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Multiple Intracerebral Hemorrhagic Lesions Complicating Minimal Cranioencephalic Trauma in A Child with Severe Hemophilia A, Trisomy 21 and Sickle Cell Trait Carrier: A Case Report

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Abstract

Intracranial hemorrhage (ICH) is a life-threatening complication in hemophilia. It often occurs during childhood after minimal head trauma. It poses a problem of diagnostic and therapeutic management, especially in Africa where CFCs are not always available.:

We report the case of an 8-year-old boy, severe hemophiliac A, carrying sickle cell trait and trisomy 21. He was admitted to emergency 6 days after a cranioencephalic trauma following by headaches with photophobia and consciousness disorders. Cerebral CT scan showed a hematoma of the posterior cerebral fossa, a left temporal hematoma, a meningeal hemorrhage and a hydrocephalus. The patient received CFCs substitution and regular neurological follow-up.

Management of ICH in hemophiliacs remains difficult in Africa because of the unavailability of treatment and the lack of knowledge of the disease. An early diagnosis and an adapted management would allow to reduce morbidity and mortality related to ICH.

Keywords: Hemophilia; intracranial hemorrhage; CFCs; sickle cell trait; case report

List of Abbreviation:

CFCs: coagulation factor concentrates

CT: Computer Tomography

CNS: central nervous system

Hb: hemoglobin

HTC: International Hemophilia Treatment Center

ICH: Intracranial hemorrhage

MRI: Magnetic resonance imaging

Introduction

Intracranial hemorrhage (ICH) is a major complication of hemophilia that occurs in childhood. In children with hemophilia, it almost always occurs in the context of mild to moderate head trauma, in contrast to adult hemophiliacs in whom it can occur spontaneously in 50% of cases [1]. It is a great challenge to the survival of severe hemophiliac patients, even in developed countries where these patients have access to high quality care. The location of the hemorrhage is roughly equally divided between subdural hematoma, intracerebral hemorrhage and subarachnoid hemorrhage [2,3]. ICH in older children was less common and had a better outcome. This survey clearly showed that most survivors of ICH suffer from some form of neurological sequelae [4]. We report the case of an 8-year-old boy who presented with ICH following a mild head injury by falling from his own height.

Patient and Observation

Patient Information: This is an 8-year-old boy not attending school due to psychological developmental delay, severe hemophiliac A, carrier of sickle cell trait and trisomy 21. He is regularly followed at the International Hemophilia Treatment Center (HTC) and was put on prophylactic treatment for 3.5 years and then stopped at the age of 6.5 years due to family constraints. He was brought by his parents to the emergency room 6 days later after a fall with a landing on his head during a game. He consulted because of consciousness disturbances that had been evolving for 2 days prior to his admission, associated with intense headache and photophobia. He reportedly had an initial loss of consciousness after the accident, 2 episodes of vomiting, and seizures.

Clinical Findings: when examined, he had pale mucous membranes, a heart rate of 95 beats/min, a blood pressure of 110/80 mmHg, a respiratory rate of 16 cycles/min and a weight of 25 kg. The neurological examination revealed: consciousness disorders with a Glasgow score of 13/15, pupils were reactive, mobilization of the 4 limbs upon stimulation, muscle tone was normal, tendon reflexes were sharp and a cerebellar syndrome. The examination of the neck performed after having ruled out a spinal instability on the CT scan revealed a meningeal syndrome. On examination of the cephalic extremity, a right occipital subcutaneous hematoma was noted. There were no other physical lesions found.

Diagnostic Assessment: the blood count revealed an anemia of 8.4 g/dl, the platelet count was normal. Screening for inhibitors could not be performed urgently, but the APTT was prolonged and corrected. We were not able to follow up the residual FVIII level in our context.

The brain CT scan with cervical scanning showed multiple hemorrhagic lesions: a bilateral cerebellar epidural hematoma as a hyperdensity more marked on the right (Figure 1), a left temporal contusion (Figure 2), a meningeal hemorrhage, and a beginning hydrocephalus (Figure 3). There was no cervical spine injury or bone damage.

The diagnosis of intracranial hemorrhage due to mild head trauma was made and he was admitted to the neurosurgical unit in Ziguinchor (455 km from the HTC).

