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Case Report

Mild hemophilia A presaged by recurrent postoperative hemorrhagic complications in an elderly patient

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Abstract

Background: Mild hemophilia without spontaneous bleeding can remain undiagnosed for a lifetime. However, intracranial hemorrhage is one of the most serious complications for patients with hemophilia. In addition, hemorrhagic complications after emergency surgery tend to arise from coagulopathy.

Case Description: An 80-year-old man was admitted with left hemiparesis and disturbed consciousness. He had no history of trauma, fever, or drug and alcohol intake. Computed tomography imaging upon admission disclosed a hemispheric subdural hematoma with a midline shift. No vascular abnormalities were identified as a source of the hemorrhage. The hematoma was removed on an emergency basis with external decompression. However, a large subcutaneous hematoma was again evident on the following day. Insufficient hemostatic maneuvers during surgery were considered the cause of this hemorrhagic complication. A second operation was performed to achieve hemostasis of the subcutaneous and muscle tissue. Thereafter, he was rehabilitated without treatment for hemophilia as he had no bleeding episodes. Cranioplasty proceeded using artificial bone at 40 days after the first operation. However, epidural hematoma developed again on postoperative day 1. His neurological status did not worsen so a repeat procedure was unnecessary. Close scrutiny uncovered a diagnosis of mild hemophilia A.

Conclusions: Accurate diagnosis is important for the management of postoperative hemorrhagic complications caused by pathologies of the coagulation system. Sufficient hemostasis of hemorrhage from subcutaneous and muscle tissue is essential even during emergency surgery to avoid postoperative complications. A diagnosis of hemophilia should be considered in the face of prolonged activated partial thromboplastin time (APTT).

Key Words: Activated partial thromboplastin time, acute subdural hematoma, diagnosis mild hemophilia A, postoperative complication

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INTRODUCTION

The incidence of hemorrhagic complications after neurosurgical surgery has declined due to advances in surgical procedures and medical instruments. However, the postoperative hemorrhagic complication rate increases in the presence of coagulopathies including hemophilia. Hemophilia is usually diagnosed during childhood because bleeding episodes usually present before the age of 5 years. Therefore, mild hemophilia may be asymptomatic and diagnosed incidentally at the time of surgery or trauma, which can result in serious complications. Intracranial hemorrhage can occur in adults with or without a diagnosis of hemophilia.[2,17,20,25] We describe an elderly patient with mild hemophilia presenting as an acute subdural hematoma (ASDH) that required emergency surgery. Here, we discuss the diagnosis of hemophilia and problems associated with postoperative complications of hemophilia among elderly patients.

CASE PRESENTATION

An 80-year-old man was admitted for left hemiparesis and disturbed consciousness at his workplace early in the morning. His medical history contained no head injuries, drug use, malignancies, blood diseases, or autoimmune diseases. He had never experienced bleeding episodes and his family history was unremarkable. His vital signs upon admission were blood pressure, 150/78 mmHg; heart rate, 105 bpm; respiration rate, 17 breaths/min; temperature, 36.7°C; and oxygen saturation, 97% on room air. Laboratory findings revealed a prolonged APTT of 39.8 (normal range, 25–35) s, but a normal prothrombin time of 92.9% (normal range, 75–125%). Other values within normal ranges included platelet count of 186 × 10^3 (normal range, 152–382) × $10^3/\mu L$; hemoglobin, 14.9 (normal range, 14.0–17.0) g/dL; and hematocrit, 44.6% (normal range, 43.0–51.0%).

Vital abnormalities were not evident at the time of presentation, however, neurological findings showed left hemiparesis including the face. Computed tomography (CT) imaging upon admission showed right thick ASDH with median deviation [Figure 1]. Computed tomography angiography (CTA) ruled out vascular abnormalities including cerebral aneurysms and vascular malformations as the cause of ASDH. The hematoma was surgically resected with external decompression due to the disturbed consciousness and left hemiparesis due to ASDH.

Postoperative CT showed a small amount of hematoma and improved median deviation [Figure 2]. However, CT performed 1 day later showed a massive subcutaneous hemorrhage appearing as acute epidural hematoma (AEDH) [Figure 3a]. The APTT was 41.3 s immediately before repeat surgery to establish hemostasis. The hemorrhagic sources were subcutaneous and muscle tissue, and hemostasis was achieved using the standard operative procedures. CT on postoperative day 1 revealed

