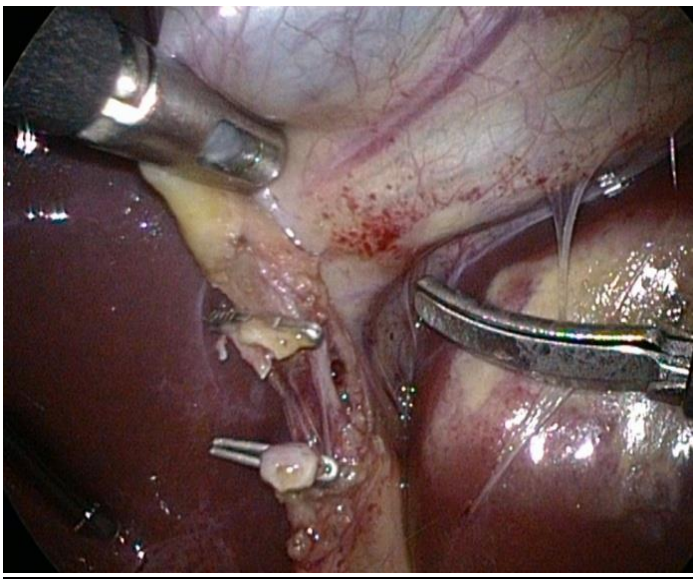

Figure 5: Clipped cystic duct and artery and resection of gallbladder from the liver bed.

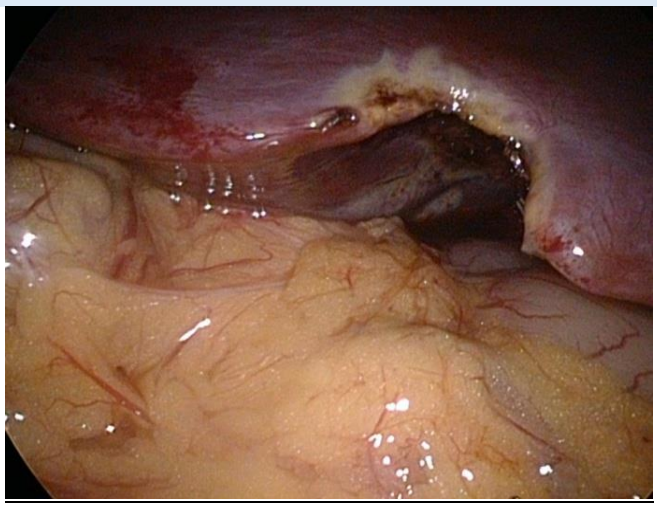

Figure 6: Resected area without gallbladder.

Laparoscopic Splenectomy:

Trocar placement:

The patient was rotated 45° degrees onto her side (Figure. 7). Additional ports were placed to facilitate the mobilisation as well as the removal of the spleen:

A 5-mm trocar was placed in the left upper quadrant. Another 5-mm trocar was placed in the left lower quadrant.

The exact placements for the five-port splenectomy were as follows:

Umbilical Port (10 mm): For the laparoscope, placed below the umbilicus.

Epigastric Port (10 mm): Midline, about 1 cm below the xiphoid process.

Left Upper Quadrant Port (5 mm): Approximately 5 cm below the costal margin, anterior axillary line.

Right Upper Quadrant Port (5 mm): Mid-clavicular line, below the costal margin.

Left Lower Quadrant Port (5 mm): Midway between the umbilicus and left iliac crest, anterior axillary line.

Figure 7: 45° rotation of patient onto side with tracer placement for splenectomy.

Procedure:

Following the cholecystectomy, the focus was shifted towards the spleen. The spleen was mobilised by dividing the splenocolic ligament followed by the short gastric vessels (Figure. 9). The splenic hilum was carefully dissected (Figure. 10). The splenic artery and vein were both clipped and divided (Figure. 11). Adhesiolysis was meticulously performed to dissect and separate fibrous adhesions between the spleen and adjacent structures to ensure the safe removal of the spleen. (Figure 12.) The significantly enlarged spleen was attempted to be extracted through the umbilical port. However, due to the unavailability of the correct specimens bag an incision was made from where the umbilical port had been placed to where the 5mm left lower quadrant quadrant port had been placed and thus the spleen was finally extracted using an open splenectomy. (Figure 13).

The entire procedure lasted 120 minutes. A drain was placed in the splenic bed and was removed on the second post-operative day.

Figure 8: Enlarged spleen in anatomical position

Figure 9: Splenocolic ligament with short gastric vessels

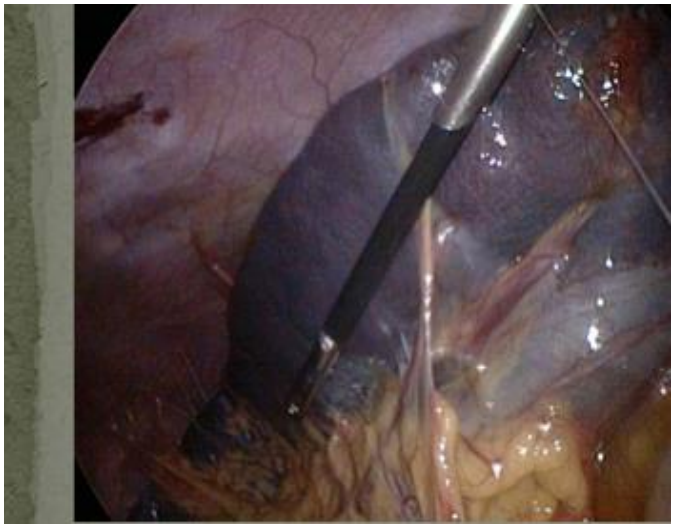

Figure 10: Dissection of splenic hilum.