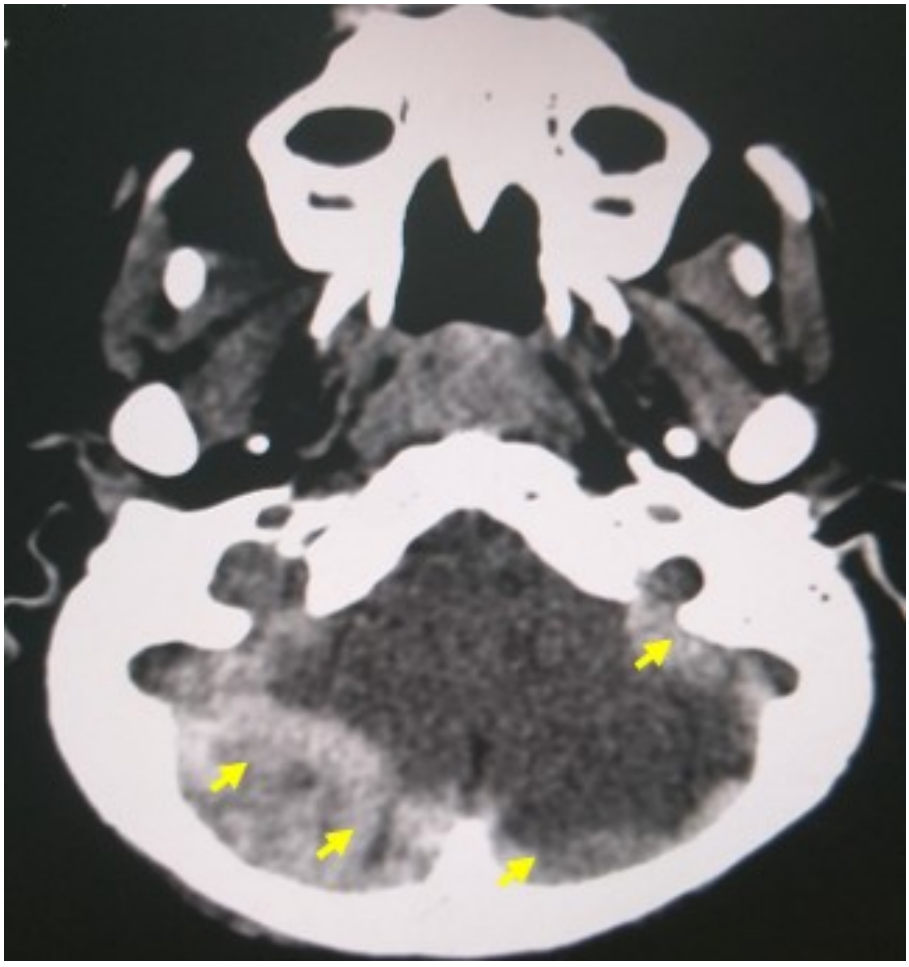

Figure 1: CT axial section: Posterior fossa haematoma, more marked on the right (yellow arrow)