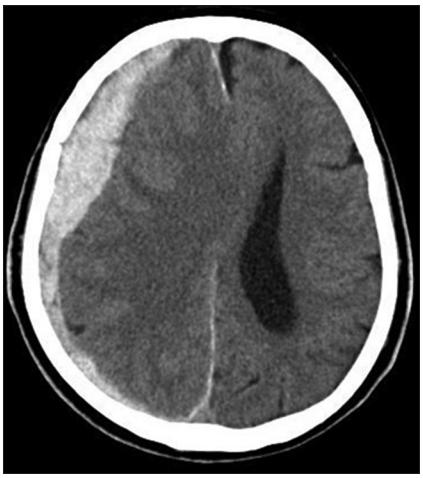

Figure 1: Initial CT imaging findings. Subdural hematoma with obviously shifted midline structure to the contralateral side

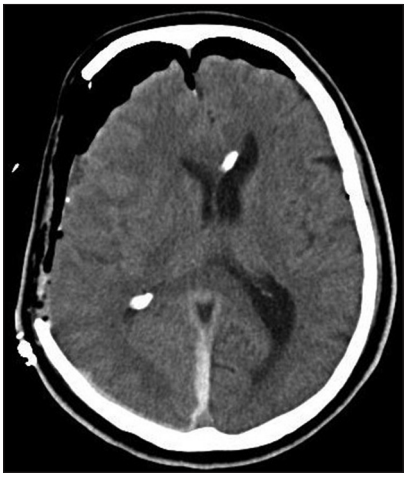

Figure 2: CT imaging findings immediately after the first surgery. Complete removal of the hematoma

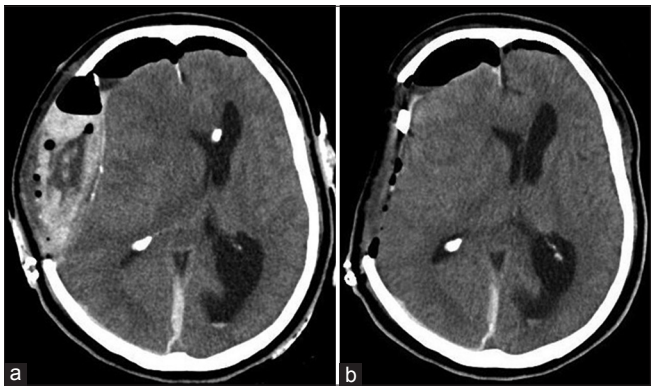

Figure 3: CT imaging findings after first and second surgeries. A large subcutaneous hematoma again appears with midline deviation on postoperative day 1 after the first surgery (a). Complete removal of the hematoma (b)

a small persistent hematoma and improved median deviation [Figure 3b]. Therefore, we judged that the hemorrhage was caused by inadequate surgical technique during the initial operation. The APTT was 42.4 s after the second procedure and his clinical course was good with rehabilitation therapy. Activities of daily life in the hospital were self-sustaining, and the value of APTT ranged between 35 and 40 s.

Cranioplasty proceeded when his general condition was stabilized at 40 days after hospitalization, although the APTT remained slightly prolonged at 35.2 s. CT imaging showed a small amount of hematoma and air immediately after the cranioplasty [Figure 4a] and a slightly thickened AEDH on the following day similar to that after the initial surgical procedure [Figure 4b]. The APTT immediately after cranioplasty was 58.6 s. Neurological findings after surgery were normal and the AEDH on CT images also gradually decreased. Another reoperation to control hemostasis was not required. However, postoperative bleeding recurred and coagulation tests including an assessment of blood coagulation factors revealed that the patient had a plasma factor VIII value of 25% (normal, 62–145%) and that he was negative for factor VIII inhibitor and had a normal value for von Willebrand factor. These findings indicated a diagnosis of mild congenital hemophilia A. According to the guidelines, we kept the bleeding tendency under observation while he underwent rehabilitation for muscle weakness of the lower extremities. Thereafter, he was discharged without neurological deficits.

DISCUSSION

The current frequency of intracranial hemorrhage in patients with hemorrhagic disease caused by the lack of factor VIII (hemophilia A) or factor IX (hemophilia B) ranges 2.7–11.2%.[3,24,28] However, the mortality rate is about 20%,[9,28,30] which is a serious risk factor,[22] particularly among younger patients with hemophilia who have a higher frequency of associated intracranial hemorrhage. The median age is 2 years.[19] Furthermore, 16–30% of all intracranial hemorrhages in patients with hemophilia are subdural hematomas.[19,23]

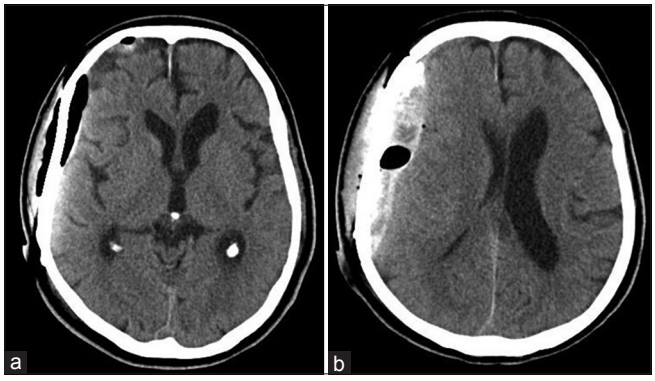

Figure 4: CT imaging findings after cranioplasty. A small amount of air is the sole abnormal finding immediately after cranioplasty (a). Epidural hematoma is evident under artificial bone at 22 hours after cranioplasty (b)

