Congenital Hemangiomas with Hypovolemic Shock, Anemia and Prolonged Jaundice in a Neonate: A Case Report.

Mansoor Aslamzai (mansooraslamzai@gmail.com)
Kabul University of Medical Sciences
Turyalai Hakimi
Kabul University of Medical Sciences
Abdul Hakim Mokhlis
Kabul University of Medical Sciences

Case Report

Keywords: Congenital Hemangioma, Shock, Anemia, Jaundice and Newborn.

Posted Date: May 18th, 2023

DOI: https://doi.org/10.21203/rs.3.rs-2886786/v1

Abstract

Background: Congenital hemangioma is an extremely rare congenital anomaly in newborn babies and may complicate life-threatening events, including bleeding.

Case Presentation: We present a very rare case of noninvoluting congenital hemangiomas along with hypovolemic shock, anemia, and prolonged jaundice in a six-day-old girl. Clinically, they were observed at birth and complicated by bleeding, hypovolemic shock, anemia, and 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 lesion. Her critical condition improved after supportive treatment, and she was discharged from the hospital. Since the index case of congenital hemangioma was associated with parental consanguinity, a genetic basis may have played a role in the pathogenesis of this anomaly. Furthermore, there will be an association between congenital hemangioma and prolonged neonatal jaundice.

Discussion and Conclusions: Congenital hemangiomas can result in life-threatening complications and may have a role in the pathogenesis of prolonged neonatal hyperbilirubinemia.

Background

Congenital hemangioma is a benign vascular tumor that is usually present at birth. They are extremely rare; however, their exact prevalence is unknown.[1,2] Congenital hemangiomas are classified into three categories: noninvoluting congenital hemangiomas (NICH), rapidly involuting congenital hemangiomas (RICH) and partially involuting congenital hemangiomas (PICH).[1-3]

Clinically, congenital hemangiomas appear 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.[1,4-6] 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.[1,6] 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 corticosteroid or beta blocker therapy, is advised for the treatment of “problematic” hemangiomas. Locations of tumors with a greater mortality rate and lesions that last until pre-school age are indications for the surgical intervention of CH.[4,6]

Case Report

A six-day-old female neonate weighing 3.5 kg was born at 38 weeks of gestation to a 22-year-old multigravida mother by vaginal delivery 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. The parents were third-degree relatives, which denoted parental consanguinity. The infant was admitted to the Neonatal Unit of the Pediatric Department due to skin lesions, bleeding, and jaundice. On general physical examination, lethargy, a rectal temperature of 34.9ºC, 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 of birth. A large, blue-red-colored lesion with a length of 14cm 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 (Figure-1). The other two smaller skin lesions with the same characteristics were observed on the dorsum of the left foot (Figure-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 gm/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 91.8fl, mean corpuscular hemoglobin of 35.8 pg), a platelet count of 317000/mm^3, a reticulocyte count of 2%, a C-reactive protein of 0.4mg/dl, a blood sugar of 89 mg/dl, a total bilirubin of 16mg/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 minutes, 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 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 (Figures -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 and was not severe, it was accepted as prolonged neonatal jaundice, and was managed by increasing the frequency of breastfeeding, but there was no indication of phototherapy in the index neonate. The hemangiomas did not regress or progress during the first four weeks of life

(Figures -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. Therefore, such management was not carried out in the index case.

Discussion

Congenital hemangioma is an extremely rare benign vascular tumor that is usually observed at birth due to an unclear etiology.[1,2,5] Based on the types of regression, CH is classified into RICH, NICH and PICH.[1-3] Usually, the diagnosis of CH is established by clinical features, and sometimes doppler ultrasonography of the lesion is used to confirm the diagnosis.[1,6] Observation care continues to be most beneficial for the majority of CH. The surgical indications for CH are tumor sites with a higher mortality rate, and those don't go away until pre-school age.[4,6]

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.[7] 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.[8-10] 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.[11,12] By the age of 4 months 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.[13]

