Journal of Anesthesiology
2013; 1(3): 24-26
Published online November 20, 2013 (http://www.sciencepublishinggroup.com/j/ja)
doi: 10.11648/j.ja.20130103.12

Congenital deficiency in factor VII of the coagulation and the anesthesia

Hicham Bakkali[1, *], Salahedine Massou[1], Fayçal Labrini[2], Khalil Mounir[1], Mustapha Bensghir[1], Hicham Azendour[1], Hicham Balkhi[1], Charqui Haimeur[1]

[1]Department of Anesthesiology and Critical Care, Military Training Hospital Med V, Rabat, Morocco
[2]Immuno-hematology department, Military Training Hospital Med V, Rabat, Morocco

Email address:

hbakkali@ymail.com(H. Bakkali), salah_masson80@hotmail.com(S. Massou), f.labrini@hotmail.fr(F. Labrini), mustaphabens_15rea@hotmail.com(M. Bensghir), hazendour@gmail.com(H. Azendour), balkhih@hotmail.com(H. Balkhi), drhaimeur@hotmail.com(C. Haimeur)

To cite this article:

Hicham Bakkali, Salahedine Massou, Fayçal Labrini, Khalil Mounir, Mustapha Bensghir, Hicham Azendour, Hicham Balkhi, Charqui Haimeur. Congenital Deficiency in Factor VII of the Coagulation and the Anesthesia. *Journal of Anesthesiology.* Vol. 1, No. 3, 2013, pp. 24-26. doi: 10.11648/j.ja.20130103.12

Abstract: The constitutional deficiency in factor VII (FVII) of the coagulation is a rare autosomal recessive disease, responsible for a hemorrhagic syndrome of variable intensity poorly correlated with plasma levels of FVII. This deficit is suspected in front of an isolated reduction in the rate of prothrombine and confirmed by the dosage of this factor. We report a case of congenital FVII deficiency in a patient aged 22 years, who underwent surgical treatment of nephrolithiasis. FVII deficiency was 22%, no replacement therapy was introduced to the patient, and no bleeding complications were observed in intraoperative or postoperative. In conclusion, the perioperative transfusion with fresh frozen plasma (FFP) or the contribution of FVII in a patient with a moderate deficit in FVII is not systematic, and it cannot be considered if there is a risk of bleeding or in case of deficiency of FVII.

Keywords: Anesthesia, Coagulation, Congenital Deficiency, Factor VII, Hemorrhage

1. Introduction

Factor VII deficiency is a congenital coagulation abnormality. It is exceptionally often acquired in connection with a tumor [1]. The hereditary deficit is transmitted through autosomal recessive mode with high penetrance. Its incidence is 1/500.000 [2]. The hemorrhagic risk during the surgical intervention is not correlated with the factor VII rate. So, the replacement therapy of this deficit is not systematic. We report a case of Factor VII congenital deficiency in a patient who underwent surgical treatment of kidney stones and who did not get any replacement therapy.

2. Observation

It refers to a patient, aged 22 years, addressed in pre-anesthetic consultation for preoperative assessment before surgical treatment of left kidney stones. The interrogation of the patient had revealed a history of isolated left renal colic, no notion of surgery before, no drug taking and a regular menstrual cycle without evidence of hemorrhage. Examination under anesthetic was without any particularity. The preoperative assessment has revealed a prothrombine rate (PR) of 47% and normal Activated Cephaline Time (ACT) of 36s/32s. Platelets rate was normal and renal assessment was without abnormalities. A hemostatic control was done and the PR was always down, 48%. A dosage of coagulation factors was requested and showed an isolated deficit in factor VII, 22%. The hypothesis of a congenital factor VII deficiency has been retained in this patient; in the absence of infectious syndrome, evolutionary tumor, system disease and the low rate of Factor VII(research of autoantibodies has not been made).Considering the moderate deficit in Factor VII, and considering the low hemorrhagic risk of surgery and the absence of hemorrhagic history, there was no setting of replacement therapy in the patient and no hemorrhagic complication was observed in per or postoperative. The postoperative PR control was 45% and the Factor VII was 24%.