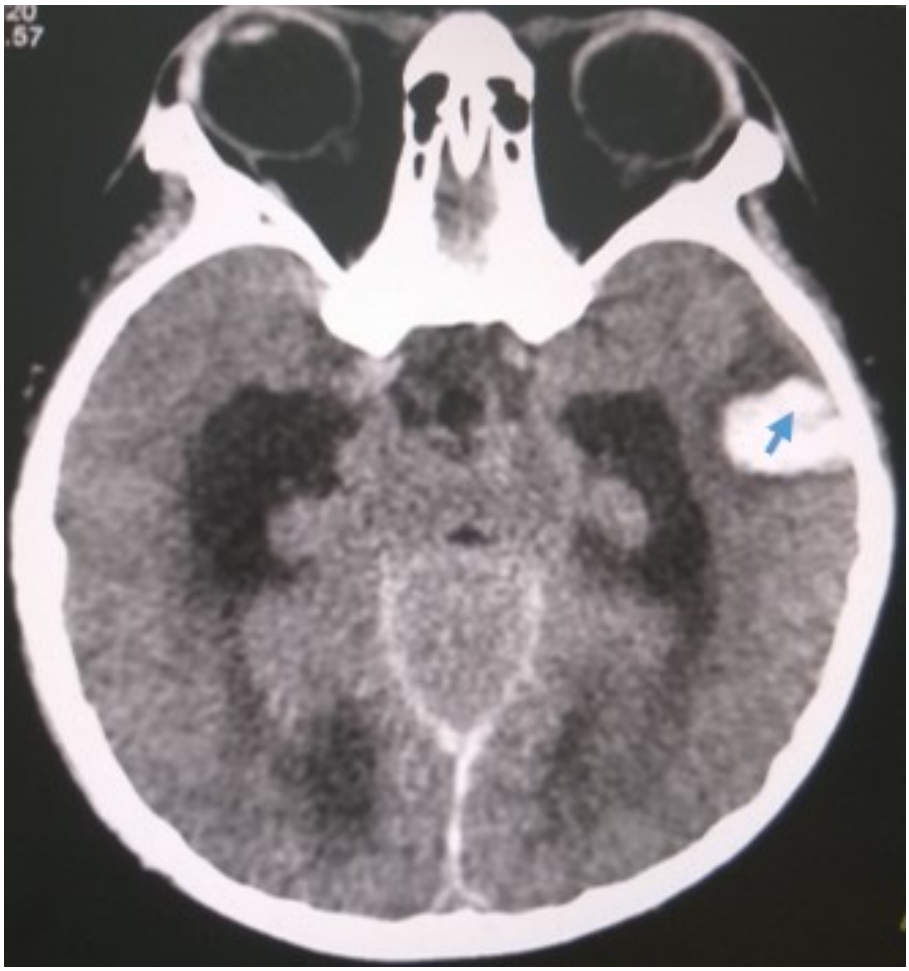

Figure 2: CT axial section: Oedemato-hemorrhagic contusion of the left temporal bone (blue arrow)

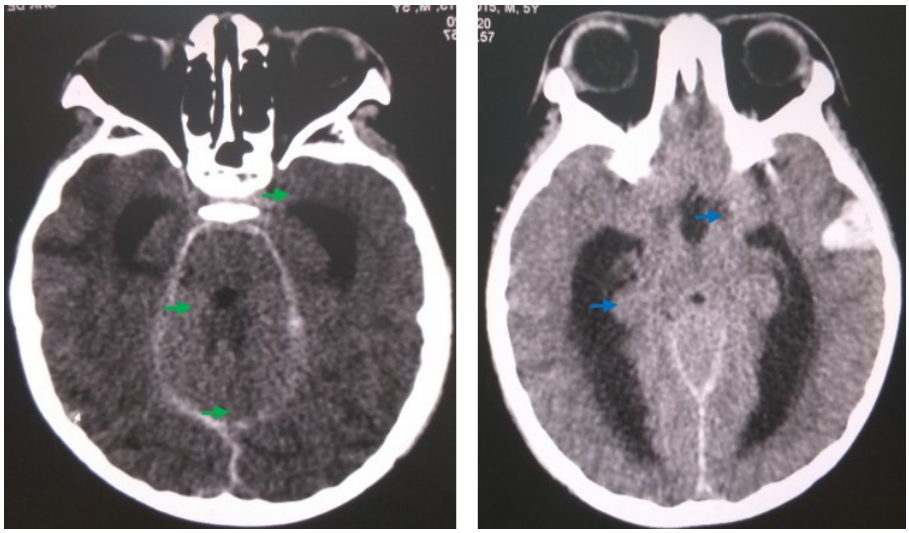

Figure 3 : CT axial section a: Meningeal hemorrhage (green arrow) b: Early hydrocephalus (blue arrow)

Therapeutic Intervention: the protocol of replacement of CFCs was written by the hematologists in the HTC. The patient received an emergency dose of CFC of 60 IU/kg, then 30 IU/kg/day for 7 days associated with tranexamic acid at 20 mg/kg every 8 hours.

He also received a transfusion of red blood cells, fresh frozen plasma at a rate of 15ml/kg/day for 3 days, analgesic and isotonic saline. A bypass valve was placed at the bedside, along with a rigorous follow-up for any neurological deterioration and/or the

appearance of new symptoms. This management was carried out in a multidisciplinary manner between the neurosurgeon, the pediatrician and the hematologist.

Follow-up and Outcomes: After two weeks, the headache and vomiting that were present at the beginning improved and clinical examination revealed a good improvement of the state of consciousness with a Glasgow score of 15/15 and a regression of the cerebellar syndrome. The follow-up CT scan showed a hematoma in the process of resorption. The child was transferred to the HTC for follow-up and screening for inhibitors. Inhibitors screening was negative and low-dose prophylaxis had been proposed to the patient but refused because of family constraints.

Discussion

Haemophilia is a rare bleeding disorder related to the X chromosome which is caused by the deficiency of clotting factors VIII (haemophilia A) and IX (haemophilia B). It is a ubiquitous disease with an incidence of 1 in 10,000 births worldwide. Hemophilia A is predominantly 85% and hemophilia B is only 15% [1].