According to the clinical and doppler ultrasonography findings, the index neonate had hemangiomas of left leg. Clinical diagnostic findings of hemangiomas were blue-red lesions with coarse and nodular surfaces and pale-pink halos (Figures -1 and 2). Doppler ultrasonography revealed superficial fast-flow vasculature within the skin lesions (Figures -3 and 4) which confirmed the diagnosis of this anomaly. The hemangiomas were identified at birth and did not regress during the first four weeks of life (Figures-5); hence, they were diagnosed as noninvoluting congenital hemangiomas. The clinical and doppler ultrasonographic features are similar to those in the literature.[1,4-6] 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.[14] 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.[1,5] 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 index 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 neonatal jaundice are Rh or ABO incompatibility, infection, and congenital hypothyroidism.[11] 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.[15] The presence of hypovolemic shock, anemia, and prolonged neonatal jaundice with CH in our case highlights a significant difference from previously reported cases.[2-4]

Conclusion

A large CH may complicate critical events, including hypovolemic shock, bleeding, and anemia. The index case of CH 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, CH may play a role in the pathogenesis of prolonged neonatal jaundice. Further analytical studies are needed to evaluate these issues.

Abbreviations

CH: Congenital Hemangioma, CRT: Capillary Refill time, CT: Computed Tomography, NICH: Noninvoluting Congenital Hemangiomas, PICH: Partially Involuting Congenital Hemangiomas RICH: Rapidly Involuting Congenital Hemangiomas.

Declarations

Ethics approval and consent to participate: This study was approved by the Department of Neonatology (Protocol no 2 dates 29/4/2023), Kabul University of Medical Sciences.

Consent for publication: 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.

Availability of data and materials: The documents used during the current study are available from the corresponding author on reasonable request.

Competing interests: The authors declare that they have no competing interests.

Funding: The authors received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Authors' contributions: MA assessed the patient's clinical features and doppler ultrasonography to make a final diagnosis, as well as prepared the manuscript. TH evaluated the newborn baby for surgical management and reviewed the manuscript. AHM performed the doppler ultrasonography. All authors read and approved the final manuscript**.**

Acknowledgements: We would like to express our gratitude to the personnel of Neonatal Unit at Maiwand Teaching Hospital, particularly Dr. Hustafa Huaryar, Dr. Mahboobulah Monshizada and Dr. Zarin for their cooperation in this study.

Authors' information: MA is a professor at the Department of Neonatology, Kabul University of Medical Sciences (KUMS). He has also been a member of the European Society for Pediatric Research since 2021. TH is an assistant professor at the Department of Pediatric Surgery, KUMS. AHM is teaching assistant at the Department of Neonatology, KUMS.

References

1. Wildgruber M, Sadick M, Müller-Wille R, Walter A. Wohlgemuth WA. Vascular tumors in infants and adolescents. Insights Imaging, 2019; 10(30). https://doi.org/10.1186/s13244-019-0718-6.
2. Braun V, Prey S, Gurioli C, Boralevi F, Taieb A, Grenier N, Loot M, Jullie ML, Léauté-Labrèze C. Congenital hemangiomas: a single-center retrospective review. BMJ Pediatr Open. 2020;4(1). http://dx.doi.org/10.1136/bmjpo-2020-000816.
3. Juan-Carpena G, Palazón-Cabanes JC, Tallón-Guerola P, Jaime MND, Betlloch-Mas I. A Case Series of 18 Congenital Haemangiomas: Clinical, Histological and Ultrasound Features, and their Relationship with Complications and Atypical Behaviour. Acta Dermato-Venereologica. 2023;103. http://dx.doi.org/10.2340/actadv.v103.3983.
4. Congenital Hemangioma. A Case Report of a Finding Every Physician Should Know. Cureus. 2018; 10(4): e2485. http://dx.doi.org/10.7759/cureus.2485.
5. Richter GT, Friedman AB. Hemangiomas and Vascular Malformations: Current Theory and Management. Int J Pediatr. 2012. https://doi.org/10.1155/2012/645678.
6. Brás S, Mendes-Bastos P, Amaro C. Rapidly involuting congenital hemangioma. An Bras Dermatol. 2017;92(6):861–3. https://doi.org/10.1590/abd1806-4841.20175608.
7. Yogen Singh Y, Katheria AC, Farha Vora F. Advances in Diagnosis and Management of Hemodynamic Instability in Neonatal Shock. Front Pediatr. 2018;6(2). https://doi.org/10.3389/fped.2018.00002.
8. Ming S, Zhang D, Chen L, Shi Y. Effects of anemia and red blood cell transfusion in preterm infants on the development of bronchopulmonary dysplasia: a propensity score analysis. All Life. 2021;14(1):830–9.
9. Kalteren WS, Bos AF, Oeveren VW. Neonatal anemia relates to intestinal injury in preterm infants. Pediatr Res. 2022;91:1452–8. https://doi.org/10.1038/s41390-021-01903-x. KooiEMW.

