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Case Report

Noninvoluting congenital hemangiomas with hypovolemic shock, anemia and prolonged jaundice in a neonate: a case report

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Abstract

Noninvoluting congenital hemangioma is an extremely rare congenital anomaly in newborn babies and may complicate life-threatening events, including hemorrhage. We present a very rare case of noninvoluting congenital hemangiomas in an Asian and Afghan six-day-old girl that was detected at birth. The noninvoluting congenital hemangiomas were accompanied by hypovolemic shock, anemia, and prolonged jaundice during the first two weeks of life. These diseases were diagnosed by medical history, physical examination, blood analysis, and doppler ultrasonography of the skin lesions. Her hypovolemic shock and hemorrhage were improved after supportive treatment, and she was discharged from the hospital in a good condition. After four weeks of life, the hemangiomas did not regress or progress, suggesting noninvoluting congenital hemangiomas. Hemangioma may complicate severe hemorrhage and shock, as well as there may be an association between congenital hemangioma and prolonged neonatal jaundice.

Keywords: congenital hemangioma, critical complications, hyperbilirubinemia and newborn

BACKGROUND

Congenital hemangioma is a benign vascular tumor that is usually present at birth, predominantly in female infants. Although it is an extremely rare anomaly, the exact prevalence is unknown [1, 2]. Based on the beginning of regression during the first days of life, congenital hemangioma is classified into three categories: noninvoluting congenital hemangioma (NICH), rapidly involuting congenital hemangioma (RICH) and partially involuting congenital hemangioma (PICH) [1–4].

Clinically, congenital hemangioma appears as a bluish-red skin eruption with less-well defined margins, pale halos, and sometimes coarse or nodular surfaces. They are fully developed at birth, and usually present as soft tissue masses or plaques on different parts of the body, such as the head, limbs, or neck [1, 4–7]. The diagnosis of a congenital hemangioma is best made based on its clinical features. In cases of doubtful manifestation, doppler ultrasonography is a useful diagnostic technique that often shows high-flow vasculature within the lesion [1, 7]. The majority of CH respond best to observational care. Since not all CHs regress, a proper follow-up is required to prevent any ulceration or bleeding from the tumor. Medical management, including beta blocker therapy, is advised for the treatment of large hemangioma. Locations of tumors with a greater mortality rate and lesions that last until pre-school age are indications for the surgical intervention of CH [1, 6]. There is a paucity of data regarding noninvoluting congenital hemangioma with hypovolemic shock, anemia, and prolonged jaundice in current literature; therefore, this case is presented.

CASE REPORT

A six-day-old Afghan female neonate was admitted to the Neonatal Unit of the Pediatric Department due to skin lesions, bleeding, and jaundice. She was born by spontaneous vaginal delivery at 38 weeks of gestation with a birth weight of 3.5 kg to a 22-year-old multigravida mother at a tertiary hospital. The mother was in good health during pregnancy and did not receive any teratogenic drugs or radiation. The baby had no history of trauma or surgery. There was no history of hemangioma in the family. The parents were third-degree relatives, which denoted parental consanguinity. On general physical examination, lethargy, a rectal temperature of 34.9°C, a respiratory rate of 68/min, a heart rate of 170/min, blood pressure of 50/30 mm Hg, an unpalpable radial pulse, a capillary refill time (CRT) of four seconds, cold extremities, weak primitive reflexes, yellowish skin up to the palms and soles, pale conjunctivas, and an oxygen saturation of 74% were detected. On local physical examination of the left leg, there were three skin lesions that were present from the time

