Case study

The Perils of Laparoscopic Cholecystectomy in a Patient with Christmas Disease

Abstract

Surgical procedures in hemophiliacs require a combined and multidisciplinary approach from both the surgeon and haematologist. This report is of a successful laparoscopic cholecystectomy in a patient with haemophilia B. We describe a 34 year old male, known case of factor 9 deficiency with gall stone disease undergoing cholecystectomy with adequate pre and post operative optimisation. Challenges in such patients include deranged clotting time, high risk of bleeding in the perioperative period. These cases should be approached with caution and performed in high volume centres with experienced team of surgeons and haematologists.

Introduction

Haemophilia B is the second most common type of haemophilia. Haemophilia A is seven times more common than haemophilia B occurring in about 1 in 30,000-60,000 male births in India [1].

Haemophilia B is inherited in X-linked recessive manner. It occurs due to the deficiency of factor IX resulting in improper clotting of blood. Bleeding is the main symptom of the disease which may occur during circumcision in children, into joints causing haemarthrosis associated with pain and stiffness, in stool and urine. Nosebleeds, prolonged bleed from cuts, tooth extraction, during surgery are other suggestive symptoms [1-8].

Diagnostic workup of such patients would reveal activated partial thromboplastic time (APTT) to be prolonged, prothrombin time and bleeding time normal, fibrinogen level normal, factor 7 is normal and factor 9 is reduced.

Hemophilia being a coagulopathy, is a relative contraindication of laparoscopic surgery. However due to its advantages such as faster recovery, reduced surgical trauma and better cosmetic results, it's role as the primary method for cholecystectomy in hemophiliacs is relevant [4].

Surgical procedures in haemophiliacs require a combined approach from both the surgeon and haematologist. This report is of a successful laparoscopic cholecystectomy in a patient with haemophilia B. Having a thorough evaluation of such patients with adequate preoperative optimisation would mitigate any further complications.

Linghor, et al. [5] reported no significant differences were in duration of surgery and drains in laparoscopically or conventionally operated hemophiliacs versus matched pairs. Complication rate also did not differ among the different groups. Their study included 109 hemophiliacs including 21 laparoscopic cases.

Georg, et al. [6] also reported similar results with differences in hospital stay in patients of post operative appendectomy, hernia repair and hemorrhoidectomy but not for cholecystectomy.

Another successful laparoscopic cholecystectomy was reported by Zhang, et al. [9] in a 29-year-old man with a history of factor V deficiency. 2 FFP transfusions 1000mg each were given to the patient at the time of admission and 6 hours prior to surgery. LC was conducted with no major or minor post-operative complication

Yoshimura, et al. [10] reported a 70-year-old male cholecystitis case with factor VII (FVII) deficiency. The patient underwent successful laparoscopic cholecystectomy with no complication, with the infusion of 1 mg of recombinant factor VIIa.

Case Presentation

Here we present the case of a 34 year old gentleman who is a known case of Factor IX deficiency and well controlled type 2 diabetes mellitus presented with

intermittent attacks of pain in the right upper abdomen associated with nausea for the past 8 months. Patient suffered the last attack of pain abdomen 7 weeks back when he complained of a sharp pain in the right upper quadrant, non progressive and lasted for a few days which relieved on oral antibiotics and analgesics. Patient did not give any history of fever, vomiting or altered bowel habits. On general physical examination no obvious abnormality was noted. Systemic examination revealed only mild tenderness in the right upper quadrant of the abdomen. An ultrasonography of the abdomen was done which revealed that the gall bladder was contracted with multiple calculi of average size 10mm-11mm within. The liver was normal in size with increased echogenicity. A diagnosis of chronic cholecystitis with cholelithiasis was made. All routine blood parameters were within normal limits. Patient was on regular follow up in the department of haematology for Haemophilia B since the past 23 years and has been receiving factor IX concentrate infusion regularly with the last infusion one month back.

On admission Prothrombin time was 11.4 seconds, Activated partial thromboplastin time was 29.3 seconds (control - 26.8 s) and international normalised ratio (INR) was 0.97. Factor VIII assay level was 68.5% (71-110%) and Factor IX assay level was 74.2% (74-110%). Patient received a total of 5 factor IX concentrate infusions. One day prior to the surgery, on the day of

the surgery and till three consecutive post operative days.

