

LETTER TO THE EDITOR

Successful control of bleeding during supracondylar amputation caused by severe compartment syndrome in patient with haemophilia A and high titre of inhibitor

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High titre of inhibitors in patients with haemophilia makes every surgical procedure complicated and potentially hazardous. Although, comprehensive care and bypassing agents are widely available, significant risk of uncontrolled bleeding still exists during surgical procedures and elective surgery needs firm justification [1–3]. Prolongation of life span and frequent significant comorbidity in patients with haemophilia and inhibitors increases the possibility of surgical emergencies. In such situations, a sufficient quantity of bypassing agents needs to be kept readily available. Compartment syndrome is a surgical emergency. The term compartment syndrome is referred to as the situation when growing haematoma increases pressure to vascular and neural structures in limited space (e.g. fascial space) causing irreversible necrosis.

Fifty-six-year-old patient with severe haemophilia A, with known high titre of inhibitor, hepatitis C virus infection and insulin independent diabetes mellitus, was admitted to hospital because of severe pains in the left leg. Pains started 2 weeks before admission to the hospital, but the patient hesitated to ask for medical care because of poor mobility. He had severe deformities of every large joint so he was chair-ridden. Two days before admission to the hospital, the pains became unbearable. The left limb, distal from the knee, was painful to the slightest touch, swollen and with signs of phlegmona. Ultrasonographic and radiological examination (Fig. 1) revealed large crural haematoma with calcification, which compressed vascular structures and caused severe compartment syndrome. Deformities of the knee and severe osteopenia could be seen, too.

Consultant vascular surgeon suggested supracondylar amputation. Inhibitor concentration in the recent months ranged from 30 BU to maximal 48 BU, immediately before operation. After patient's consent to the procedure, by-passing agent, activated recombinant factor VII (rFVIIa, NovoSeven amp.), was administered ($90 \mu\text{g kg}^{-1}$ of body weight) intravenously, as a bolus injection, 1 h before amputation. The procedure was completed without any complications. Tourniquet was not used and subfascial drain was inserted. The surgeon noticed that intraoperative blood loss was slight-to-moderate. Haemoglobin level dropped from 130 g L^{-1} to 90 g L^{-1} and patient received one unit of packed red cells during the first postoperative day. Postoperatively, rFVIIa was administered in doses of

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Anchor Name: surgery

[VJYA (Vijay A)]

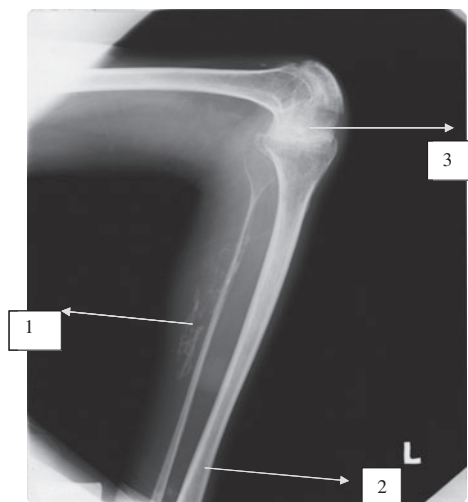


Fig. 1. 1 – encapsulated chronic haematoma with calcification 2 – osteopenia 3 – ankylated knee joint.

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Fig. 2. Postoperative day 1.



Fig. 3. Postoperative day 15.

90–180 $\mu\text{g kg}^{-1}$, every 2 h, as intravenous bolus. Oozing, while gradually decreasing, lasted for days (Fig. 2).

After 5 days, the intervals between rFVIIa bolus were extended to 3 h and then, gradually up to 6 h. Administration of rFVIIa extended up to 15 days. Although there is no test for monitoring the activity of rFVIIa, the aim was to keep prothrombin time under 10 s. Antifibrinolytic agent (tranexamic acid, 6–8 gr., orally, per day) was also included since postoperative day 1, as well as everyday wound toilette and dressing. The patient was discharged fully recovered (Fig. 3).

Discussion

There are several recorded cases of patients with haemophilia and high titre of inhibitor, who have undergone limb amputation [2,4,5]. To our knowledge, our haemophilia patient is unique. The patient had high concentration of inhibitor, diabetes mellitus and hepatitis C infection and undergone limb ampu-

tation because of compartment syndrome and phlegmona. During his treatment, only activated recombinant factor VII was used, in bolus injections, with an extension of intervals between them. It seems that the dose of 90 $\mu\text{g kg}^{-1}$ BW of rFVIIa was sufficient immediately after the operation, but not completely after 24 or 48 h postoperatively. That dosage needed to be elevated up to almost 200 $\mu\text{g kg}^{-1}$ BW. Maybe, the explanation for that could be local accumulation of fibrin degradation products. So, it would be advisable to start with higher doses of rFVIIa and then to decrease to 90 $\mu\text{g kg}^{-1}$ BW and to use antifibrinolytic agent. Although oozing from the wound lasted for days and healing was slower, there were not too many differences in comparison to other diabetic patients. Other authors [3,5] used combination of NovoSeven and FEIBATM or used continuous infusion of rFVIIa. Combination of tranexamic acid and rFVIIa did not produce thrombotic complication in our patient. It seemed to be effective and safe.

Disclosures

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

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