Case Report

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Acquired Esotropia, Focal Seizure, and Subdural Hemorrhage (SDH) in a newly diagnosed moderate hemophilia B child: a case report

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ABSTRACT

Introduction*: This case report is relevant to clinicians across multiple disciplines, especially in resource-limited settings, as it illustrates how careful non-surgical management can lead to favorable outcomes in complex bleeding conditions. It could guide future case management, especially for pediatricians and hematologists.*

Case Presentation*: We present the case of a 2-year and 4-month-old male Ethiopian child who was newly diagnosed with moderate hemophilia B and developed esotropia, focal seizures, and subdural hemorrhage after a fall. The surgery was deferred because of the high risk of bleeding, and he was successfully managed with factor IX (FIX), and the child was discharged with a full recovery. Currently, the child is 4 years and 6 months old, and he is active without any apparent focal neurological deficits or seizures. The child is on regular follow-up and does not require any further interventions except for factor IX (FIX) transfusion.*

Conclusions*: This is the first case report describing a newly diagnosed child with Hemophilia B presenting with acquired esotropia, subdural hemorrhage, and focal seizure. Despite the presence of mass effects, the patient was successfully treated with factor IX replacement therapy, avoiding the need for neurosurgical intervention.*

Keywords: Esotropia, Seizure, Subdural Hemorrhage, Hemophilia

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1. Introduction

Hemophilia is a rare genetic disease that results from mutations in the genes that code for proteins necessary for normal blood clotting [1]. Worldwide, the prevalence per 100,000 males is 17.1 cases for hemophilia A, 3.8 cases for hemophilia B [2]. In Africa, the overall prevalence of Hemophilia A was 6.82 cases per 100,000 persons [3]. In Ethiopia, the prevalence of Hemophilia is 2.2 to 9.6 cases per 100,000 persons [4]. There are three types of Hemophilia caused by a deficiency of coagulation factors: hemophilia A, hemophilia B, and hemophilia C. [5]

Hemophilia A is caused by a deficiency of clotting factor VIII and is the most common type. A deficiency in clotting factor IX causes Hemophilia B (also known as Christmas disease) [6]. Hemophilia C (also known as Rosenthal syndrome) is a deficiency of clotting factor XI. It is a sporadic form of hemophilia that is found equally in men and women and is most common in certain ethnic groups, especially people of Ashkenazi Jewish descent (occurring in up to 8%). Most patients with hemophilia C have less severe bleeding problems than those with factor VIII or IX deficiency and do not experience hemarthroses. The risk of bleeding does not correlate with the Factor level in patients with hemophilia C. [7]

In Hemophilia A and B, severity is classified into three categories according to the level of circulating clotting factors. Mild hemophilia is defined by greater than 5% to 40% factor activity. Moderate hemophilia has 1% to 5% factor activity. Severe hemophilia has less than 1% factor activity. Patients with mild hemophilia tend to experience abnormal bleeding only in response to surgery, tooth extraction, or injuries. Conversely, patients with moderate hemophilia experience prolonged bleeding responses to relatively minor trauma, and patients with severe hemophilia experience frequent spontaneous bleeding, especially recurrent hemarthroses and soft-tissue hematomas, leading to severe arthropathy, joint contractures, and pseudotumors and, consequently, to chronic pain, disability, and reduced quality of life.[8]

Over time, Hemophilia A and B are considered clinically indistinguishable from each other. Recent evidence, however, suggests that patients with hemophilia B have a less severe bleeding phenotype, a lower bleeding frequency, and better long-term outcomes (a lower likelihood of joint arthroplasty).[9] This is the first case report globally of acquired esotropia, focal seizures, and subdural hemorrhage in a newly diagnosed child with type B hemophilia following an accidental fall. This case report is unique because of its multifaceted clinical presentation, the challenges posed by Hemophilia B, and the implications for treatment strategies across different specialties.

2. Case report

History of presenting illness

This was a 2-year and 4-month-old male Ethiopian child who presented to the pediatric emergency department with abnormal body movement, which was characterized by rhythmic jerking of his right hand with lip-smacking. He had three attacks, each lasting 4 to 5 minutes with drooling of saliva, but he gained consciousness between each episode. In association with this, he had a history of high-grade intermittent fever and decreased mental acuity. For this, he visited a nearby governmental hospital and was admitted for 4 days with the assessment of complicated meningitis + focal seizure. The patient was on ceftriaxone, vancomycin, and phenytoin. A brain MRI revealed a subdural hemorrhage, so the patient was referred to Tikur Anbesa Specialized Hospital (TASH) for neurosurgical evaluation. At TASH, a detailed history revealed that the child's maternal uncle had hemophilia and that the child had a history of easy bruising but no bleeding during circumcision.

