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Case Report

Hemophilic Neonate with Massive Subdural Hematoma Rescued Using Life-Saving Management: A Case Report

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Abstract

Hemophilia is a complex gene mutation blood disorder, that does not allow the clotting of the blood in the typical way it should, due to the insufficiency of blood clotting proteins also known as clotting factors. Hemophilia A is caused by the deficiency of clotting factor VIII (FVIII) and Hemophilia B is caused by the deficiency of clotting factor IX (FXI), which is present in the X chromosome. These are the key factors that help in the clotting of the blood. We could say that hemophilia A is a blood disorder that is genetic. The treatment is mainly blood clotting factor replacement therapy. We present a case of a 5-day-old neonate male with Hemophilia A, subdural hematoma with no known parental history of Hemophilia. Also highlights how the timely initiation of factor VIII (FVIII) transfusion along with other immediate interventions like the decompression of the subdural space could positively impact the outcome.

Keywords: *Neonate; Hemophilia A; Intracranial Hemorrhage (ICH); Subdural Hematoma (SDH); Factor VIII (FVIII); External Ventricular Drain (EVD)*

Introduction

Hemophilia A is one of the most common blood disorders in the world. It has been reported that over 1.2 million people have been diagnosed with Hemophilia A worldwide [7]. They occur among all, irrespective of race and nationality, and can be found among 1 in every 4,000 to 1 in every 5,000 males [4,5]. Hemophilia A can be classified into 3 categories where the FVIII would be 6% - 49% in mild, moderate with 1% to 5% and severe being less than 1% [1-3,5,6].

Intracranial hemorrhage (ICH) occurring spontaneously in Haemophilia is highly uncommon and is around 3.5% to 5.5% at birth and mostly follows after a traumatic delivery [1,7]. It has been reported that around 1% - 4% of ICH with hemophilia is commonly found among neonates [3,5]. This is a serious event with mortality rates of up to 20% despite the management of such conditions being highly advanced these days [1]. With early diagnosis and intervention, it has been observed that people with Hemophilia A can lead a normal life.

We would like to present a case about the efficient management of a neonate with hemophilia A having SDH, which is one kind of ICH.

Case Presentation

This case presents a 5-day-old male neonate that was born to a gravida 2 para 1 live 1 mother, by normal vaginal delivery at 38 + 6 weeks gestation with a birth weight of 3.19 kg, head circumference of 34 cm, and blood group - O+ve. The baby cried soon after birth,

with an Apgar score of 8/9, and with the perinatal and post-natal period for the baby being uneventful. On the 5th day of life, the baby was admitted to the hospital with complaints of poor feeding and abnormal movements. On examination, the baby was found to have pallor, multiple aponic episodes, and seizures. Due to multiple ongoing seizures and respiratory depression, the baby was immediately intubated, ventilated, and started on antiepileptic medications immediately.

The baby's vitals were unstable and the CBG results were suggestive that the baby was having respiratory acidosis. Because of a weak respiratory effort by the baby, a CXR was performed which is suggestive of normal findings.

The neurological examination of the infant displayed poor activity, posturing, and reduced reflexes. The examination of the pupil revealed sluggish pupillary reflex and gross asymmetry in size. Given the above abnormal neurological findings and refractory seizures, CT imaging of the brain was performed. The CT brain (Figure 1) was suggestive of massive SDH with compression of the brain and gross midline shift (> 20 mm), which was one of the causes of the recurrent refractory seizures. The antiepileptic medications (Inj. Phenobarbitone and Inj. Midazolam) helped in subsiding the seizures.

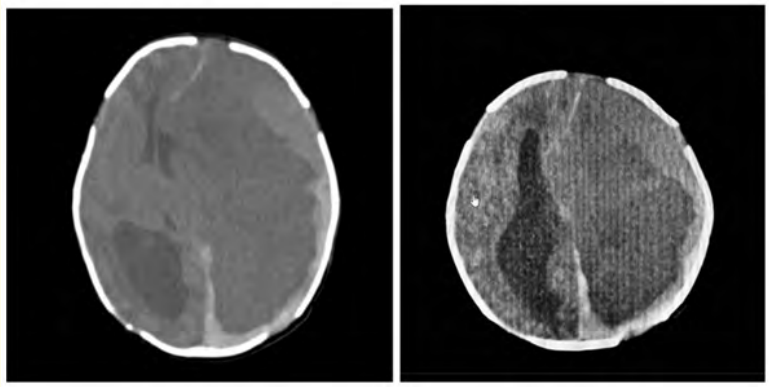

Figure 1: *CT of the brain showing a massive subdural hematoma with a 20 mm midline shift to the right and right ventricle dilation.*

The Preliminary hematological investigations were suggestive of severe anemia, and prolonged aPTT of 105 seconds. A sample for factoring levels was sent as we strongly considered the possibility of hemophilia. After the clinical diagnosis of hemophilia A (FVIII), the neonate was administered PRBC, FFP, and Cryoprecipitate transfusion immediately. On retrospective analysis of the history, no history of hemophilia or any other blood disorders was found in the family.

