

A Case of von Willebrand Disease Discovered during Treatment of a Sacral Pressure Ulcer

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Abstract

A sacral pressure ulcer developed in a patient hospitalized for cerebral infarction. Each time necrotic tissue was debrided from the ulcer, pressure hemostasis was necessary to stop the bleeding. As treatment continued, the pressure required to stop the bleeding caused the ulcer to worsen, leading to a downward spiral in the patient's condition. While trying to determine the cause of this problem, we discovered that the patient had von Willebrand disease. Medication controlled the bleeding, and the pressure ulcer began to heal at the same time. It was clear to us that conservative treatment would lead to a complete cure but that the healing process would take a long time and require continued administration of an expensive drug. We decided, therefore, to close the wound with a fasciocutaneous flap so that the patient could be quickly transferred to a rehabilitation hospital. About 1 month after surgery, epithelialization was complete, we were able to discontinue medication, and the patient was discharged. This experience demonstrates the importance of determining the cause of any deviation from the normal course of healing in pressure ulcers. It also indicates that the use of fasciocutaneous flaps, which involve little intraoperative bleeding in short surgeries, is appropriate in cases like this one.

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Key words: pressure ulcer, von Willebrand disease, bleeding tendency, fasciocutaneous flap

Introduction

We treated a patient with a sacral pressure ulcer which deviated from the regular healing course. A search for the cause of the problem revealed that the patient had von Willebrand disease. We present here the case history and the findings we obtained.

Case Report

Patient: a 57-year-old man
Medical history: hypertension, external hemorrhoids
History of present illness: The patient was hospitalized in Chikusei City Hospital for cerebral

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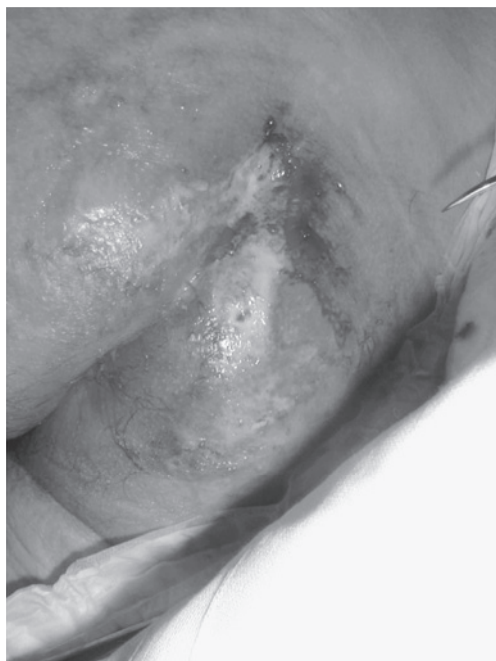


Fig. 1 Incipient stage of the sacral pressure ulcer (stage II)

infarction in the territory of the left anterior cerebral artery; craniotomy and hematoma evacuation, trapping, and external decompression were performed for hemorrhagic infarction accompanied by impaired consciousness. The patient's general condition improved, but a sacral pressure ulcer developed during the course and worsened to stage III (**Fig. 1**). Necrotic tissue was repeatedly debrided from the ulcer at the bedside, but we had difficulty stopping the bleeding each time and had no choice but to use pressure hemostasis. This led to a worsening of the pressure ulcer in a downward spiral (**Fig. 2**). Efforts to determine the cause of the bleeding revealed that the patient had von Willebrand disease (von Willebrand factor activity, 8%; von Willebrand factor antigen quantitative, 12%; coagulation factor VIII activity, 64%; Multimer pattern, normal). A factor VIII preparation was administered every day (Confact F[®] Chemo-Sero-Therapeutic Research Institute, Kumamoto, Japan: 500 unit/day), and improved the von Willebrand factor activity to 31%, leading to a decline in bleeding, and allowing preparation of an adequate wound bed. A decision was made to close the wound with a fasciocutaneous flap including the gluteal perforators which were

