

Case Report

Successful laparoscopic common bile duct exploration in a patient with factor V deficiency, a case report and review of literature

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Abstract: Factor V deficiency is a rare bleeding disorder. Fresh frozen plasma (FFP) is the only source of factor V because factor V concentrates is not available now. We present here a patient had concomitant gallbladder and common bile (CBD) stones with factor V deficiency. The patient is successfully treated by laparoscopic CBD exploration and cholecystectomy with perioperative fresh frozen plasma transfusion. To best of our knowledge, this is the first report of laparoscopic surgery successfully performed in a factor V deficiency patient. Our result suggest that laparoscopic surgery in a factor V deficient patient can be performed safely if normal coagulation profile is achieved after injection of FFP. Our experience in this case also indicate that the incidence of delayed bleeding after surgery is low once hemostasis is successfully obtained during operation and there is no need to continue FFP infusion beyond day 2 postoperative.

Keywords: Laparoscopic, surgery, factor V deficiency, FFP transfusion

Introduction

FV deficiency (parahemophilia) is a rare bleeding disorder affects an estimated 1 in 1 million people [1]. Variety of symptoms related with disorder of coagulation could be obtained while the most dangerous complication is excessive bleeding following surgery, trauma, or childbirth. FFP is the only source of FV because FV concentrates is not available now. We present a case of cholangiolithiasis with FV deficiency that underwent laparoscopic cholecystectomy and common bile duct stone extraction (LC + LCBDE).

Case report

A 29-year-old male patient was admitted for complaints of pain in the right hyperchondrium, which was intermittent and colicky for duration of two years with intermittent fever. The patient had a history of FV deficiency. However, he experienced few bleeding problem in daily life and had history of dental extraction without complication. Physical examination of the

patient revealed nothing significant, with no evidence of jaundice. There was tenderness to palpation in the right upper quadrant of the abdomen, but no rebound tenderness was documented. Laboratory tests showed: white blood cells, $9.92 \times 10^9/L$; neutrophils, 57.6%; hemoglobin, 145 g/L; total bilirubin, 29.3 mol/L; aspartate aminotransferase, 148 U/L; alanine aminotransferase, 27 U/L; serum alkaline phosphatase, 382 U/L; and γ -glutamyl transpeptidase, 152 IU/L. MRCP (**Figure 1**) and ultrasonography showed multiple stones in the gallbladder and excessively dilated CBD. Admission TEG showed the patient is lack of coagulation factors (TEG CI -5.4), FV C: 2.4%. To evaluate the effect of FFP transfusion, a FFP test was carried out. After he administered 1000 mL (16.7 mL/kg) of FFP, TEG was in normal range (TEG CI -0.2). The change of coagulation parameters (PT and APTT) was show in **Figure 2**.

Depending on this finding, we decided to do LC + LCBDE. Another 1000 ml of FFP was administered 6 hours before operation. Preoperative PT and APTT was 12.8 and 34.7 respectively

Successful laparoscopic CBD exploration in a patient with FV deficiency

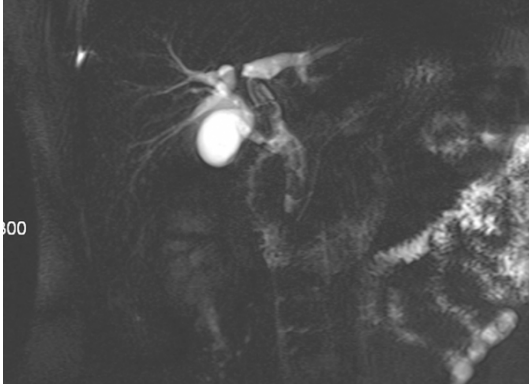


Figure 1. MRCP show there are multiple stones in common bile duct.

(**Figure 1**). LC + LCBDE was performed. The operation went smoothly, there was nearly no bleeding during operation. After confirming the CBD was free of stone via choledochoscope, a T-tube was inserted into the CBD. The patient received another 1000 mL of FFP the first day after operation. The PT and APTT were 14.7 and 36.6 respectively at the first day postoperative and 15.7 and 40.0 at the second day postoperative (**Figure 2**).

He had an uneventful postoperative course. No postoperative morbidity such as bleeding or bile leakage was documented. The peritoneal drainage was removed the third day postoperative. The cholangiogram via T-tube was performed 1 week postoperative and no residue stone was found (**Figure 3**). He was discharged after the cholangiogram and the T-tube was removed 4 weeks after operation.

Discussion

FV deficiency was first described in 1947 [2]. It was rare which was usually caused by mutations in the F5 gene. F5 gene mutations prevent the production of a functional protein called coagulation FV, which was a protein present in the human plasma, as well as in platelets bound to the binding protein multimerin. People with this disease usually had less than 10 percent of normal levels of FV in their blood. The signs and symptoms of this condition could begin at any age with a variety of clinical characteristics caused by disorder of coagulation [3]. Three most frequent bleeding sites were the gastrointestinal tract (38%), genitourinary tract (34%) and the surgical site

(18%), which could be life-threatening [4]. Biological profile usually included prolonged PT and APTT. As few publications dealt with laparoscopic surgery in patients with FV deficiency, we report herein the perioperative management of a severe FV deficient patient undergoing laparoscopic common bile duct exploration, and to our knowledge it is the first publication detailing management of a laparoscopic surgery in the setting of FV deficiency.

