Subacute Subdural Hematoma in Hemophilia B: A Case Report of Successful Treatment

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Abstract

The authors report a case of hemophilia B complicated by subacute subdural hematoma after minor head injury, which was successfully managed by surgery and the use of concentrates of plasma factor IX (Konÿne).

Key words: Hemophilia B, subdural hematoma, replacement therapy, surgical therapy

Introduction

Because of the difficulty of hemostasis during and after surgical intervention in the hemophiliac, the morbidity and mortality in such patients have been greater than in those without this kind of disorder. Deficiency of plasma factor IX (plasma thromboplastin component, PTC) is called hemophilia B or Christmas disease, the incidence of which is about one fifth of classic hemophilia (hemophilia A, deficiency of plasma factor VIII or HAHG).^{1,2)} The recent development of potent, low volume concentrates of factor IX from whole blood has improved the management of bleeding in patients with hemophilia B, especially those with an intracranial hematoma, which had been frequently fatal.^{1,1)}

We would like to present here a case of subacute subdural hematoma with hemophilia B, which was successfully managed by surgery and administration of the defective factor.

Case Report

A right-handed, 4-year-old boy had had repeated episodes since infancy of ecchymosis in the extremities, caused by even light trauma. He

visited Nara Medical College on Feb. 9, 1971, and was documented as a hemophilia B due to selective absence of factor IX in the plasma and serum (factor VIII; 1-stage: 120%, 2-stage: 175% and factor IX; 1-stage (plasma): 0%, 1-stage (serum): 0%, 2-stage (serum): 0%). His brother was also documented as a hemophilia B.

The patient had been in good health until Dec. 22, 1972, when he fell and struck his head without scalp injury or loss of consciousness. In the ensuing four days, a headache and then a clonic convulsive seizure in the upper right extremity developed, during which time, however, consciousness was clear. He was transferred to our clinic by ambulance on Dec. 26, 1972.

Positive findings on admission were inattentiveness, questionable motor aphasia, hemiparesis on the right and slight nuchal rigidity. No fracture was found in the plain skull X-ray films. For the management of the hemophilia, 200 ml of fresh whole blood were transfused on the day of admission, and he was placed under close observation. EEG and echoencephalogram suggested an intracranial space-taking lesion on the left. A vial of Konÿne* was given intravenously on Dec. 28, 1972. However, consciousness level gradually deteriorated and he became semicomatose around midnight on Dec. 28. A left carotid angiography was performed the next morning, revealing an avascular area in the left convexity (Fig. 1).

*Content of factor IX: 564 ± 47 unit/V., Midori Juji Co., Japan

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Fig. 1 Left carotid angiograms. Note an avascular area in the convexity on the left associated with shift of the anterior cerebral artery to the right and downward displacement of the Sylvian point.

Operation was performed under general anesthesia after angiography. At first a burr-hole was made in the left parietal bone and the dura was incised. A subdural collection of clotted blood was confirmed under a very fine membrane. Then a free bone flap craniotomy was carried out to expose the whole hematoma. Blood coagula protruded out through the dural opening under high pressure. The hematoma was 1.0 cm thick at the center of the parietal region. After removal of the blood clot, the brain remained quite tense, and the dural incision could not be closed. The dural opening was covered by fibrin film trimmed suitably and the bone flap was replaced with loose fixation, allowing a space for brain swelling. The patient received two vials of Konÿne preoperatively, 200 ml of fresh whole blood during surgery and one vial of Konÿne just after surgery. The patient tolerated the procedures well and returned to the ward in essentially unchanged condition from the neurological aspect. Postoperative transfusions of two vials of Konÿne was administered daily.

Although the patient remained semicomatose

through the second postoperative day, improvement continued and on the tenth postoperative day, he became alert and nearly normal except for a mild hemiparesis on the right. The healing of the scalp incision was uneventful and the sutures were totally removed on the fifth postoperative day. Because of a slight bulging of the wound after the removal of the sutures, the transfusion of Konÿne was increased from 2 to 3 vials a day for the following three days and was maintained at 1 or 2 vials a day, depending on need, until the 14th postoperative day. Serial measurements of Kaolin PTT and recalcification were carried out to determine the dosage of Konÿne. Intermittent assays of the plasma factor IX level were performed pre-, intra-, and postoperatively every day or two, by courtesy of Midori Juji Co., Japan. The level of plasma factor IX was 1.7% just before the first transfusion of Konÿne, 17 hours after the first transfusion of fresh whole blood. By repeated transfusion of Konÿne, the level of plasma factor IX was maintained over 30% during the replacement therapy.

The patient was discharged on Jan. 26, 1973,

with a very slight hemiparesis on the right. Three years after the operation he had only a questionable hemiparesis on the right and was performing well at school.

Discussion

It is widely known that hemophilia is a sex-linked, recessively inherited disorder which affects males, and is transmitted by apparently normal females. Aggeler, et al.¹⁾ Biggs, et al.³⁾ separated hemophilia B from classic hemophilia, however the treatment of the former has been preceded by that of the latter with the concentrates of factor VIII such as HAHG (human antihemophilic globulin, Cohn's fraction I).^{4,9)} In 1965 the concentrates of factor IX were reported in the work of Tullis, et al.¹¹⁾ The availability of the concentrates has reduced the risks of surgical procedure in patients with hemophilia B. Despite many reports relating to intracranial surgery of patients with classic hemophilia,^{6,7,8,10)} there have been only a few reports of successful intracranial operation on such patients.²⁾

The patient in this report was evaluated to be lacking factor IX and had the following symptoms and findings: a lucid interval for about 4 days, neurological lateralities, a remarkable asymmetry of EEG and an abnormal echoencephalography, which suggested a unilateral intracranial hematoma. We performed a carotid angiography followed by an operation. Under the use of Konÿne, no troublesome bleeding occurred during the procedures of angiography or of the operation. Daily transfusion of Konÿne was maintained for 14 days after the operation, and the sutures were removed on the 5th postoperative day. The level of plasma factor IX was ascertained and maintained at over 30%.

As to the management of surgical cases with hemophilia; how long and what level of defective plasma factor VIII or IX should be maintained during operation and postoperative state have been controversial. Mazza, et al.⁷⁾ reported their experience with the use of factor VIII concentrates during surgery in patients with hemophilia A and suggested that such patients should have received prolonged postoperative administration of factor VIII. They also states that none of their patients showed

evidence of hemorrhage when factor VIII was above the level of 25%. It would, therefore, seem reasonable to maintain the factor VIII at this level, or higher until the wound heals completely. On the other hand, George, et al.⁵⁾ experienced ten patients with classic hemophilia and four with hemophilia B, and suggested that a less prolonged usage (two to five days after operation) of the concentrates might be equally effective in providing hemostasis. In our case, the bulging of the scalp wound remained for several days even after the removal of the sutures. Therefore, the transfusion of Konÿne was maintained up to the 14th postoperative day, resulting in no episode of rebleeding. The transfusion of the concentrates should be continued in such a case until the healing of the operative wound becomes complete.

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