Extraction of four wisdom teeth in a patient with congenital factor V deficiency hemophilia

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Congenital factor V deficiency was first described by Owren in 1947.1 It is thought to be transmitted by an autosomal recessive gene (q23-24)2 found in 1 out of every 1 million population and usually with no gender or race correlation.3 To date, 150+ cases have been reported in the world literature.3 The description of rare case of this disease is justified, because they may add further information about the condition of the hemorrhagic tendency. The object of the present paper was to report the bleeding control for extraction of 4 wisdom teeth with congenital factor V deficiency hemophilia and review the literature. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;112:e1-e3)

The patient was a 37-year-old Japanese man diagnosed with congenital factor V (FV) deficiency hemophilia by internist at medical department of Nagasaki University who came to our clinic for extraction of wisdom teeth. No other main features usually presented by the patients have been onset without previous spontaneous bleeding from the gingiva. The FV level in the patient’s plasma was 1% of normal. His activated partial thromboplastin time (APTT) and prothrombin time (PT; international normalized ratio) were 20% of normal (Fig. 2). Other platelet and vascular tests were within normal limits. Bleeding tendency was usual throughout his life, and no hemarthrosis ever occurred.

Panoramic x-ray picture of the patient revealed the 4 wisdom teeth (Fig. 1, A). Extraction of 4 wisdom teeth carried out under local anesthesia (2% xylocaine 6 mL with 1:20,000 epinephrine) with fresh frozen plasma (FFP; 450 mL) intravenous transfusion (Fig. 1, B). Operation required 40 minutes and each socket was sutured. Bleeding from the sockets endured for 8 hours after the extraction and stopped the next morning (Fig. 1, C-F).

The plasma level of FV was increased from 1% to 11% during the transfusion of 450 mL FFP and decreased to 6% next morning. No more FFP was indicated in the patient after extraction, because oozing from the socket stopped. Laboratory reports revealed that APTT and PT were 20% of normal level at the before operation and corrected to 50% the next morning (Fig. 1).

Sutures were removed 3 weeks after the extraction, and no other complaint was reported.

Factor V deficiency (parahemophilia), thought to be an autosomal recessive inheritance, has been described in both genders and occurs without geographic or ethnic preference.4 Cui et al.5 reported that approximately one-half of homozygous mouse embryos deficient in FV (FV−/−) die at embryonic day 9-10. The remaining FV−/− mice progress normally to term, but die from massive hemorrhage within 2 hours of birth. These data suggested direct evidence for the existence of other critical hemostatic functions for thrombin in addition to fibrin clot formation, and they identified a previously unrecognized role for the coagulation system in early mammalian development.5

Clinical manifestations of FV deficiency are varied in type and severity. In later life, these are usually seen as mucous hemorrhages, most frequently epistaxis in men and menorrhagia in women.6 The present case was diagnosed after spontaneous bleeding from the gingiva. FV acts as a procoagulant cofactor; it plays an indirect role in fibrin formation by the formation of prothrombin, which is subsequently converted to thrombin, leading to the conversion of fibrinogen to fibrin. The plasma anticoagulant factors protein S and protein C, as part of this well balanced system, will then turn off FV, thus limiting the coagulation cascade. This limiting action reduces the risk of excessive clot formation.7

In FV deficiency, correlation between plasma and factor V levels and bleeding symptoms is not always

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consistent, and patients without significant manifestations have been reported.\(^6\) The relative contribution of plasma versus platelet FV to FVa (factor Y)–binding interactions in the prothrombinase complex are not clearly defined.\(^8\) Even though FV deficiency is a rare bleeding disorder, it follows the pathophysiologic complexity of defects in the coagulation pathways.\(^9\) Oral surgeons should keep in mind that prothrombotic

Fig. 1. A, Pre- and, B, Post-extraction panoramic x-rays of the patient. C-F, Sockets 1 day after the extractions. Bleeding from the wounds had already stopped (24 h).

Fig. 2. The patient’s value of clinical blood coagulation analysis before and after extractions (24 h). A, Changes of factor V before and after extraction. The level was increased 11% of normal during infusion of fresh frozen plasma (FFP; 450 mL), activated partial thromboplastin time (APTT) (B), prothrombin time (PT) (C), and PT–international normalized ratio (INR) (D), were estimated as 20% of normal level before extraction. The levels of APTT and PT corrected to 50% of normal after infusion of the FFP but still far from the normal level.
agents used preoperatively may increase the risk for acute myocardial infarction in the presence of the factor V Leiden mutation before a tooth extraction. Furthermore, single-chain FV acts as a cofactor contributing to the acceleration of inactivation of factor VIIIa by activated protein C–protein S complex (anticoagulant).

There are few reports dealing with the extraction at surgery levels involving deficiency of FV (parahemophilia). Extraction of 4 wisdom teeth were carried out at 1 time under the level of FV within 10% of normal levels during operation. The complicated bleeding diathesis that survived longer appeared to reflect platelet, rather than plasma, FV activity. These results suggest that platelet factor V is an essential component in maintaining stable and prolonged hemostasis after trauma. Because it appears that there is enough FV available on the unperturbed platelet surface to provide an optimal contribution to prothrombinase complex, assuming its maximal activation, these observations would suggest some inherent advantage of platelet-released FVa over that in the plasmatic environment of the platelet. Most likely, the recruitment and concentration of platelets to a site of vascular injury provides an excess of local platelet FVa receptor sites that require the release of the localize, stored platelet FV.

CONCLUSION
A patient whose coagulation FV decreased below 1% of normal level underwent extraction of the 4 wisdom teeth under transfusion of FFP (450 mL).

REFERENCES

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