10. Aslamzai M, Froogh BA, Mukhlis AH, Faizi OA, Sajid SA, Hakimi Z. Factors associated with respiratory distress syndrome in preterm neonates admitted to a tertiary hospital in Kabul city: A retrospective cross-sectional study. Global Pediatr. 2023;3. https://doi.org/10.1016/j.gpeds.2023.100035.
11. Giannattasio A, Ranucci G, Raimondi F. Prolonged neonatal jaundice. Ital J Pediatr. 2015;41(Suppl 2):A36. https://doi.org/10.1186/1824-7288-41-S2-A36.
12. Tan HS, Balasubramaniam IS, HSS AS, Balasubramaniam TS, Yeong ML, Singh CC. Impact of a standardized protocol for the Management of Prolonged Neonatal Jaundice in a regional setting: an interventional quasi-experimental study. BMC Pediatr. 2019;19:174. https://doi.org/10.1186/s12887-019-1550-3.
13. Maisels MJ. Managing the jaundiced newborn: a persistent challenge. CMAJ. 2015;187(5):335–43. https://doi.org/10.1503/cmaj.
14. Wojcik MH, Agrawal PB. Deciphering congenital anomalies for the next generation. Cold Spring Harb Mol Case Stud. 2020,7;6(5). https://doi.org/10.1101/mcs.a005504.
15. Jung HL. Update on infantile hemangioma. Clin Exp Pediatr. 2021;64(11):559–72. https://doi.org/10.3345/cep.2020.02061.