Hemophilia is classified as mild, moderate, or severe according to whether factor VIII levels are 6–25%, 2–5%, or <1%, respectively.[12,13] Bleeding symptoms appear more frequently in patients with moderate and severe hemophilia, and occasionally in those with mild disease. Spontaneous bleeding occurs in the joints and muscles of patients with severe disease and occasionally in those with moderate disease, but it can also arise after trauma.[13,32] Individuals with mild hemophilia do not bleed spontaneously and can hemorrhage during major trauma and surgery.[13] The characteristics of intracranial hemorrhage of hemophilia precede head injury in children, but half of all adult patients with hemophilia do not experience head trauma.[5,13,22]

Our patient did not develop bleeding symptoms due to trauma or surgery; the diagnosis of mild hemophilia was derived from scrutiny of the coagulation system. The possibility of hemophilia in ASDH without trauma history should be considered. Our patient required emergency surgery for ASDH, but the diagnosis of hemophilia was delayed because of the following reasons: first, the cause of rebleeding after the initial emergency procedure had to be evaluated, and second, the underlying cause of the mild prolonged APTT had to be determined.

Postoperative hemorrhagic complications after neurological surgery should be considered. The clinical rate of deterioration is about 0.77–6.9% among patients with postoperative hematoma complications.[4,6,10,18]

Desai noted that 1.9% of postoperative hemorrhage requires reoperation for intracranial pressure control (10). Furthermore, coagulopathy might increase postoperative bleeding if the cause of ASDH is traumatic accidents.[1,8,10,16]

The massive hematoma arising from subcutaneous and muscle tissue in our patient after the first surgical procedure was associated with a cerebral hernia. Therefore, reoperation was necessary to control intracranial pressure and prevent rebleeding. We initially considered that inadequate surgical technique and procedures related to hemostasis after the initial emergency surgery caused the hematoma.[11,18] However, postoperative hematoma appeared again as AEDH after cranioplasty. Hemorrhagic complications can recur despite sufficient hemostatic procedures during surgery. Therefore, we conducted a blood coagulation scrutiny of mild prolonged APTT for diseases associated with coagulopathy, including a deficiency of factor VIII.[15,16]

We investigated the status of APTT and the clinical course of patients with mild hemophilia A. The initial

APTT in our patient after craniotomy was mildly prolonged at 39.8 s, but the change in APTT did not parallel the clinical hemorrhagic symptoms. Some reports describe patients with mildly prolonged or normal APTT.[27,29,31]

The measured value should be carefully considered because various causes could prolong the APTT including the current status of the patient and the anticipated clinical course.[26,31]

Stieltjes *et al*. found that 50% of cerebral hemorrhagic episodes occurred in patients aged >15 years with mild hemophilia, and about 33% of cerebral hemorrhages occurred in patents with moderate or mild hemophilia.[28] Ljung noted a higher frequency of cerebral hemorrhage in adult patients with mild hemophilia, and that many of them develop sequelae.[21]

The general incidence of cerebral hemorrhage is higher in the elderly than in young persons, and hypertension is considered a risk.[21,28] Although different from our patient, intracranial hemorrhage has recently been associated with acquired hemophilia caused by autoantibodies against factor VIII. The causes of acquired hemophilia comprise malignant tumor, autoimmune disease, pregnancy, and advanced age.[14,28] Therefore, hemophilia should be considered when the source of intracranial hemorrhage in an elderly patient remains obscure.[7,25]

Our experience with the present patient has highlighted an important consideration: coagulation should be appropriately assessed if postoperative hemorrhage occurs in an emergency situation even when surgical technique is appropriate. Thus, hemophilia and other diseases associated with blood coagulation factors can be diagnosed.

CONCLUSIONS

Abnormal bleeding can develop in patients with mild hemophilia because of surgery or tooth removal, but rarely during activities of daily living. Therefore, mild hemophilia determined only by a general blood coagulation examination and medical practice is insufficient. That is, neurosurgeons should become more aware of surgical treatment for blood coagulopathies including hemophilia regardless of age.

Consent/assent

The patient provided written, informed consent/assent to the publication of this case report.

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Conflicts of interest

The authors have no conflicts of interest concerning the materials or methods used in this study or the findings described herein.

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