After all the pre-anaesthetic work up, patient underwent laparoscopic cholecystectomy under general anaesthesia. Pneumoperitoneum was created using Verres technique. Intraoperatively, gall bladder was found to be contracted with minimal adhesions. Calots dissection was done and cystic artery and cystic duct was identified, clipped, and cut (as shown in Figure *1*). The gall bladder was dissected off the GB fossa and specimen was retrieved. Haemostasis was achieved. Port sites were closed. The gall bladder specimen contained multiple pigment stones with the largest being 10mm in size (as shown in Figure *2*). Immediate post operative period was uneventful. Coagulation profile done on post operative day 1 showed INR to be 0.84, Factor 8 assay level of 72.1% and Factor IX assay level of 80.5%. No complications were noted in the post operative period. Patient was discharged with advice on post operative day 4. The histopathological examination of the specimen was consistent with chronic cholecystitis. On follow up after 4 weeks, patient is doing well.

Discussion

Laparoscopic cholecystectomy is the preferred treatment option in cholelithiasis and cholecystitis. Matzsch et al performed the first laparoscopic cholecystectomy in a patient of haemophilia B and

reported longer operating time due to presence of an intrahepatic gall bladder but no bleeding or major complications [4]. They suggested laparoscopic cholecystectomy be adapted as the treatment of choice in haemophiliacs. And that the surgery be conducted with a team of haematologists and adequate coagulation laboratory facilities.

Another case report [9], reported intra op bleeding of 500ml and to manage this case, patient received recombinant factor VII concentrate, every 12h, 24h before the surgery and 4 days after the surgery. Furthermore 1g tranexamic acid was given every 8h during this period. Surgeon's expertise in laparoscopic cholecystectomy plays an impact on the outcome and hospital stay.

Linghor et al. [5] conducted a comparative study on haemophiliac patients who underwent open or laparoscopic surgery. They enrolled 109 haemophiliacs among which twenty one were cases requiring surgery. Fifteen underwent laparoscopic surgery and nine had open surgery. They reported no difference between laparoscopic and open surgery in terms of hospitalisation length and complications [6].

The patient in our report had normal levels of clotting factors 7, 8 and 9 preoperatively. There were no bleeding complications intra operatively. Post operative clotting factor assays were also normal. Haematologist was consulted for both pre op preparation and discharge

advice. Patient was discharged on post op day 4 and followed up after a month. He reported no complication or any bleeding manifestation.

Zabihi et al [4] published a case report of laparoscopic cholecystectomy on a patient with hemophilia A. The patient underwent LC with no complications and required 5 transfusions of factor VIII concentrate. They also suggested laparoscopic cholecystectomy as the preferred method in such patients.

Mallen, et al. [11] reported a case of an elective cholecystectomy in a patient with von Willebrand's disease. The patient underwent laparoscopic cholecystectomy with adequate preoperative evaluation. They described the laparoscopic method as the better choice as it requires lesser cutting of the abdominal musculature and better magnification to achieve hemostasis.

Conclusions

Hemophilia B is a rare condition and further research is required in describing the challenges encountered while operating such patients. In our case report the patient underwent successful laparoscopic cholecystectomy and required 5 post op factor IX transfusions. Laparoscopic Cholecystectomy can be used as the treatment of choice in haemophiliacs. Careful monitoring of the patient during the post operative period is vital to prevent adverse bleeding reactions. Such cases should be

operated in tertiary care centers and by experienced surgeons. A thorough preoperative check up must be done which should include routine coagulation parameters and factor IX assays, to predict any complications that the surgeon may encounter. Haematologists should be consulted and provisions for safe bleeding management should be ensured.

