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CASE REPORT

Persistent surgical wound bleeding: A rare condition related to acquired hemophilia A

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Summary Acquired hemophilia A (AHA) is a rare condition that predisposes affected patients to a bleeding tendency, even after a trivial physical insult. We present our experience with a 45-year-old male patient who was referred to our institute because of persistent bleeding from a left forearm surgical wound after fasciotomy. He was diagnosed as having AHA. Surgical treatment in combination with recombinant activated factor VII (rFVIIa) led to a satisfactory result. Clinical awareness and multidisciplinary professional connections are necessary in the treatment of AHA. Acquired hemophilia should be considered in the differential diagnosis of patients with uncontrolled bleeding episodes.

1. Introduction

Acquired hemophilia A (AHA) is a condition in which inhibitor autoantibodies are produced against coagulation factor VIII.[1] It is a rare condition, but the associated morbidity and mortality rates are significant.[2] Furthermore, although autoantibodies for other coagulation factors may also develop, inhibitors of factor VIII are the

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most common.[2] The incidence of AHA is estimated between 1.34 and 1.48 cases per million individuals each year, and most (>85%) individuals with AHA are older than 60 years.[2] Some predisposing factors that have been reported for AHA are postpartum status, underlying autoimmune disease, and hematologic or solid cancer.[3] Approximately one-half of AHA patients have no obvious predisposing factors.[4] The most common predisposing conditions for AHA that have been reported are interestingly autoimmune disease and malignancy, which have prevalence rates of approximately 10% and 12%, respectively.[5] In this report we present a relatively young patient who had only a mild physical insult and a dramatic consequence.

2. Case report

A 45-year-old male patient had a medical history of underlying gouty arthritis and cervical spine stenosis that were surgically treated 5 years previously. He had a 2-month history of repeated episodes of ecchymosis around both forearms after minor contusions; however, these symptomatic episodes usually subsided spontaneously. Nothing of significance was noted after the evaluation of his clinical history, except that he had taken an unknown herbal medication for approximately 3 months prior to hospitalization. His current problem followed an insult to his left forearm, which was hit when playing with his son. Progressive swelling, pain, and ecchymosis developed the next day and did not subside. He was admitted to a local hospital for treatment.

Progressive numbness, increasing swelling, and persistent sharp pain occurred after hospitalization. In addition, weak radial arterial pulsation was also noted (this was documented in the referring summary, which was maintained among the medical records of our institute). Because of suspected compartment syndrome, he underwent emergency fasciotomy on day 2 of hospitalization. His preoperative international normalized ratio (INR) was 0.98, and his activated partial thromboplastin time (APTT) was more than 120 seconds (because of the emergency situation, the result of the prothrombin/APTT [PT/APTT] test had apparently been reported after he had been sent to the operating room). However, persistent postoperative bleeding was encountered at the surgical site. Additional attempts to achieve hemostasis produced no obvious benefit, and the patient's clinical condition deteriorated. On postoperative day (POD) 1, he had hypovolemic shock, changes in consciousness, and a hemoglobin level of 4.9 g/dL. Emergency resuscitation such as transfusion and fluid challenge were persistently maintained. On POD 2, the patient was transferred to our institute for further management.

At our emergency department (ED), his initial vital signs, including his level of consciousness, were documented as follows: body temperature of 36°C, pulse rate of 138 beats per minute, respiratory rate of 26 breaths per minute, systolic/diastolic blood pressure of 110/81 mmHg, and alert. As Fig. 1 shows, the left forearm has a large wound with diffuse oozing. Computed tomography was performed at our ED. Active arterial bleeding was noted at the elbow area (Fig. 2). The on-duty plastic surgeon was consulted immediately. However, no definite bleeding site could be identified for suture and ligation. Epinephrine gauze, elastic bandages, and a tourniquet system were all applied for hemostasis. The patient was immediately admitted to the intensive care unit (ICU) for close observation and correction of coagulopathy.

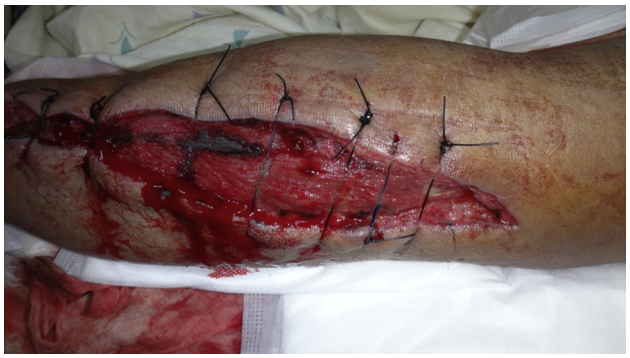

Figure 1 The left forearm on the third day after admission to our hospital. There were no obvious bleeders, but diffuse wound oozing is present.

During the ICU admission, repeated episodes of massive bleeding were noted, requiring multiple blood transfusions. In addition, bedside suture ligation was performed several times to control bleeding. Because of persistent bleeding with coagulopathy, we consulted a hematologist for further evaluation. The hematologist recommended several surveys to assess the coagulopathy, including surveys that evaluated mixing APTT, factor VIII, markers for autoimmune conditions, and possible malignancy.

On account of a prolonged mixing APTT (60.8 seconds) and a low factor VIII level (1.5%), the hematologist recommended a supplemental factor VIII prescription and measurement of factor VIII inhibitor titers. However, even after supplementation of factor VIII, diffuse oozing persisted. The factor VIII inhibitor titer was later reported to be 23.52 Bethesda units (BU). A diagnosis of AHA was established because of the high titer of factor VIII inhibitor and low concentration of factor VIII. On POD 8, recombinant activated factor VII (rFVIIa; NovoSeven RT; Novo Nordisk Inc., West, Princeton, NJ, USA) was prescribed (using 1-mg vials) to activate the extrinsic hemostasis pathway, and prednisolone and cyclophosphamide were administered to suppress factor VIII inhibitors.

Recombinant activated factor VII was administered by intravenous infusion with seven vials every 2 hours for three doses and was then changed to three vials every 6 hours for 2 days. After completing the first treatment cycle, mild oozing of the wound persisted. We then initiated another cycle of rFVIIa treatment, which was completed on POD 16. The patient's later hospital course was uneven (he had an episode of recurrent bleeding), however, he was fully recovered on POD 31 and was discharged with oral immunosuppressant agents. Fig. 3 summarizes the serial hematological data and his coagulation profile during the hospital stay.

This patient was then followed-up at the outpatient departments of the trauma surgery and hematology departments. Three months after the hospital discharge, the factor VIII level had recovered up to 48% (the reference is above 50%) and the immunosuppressant therapy was discontinued.