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Functional medicine

Prostate surgery in severe congenital factor VII deficiency: A case report

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Introduction

Congenital factor VII (FVII) deficiency is an autosomal recessive disorder with an estimated prevalence of 1/500,000 individuals without ethnic or gender predilection, the gene for factor VII is found on chromosome 13, adjacent to the gene for factor X.[1] Factor VII deficiency can cause bleeding particularly in cases where factor VII is extremely low, but a few cases where factor VII function is lacking entirely or sub-totally may not present with a history of bleeding. Patients with congenital Factor VII deficiency who require surgery can be treated efficiently and safely with rFVIIa or antifibrinolytic agents. Most reports have recommended replacement therapy for patients with congenital factor VII deficiency that must undergo surgery.[2] It is generally believed that substitution therapy should be administered to patients undergoing surgery and having FVII activity of less than 10% and to patients with a history of recurrent bleeding.

Case report

A 68-year old heavy smoker man diagnosed at the age of 4 years as having congenital Factor VII deficiency presented to the emergency department for acute urinary retention. History goes back to a few years ago when he started to complained of severe frequency, urgency and nocturia. He was diagnosed with benign prostatic hyperplasia and was accordingly given medical treatment with no further improvement, he also reported more than 20 visits to the emergency department complaining of urinary retention during the last year and underwent catheterization several times.

He consulted his urologist who refused to proceed with surgical treatment for BPH due to his hematologic condition. Examination revealed normal blood pressure, respiratory rate, temperature and heart rate of 105, Oxygen saturations were 93% on room air. Examination of the heart, chest and, abdomen was normal except for palpable bladder in the lower abdomen. A Foley catheter was inserted after several attempts that resulted in severe hematuria. Initial blood tests included a white blood cell count of 6×10^9/L, Hb level 10g/dl, platelet count of 220*109/L (normal range, 149–319*109/L), Coagulation studies showed prothrombin time PT-INR of 4.9 and an activated partial thromboplastin time (aPTT) of 25.8 sec (normal, 22–33 sec), factor VII level was 2% of normal.

Ultrasound of the prostate was done and it showed a prostate gland of 100g with post void residue of 200ml (Fig. 1). The patient was admitted to the hospital and an initial 25 μg/kg dose bolus infusion of rFVIIa was given immediately before starting surgery.

A Greenlight Laser/180w Prostatectomy was done (Fig. 2) and it took 1h 20min.After 3 hours of administration of rFVIIa, FVII activity was 210% of normal and PT-INR was normalized to 0.8. During surgery, no bleeding problems were noted and postoperatively the urine was pinky. Postoperative day 1 the urine turned bloody, PT-INR was 2.8 and, FVII activity was 3% Therefore, rFVIIa was repeatedly administered (15μg/kg) every 8 h until POD 3 until urine is clear, Foley was removed and the patient was discharged on postoperative day 3.

Discussion

Factor VII deficiency presents with a wide spectrum of symptom severity that sometimes correlates poorly with FVII levels, a number of patients with undetectable FVII being totally asymptomatic. On the other hand, Intracranial bleeding was reported being frequent and severe after birth in a series of FVII-deficient patients.[1]

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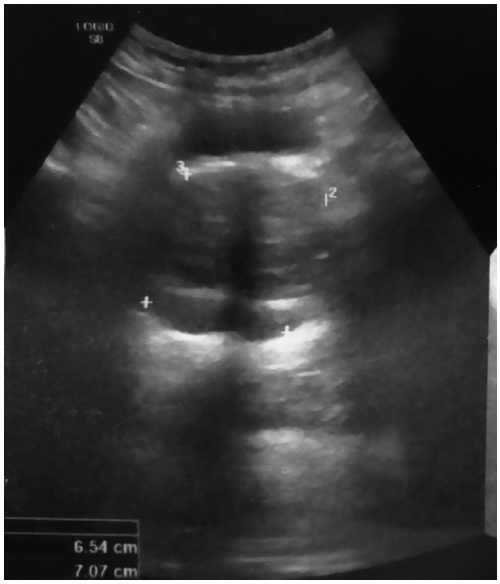

Fig. 1. Ultrasound of prostate showing enlarged prostate gland with estimated volume of 100g.

