

(CASE REPORT)



A spontaneous cerebral hemorrhage case in the right temporal lobe in a patient with hemophilia C (XI factor deficiency)

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Abstract

A 28-year-old man was taken to our hospital with sudden onset of severe headache. The computed tomography (CT) scan revealed subcortical hemorrhage of the right temporal lobe. The patient has past medical history of hemophilia C (XI factor deficiency). On neurological examination no deficit was observed (Glasgow Coma Scale (GCS) -15). 10 hours after admission, his GCS and CT scan of the brain were unchanged. After consultation with the hematologist, he received fresh frozen plasma, mannitol, tranexamic acid during the treatment. But 36 hours after admission, the neurological status was worsened (GCS -12). A decision was made to immediately perform a temporal craniotomy with evacuation of intracerebral hematoma. Surgery and the postoperative period were uneventful. The patient was discharged without neurological deficit. This case highlights the importance of adequate preparation of the patient presurgically as much as possible to minimize the risk of complications (rebleeding) during the operation and in the postoperative period.

Keywords: Spontaneous cerebral hemorrhage; Factor XI deficiency; Surgery; Right temporal lobe

1. Introduction

Hemophilia C (HC) is a rare, inherited bleeding disorder where the blood does not clot properly. In patient with Hemophilia C (also called Rosenthal Syndrome) the blood is deficient in factor XI. Spontaneous intracerebral hemorrhage is a rare and challenging condition to treat, especially in hemophiliacs [1]. Intracranial hemorrhage (ICH) is the leading cause of deaths, in patients with hemophilia and accounts for 20% of non-infections deaths. Neurosurgical intervention in the acute setting of intracranial hemorrhage in patients with HC is often reserved for those with a deteriorating neurological exam and /or/ clinically significant hemorrhage with mass effect. Beyond prompt surgical intervention the perioperative management of HC is of paramount importance to optimize outcomes [2,3]

2. Illustrative Case

A 28-year-old man was taken to our hospital with sudden onset of severe headache. Complaints were available for about 2 days. The patient had no history of trauma, except for consumption of alcohol. The patient has 22 years history of hemophilia c (XI factor deficiency). The computed tomography (CT) scan revealed subcortical hemorrhage of the right temporal lobe (Fig 1).

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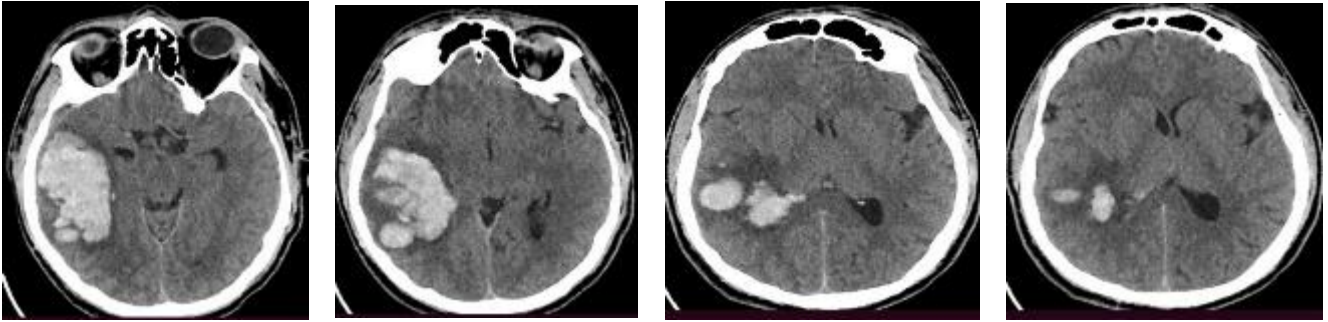


Figure 1 Non-Contrast Axial CT the head demonstrating a 7.6x3.8x2.6 cm acute intraparenchymal hematoma centered in the right temporal lobe with an associated 7 mm midline shift

His GCS at admission was 15. The patient had no neurological deficit. The patient was admitted to intensive care unit. An urgent consultation with the treating hematologist was also performed, and a decision was made to manage the patient conservatively (fresh frozen plasma 4 units, mannitol 10% 1000 mg, tranexamic acid 1000 mg). 10 hours after admission, his GCS and CT scan of the brain were unchanged (Fig 2). We have also performed CT angiography for exclude any vascular pathology.

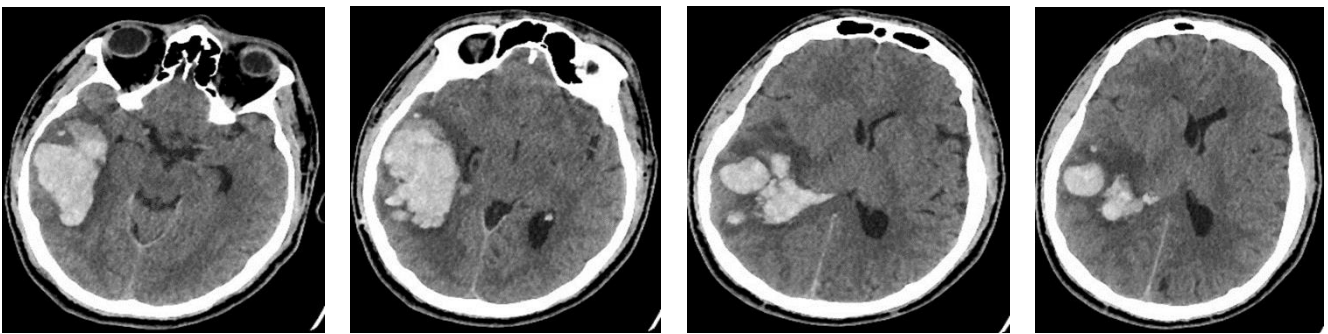


Figure 2 Non-Contrast Axial CT the head 10 hours after admission demonstrating a stable acute intraparenchymal hematoma centered in the right temporal lobe with an associated 7 mm midline shift