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of birth. A large, blue-red-colored lesion with a length of 14 cm and a width of 8 cm was visible on the left anterolateral thigh up to the middle of the calf. The surface of the lesions had a coarse and nodular appearance with pale-pink halos (Fig. 1). The other two smaller skin lesions with the same characteristics were observed on the dorsum of the left foot (Fig. 2). On admission, bleeding was observed from a ruptured nodular telangiectasia of the large skin lesion that had started three hours before her arrival at the hospital. Blood investigations revealed a hemoglobin of 9 g/dl, a total leucocyte count of 9000/mm^3 (polymorphs 45%, lymphocytes 50%, eosinophils 2%, monocytes 2% and basophil 1%), a red blood cell count of 2.8 million/mm^3 (mean corpuscular volume of 91.8 fl, mean corpuscular hemoglobin of 35.8 pg), a platelet count of 317 000/mm^3, a reticulocyte count of 2%, a C-reactive protein of 0.4 mg/dl, a blood sugar of 89 mg/dl, a total bilirubin of 16 mg/dl, indirect bilirubin of 15 mg/dl, a baby blood group of ARh positive, and a mother blood group of ARh positive. Prothrombin time, INR, activated partial thromboplastin time, alanine aminotransferase, aspartate aminotransferase, and thyroid and renal function tests were within normal limits. Abnormal clinical findings such as tachypnea, tachycardia, hypothermia, hypotension, a prolonged CRT, an unpalpable radial pulse and cold extremities, as well as a history of bleeding from the large skin lesion for three hours, were used to diagnose hypovolemic shock. Neonatal anemia and indirect hyperbilirubinemia were diagnosed according to low blood hemoglobin and high indirect bilirubin, respectively. Initially, the patient was given 35 ml of intravenous normal saline during the first 20 min, oxygen therapy, and a local dressing to stop bleeding. This was followed by the administration of a 70 ml fresh blood transfusion during the next one hour for the management of hypovolemic shock and bleeding. After the management mentioned, the baby became stable with normal vital signs and enough breastfeeding. On the second day of admission, doppler ultrasonography of the skin eruption and a second blood investigation were advised. The longitudinal and cross-sectional images of doppler ultrasonography demonstrated superficial fast-flow vasculature within the skin lesions (Figs 3 and 4). Congenital hemangiomas were diagnosed by the presence of typical skin lesions and doppler ultrasonographic findings in the baby. The second blood investigation revealed normal parameters except a hemoglobin level of 11.5 g/dl with normocytic normochromic red blood cells, and a total bilirubin level of 16.5 mg/dl which denoted neonatal anemia and hyperbilirubinemia, respectively. On the third day of admission, petrolatum ointment was applied to the surface of the lesion three times daily to prevent ulceration, as well as propranolol (7 mg) and iron sulfate (8 mg) twice daily were given orally to treat hemangiomas and neonatal anemia, respectively. As indirect hyperbilirubinemia of 11 mg/dl lasted up to the age of 17 days, it was accepted as prolonged neonatal jaundice. For the diagnosis of breast milk jaundice, breastfeeding was discontinued for 48 h with no response. The prolonged neonatal jaundice was managed by increasing the frequency of breastfeeding, but there was no indication of phototherapy in the index neonate. The third blood investigation on day 25 of life showed a hemoglobin level of 13 g/dl with normocytic normochromic red blood cells, and a total bilirubin level of 4 mg/dl which indicated the improvement of neonatal jaundice and anemia. The hemangiomas did not regress or progress during the first four weeks of life (Fig. 5), hence, they were diagnosed as noninvoluting congenital hemangiomas. The surgical indications for CH include tumor sites with higher death rates and lesions that persist until pre-school age.

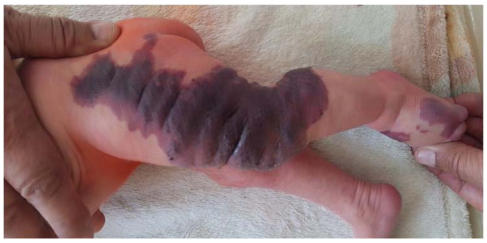

Figure 1. A large blue-red colored lesion is visible on the left anterolateral thigh up to the middle of calf. Two other lesions are seen on the dorsum of left foot. The surface of lesions had coarse and nodular appearance with pale-pink halos. These findings are typical for congenital hemangiomas.