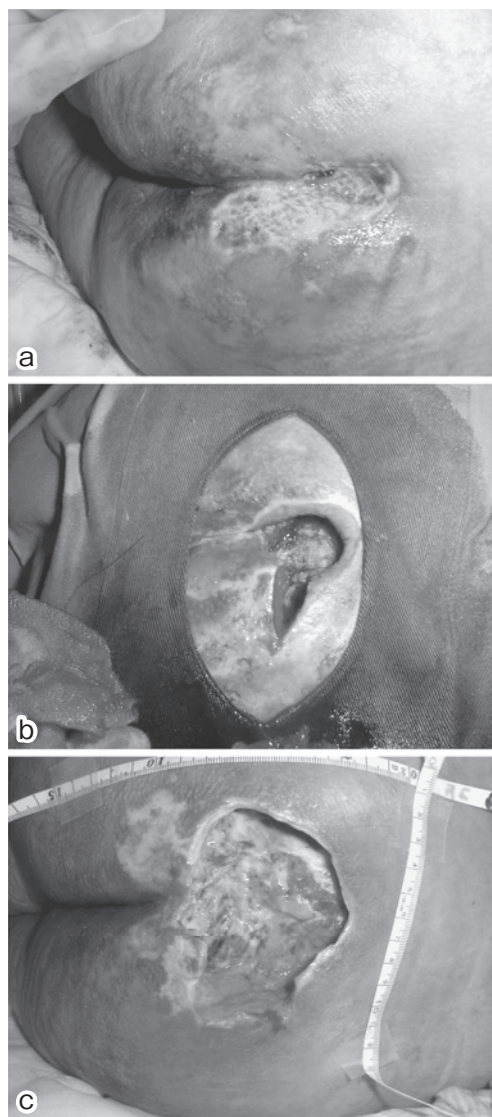


Fig. 2 The pressure ulcer that was exacerbated by pressure hemostasis over one month
a. February 23 b. March 1 c. April 2

preoperatively examined with a Doppler flowmeter in the pedicle. The aims of the treatment were to quickly transfer the patient to a rehabilitation hospital and to discontinue administration of the expensive factor VIII preparation that would otherwise have had to be used for some time on the wound surface. The operation time was 3 h 35 min, and blood loss was 590 g. The flap survived completely; the postoperative course was uneventful, the suction drain was removed after 8 days, and the medication was discontinued following complete epithelialization. About 1 month after surgery, the patient was transferred to a facility specializing in rehabilitation (**Fig. 3**).

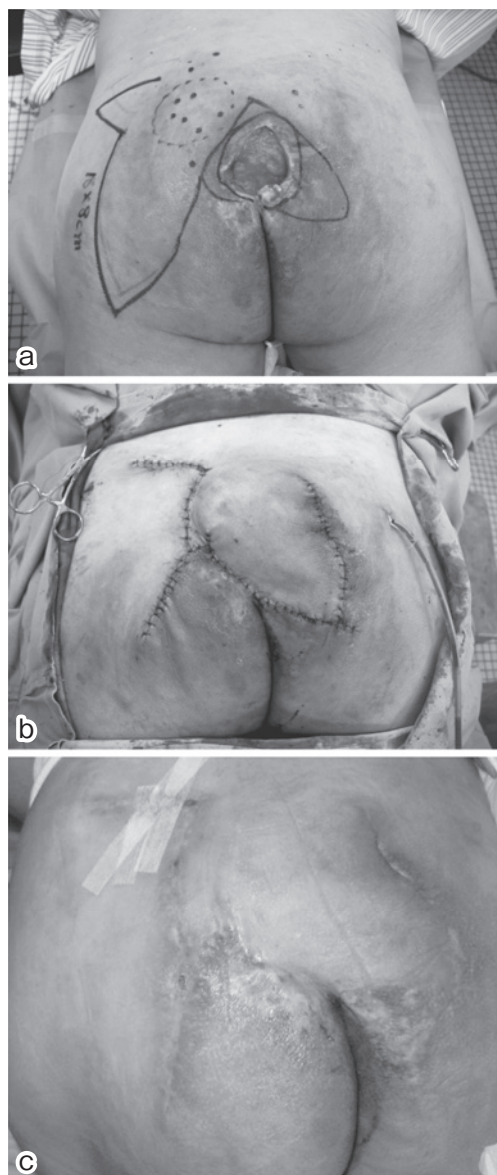


Fig. 3 a. Preoperative view of the sacral pressure ulcer (stage III) and design of the flap
 b. After transposition of the flap
 c. One month after operation

Discussion

In von Willebrand disease, the most commonly inherited coagulopathy, bleeding results from a congenital lack of von Willebrand factor, the coenzyme of factor VIII¹. In the present patient, a pressure ulcer rapidly worsened despite appropriate treatment, but then rapidly improved after von Willebrand disease was diagnosed and treated. This case confirms the importance of quickly identifying the factors causing pressure ulcers to deteriorate and actively tackling them. It should always be

borne in mind that patients with pressure ulcers that do not follow the regular healing course may have rare complicating diseases. This was true of the present patient, who was treated in the Department of Neurosurgery without adequate review of his medical history because of the urgency of treating the cerebral infarction. In later questioning, however, the patient was found to have a history of postoperative bleeding following surgery for hemorrhoids, suggesting that earlier detection of von Willebrand disease might have been possible.

It was clear to us that conservative treatment would lead to a complete cure in this patient. However, early transfer to a rehabilitation hospital was conditional on closure of the wound, and application of an expensive factor VIII preparation was required for the period until the wound closed. Therefore, we decided to operate, even though some risk was present with respect to intraoperative bleeding. The selected reconstruction procedure involved the use of a fasciocutaneous flap, including the gluteal perforators in the pedicle, which could be easily and quickly elevated to minimize bleeding²⁻⁴. During surgery, more time was needed for hemostasis than is normal for this kind of operation, with the result that the operative time and amount of bleeding both exceeded predictions. However, no blood transfusion was required and the postoperative course was good, indicating that the selected procedure was appropriate.

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