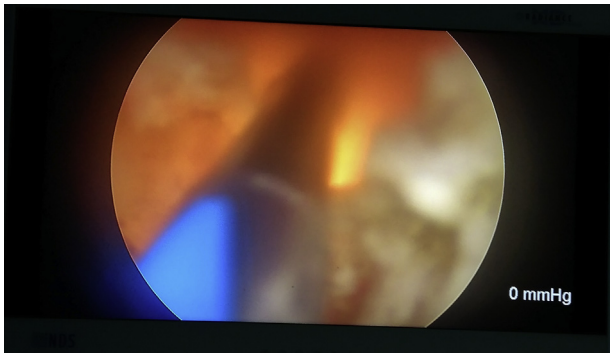

Fig. 2. GreenLight vaporization of the prostate.

A recent retrospective study has suggested that FVII activity of less than 10% is a risk factor for bleeding complications associated with a surgical procedure, in this study one prostatectomy and one transurethral resection of the prostate for cancer were performed on two asymptomatic patients with FVII:C levels of 8% and 12% respectively, none of these patients experienced excessive bleeding[3] the number of patients was too small to have any conclusions. For those surgical procedures that carry a high risk of bleeding several studies recommend using FVII levels of 10% as a threshold in combination with a full clinical history to determine the need for replacement therapy.[3]

However, after a review of 13 cases by Yorke and Mant, it was suggested that routine preoperative clotting factor replacement may not be required in congenital factor VII deficiency. It is proposed that replacement therapy should be available for use if required, but its routine preoperative use is probably unnecessary in this disorder as more than 50% of these 13 cases were considered a minor nonaggressive operation.[2]

In our case, since preoperative FVII activity and PT-INR were 2% and 4.9 respectively, we planned to give FVII replacement as rFVIIa to increase plasma FVII.

Administration of rFVIIa has become the most acceptable option for patients, with congenital FVII deficiency, which underwent surgical procedures.

Factor VII deficiency requires substitution therapy with only relatively small amounts of rFVIIa. The recommended dose of 15–30 $\mu g\,kg^{-1}$ every 4–6h until homeostasis.[4] At least 10% of FVII activity would be needed for homeostasis.[3] Accordingly, in our case bolus administration of 15 $\mu g\,kg^{-1}$ of rFVIIa was sufficient to maintain FVII activity above 10%, and additional administration of rFVIIa was needed after surgery when bleeding occur. Several cases of thromboses in relation to the use of rFVIIa have been reported, close monitoring for signs and symptoms of thrombotic events is warranted in all patients who were treated with rFVIIa (or any pro-hemostatic agent), particularly in the case of elderly and any other patients with concomitant conditions and/or predisposing risk factors for thrombosis.[5]

Conclusion

Inherited factor VII deficiency is a rare autosomal disorder. Patients with congenital FVII deficiency who require surgery can be treated efficiently and safely using rFVIIa. There are no guidelines for hemostatic control during Urologic surgery in patients with congenital factor VII deficiency. The use of replacement therapy is indicated in those patients who are planned to undergo an aggressive urological procedure with a high risk of bleeding. rFVIIa achieved hemostatic control, as shown by our case.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2018.09.024.

References

1. Mannucci PM, Duga S, Peyvandi F. Recessively inherited coagulation disorders. *Blood.* 2004;104:1243–1252.
2. Yorke AJ, Mant MJ. Factor VII deficiency and surgery: is preoperative replacement therapy necessary? *J Am Med Assoc.* 1977;238:424–425.
3. Benlakhal F, Mura T, Schved JF, et al. A retrospective analysis of 157 surgical procedures performed without replacement therapy in 83 unrelated factor VII-deficient patients. *J Thromb Haemostasis.* 2011;9:1149–1156.
4. Mariani G, Konkle BA, Ingerslev J. Congenital factor VII deficiency: therapy with recombinant activated factor VII—a critical appraisal. *Haemophilia.* 2006;12:19–27.
5. Neufeld EJ, Negrier C, Arkhammar P, et al. Safety update on the use of recombinant activated factor VII in approved indications. *Blood Rev.* 2015;29(Suppl 1):S34–S41.