Figures

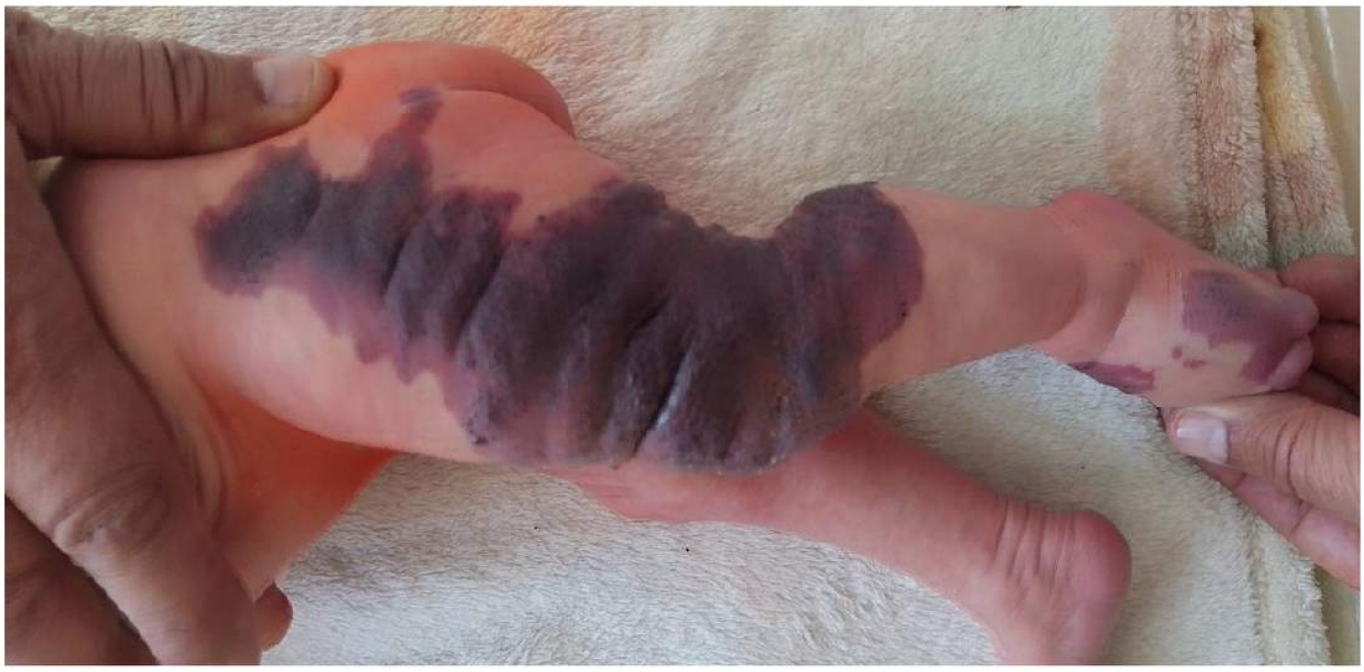

Figure 1

A large blue-red colored lesions are 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.