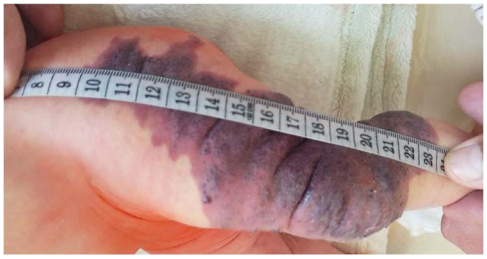

Figure 2. A large blue-red colored lesion is visible on the left anterolateral thigh up to the middle of calf. Two other lesions are seen on the dorsum of left foot. The surface of lesions had coarse and nodular appearance with pale-pink halos. These findings are typical for congenital hemangiomas.

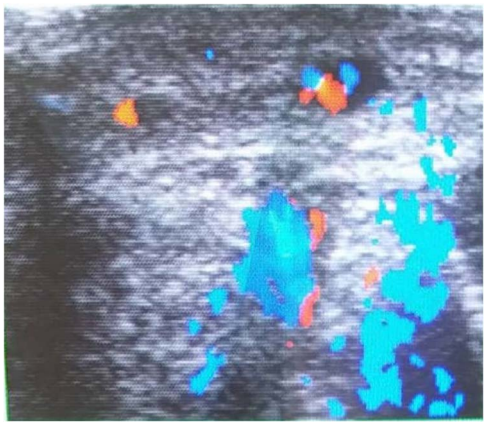

Figure 3. The cross section image of doppler ultrasound demostrates superficial vascular structure in the skin lesion of the neonate.

Therefore, such management was not carried out in the present case.

DISCUSSION

Congenital hemangioma is an extremely rare benign vascular tumor that is usually observed at birth with a female gender predominance and an unclear etiology [1, 2, 4, 5]. Based on the types of regression, CH is classified into RICH, NICH and PICH [1–3]. Usually, the regression of CH begins within a few days of birth, and the diagnosis is established by clinical features. Sometimes, doppler ultrasonography of the lesion is used to confirm the diagnosis [1, 4, 7]. Observation care continues to be most beneficial for the majority of CH. Propranolol in a dose of 2-3 mg/kg/day for six months may reduce the size of large CH in 50% of cases. The

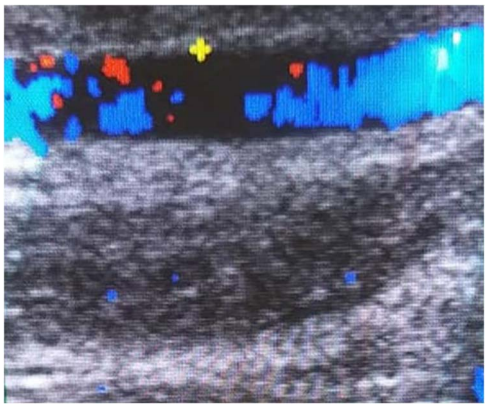

Figure 4. The longitudinal view of doppler ultrasound shows high-flow superficial vasculature within the skin lesion in the index baby.

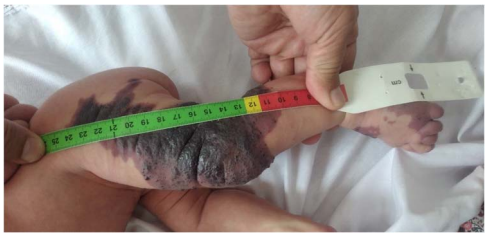

Figure 5. After four weeks of management, the skin lesions of the infant did not regress which denote noninvoluting congenital hemangiomas.

surgical indications for CH are tumor sites with a higher mortality rate, and those don't go away until pre-school age [1, 6].

