Transfusion Medicine and Hemotherapy

Case Report · Kasuistik

Transfus Med Hemother 2009;36:138–139
DOI: 10.1159/000205405

Received: August 15, 2007
Accepted: September 24, 2008
Published online: March 6, 2009

Successful Use of Factor VIII Concentrate and Fresh Frozen Plasma for Four Dental Extractions in an Individual with Combined Factor V and VIII Deficiency

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Key Words
Combined factor V and VIII deficiency · Dental extraction · Factor VIII concentrate · Fresh frozen plasma

Summary
Background: Combined factor V and VIII deficiency (CF5F8D) is a rare autosomal recessive disorder, with an estimated prevalence of about 1:100,000 in the Jewish population. Affected individuals have between 5 and 30% of normal levels of factor V and VIII, whereas the levels of other plasma proteins are not altered. This bleeding disorder has been treated by replacement therapy with plasma infusion and FVIII concentrate as source of factor V and VIII, respectively, depending on the severity of the haemorrhage. **Case Report:** The individual with CF5F8D was a 22-year-old man with APTT of 51 s (control 33 s), PT of 27 s (control 12 s), factor V of 13%, and factor VIII of 21%. He had four untreatable carious teeth, including the maxillary first premolars and the mandibular first and second right molar. Factor VIII concentrate infusion began 12 h before the first dental extraction procedure and was continued in 8 h intervals. Moreover, 10 ml/kg of fresh frozen plasma were given for compensating factor V deficiency. No remarkable haemorrhage occurred during and after the procedures. The levels of factor V and VIII post procedures were 64 and 75%, respectively. **Conclusion:** It seems that plasma levels of 60–75% for both factor V and VIII may be sufficient for major dental procedures.

Schlüsselwörter
Kombinierte Faktor-V/Faktor-VIII-Defizienz · Zahnextraktion · Faktor-VIII-Konzentrat · Frischgefrorenes Plasma

Zusammenfassung
Hintergrund: Die kombinierte Faktor-V/Faktor-VIII-Defizienz (CF5F8D) ist eine seltene autosomal rezessive Erbkrankheit mit einer Prävalenz von ca. 1:100 000 in der jüdischen Bevölkerung. Die betroffenen Personen haben nur 5–30% der normalen Faktor-V- und Faktor-VIII-Plasmakonzentrationen; andere Plasmaproteine zeigen normale Werte. Abhängig von der Schwere der Hämorrhagie wird diese Blutungsstörung mithilfe von Plasmainfusionen und Faktor-VIII-Konzentraten zur Kompensation des Faktor-V- und Faktor-VIII-Mangels behandelt. **Fallbericht:** Ein 22 Jahre alter Mann mit CF5F8D, bei dem vier kariöse Zähne (die ersten maxillären Prämolaren sowie der erste und zweite mandibuläre Molar) extrahiert werden mussten, wurde in unsere Klinik eingeliefert. Seine Blutwerte waren wie folgt: aPTT 51 s (normal 33 s), PT 27 s (normal 12 s), Faktor V 13% und Faktor VIII 21% der Normalwerte. Mit der Infusion von Faktor-VIII-Konzentrat wurde 12 h vor der ersten Zahnextraktion begonnen. Die Infusion wurde in 8-Stunden-Intervallen fortgesetzt. Darüber hinaus wurden 10 ml/kg frischgefrorenes Plasma zur Kompensation der Faktor-V-Defizienz appliziert. Während und nach den Eingriffen kam es zu keinen nennenswerten Blutungen. Die Plasmakonzentrationen für Faktor V bzw. VIII nach den Zahnextraktionen waren 64 bzw. 75%. **Schlussfolgerung:** Plasmakonzentrationen für Faktor V und VIII von 60–75% der Normalwerte scheinen ausreichend zu sein, um größere kieferchirurgische Eingriffe vornehmen zu können.

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Introduction

Combined factor V and VIII deficiency (CF5F8D, also known as Mendelian inheritance # 227310 in online databases) is a rare autosomal recessive haemorrhagic disorder that was first described by Oeri et al. [1] in 1954. Most reported families are from the Middle East. About 147 cases have been reported worldwide until May 2006 [2–6]. Affected individuals have plasma levels of factor V and VIII in the range of 5–30% of normal limits and show a moderate haemorrhagic tendency. The most common, frequent, spontaneous bleeding manifestations among individuals with CF5F8D include epistaxis, and the most frequent, traumatic bleeding symptoms are post-dental extraction bleeding, cut-related bleeding, and haemorrhage after circumcision in males. In the current survey, we report four dental extractions using factor VIII concentrate and fresh frozen plasma (FFP).

Case Report

The affected person was a 22-year-old member of a family in north-eastern Iran with three known cases of CF5F8D. The parents of the patient showed first-degree consanguinity. The patient's haemostatic levels were as follows: APTT 51 s (control 33 s), PT 27 s (control 12 s), factor V 13%, and factor VIII 21%. The other coagulation factor levels were unremarkable. No factor VIII inhibitors were detected. The patient had four untreatable carious teeth, including the maxillary first premolars and the mandibular first and second right molar.

