Neonatal Coagulopathy Presents as Unusual and Severe Subgaleal Hematomas After Vacuum Delivery

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Neonatal subgaleal hematomas (SGHs) are infrequent but may be underdiagnosed collections of blood beneath the galea, often caused by vacuum delivery. With massive bleeding into the subgaleal space, exsanguination and hypovolemic shock have caused death in 20–60% of newborn infants [1]. SGHs are mostly caused by vacuum delivery, and sometimes by neonatal coagulopathy. We report a case of a newborn who suffered from severe SGHs and anemia on the third day after delivery.

A 26-year-old, gravida 1, para 0, mother, who had regular prenatal care at our hospital with no abnormal laboratory data or abnormal sonographic findings, was admitted to our delivery room at 38 weeks' gestation for labor. The first and second stages of labor took about 15 hours and 6 hours, respectively. Vacuum extraction was used because of maternal exhaustion and prolonged second stage. Third-degree perineal laceration was found after delivery. The male baby was active and irritable after delivery, with Apgar scores of 9 and 10 at 1 and 5 minutes, respectively. A small caput succedaneum occurred as usual, on the posterior fontanel. Unfortunately, the caput succedaneum seemed to progress until a big cephalohematoma was noted on the third day of delivery. It crossed the midline (sagittal suture) and covered the whole occipital area down towards the posterior neck, even though we packed it with an elastic cap. It continued to grow toward the anterior neck region and then the cheek. Patches of ecchymosis were noted over the eyelids (Figure), anterior neck, and ears. Because of this unusual appearance, the complete blood count was checked and revealed an anemic and coagulopathy status (red blood cells $1.81 \times 10^6/\mu L$; hemoglobin 6.8 g/dL; hematocrit 19.5%; platelet $337 \times 10^3/\mu L$; activated partial thromboplastin time > 200 seconds). The factor IX assay decreased markedly to 1.1%. Brain ultrasound was arranged, and it showed no obvious intracranial hemorrhaging or lesions.

Figure. Ecchymosis over both eyelids.

The hematoma and hemorrhaging seemed to be established after the blood transfusion with packed red blood cells of 60 mL, and resolution was noted 10 days after delivery. Thus, under the impression of factor IX deficiency, we sent the blood sample to the National Taiwan University Medical Center where the diagnosis was confirmed. The baby fed well and no neurologic deficits were noted after delivery. Unfortunately, the patient and her baby were lost to follow-up after discharge.

Hemophilia B is a deficiency in the clotting factor IX. It is an X-linked recessive coagulation disorder, which occurs in one out of every 25,000–30,000 male births and requires even rarer genetic circumstances for phenotypic expression in females [1]. It presents as the

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impairment of blood clotting ability and prolonged bleeding time. The outcomes are good with early treatment and management.

The severity of hemophilia B symptoms depends on how a particular gene abnormality affects the activity of the factor IX. When the activity is less than 1% of normal, episodes of prolonged bleeding may occur for no apparent reason. Severity of symptoms can vary, but severe forms become apparent soon after birth. Prolonged bleeding is the disease's hallmark. Additional bleeding manifestations make their appearance when the infant becomes mobile. People whose clotting activity is 5% of normal may have only mild hemophilia. They rarely have unprovoked bleeding episodes, but surgery or injury may cause uncontrolled bleeding, which can be fatal (1.1% of infants suffered from severe subgaleal hematomas after the instrumental delivery, like our case). Milder hemophilia may not be diagnosed at all, although some people, whose clotting activity is 10–25% of normal, may bleed excessively after surgery, dental extractions or major injuries.

Neonatal SGHs are diagnosed when collections of blood are noted beneath the galea, which are often caused by vacuum delivery [2]. It is a rare but potentially lethal condition found in newborns. Optimizing the outcomes for babies with SGH requires early diagnosis, careful monitoring, and prompt treatment. Diagnosis is made by history taking and physical examination. Monitoring includes a minimum of 8 hours of observation for all babies following difficult vacuum extractions or forceps deliveries, regardless of Apgar scores or need for resuscitation. This observation should include at least hourly recording of vital signs. Examination of the head, including the circumference of the head and assessment of the location and characteristics of any swelling, should be repeated hourly if concerns are present. The presence of fluctuance early on, whether or not the swelling is progressive, is an important distinguishing feature of SGH. Because blood spreads through a large tissue plane, blood loss may be massive before hypovolemia becomes evident (as in our case).

When SGH is suggested, hemoglobin measurements should be performed as soon as possible and should be monitored every 4–8 hours, as should coagulation studies [2]. Kilani and Wetmore [3] revealed the presentations and outcomes of 34 infants with SGHs, and the mortality rate was 11.8%. Those who died had significant volume loss with anemia, coagulopathy and shock, requiring large volumes of blood and blood product transfusions. The presence of intracranial hemorrhage did not correlate with the severity of SGH or death, but the severity of SGH correlates with death. Minor neurologic abnormalities were noted in only 11% of infants [3]. For the treatment of SGHs, Amar et al [4] presented two cases of neonatal SGHs, who had radiographic features indicative of elevated intracranial pressure as well as neurologic decompensation. The first patient received scalp incision initially, and the second patient was too unstable for operative intervention and died. This surgical evacuation of neonatal SGH has not been described previously. Extracranial cerebral compression represents another way by which neonatal SGH may jeopardize the infant's life. Management should consist of measures to correct hypovolemic shock and disseminated intravascular coagulation, as well as surgical intervention to control elevated intracranial pressure [4].

In our case, neither parents had obvious family history of bleeding. Hemophilia or other bleeding and clotting disorders were suggested, as there may be marked hematoma from delivery, especially after a vacuum extraction. For the diagnosis of hemophilia B, there are several laboratory abnormalities, such as prolongation of activated partial thromboplastin time (variable with degree of hemophilia), decreased factor IX activity, and normal bleeding time and thrombin time. In addition, imaging of the central nervous system may be in order, especially when there have been neurologic signs. Once the defect has been identified, other family members will need less testing to diagnose the disorder.

Hemophilia B is typically treated by infusing the missing clotting factor. The amount infused depends upon the severity of bleeding, the site of the bleeding, and the size of the patient. A hepatitis B vaccine is recommended for individuals with hemophilia B, because they are at increased risk of developing hepatitis due to exposure to blood products. During a bleeding episode, more clotting factors are needed. To prevent a bleeding crisis, people with hemophilia and their families can be taught to administer factor IX concentrates at home at the first signs of bleeding. People with severe forms of the disease may need regular prophylaxis infusions two to three times a week.

Both parents of the child with hemophilia should receive genetic counseling. A woman who gives birth to a child with hemophilia often has other male relatives who also have hemophilia. Sometimes, a baby will be born with hemophilia when there is no known family history. This means that either the gene has been "hidden" (i.e. passed down through several generations of female carriers without affecting any male members of the family) or the change in the X chromosome is new

(a "spontaneous mutation"). All daughters of a man with hemophilia will be carriers, and none of his sons will have hemophilia. Unfortunately, the parents of this baby were lost to follow-up after delivery.

Neonatal coagulopathy is not common, but it should be alerted when abnormal progression of cephalohematoma to subgaleal hematoma after instrumental delivery or even after spontaneous delivery is observed. Early management is necessary for good outcomes. Genetic counseling for the parents should be carried out in order to determine the risks of hemophilia in the future.

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