Figure 11: Clipped and divided splenic artery and vein.

Figure 12: Adhesions dissected and separated.

Figure 13: Cavity after spleen had been resected.

Figure 14: Spleen extracted through open splenectomy.

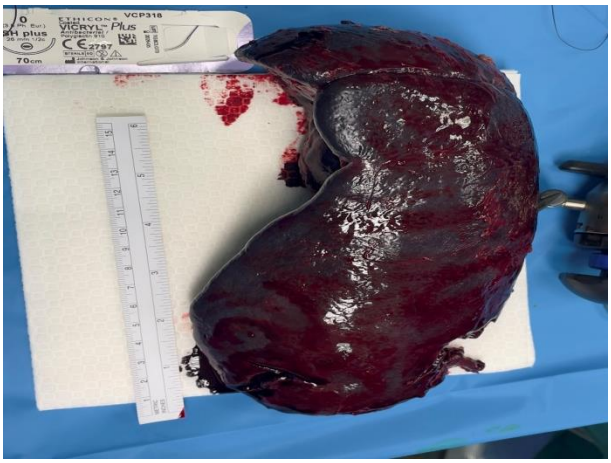

Figure 15: Extracted spleen measuring 177cm in length.

Figure 16: Surgical drain inserted and post operative result.

Post-operative course:

The patient's post-operative course was satisfactory and she was mobilised on the first post-operative day. She tolerated oral intake well. A haematological follow-up revealed no signs of jaundice or anaemia. The patient remained stable and was discharged on the fifth post-operative day.

Figure 17: Fifth post-operative day result, day of discharge.

Discussion:

In paediatric patients a combined laparoscopic cholecystectomy and splenectomy approach in

the management of CHS is a strategic and beneficial method. It coincides with the well-documented advantages of laparoscopic surgery, incorporating a faster return to normal activities, minimal trauma, and reduced postoperative pain.[4]

Laparoscopic procedures have expanded in numerous surgical fields due to these benefits. The first case of laparoscopic splenectomy, reported by Delaitre and Maignien,[5] lay the groundwork for its widespread adoption in treating hematologic diseases. Thus, laparoscopic splenectomy has now become the gold standard for elective hematologic spleen diseases.This includes cases of splenomegaly. It is safely performed at several referral centres.

Cholelithiasis is often associated with splenic disorders in patients who are in need of a splenectomy. However, reports on a combined laparoscopic splenectomy and cholecystectomy are scant. There is also limited data on the specific outcomes of these concurrent procedures. It is pivotal to understand the feasibility as well as safety of these combined surgeries. Especially when advocating for prophylactic splenectomies during cholecystectomies in patients with CHS. Not performing a prophylactic splenectomy during cholecystectomy for cholelithiasis could ultimately lead to an 18% lifetime risk of subsequent therapeutic splenectomy for these patients.[6]

The technique for these combined procedures varies in the literature as there are differences in the order of operations as well as patient positioning. For example, Yamagishi and Watanabe,[7] reported a method involving a 1-cm incision below the umbilicus for the laparoscope and three 5-mm trocars in the sub-xiphoid and subcostal regions. After securing the splenic hilum, the spleen was removed through an enlarged umbilical incision. A cholecystectomy was then performed in a standard laparoscopic manner.

Sasaki et al.[8] used a 4-trocar technique for a laparoscopic splenectomy in a semi-lateral position. This was followed by a cholecystectomy in the supine position. They emphasised a careful dissection to avoid any pancreatic injury. They also used a retrieval bag for the excised spleen.

In Nobili et al.'s series,[9] they first performed a cholecystectomy with the patient in the supine position, followed by a splenectomy in a semi-lateral position. They used the 5-trocar technique, which provided a good view of the operative field. This approach also facilitated with the dissection and ensured no complications. This aligns with the technique used in our case, where the combined approach proved effective and safe. Ultimately resulting in minimal postoperative complications and a satisfactory and short recovery.

Blood loss and conversion rates are critical metrics in evaluating the success of any procedure, especially in a joint cholecystectomy and splenectomy. Nobili et al. reported blood loss ranging from 10 to 700 mL, with a mean of 40 mL, and a conversion rate of 3% to 8%.[9] In our series, the blood loss was minimal. The patient also dud not require a blood transfusion. The conversion rate was 8.3%, comparable to literature data.

Operative times for combined procedures such as a cholecystectomy and splenectomy usually range from 70 to 300 minutes, with increased experience reducing the mean operative time to approximately 100 minutes. Hospital stays typically range from 3.5 to 15 days.[10] Our patient had a postoperative stay of 5 days, aligning with these findings.

Portal vein thrombosis (PVT) is an extreme and severe complication post-splenectomy, especially in patients with thrombocytosis, splenomegaly, and congenital thrombophilia disorders. Laparoscopic surgery might increase the risk of PVT due to pneumoperitoneum but could also prevent it by minimising postoperative coagulation changes.[11] With our patient no signs of PVT were reported.

Conclusion:

This case highlights how pivotal a multidisciplinary approach is in managing complex cases of CHS. A successful cholecystectomy and splenectomy provided an all-encompassing solution to the patient's haematological and biliary pathology. The importance of laparoscopic surgery in simultaneously treating gallbladder complications and ailments as well as those of the spleen in a single operation should be emphasised. These integrated procedures not only reduce overall morbidity but also optimise recovery, accentuating the importance of this approach in paediatric patients with CHS. Further studies as well as long-term follow-ups are essential to validate the benefits and safety of this combined surgical approach in the paediatric population.

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