But 36 hours after admission, the neurological status of the patient was worsened. His GCS dropped to 12. A decision was made to immediately perform a temporal craniotomy with evacuation of intracerebral hematoma. Surgery and the postoperative period were uneventful. There was no need for blood transfusion during the operation.

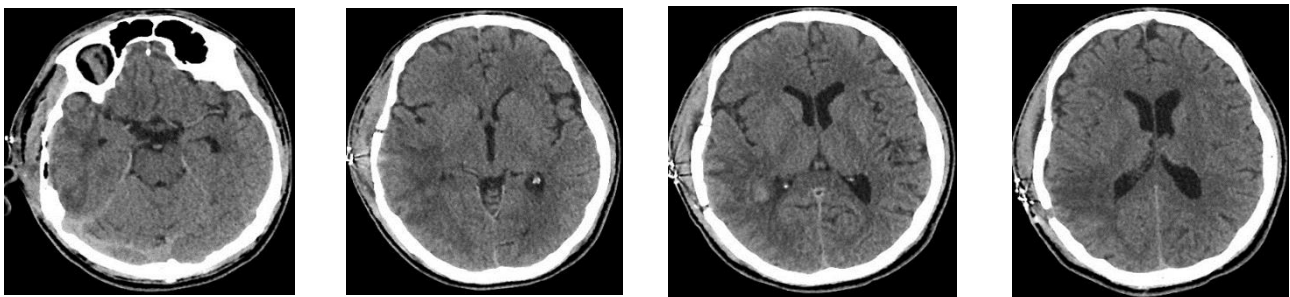


Figure 3 Non-Contrast Axial CT of the head 8 days after operation demonstrating an absence of intraparenchymal hematoma and no evidence of midline shift. Intraparenchymal edema in the right temporal lobe

The next 2 days after the operation patient received 4 units of fresh frozen plasma, tranexamic acid 1000 mg and mannitol 10% 1000 mg per day. Blood tests were performed and 7 days after operation we have a good result (APTT 29.2, INR 1.2, XI factor 115, XIII Factor 82). Post operation CT of the brain shows also a good result (Fig 3). The patient was discharged from the hospital 9 days after admission without neurological complications. 7 days after discharge; the patient still had visual disturbances in the right eye. The tests revealed right upper temporal quadrants, which, however, recovered over time. One month after discharge, the patient's visual disturbances were resolved, and a head

CT scan revealed no significant pathology (Fig 4). The patient returned to his normal life and was periodically observed by hematologist.

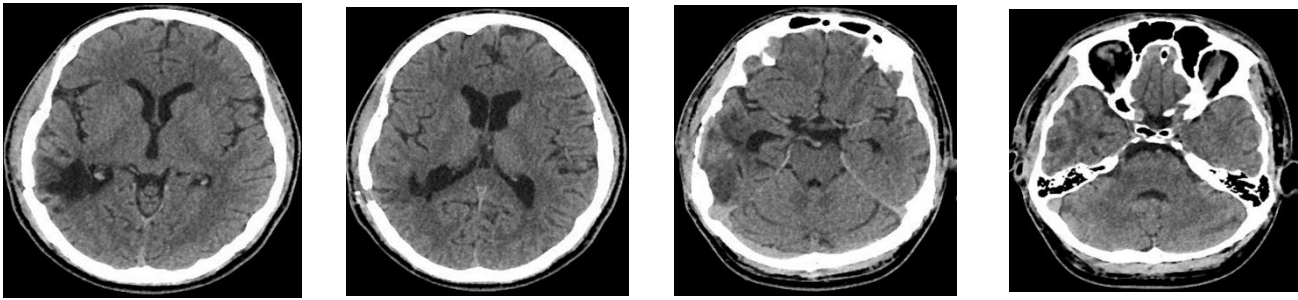


Figure 4 Non-Contrast Axial CT the head 40 days after operation demonstrating an absence of intraparenchymal hematoma and no evidence of midline shift

3. Conclusion

ICH is the leading cause of deaths, in patients with hemophilia and accounts for 20% of non-infections deaths.

Spontaneous intracranial hemorrhage is a serious and potentially fatal sequela of Hemophilia. Correction of clotting factor deficiency is of paramount importance prior to surgery and in the immediate post-operative period. However, we would like to recommend the following:

- Delay surgery as much as possible.
- Conduct conservative treatment, especially correction of the coagulation factors
- Multidisciplinary management of those patients is important

The fulfillment of the above-mentioned points allows avoiding intra- and early postoperative complications, in particular rebleeding, recurrence of intracranial hematomas.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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