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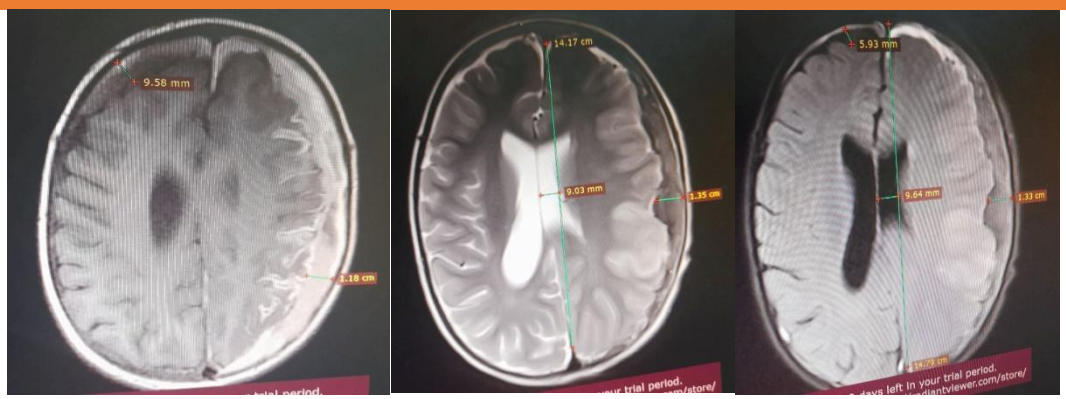

Figure 1: Brain MRI with contrast: Left holocerebral convexity and right occipital SDH (acute to subacute) with subfalcine herniation plus left front oparietal gyriform restriction likely postictal

Past Medical History

A month back, he experienced a fall on the left side of the head while he was playing in the house. The next morning, he started to experience frequent episodes of vomiting of ingested matter, and he developed inward deviation of his left eye. The vomiting stopped with ondansetron. For the eye complaint, the patient visited an ophthalmology clinic, and on retinoscopy examination, there was a slightly blurry disc margin. With the assessment of acquired esotropia, he was managed with eye patching therapy. The ophthalmologist suggested brain imaging, but it was not performed at that time.

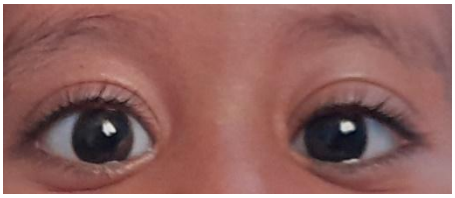

Figure 2: Patient picture: Patient image showing left eye inward deviation (esotropia). Posted with parental consent

Physical examination

The patient was irritable in general appearance. The vital signs were unremarkable except for the fever (38.3 degrees centigrade). HEENT evaluation = Inward deviation of the left eye, Pale conjunctiva, 2x3 cm ecchymosis over the left mandibular area. Central nervous system evaluation, Pediatric coma score =14/15 (E4M6V4), pupils were midsized and reactive bilaterally with no signs of lateralizing.

Investigations

On CBC, WBC=13.2 K With a neutrophil count of 82.8%, hematocrit=22.3, hemoglobin=7.4 and platelet=300k

Factor IX =1.8%, Factor VIII =293.9%, aPTT =190 seconds, INR=1.08 and PT=10.7.

Management

For this, he started Factor IX (FIX) therapy by using Factor IX replacement formula (according to World Federation of Hemophilia and other standard guidelines): FIX dose (IU) = (Target F IX levels – Factor FIX baseline levels) X body weight (kg) X 1 unit/kg. In this case, the target was 100% because of intracranial bleeding.

Outcome

After 3 days of initiation, the esotropia disappeared, the child became active, the seizures and fever were controlled, the aPTT normalized (24.4 s), and the repeated MRI revealed a significant reduction in the midline shift.

The surgery was deferred due to the high risk of bleeding, and there was great clinical improvement with the factor replacement. With an 8-day total stay in the pediatric ward, the child was discharged in full recovery and assigned to the OPD of hematology, neurology, and neurosurgery. After 4 months of continued phenytoin, an electroencephalogram (EEG) was performed, which revealed a normal wake EEG tracing. Thus,

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the neurology team decided to taper the phenytoin and discontinued it. Currently, the child is 4 years and 6 months old, and he is active without any obvious focal neurological deficits or seizures. The child is on regular follow-up and does not require any further interventions except for factor IX (FIX) transfusion.