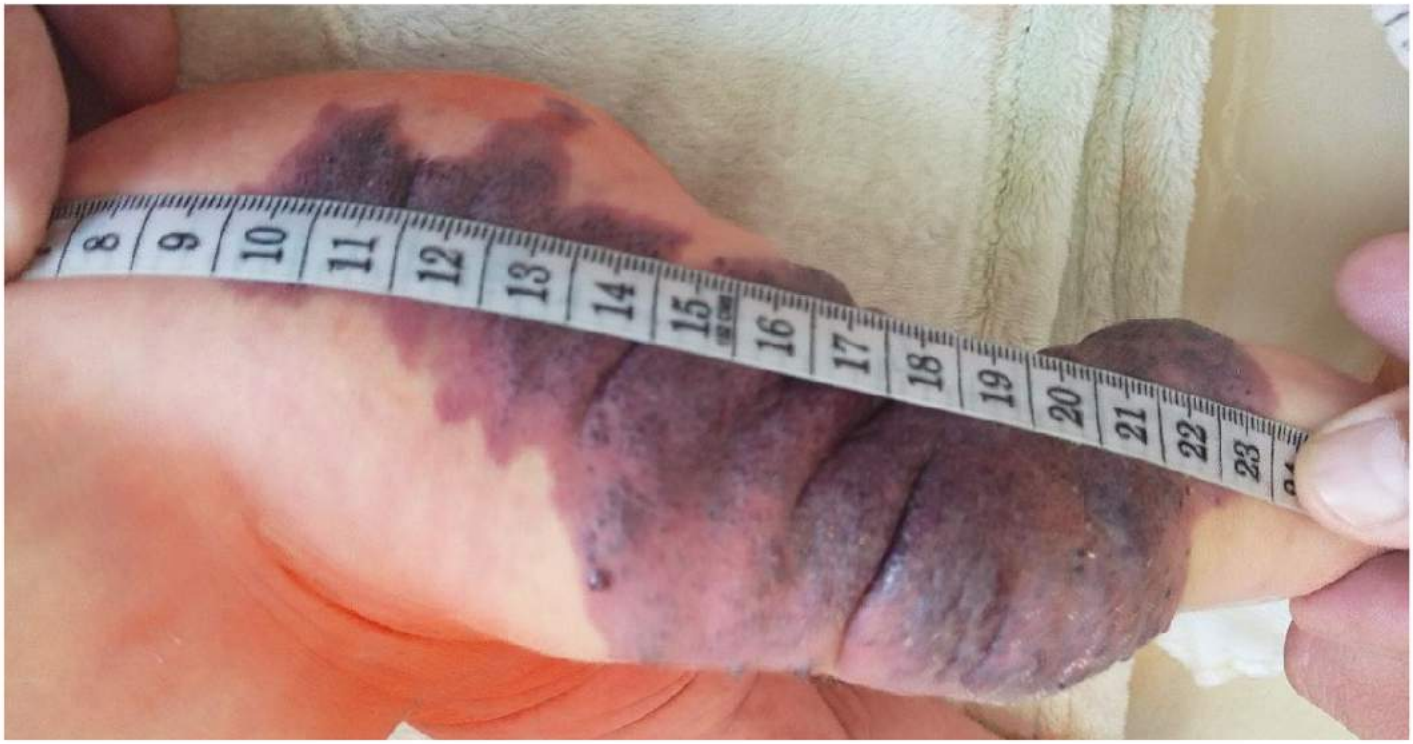

Figure 2

A large blue-red colored lesions are 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.