On February 25, 2007, the patient was hospitalised, and factor VIII concentrate (Koate-DVI®; Bayer Corporation, Elkhart, IN, USA) was infused 12 h before the first dental extraction procedure. The infusion was continued with 8 h intervals. The affected individual weighed 54 kg and had no history of response to desmopressin. Thus, 10 ml/kg of FFP for compensating factor V deficiency were given [7] and 2,000 IU (40 IU/kg) factor VIII concentrate to raise factor VIII level over 90%. For minimal exposure to the direct blood product, two bags of FFP were infused half an hour before the extraction of the upper premolars on the 2nd day. After the procedure, factor VIII concentrate infusion was continued for 24 h with 20 IU/kg. Two further bags of FFP and 40 IU/kg of factor VIII concentrate were infused on the 3rd day when the two other carious teeth (mandibular first and second right molar) were extracted. Infusion of factor VIII was continued once again with 20 IU/kg 12 h after the procedure.

No remarkable haemorrhage occurred during and after the procedures. The levels of factor V and VIIII post procedures were 64 and 75%, respectively. In conclusion, there was no necessity to prescribe antifibrinolytic agents such as tranexamic acid due to perfect control of the haemorrhage. The patient was discharged on March 1, 2007.

Discussion

Inherited deficiency of factor V and VIII represents the most prevalent, combined coagulation defect [8]. It seems more common among Jews and Iranians, where consanguineous marriages are frequent [9]. To our knowledge, this is the first case with major dental extraction reported. We chose to prescribe a combination of factor VIII concentrate and FFP before dental extraction due to the pronounced bleeding tendency of the disease and the number of carious teeth, although plasma therapy by means of plasma exchange or FFP and desmopressin has been proposed elsewhere for other surgical procedures [10]. Takeuchi et al. [11] reported a patient's right mandibular and first molar extraction using transfusion of FFP combined with local haemostatic treatment.

Additional case coverage is necessary to establish valuable guidelines for haemorrhagic, invasive, and surgical procedures in individuals with combined factor V and VIII deficiency.

Disclosure

The authors declare no conflict of interest.

References

1 Oeri J, Matter M, Isenschmid H: Angeborener Mangel an Faktor V (Parahaemophilie) verbunden mit echter Haemophilie A bei zwei Brüdern. Med Probl Paediatr 1954;1:575–588.

2 Farah RA, de Moerloose P, Bouchardy I, Morris MA, Barakat W, Sayad AE, Neerman-Arbez M: Combined factor V – factor VIII deficiency (F5F8D): compound heterozygosity for two novel truncating mutations in LMAN1 in a consanguineous patient. Thromb Haemost 2006;5:893–895.

3 Louanchi L, El Fegoun Sorary B, Meriem B: Clinical aspects and management of rare hereditary hemostasis disorders. Haemophilia 2004;10(suppl 3): 127–131.

4 Mansouritorghabeh H, Rezaieyazdi Z, Pourfatholah AA: Combined factor V and VIII deficiency: a new family and their haemorrhagic manifestations. Haemophilia 2006;12:169–171.

5 Mohanty D, Goshi K, Shetty S, Menegatti M, Duga S, Peyvandi F, Mannucci PM: Genotype analysis of rare coagulation factor deficiency cases from India. Haemophilia 2004;10(suppl 3):127–131.

6 Pathare A, Al Kindi S, Al Abri Q, Al Haddabi H, Gravell D, Dennison D: Spectrum of inherited bleeding disorder from sultanate of Oman. Haemophilia 2004;10(suppl 3):127–131.

7 Sallah SA, Angchaisuksiri P, Roberts HR: Use of plasma exchange in hereditary deficiency of factor V and factor VIII. Am J Hematol 1996;52:229–230.

8 Peyvandi F, Tuddenham EG, Akhtari A, Lak M, Mannucci PM: Bleeding symptoms in 27 Iranian patients with the combined deficiency of factor V and VIII. Br J Haematol 1998;100:773–776.

9 Mansouritorghabeh H, Rezaieyazdi Z, Pourfatholah AA, Rezai J, Esmaili HA: Haemorrhagic symptoms in patients with combined factors V and VIII deficiency in north-eastern Iran. Haemophilia 2004;10:271–275.

10 Bauduer F, Guichandut JP, Ducout L: Successful use of fresh frozen plasma and desmopressin for transurethral prostatectomy in a French Basque with combined factor V + VIII deficiency. J Thromb Haemost 2004;4:675.

11 Takeuchi M, Shikimori M, Kaneda T: Dental extraction in a patient with congenital deficiencies of factors V and VIII. J Oral Maxillofac Surg 1984;42:327–330.