Shock is an acute process characterized by the body's inability to deliver adequate oxygen to meet the metabolic demands of vital organs and tissues. In the neonatal period, a common type of this disorder is hypovolemic shock due to bleeding. The diagnosis of shock in neonates is made by the presence of several indicators of inadequate circulatory functions, consisting of hypotension, a prolonged CRT, an unpalpable radial pulse, cold extremities, tachypnea, tachycardia and hypothermia. Oxygen therapy and the administration of isotonic intravenous fluid or blood transfusion are the mainstay treatments for hypovolemic shock [8]. Neonatal anemia is defined as a hemoglobin level of less than 13 gr/dl. Hemorrhage is the main cause of anemia in neonates, and for anemic newborn babies with respiratory support, blood transfusion is indicated when the hemoglobin level falls below 11.3 gr/dl [9–11]. Jaundice that lasts more than 14 days in fully-term infants is defined as prolonged neonatal jaundice. Rh or ABO incompatibility, infection and congenital hypothyroidism are the main causes of prolonged unconjugated hyperbilirubinemia [12, 13]. By the age of 4 days in healthy newborns, the total serum bilirubin level of up to 15–17.5 mg/dl is generally harmless and manageable with frequent breastfeeding. Phototherapy and exchange blood transfusion are advised for severe hyperbilirubinemia [14].

According to the clinical and doppler ultrasonography findings, the mentioned neonate had hemangiomas of left leg. Clinical diagnostic findings of hemangiomas were blue-red lesions with coarse and nodular surfaces and pale-pink halos (Figs 1 and 2). Doppler ultrasonography revealed superficial fast-flow vasculature within the skin lesions (Figs 3 and 4) which confirmed the diagnosis of this anomaly. The hemangiomas were identified at birth in a girl and did not begin regress during the first four week of life (Fig. 5); hence, they were diagnosed as noninvoluting congenital hemangiomas. The clinical and doppler ultrasonographic features are similar to those in the literature [1, 4–7]. The parental consanguinity was positive in the current case, suggesting a possible role of genetics in the development of CH. This hypothesis is consistent with Wojcik and Agrawal's finding that congenital abnormalities can result from a variety of genetic variations [15]. Infantile hemangiomas may be considered in the differential diagnosis of CH. They typically manifest as minimally elevated red papules or nodules after birth. In contrast to CH, infantile hemangiomas start to proliferate after a silent period of 1–3 weeks and begin rapid growth during the first few months of life [1, 6]. Based on the abnormal clinical findings, including hypotension, prolonged CRT, unpalpable radial pulse tachypnea, tachycardia, hypothermia, and cold extremities, as well as a history of bleeding from the nodular telangiectasia of a large skin lesion for three hours, the diagnosis of hypovolemic shock was made. The current case was accompanied by indirect hyperbilirubinemia and normocytic-normochromic anemia on day 17 of life, which denoted prolonged neonatal jaundice and anemia due to blood loss. The common causes of prolonged indirect neonatal jaundice are Rh or ABO incompatibility, infection, breast milk jaundice and congenital hypothyroidism [12]. Since none of these disorders were detected, CH may play a role in the pathogenesis of prolonged jaundice. This hypothesis is supported by the findings of Jung that hemangioma may cause jaundice due to hemolysis [16]. The presence of hypovolemic shock, anemia, and prolonged neonatal jaundice with NICH in our case highlights a significant difference from previously reported cases [2–5].

CONCLUSION

A large NICH may complicate critical events, including hypovolemic shock, bleeding, and anemia during neonatal period. This case of NICH on the left leg was associated with parental consanguinity. Therefore, in the pathogenesis of this anomaly, a genetic basis may be implicated. Since the current case was accompanied by prolonged neonatal jaundice, NICH may play a role in the pathogenesis of prolonged neonatal jaundice. Further analytical studies are needed to evaluate these issues.

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CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

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ETHICS APPROVAL

This study was approved by the Department of Neonatology (Protocol no 2 dates 29/4/2023), Kabul University of Medical Sciences.

CONSENT

Written informed consent was obtained from the patient's mother for the publication of this case report and accompanying images. The Helsinki Declaration was taken into consideration. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

GUARANTOR

Mansoor Aslamzai, MD, Professor of Neonatology.

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