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Continuous infusion of human prothrombin complex in a patient with congenital factor VII deficiency undergoing laparoscopic cholecystectomy: A case report from China



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ABSTRACT

INTRODUCTION: Factor VII deficiency is a rare cause of haemorrhagic syndrome. **PRESENTATION OF CASE:** The authors describe a case of a 48 years old patient with congenital factor VII deficiency suffering abdominal discomfort diagnosed as gallstone, that successfully underwent laparoscopic cholecystectomy with continuous infusion of Human Prothrombin Complex (PPSB) around the procedure.

CONCLUSION: The usage of PPSB solved the clotting problems enabling the surgical procedure, without risks for the patient.

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1. Introduction

Factor VII (FVII) is a vitamin K-dependent clotting factor that is part of the extrinsic pathway of blood coagulation [1]. Congenital FVII deficiency is a rare bleeding disorder. In patients with congenital FVII deficiency, bleeding manifestations and clinical findings vary widely, ranging from asymptomatic subjects to life threatening bleeding [1–3]. However, severe and life-threatening hemorrhaging is rare in general (about 5%) and occurs most frequently during the first six months of life [1,2]. Treatment has traditionally involved FVII replacement therapy using fresh frozen plasma, prothrombin complex concentrates or plasma FVII concentrates [2,3]. Intravenous administration of recombinant FVIIa is now widely used for the treatment of FVII deficiency.

As FVII deficiency is a rare disease, experience with surgery in FVII-deficient patients is limited. There have been some reports describing the use of recombinant activated FVII (rFVIIa) during surgery in patients with congenital FVII deficiency [4–6]. Human Prothrombin Complex (PPSB) [7] contains coagulation factors II, VII, IX, X, so we thought it could also be used as a substitute for factor VII deficiency patients.

In this report, we describe a case of successful replacement treatment with PPSB, without episodes of bleeding and thrombosis. This

is the first report of the patient with congenital FVII deficiency undergoing laparoscopic cholecystectomy with repeated administration of PPSB.

2. Case report

The patient was a 48-year-old man (170 cm, 70 kg), he came to our hospital suffering abdominal discomfort. After examination of abdominal ultrasound and CT scan, gallbladder stone was confirmed. His physical examination was negative. He had no significant past medical history, he had never experienced a significant bleeding episode. His family history was unremarkable.

We discussed treatment options with the patient, and planned to perform the laparoscopic cholecystectomy. During pre-surgical preparation, there were unexpected blood coagulation test findings. The prothrombin time (PT) was 32.1 s (reference range, 10.0–16.0 s) and the activated partial thromboplastin time (APTT) was 26.5 s (reference range, 27.2–41.0 s). Other blood test results, including those for liver and renal function, total protein were all within normal range, and test for hepatitis B and C was negative. As the PT was abnormal, while other tests were basically normal, the consulting hematologist recommended more tests to confirm the diagnosis. FVII activity (FVII:C) was 2.7%, with no FVII inhibitor activity, so he was diagnosed with congenital FVII deficiency.

We worked out a plan to inject PPSB perioperatively, according to the instruction for PPSB [7]. We first administered PPSB 1800U (20–25U/kg [7]) intravenously before the surgery, and checked PT and FVII:C. At that time, PT was 11.8 s, FVII:C was 51.4%, within normal range and hematologist thought that was enough, so we started

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Table 1

Parameters of coagulation function during perioperative period.

Date	2015–5–25	2015–6–5	2015–6–9 day of surgery	2015–6–11
PT (s)	32.1	29.5	11.8	16.4
APTT (s)	26.5	23.2	24.6	25.4
FDP (ug/mL)	2.5	3.92	3.23	2.52
D-dimer (mg/L)	0.2	0.1	0.1	2.4
INR	2.64	2.54	1.03	1.44
PLT (10 ⁹ /L)	138	234		215
FVII:C	2.7	3.2	51.4	

the operation. Laparoscopic cholecystectomy proceeded smoothly and took 20 min with a total blood loss of 30 mL 900 U PPSB was administered intravenously every six hours [7] postoperatively for the first 48 h, cause the half-time of factor VII is 6–8 h [2,3]. PT, APTT, and levels of fibrin degradation product (FDP) and D-dimer during the clinical course are summarized (Table 1).

The patient was recovering well, and discharged from the hospital two days after surgery without episodes of bleeding and thrombosis. He is satisfied, and followed up by the surgeons at our hospital.

3. Conclusion

Here we described a very rare case of a FVII-deficient patient requiring PPSB for the laparoscopic cholecystectomy. Unfortunately, the optimal dose and method of administration for PPSB has not been established for various surgeries due to its rarity. The accumulation of cases of this disease in various fields is needed to determine the most appropriate replacement therapy.

Conflict of interest statement

None declared. All of the authors stated that they had no interests which might be perceived as posing a conflict or bias.

Disclosures

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. Our case report is compliant with the SCARE Guidelines [8].

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Ethical approval

Approval has been given, ethics committee of Luwan Branch, Ruijin Hospital, Shanghai Jiaotong University School of Medicine, Number 252675.

Consent

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Author contribution

Ru Zhou: study concept, data collection, writing the paper.
Qiaofeng Chen: study design.
Xunbo Huang: Clinical treatment.
Mingliang Wang: Clinical treatment.

Guarantor

Ru Zhou, Qiaofeng Chen.

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