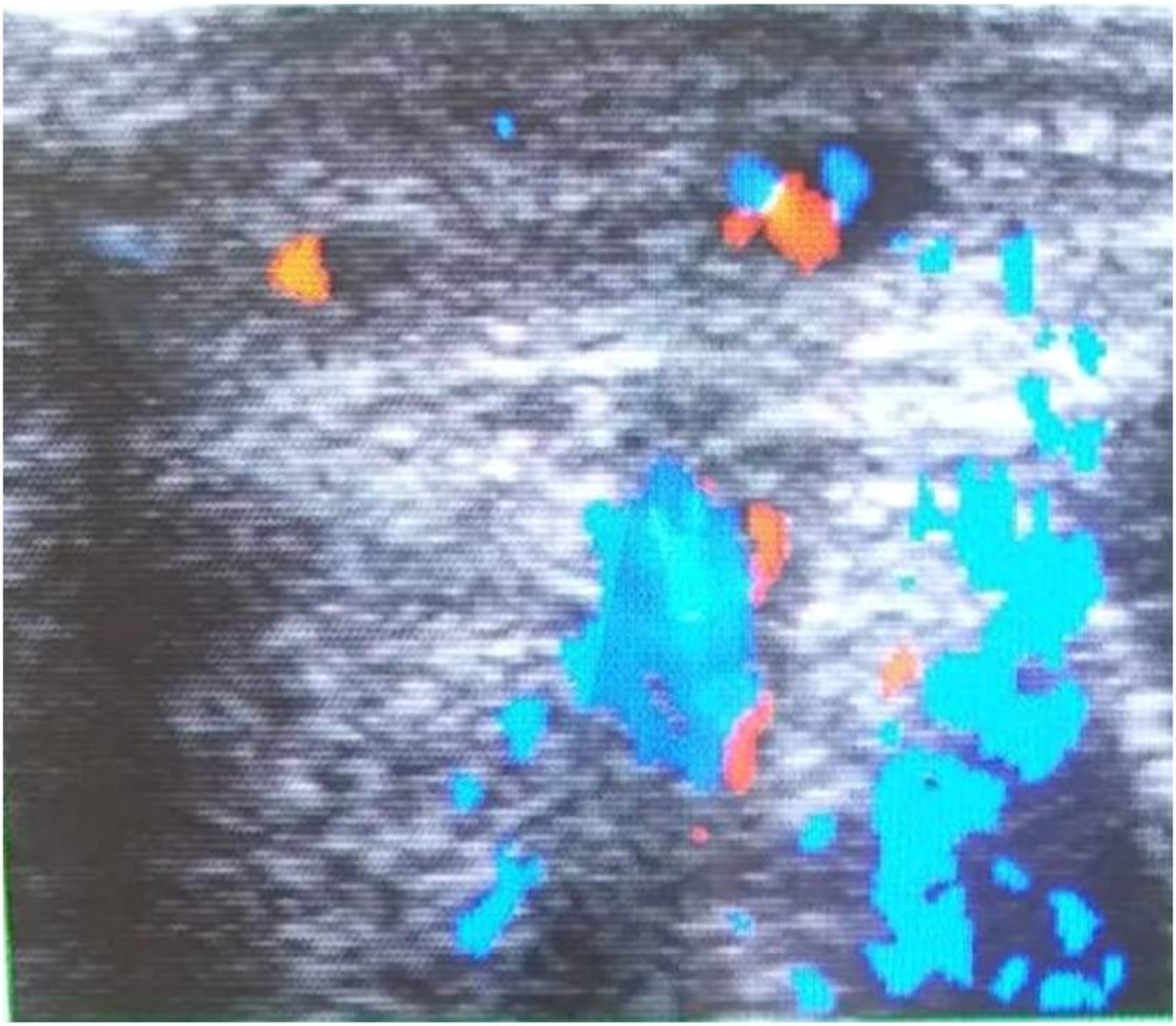

Figure 3

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

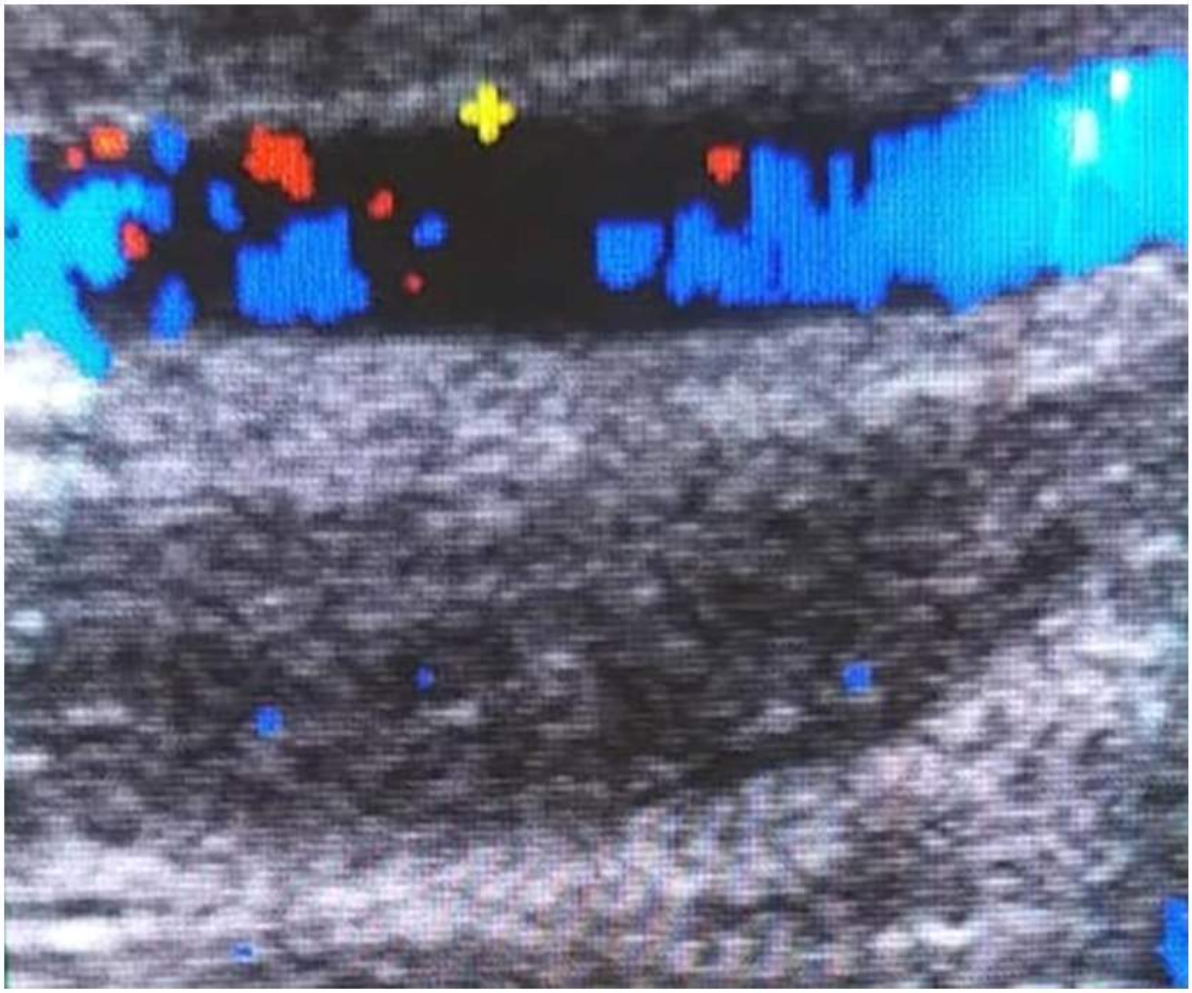

Figure 4