3. Discussion

FVII is a dependent vitamin k factor, synthesized by hepatocytes. It is present in the plasma with a weak concentration (0.5g/l) [3]; its half-life is very short: 3 or 4 hours. The acquired deficit is frequent in different circumstances: Hepatic insufficiency, vitamin k deficiency or the setting of treatment with vitamin K antagonists. The acquired deficits associated with bone marrow transplantation either to antibodies (immunological and lymphoproferative diseases) or accrued during severe bacteremic episodes are anecdotal. The constitutional deficit FVII is a rare autosomal recessive disease; its prevalence is estimated at 1/500 000 in general population [2]. It reaches both sexes in general. The quantitative, qualitative, or mixed deficits are differentiated in correlation with activity coagulant Factor VII (VIIc) in one hand and antigenic concentration (VIIAg) in the other hand. Clinically, the hemorrhagic manifestations are similar to the ones of the hemophilia [4]. Hemorrhagic syndrome associated with constitutional deficits in FVII is extremely variable. It is poorly correlated with the biological phenotype particularly on FVIIc activity [5]. We distinguish in homozygous, whose Factor VII rate is less than 10%, three levels of gravitates: the severe form when the Factor VII is less than 3%, the moderate form refers to Factor VII, starting from 3 to 5%, and the mild form, when it is greater than 5%. The heterozygosis whose Factor VII is between 20% and 60%, corresponds to minor form of the disease [10]. It could therefore not be direct relationship between Factor VII rate and surgical bleeding. In addition to hemorrhagic manifestations, some cases of spontaneous thrombosis have paradoxically been reported in this category of patients [6, 7]. The Factor VII deficit increases the hemorrhagic risk during the surgical intervention although its importance is not always correlated with plasma concentration [8]. On a therapy level, prevention of hemorrhagic risk during the surgical operation is based on administration of Factor VII [9]. However, this replacement therapy is not systematic except in case of severe or moderate spontaneous hemorrhagic syndrome history, or in case of hemorrhagic surgery. The replacement therapy needs frequent iterative injections of the short half-life of Factor VII (4 hours). Other authors suggested continuous perfusion [11]. The necessary minimum rate of Factor VII for surgical hemostasis would be at about 10 to 15 % [12]. The number of Factor VII to administer is between 20 and 40 IU/kg according to duration of the intervention (1ml of Factor VII corresponds to 25 IU). Another recommended therapeutic alternative is the administration of frozen plasma (FP)(1ml of FP contains 1unit of Factor VII) or by iterative injections, or by continuous perfusion(11). The replacement therapy is essential in case of moderate or severe hemorrhagic history, the dose to administer is between 20 and 30 IU/kg if it is of short and little hemorrhagic intervention and 40 IU/kg every eight hours, in the other cases as long as the hemorrhagic risk is still there.The verification of the efficiency is thanks to biological assays of FVII. See Quick time every two hours till the twelfth hour, then, every four hours till the twenty-fourth hour in order to maintain FVII rate about 10 to 15% and PR of 35%.

In our case, Factor VII deficit is very moderate (22%) in a patient without hemorrhagic history and who underwent some hemorrhagic surgery. Consequently, no replacement therapy was administered to that patient. No hemorrhagic or thrombotic event has been noted in postoperative and postoperative control of FVII was 24%.

4. Conclusion

FVII congenital deficit is a rare hereditary hemorrhagic disease. Clinically, the hemorrhagic manifestations are similar to the ones of the hemophilia. The hemorrhagic risk after one surgery is hardly (with difficulty) predictable and perioperative transfusion by FVII or by frozen plasma to a patient presenting a moderate FVII deficit is not systematic.

Conflict of Interest

No conflict of interest to be declared by the authors

References

[1] Meaudre E, Kenane N, Kaiser E, Gaillard PE, Saillol A, Cantais E, Palmier B. Isolated acquired factor VII deficiency in patient with severe head trauma: use of factor VII (factor VII-LFB). Ann Fr Anesth Reanim 2005;24:1383–6.

[2] Ragni MV, Lewis JH, Spero JA, Hasiba U. Factor VII deficiency. Am J Hematol 1981; 10: 79-88.

[3] Perry DJ. Factor VII deficiency. Br J haematol 2002;118 (3):689-700.

[4] Doran SE, Henry TR, Bockenstedt PL, Ross DA. Uncomplicated stereotactic and open surgical procedures in patients with factor VII deficiency. Surg Neurol 1994 ; 42 : 79 – 82.

[5] Mathilde HB, Guillin MC, Kenneth AB. Les déficits constitutionnels en facteur VII de la coagulation et les mécanismes moléculaires qui en sont responsables. Hématologie 1999; 5: 199-205.

[6] Park HK, Horowitz M, Jungreis C, Genevro J, Koebbe C, Levy E, Kassam E. Periprocedural morbidity and mortality associated with endovascular treatment of intracranial anevrysms. AJNR Am J Neuroradiol 2005; 26: 506–14.

[7] Norback O, Gal G, Johansson M, Solander S, Tovi M, Persson L, Ronne-Engström E, Enblad P. The establishment of endovascular aneurysm coiling at a neurovascular unit: report of experience during early years. Neuroradiology 2005; 47: 144–52.

[8] Ranchère JY, Ménart C, Lienhart A, Attali O. Factor VII deficiency and surgery. Ann Fr Anesth Réanim 1999; 18 :772-775.

[9] Roberts HR, Monroe DM, Escobar MA. Current concepts of

hemostasis: implications for therapy. Anesthesiology 2004; 100:722– 30.

[10] Saint-Raymond S, Greffe B, Carré J, Pujante C, Goguel A . Practical approaches for surgical procedures in congenital factor VII deficiency. Ann Fr Anesth Réanim.1989, 8 :518-521.

[11] Muleo G, Santaro R, Iannacaro PG, Papaleo P, Leo F. The use of recombinant activated factor VII in congenital and acquired factor VII deficiency. Blood Coag Fibrinolysis 1998; 9: 389-90.

[12] Bolton-Maggs PH, Perry DJ, Chalmers EA, Parapia LA, Wilde JT, Williams MD, Collins PW, Kitchen S, Dolan G, Mumford AD. The rare coagulation disorders--review with guidelines for management from the United Kingdom Haemophilia Centre Doctors' Organisation. Haemophilia 2004; 10:593–628.