ICH is among the leading causes of death in hemophilia patients and accounts for 75% of neurological sequelae in survivors. In children with hemophilia, minor head trauma is the main cause of ICH and is often overlooked. However, spontaneous ICH can occur in patients with severe hemophilia [3, 5, 6]. This can be explained in our case by the fact that our patient had a delay in psychomotor development that could lead to falls during activities (games).

In addition, our patient consulted us late because of the isolation of his village. The symptoms most frequently found in ICH are headaches (44.8%), vomiting (44.8%) and lethargy (41.3%), often in combination. Other neurological manifestations can be observed such as convulsions (10.3%) and coma (10.3%) [7].

Prevention of these head injuries is possible by limiting movements and by using a soft helmet. In patients with hemophilia, the diagnosis of ICH should not be ruled out on the basis of the absence of a history of head injury or the absence of clinical signs [2]. For patients with moderate or minor hemophilia, the risk of intracranial hemorrhage is less than 50% of that of patients with severe hemophilia [8].

ICH, which has an incidence of 3-12%, accounts for more than 30% of deaths in hemophiliacs [6]. Mortality is more closely related to the location of the hemorrhage than to the severity of the hemophilia. Subdural hematoma and subarachnoid hemorrhage have a better prognosis [2].

Cranial CT or MRI is very useful and performed in suspected cases to diagnose and confirm ICH, but should not delay CFC administration under any circumstances. This imaging allows visualization of the site, amount of bleeding, and prediction of prognosis [1].

The coexistence of sickle cell trait and hemophilia is an uncommon association and rarely described in the literature, even in predominantly black populations, where the prevalence of both conditions is higher [9]. Few cases have been described in the literature: one reported the simultaneous presence of hemophilia A and homozygous sickle cell disease Hb SS in a 30-year-old black man and the second reported an association of sickle cell disease Hb SS and hemophilia B, in a 15-year-old African-American man [10]. Many authors consider sickle cell disease to be a hypercoagulable prothrombotic state and hypothesize that the association of sickle cell disease with hemophilia should alter the thrombohemorrhagic balance and positively improve hemorrhagic hemophilic phenotypes [11].

Bleeding disorders are contraindications for intracranial pressure monitoring [2]. However, given the multitude of hemorrhagic lesions, the absence of a trained anesthesiologist, the lack of an on-site hematologist, and the inability to evacuate the child

urgently to the HTC, the patient was a candidate for neurologic monitoring.

In addition, the availability of sufficient quantities of clotting factor concentrates must be ensured before major surgery is undertaken. Some authors suggest that intracranial bleeding can be managed conservatively in most patients with hemophilia. It is recommended in the literature to saturate at 100% at least during the first 5 days to ensure very rapid resorption of bleeding [1]. However, in our patient, we started CFC with an initial dose of 60 IU/kg and then continued with a dose of 30 IU/kg.

A multicenter retrospective study of 1515 children with hemophilia A showed that children on regular and frequent prophylaxis have a low risk of ICH compared with those on irregular prophylaxis or those treated on demand [7].

Our patient was on low dose prophylaxis [12] but secondarily interrupted because of family constraints (moving to live with his grandparents in a very remote village with no access to electricity). This case illustrates the difficulties of following prophylaxis in a rural environment with the discontinuation of treatment and the occurrence of a CNS hemorrhage [13].

Nevertheless, this case shows the effectiveness of the management of a major hemorrhage, even at 450 km from the HTC of Dakar with conservative treatment.

Patient Perspective: Both the mother and the child were very pleased and satisfied with the care received and especially with the absence of neurological sequelae.

Informed Consent: the mother, the child's legal guardian, was informed of the rare and particular case and the authors' interest in publishing his case. Free and informed consent was obtained from the mother to use the CT scans.

Conclusion

Intracranial hemorrhage is a serious complication of hemophilia with a high morbidity and mortality. Its management is multidisciplinary, involving the hematologist and the neurosurgeons, and constitutes a real challenge in a neurosurgical unit that does not have an adequate technical platform.

Competing Interests

The authors declare no competing interest.

Authors' contributions

SAT and YC drafted the manuscript. SAT, MS and SD were responsible of hematological management and wrote the CFCs substitution protocol. YC and AD were responsible of the neurological management and monitoring of the patient. MS and SD reviewed and corrected draft. SD and SAB were the scientific advisors. All authors read and approved the final manuscript.

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