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

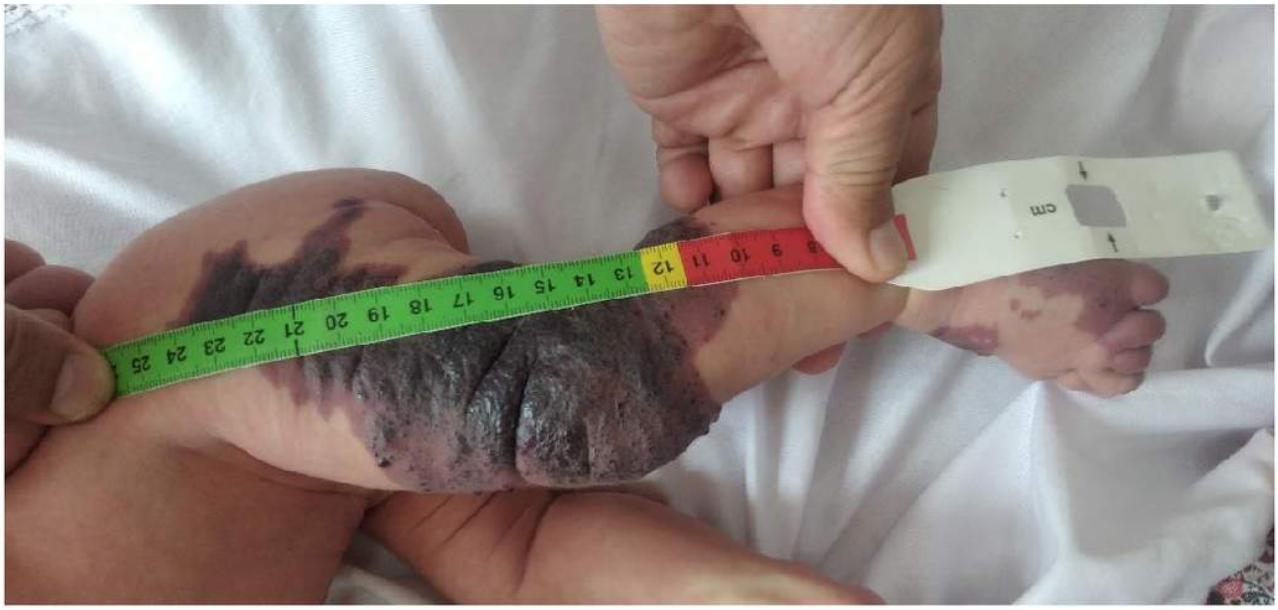

Figure 5

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