References

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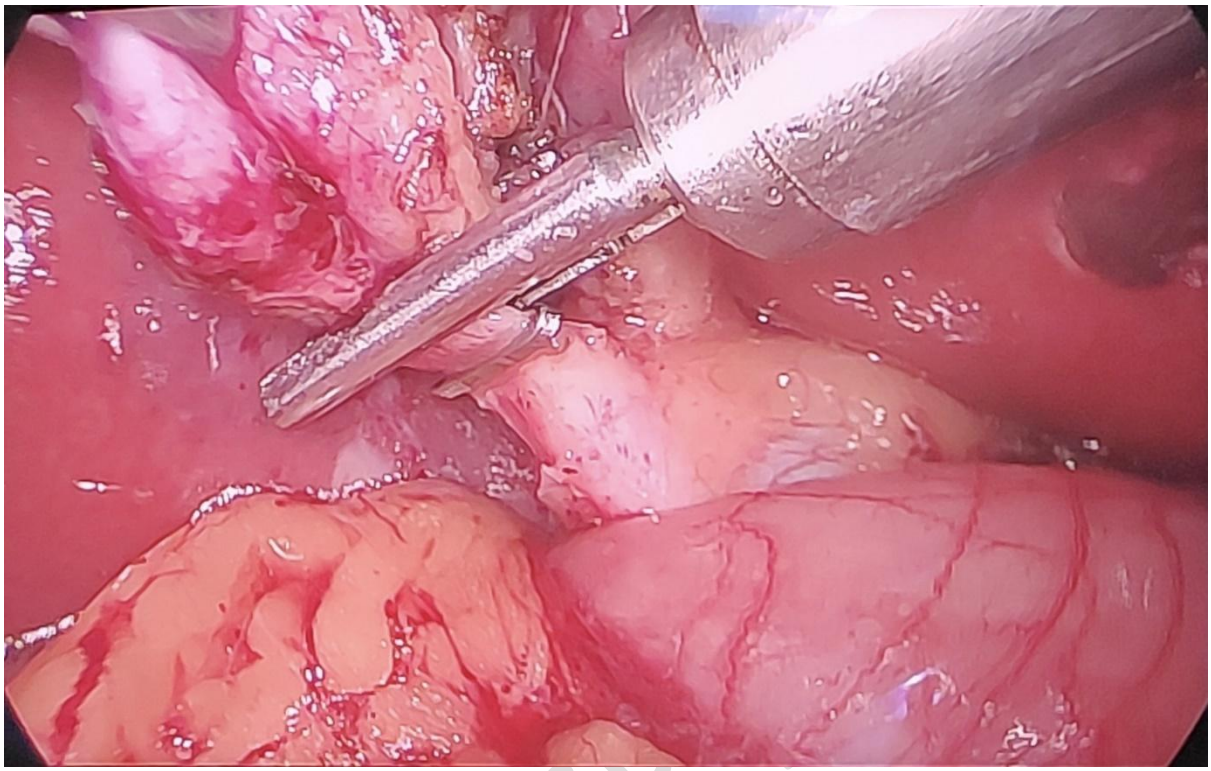

Figure 1: Cystic duct identified and clipped

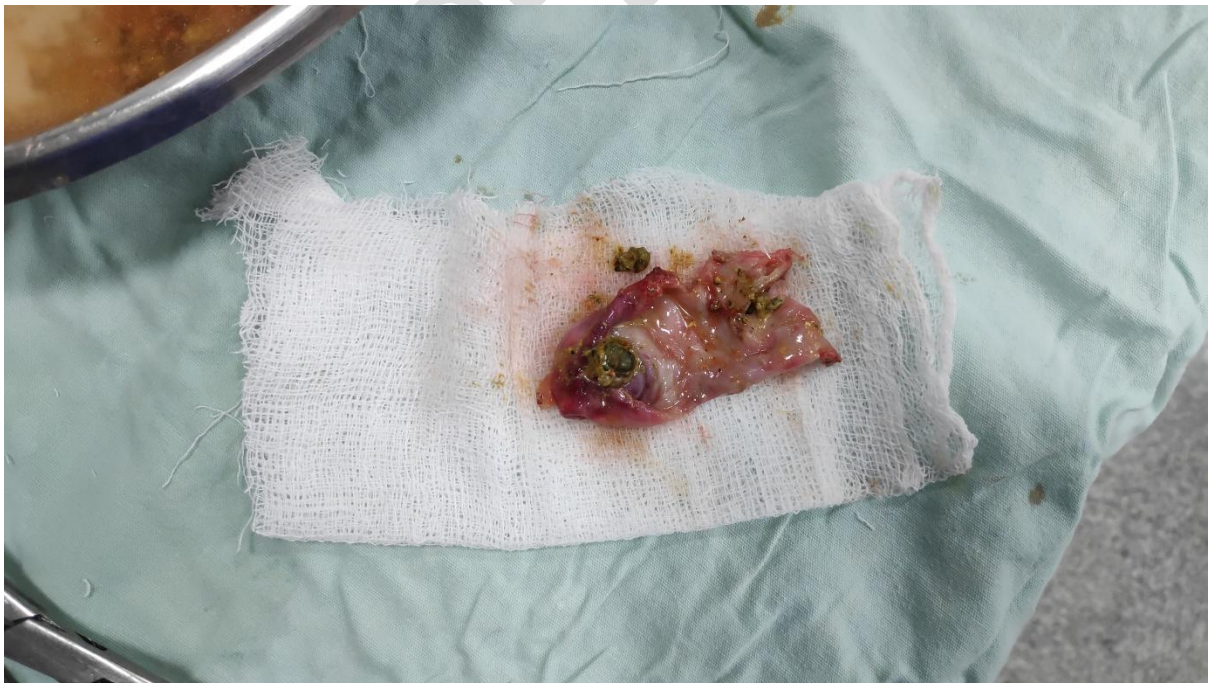

Figure 2: Gall bladder specimen contained multiple pigment stones