The goal of hematologic management for patients with FV deficiency in the perioperative period is to maintain sufficient levels of FV, which is thought to be 25% to 33%, for a sufficient period of time to prevent bleeding complications [5]. PT and APTT results are the main guidance to the treatments including surgery because there is no rapid FV assay available now. Here we suggest for those patients, FFP test is important indicator for the safety of surgery. The coagulation profile of our patient became normal after FFP test, which indicate it is safe to perform the operation.

Currently, there is no defined protocol of these patients. The half-life of FV is 12-36 hours. Tanis et al. treated a 12-year-old boy with congenital FV deficiency with continuous FFP infusion (750 ml/day for 10 days postoperatively) during uncomplicated surgery for a pseudotumor [6]. Mapp et al. treated a mitral valve regurgitation patient with Approximately 20 mL/kg, 10 mL/kg and 10 mL/kg FFP pre- and 8, 16 hours post-operative respectively [7]. Chava et al. operated a splenectomy and proximal lienorenal shunt with FV deficiency with 1500 ml of FFP the day before operation, 900 ml of FFP over 3 hours just before the operation, 600 ml of FFP every 12 hours for the first 48 hours followed by 450 ml of FFP every 12 hours for the next 48 hours postoperative [8]. Yotsumoto et al. reported quadruple coronary artery bypass in a FV deficient patient. Their strategy included pre- and intraoperative FFP transfusion which was continued until postoperative day 3 [9]. There is no consensus about the discontinuation of FFP infusion in the postoperative period. Most authors recommend continuing FFP transfusion for 3 days or more postoperatively [6, 8, 9]. In our case, the patient received FFP transfusion 16.7 mL/kg 6 hours preoperatively and the first day postoperative respectively. The results of coagulation profile and outcomes

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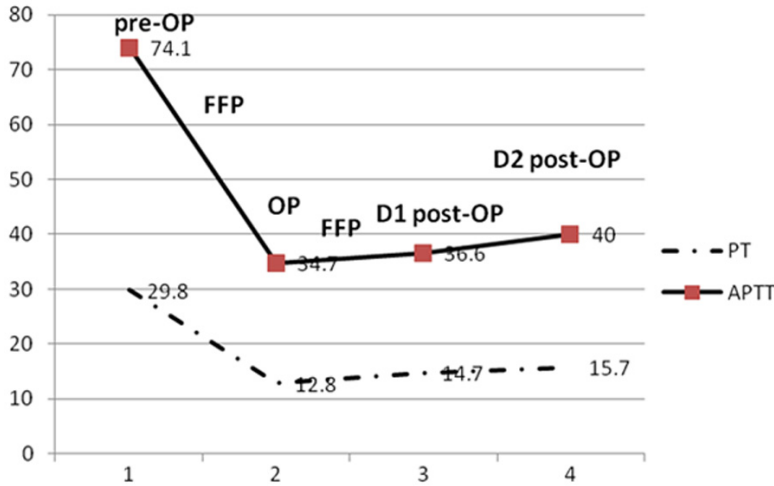


Figure 2. Coagulation parameters (PT and APTT) with time. OP: operation, D1: day 1, D2: day 2.

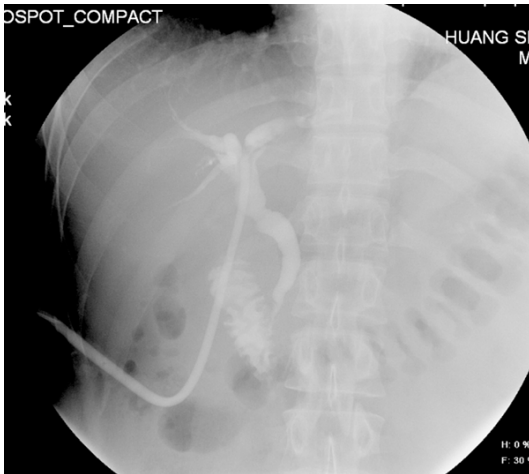


Figure 3. Postoperative cholangiogram via T-tube proved there is no residue stone in common bile duct.

in the perioperative period indicate that it is enough for hemostasis. Our results suggest that FFP transfusion (15-20 mL/kg) for 1 day postoperative is enough for minimal invasive surgery if the FFP test shows favorable results.

Conclusion

We surmise that laparoscopic surgery in a FV deficient patient can be performed safely if normal coagulation profile (PT and APTT) is achieved in the peri-operative period after injection of FFP (FFP test). FFP test is important for those FV deficiency patients who going to have surgery. In our opinion, the PT and APTT

level were recommended to be normal by loading with 15-20 ml/kg of FFP for surgery. Our experience in this case also indicated that the incidence of delayed bleeding after surgery appeared to be low once hemostasis was successfully obtained during operation. We felt that there is no need to continue FFP infusion beyond day 2 postoperative.

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