A 51-year-old man with hemophilia B presented to our hospital with acute urinary retention after experiencing severe proctodynia for seven months. Neurologic examination revealed hypesthesia of the right L5-S1 nerve roots distribution and weakness of the bilateral tibialis anterior and gastrocnemius muscles. The patient was receiving albutrepenonacog alfa, a factor IX/albumin fusion protein, for hemophilia treatment once a week. Upon admission, blood biochemical tests revealed that factor IX activity was within the normal range (107%; reference range, 70%-149%), whereas the activated partial thrombosis time (APTT) was slightly prolonged (41.5 sec; reference range, 25-36 sec). Magnetic resonance imaging (MRI) was performed in the lateral decubitus position; the supine position was too painful for the patient. Neurologic examination and MRI (Fig. 1, 2) revealed paralysis caused by intratumoral hemorrhage in the cauda equina region. On the day of admission, we performed an emergency laminectomy and tumor extirpation. Intraoperative findings included intratumoral hemorrhage and association of the tumor with a nerve root of the cauda equina (Fig. 3). Postoperatively, the patient’s neurological symptom immediately improved. The pathological diagnosis was schwannoma (Fig. 4).

We conducted a factor IX replacement therapy while monitoring APTT according to the protocol for perioperative treatment for hemophilia. Severe intraoperative bleeding did not occur, and the surgical drainage tube was removed on postoperative day two because the bleeding rate from the tube decreased from 60 to 30 mL/day. However, after removing the drainage tube, there was continuous bleeding from the drainage hole; therefore, the patient received a second dose of albutrepenonacog alfa (25 IU/kg) on postoperative day two. Bleeding from the drainage hole continued on postoperative day three; additional 50 IU/kg of albutrepenonacog alfa was administered, and hemostasis was finally confirmed on postoperative day four. The total bleeding volume after surgery reached 1600 mL, although the activity of perioperative factor IX exceeded 100%, and the postoperative APTT was within the normal range. Despite the unexpected drainage tube issue, the postoperative course was favorable with almost complete neurological recovery except for residual lower limb numbness.

Hemophilia is an inherited bleeding disorder characterized by improper blood clotting and intraarticular and intramuscular bleeding\(^1\). Although intracranial hemorrhage has been estimated to occur in about 3%-10% of hemophilia cases\(^2\), spinal intratumoral hemorrhage is rare\(^3\), and intraspinal hemorrhage is even rarer (0.001% of hemophilia cases\(^4\). Spinal schwannomas often accompany minor hemorrhages; however, it is rare for hemorrhage schwannomas to exacerbate neurological symptoms\(^5\). There is only one previously reported case of schwannoma with intratumoral hemorrhage due to hemophilia\(^6\).

Factor IX replacement therapy is required to minimize bleeding and maintain hemostasis during surgery in patients with hemophilia B. The World Federation of Hemophilia guidelines recommend that factor IX activity levels should be within the 60%-80% range in patients with hemophilia B prior to major surgery\(^7\). The patient in the current study was receiving albutrepenonacog alfa treatment once a week, and his preoperative factor IX activity was 107%. We conducted factor IX replacement therapy according to the protocol for
perioperative treatment for hemophilia. Although factor IX activity remained within the recommended range and APTT was within the normal range during acute therapy, continuous bleeding occurred from the drainage hole, and the reason for bleeding after drainage tube removal is unclear. Possible explanations are that a small artery was damaged when the drainage tube was inserted, or the first additional dose of factor IX after the operation was insufficient. We were not able to find any reports describing drainage tube-related issues in hemophilia patients. Thus, we recommend that healthcare professionals should keep in mind the possibility of this unexpected complication.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Ethical Approval: R1-17-01

Author Contributions: Shingo Imabeppu and Satoshi Shimizu wrote and prepared the manuscript. Yukihiro Matsuyama and Yu Yamato revised the drafted paper. Keiichi Nakai and Satoshi Shimizu performed emergency operation.
All authors approved the manuscript to be published.

**Informed Consent:** Informed consent was obtained from the patient in our report.

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