Because of the critical clinical condition of the infant, IV antibiotics were initiated, which were later discontinued as the blood culture was sterile. Only Inj. phenobarbitone was given as maintenance to prevent the baby from seizing further.

It was definite that if an immediate intervention for the baby was not done, there could be an 80% - 90% risk of sudden death. Given the worsening clinical conditions and abnormal imaging studies, it was suggestive of SDH with a gross midline shift (> 20 mm), a decision for early surgical intervention to decompress the brain was made by the neurosurgeon. So, with the consent from the parents, a decision was made to drain the SDH. The baby would not be able to tolerate an invasive procedure like craniotomy considering the high chances for coagulopathy. So as a life-saving measure, a burr hole was made at the frontal bone in the coronal suture line to drain the hemorrhagic fluid. The intervention was conducted in the NICU itself as the baby was coning rapidly. Evacuation of the SDH was done and around

50 to 70 ml of hemorrhagic fluid was drained from the subdural space. We noticed that the blood drained had no clots owing to the coagulation disorder. An additional external ventricular drain (EVD) was inserted into the subdural space *in situ* to further facilitate the decompression and drainage.

In time the baby stopped seizing with signs of improvement in the neurological status and consciousness levels. The neonate was administered with clotting FVIII transfusions for the next 14 days. Meanwhile, we could confirm the diagnosis as severe hemophilia as the factorate level is 1%. Currently, the baby is 4 months old and is active without any obvious focal neurological deficit. The baby is on regular follow-up with us and did not require any further interventions except for factor VIII (FVIII) transfusion.

Discussion

Hemophilia A is a genetic blood coagulation disorder that happens due to the deficiency of clotting factor VIII (FVIII). When it comes to cases of severe hemophilia A, we can see that bleeding episodes can occur even without any obvious reasons of injury or trauma and are mostly seen in children aged below 2 years [1-3,5,6]. It is also seen that around 50% of males who are diagnosed with hemophilia have mothers who are carriers of hemophilia and around 30% of neonates have a negative family history of this disease [6].

Here we would like to highlight the presentation of hemophilia A linked with SDH which is very uncommon and can go highly unnoticed and be spontaneous without any known triggers such as injury during/uneventful birth or any other head traumas. Especially when there is no known family history of clotting disorders the diagnosis of such cases can be tricky. In these types of crucial cases, a quick diagnosis and application of apt and immediate interventions are fundamental.

In our case, the neonate was presented with pallor, anemia, respiratory depression, and multiple refractive seizures which are very commonly seen in hemophilic patients with intracranial bleedings [4,5]. The neonate had no known parental history of any clotting disorder. It has been recorded that in such cases intracranial bleeds are usually seen in the first week of life [5]. The CT of the brain revealed a massive SDH with compression of the brain and gross midline shift (> 20 mm). And the hematological investigations showed anemia with prolongation on the aPTT of 105 seconds which is commonly seen in hemophilia [5]. The clotting FVIII test revealed severe Hemophilia A with FVIII being just 1%.

Here we would like to emphasize how with the early diagnosis of hemophilia A it is possible to find the underlying cause for the deterioration of the baby's condition. The timely intervention with the initiation of Factor FVIII transfusions, PRBC, FFP, cryoprecipitate, and neurosurgical procedures helped save the child from severe neurological deficits. Our early neurosurgical intervention not only could put a hold on the progressive worsening of this fatal condition but also could result in a remarkable improvement in the clinical status of the baby like improvement in the sensorium, seizure control, general vital parameters, and long-term neurological outcome.

Conclusion or Summary

Early diagnosis of Haemophilia A and early treatment with factor VIII is important for the prevention of complications related to the disease. For hemophilic babies who present with massive intracranial hemorrhage, early interventions like drainage of SDH are life-saving, it is also crucial for symptom control and improving the long-term outcome.

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