

LETTER TO THE EDITOR

Successful use of recombinant FVIIa (Novoseven[®]) in the management of uncontrolled bleeding after emergency fasciotomy

T. SIPAHI* and A. KUYBULU†

*Pediatric Hematology Unit, Department of Pediatrics, Faculty of Medicine; and †Department of Pediatrics, Faculty of Medicine, Süleyman Demirel University, Isparta, Turkey

Dear Editor,

There are some reports about using recombinant factor VIIa (rFVIIa) in the management of different bleeding situations in children [1]. As per reports in the literature, rFVIIa concentrates were used in a very limited number of patients with congenital or acquired haemophilia who developed inhibitors.

In the beginning, rFVIIa was used for the treatment of bleeding associated with inhibitors to factor VIII and IX [1]. Recently, it has been used successfully in the treatment of paediatric coagulopathies and platelet function defects including inherited and acquired deficiencies of factor VII, Glanzmann's thrombasthenia and Bernard–Soulier syndrome [2].

In this report, we present a patient who developed a severe right forearm compartment syndrome that required emergent fasciotomy. He was treated with rFVIIa successfully.

A previously healthy 1.5-year-old boy was presented to our department with a 2-day history of swelling and ecchymoses of his right arm and elbow. His medical history revealed that occasionally he had been having ecchymoses on his extremities but had never been to a doctor. There was no known family history of bleeding disorders.

On admission, his physical examination revealed remarkable tachycardia, marked pallor, ecchymoses and severe swelling of his right forearm and right hand including all his fingers at the right side. He could not move his fingers and his right radial pulse could not be sensed.

His haemoglobin level was found to be 5.6 g dL⁻¹, haematocrit 18.2%, white blood cell count

14.7 × 10⁹ L⁻¹, platelet count 510 × 10⁹ L⁻¹, prothrombin time 14.6 s and, activated partial thromboplastin time 94.1 s. The other laboratory parameters were in the normal range. But other coagulation tests and factor levels were not studied, as they are not studied in our laboratory during the weekend under normal circumstances.

The patient developed compartment syndrome and emergent fasciotomy was required (Fig. 1). The peripheral circulation of the right forearm had severely decreased and if it continued like that, amputation would have been the only option. Fresh-frozen plasma with a dose of 20 mL kg⁻¹ and erythrocyte suspension were administered, urgently. Despite several packed red cell and fresh-frozen plasma transfusions, bleeding continued. Then we decided to administer rFVIIa for stopping the massive bleeding. Recombinant FVIIa (Novoseven[®]) was initiated at a dose of 30 µg kg⁻¹ every 2 h. After the second dose of rFVIIa,



Fig. 1. Patient with compartment syndrome requiring emergent fasciotomy.

Correspondence: Prof. Dr Tansu Sipahi, Angora Evleri, Kediseven Caddesi, No:92, Beysukent, Ümitköy, Ankara, Turkey.
Tel.: +90 50 58161444; fax: +90 31 24196677;
e-mail: tansusipahi@hotmail.com

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bleeding stopped. The next day, coagulation study results were obtained. Factor VIII, IX and XI levels were 4%, 70% and 69% (normal = 70–120%), respectively. Von Willebrand factor antigen, ristocetin cofactor activity, factor VII and factor XII levels of the patient were in the normal range. No antibody was detectable against factor VIII. Then was diagnosed as having haemophilia A, and recombinant FVIII replacement therapy was started. Homeostasis was maintained with repeated doses of recombinant factor VIII every 12 h. On the sixth day of hospitalization, skin graft was taken from his leg and applied to his forearm on fasciotomy areas. Replacement therapy with rFVIII was continued for 10 days. He was also treated with empiric intravenous antibiotics. He was discharged after complete healing. After 6 months of follow up, only a little scar residue remains on the fasciotomy area, and otherwise, he can use his right arm without any disability (Fig. 2).

Haemophilia A is a hereditary X-linked bleeding disorder caused by deficiency of coagulation factor VIII in plasma, occurring in one of 5000 males born. The clinical picture of the disease is related with plasma coagulation factor levels that define the degree of haemophilia as mild, moderate and severe (6–30%, 1–5% and <1%, respectively) [3]. The severity of the disease may influence the age at which it is diagnosed. Although our patient had some symptoms, he had never consulted a doctor.

Compartment syndrome is a very rare and life-threatening complication of an inherited bleeding disorder. For treatment of compartment syndrome, emergent fasciotomy is usually required. However, factor levels >50% is necessary at the time of process [3]. That he was a haemophiliac was unknown during his presentation. However, his medical his-



Fig. 2. After 6 months, the aspect of the forearm.

tory, physical examination and laboratory findings revealed an inherited congenital bleeding disorder, i.e. haemophilia. Moreover, clinical findings required an emergency fasciotomy. Although we administered packed red cell and fresh-frozen plasma many times, we could not control bleeding.

We administered two doses of rFVIIa, at a dosage of 30 $\mu\text{g kg}^{-1}$ and after the second dose of rFVIIa, the bleeding stopped and his haemoglobin increased. rFVIIa can be used at 35–70 $\mu\text{g kg}^{-1}$ per dose in every 2–3 h. But when it is used at a dose of 70–120 $\mu\text{g kg}^{-1}$ (especially 90 $\mu\text{g kg}^{-1}$), the efficacy rate was 92% [4,5]. We especially want to emphasize that NovoSeven can be used successfully in a bleeding patient without a confirmed diagnosis. Luckily, we controlled the massive bleeding without any side-effect at a lower dose than recommended.

In conclusion, rFVIIa is a universal and life-saving haemostatic agent and it might be used for unexpected refractory life-threatening haemorrhage, which has failed to respond to conventional therapy.

Disclosures

The authors stated that they had no interests which might be perceived as posing a conflict or bias.

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