Figure 3: Patient Picture: After esotropia disappeared with treatment

3. Discussion

The strengths of this case report include the Novelty of the case, a comprehensive management approach, and multidisciplinary relevance. As a single case report, the findings may not be generalizable to all patients with hemophilia. Our case illustrates that conservative management with factor replacement without neurosurgical treatment may be considered and may result in good outcomes in hemophilic children with complications such as esotropia, seizures, and subdural hematomas. Intracranial hemorrhage (ICH) is relatively rare, with an incidence ranging from 2 -15%, but it is one of the most dangerous and life-threatening events in individuals with hemophilia.[10] Several risk factors are associated with ICH in patients with hemophilia, including a history of trauma, severe disease, the presence of factor inhibitors, age over fifty years, age two years or younger, and not receiving prophylactic treatment regimens.[11]

ICH patients present with a wide spectrum of manifestations. Among these, the patient will have focal neurologic signs. The specific presenting symptoms may vary according to the location of the bleeding overlying the brain structures impacted. For example, if the bleeding involves the frontal lobe, the patient will have hemiparesis and speech impairment, and if the posterior fossa is involved, the patient will experience headache, vomiting, anisocoria, nuchal rigidity, ataxia, and cranial nerve palsies. Among the cranial nerve palsies, abducens nerve palsy (cranial nerve 6) is responsible for acquired esotropia. Esotropia is a type of strabismus or misalignment. In esotropia, the eyes are crossed; that is, while one eye looks straight ahead, the other eye is turned in toward the nose. Without proper treatment, esotropia poses increased risks of injury, irreversible vision loss, and decreased functional ability, ultimately significantly reducing one's socioeconomic status. [12, 13]

Hemophilia is a type of secondary hemostasis disorder that manifests as increased activated partial thromboplastin time (aPTT). The platelet count and prothrombin time (PT) are normal in hemophilia patients. Measurement of the factor activity level revealed a reduced level (less than 40%). Neuroimaging, particularly noncontrast head CT, is a vital tool for diagnosing intracerebral hemorrhage. However, factor administration should not be delayed while awaiting neuroimaging; an immediate dose should be given to raise the levels of FVIII or FIX to 80-100% in severe and life-threatening CNS bleeding.[14] For hemophilia A, an initial acute dose of approximately 50 IU/kg of FVIII, followed by repeated bolus dosing every 8–12 h or continuous infusion. In hemophilia B, the initial dose, in general, is 100–120 IU/kg followed by bolus dosing every 12–24 h or continuous infusion. As per the World Federation of Hemophilia Guidelines, for confirmed CNS bleeding, appropriate factor levels need to be maintained for a period of up to 14 days. [15]

The mainstay of treatment for hemophilia B involves replacing the missing blood coagulation factor FIX when bleeding episodes occur (on-demand treatment) or by scheduled infusions several times per week (prophylaxis treatment). Both plasma-derived (PD) and recombinant (r) FIX

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clotting factor concentrates are suitable for these different strategies of hemophilia B management. [16]

Owing to the economic constraints associated with its procurement, bleeding episodes are regularly dealt with fresh frozen Plasma (FFP) or cryoprecipitate in low-resource countries. Compared with FFP, Cryoprecipitate is better owing to rapid correction of the coagulation fraction, which leaves a lower chance of volume overload and minimizes the likelihood of recipient leucocyte-mediated nonhemolytic febrile transfusion reactions. However, FFP is a readily available and affordable option. [17] Studies have shown that simultaneous treatment with TXA and rFVIII significantly improves clot stability in patients with hemophilia A. [18] Desmopressin (DDAVP) is a treatment option for some patients with mild factor VIII deficiency. DDAVP causes the release of vWF and factor VIII stores from endothelial cells. A DDAVP challenge is performed to prove the response. If a patient has an adequate response to DDAVP, it can be used as a treatment for acute bleeding and prophylactically for tooth extraction in patients with mild disease. DDAVP is not effective in patients with moderate or severe hemophilia A. [19]

The use of surgery for subdural hematomas in hemophiliac children is debatable. The general recommendation for hemophilia patients with intracranial hemorrhage is conservative and is based on factor replacement. Neurosurgery in a child with hemophilia may be considered only when the patient has life-threatening central nervous system bleeding. [20]

4. Conclusion

This is the first case report describing a newly diagnosed Ethiopian child with Hemophilia B presenting with acquired esotropia, subdural hemorrhage, and focal seizure. Despite the presence of mass effects, the patient was successfully treated with factor IX replacement therapy, avoiding the need for neurosurgical intervention.

Abbreviations

aPTT -activated partial thromboplastin time
DDAVP- Deamino-8-D-arginine vasopressin
EEG-Electroencephalogram
FFP-Fresh frozen Plasma
SDH -Subdural hematoma
ICH-Intracranial hemorrhage
PD-Plasma-derived
PT-prothrombin time
TASH -Tikur Anbesa Specialized Hospital

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Competing interests

The authors declare that they have no competing interests.

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