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Scarlet Enigma: A Case of Diffuse Neonatal Hemangiomatosis

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ABSTRACT

Infantile hemangioma is the most common vascular tumor in the neonatal stage, characterized by the proliferation of endothelial cells and generally exhibiting benign behavior. However, the presence of more than five skin lesions may indicate involvement of other organs, particularly the liver, where the prognosis can worsen due to associated cardiac, thyroid, and hematological complications. This study involves a case analysis and a review of current literature. The clinical case discusses a 39-day-old female infant with a history of NICU management, presenting with multiple generalized cutaneous hemangiomas from birth and an episode of gastrointestinal bleeding. Imaging revealed numerous hepatic hemangiomas and significant hepatomegaly, while laboratory tests showed coagulation abnormalities and thrombocytopenia. A diagnosis of diffuse neonatal hemangiomatosis (cutaneous and hepatic) was made, and treatment with propranolol was initiated. The prognosis for diffuse hemangiomatosis is severe due to the high risk of hemorrhagic complications if treatment is not promptly started. Therefore, we recommend initiating propranolol treatment upon evidence of such lesions, even before beginning the diagnostic protocol.

ARTICLE DETAILS

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INTRODUCTION

Infantile hemangioma is the most common vascular tumor in the neonatal stage; it is characterized by the proliferation of endothelial cells and generally exhibits benign behavior. It tends to predominate during the first two months of life, with a prevalence of 1-3% in the first days, 90% in the first month of life, and 10% in the first year, with an average age at diagnosis of 2.7 months, occurring more frequently in girls with a 4:1 ratio compared to boys. Significant risk factors for alterations in this pathology include perinatal hypoxia, maternal smoking, alcohol use, in vitro fertilization, advanced maternal age, multiple gestation, placenta previa, preeclampsia, gestational diabetes, as well as low birth weight and prematurity, with the latter being the most relevant. Lesions begin to develop during the first trimester of pregnancy, between weeks 6 and 10 of gestation, forming masses of rapidly dividing endothelial cells with or without lumens, with a multilaminated basal membrane. During the initial phase, this lumen will dilate, and the endothelial cells will thin, depositing fibrous tissue. Those that involute will contain capillaries and veins with a flattened endothelium in a stroma of fibrous tissue, collagen, and reticular fibers. Pluripotent cells (CD13 and CD4) are generated, which proliferate and differentiate into endothelial progenitor cells as a result of their abnormal and disordered proliferation. This vascular growth interacts with estrogens and progesterone in various vascular anomalies, which explains the predominance in females. Hemangiomas can be classified by depth as superficial, deep, or mixed, by distribution pattern as focal, multifocal, segmental, or indeterminate, and by the risk of sequelae and complications as high, intermediate, or low, with the most commonly

affected sites being the trunk, head, and neck, although they can also include mucous membranes and other organs. Superficial hemangiomas may be only in the superficial dermis, deep ones may extend to subcutaneous tissue, and mixed ones include both types together. Generally, they exhibit benign behavior; however, the presence of more than five skin lesions may indicate involvement of other organs, especially the liver, where the prognosis can worsen due to associated cardiac, thyroid, and hematological.

CLINICAL CASE

A female newborn, late preterm at 35 weeks of gestation and 24 days of extrauterine life, with a history of neonatal intensive care unit management due to neonatal respiratory distress syndrome, ABO incompatibility, late neonatal sepsis, suspected aortic coarctation, and thrombocytopenia.

The infant was the product of the second gestation with an uncomplicated delivery, appropriate weight for gestational age, and subsequent onset of progressive respiratory distress requiring ventilatory support until orotracheal intubation at 15 days of life, along with antibiotic treatment with vancomycin, caspofungin, and meropenem due to suspected sepsis.

Upon admission, more than 20 bright red maculopapular dermal lesions of varying sizes were observed, the largest measuring 13 x 8 mm, with well-defined borders, distributed including palms and soles. Their presence was noted since birth with apparent gradual growth and the appearance of new lesions.

As part of the approach to neonatal hemangiomatosis, an abdominal ultrasound was requested to check for visceral involvement, reporting countless hemangiomas, the largest measuring 20 x 13 mm, along with significant hepatomegaly extending to the right hypochondrium with a mass effect on the spleen and stomach, without evidence of hemangiomas in the kidneys, spleen, or gallbladder. Liver function tests showed normal transaminases, elevated bilirubins with a cholestatic pattern, and elevated alkaline phosphatase and lactate dehydrogenase, leading to the initiation of ursodeoxycholic acid.

Since hospital admission, the patient presented with significantly prolonged coagulation times (PT 15.3 sec, aPTT 64.6 sec), requiring transfusion of fresh frozen plasma on two occasions, as well as vitamin K, and persistent thrombocytopenia despite antimicrobial treatment including antifungal, requiring transfusion twice. Due to persistent thrombocytopenia (65,000), studies were conducted to rule out infectious causes such as TORCH, which were negative. A note of referral mentioned an episode of coffee-ground bleeding through an orogastric tube at 48 hours of life, managed with omeprazole, with no new bleeding episodes or hemoglobin decrease.

Chest X-rays showed cardiomegaly with a thoracic index of 0.65, with apparent right chamber enlargement, and on physical examination, a grade III plurifocal murmur was auscultated, prompting a pediatric cardiology evaluation. An echocardiogram concluded mild pulmonary stenosis, with the rest of the heart structurally sound, not requiring treatment at the moment.

The patient was referred to a tertiary care unit for multidisciplinary management and suspected visceral involvement. During the stay, a new pediatric cardiology evaluation reported a patent foramen ovale with preserved ventricular function and mild pulmonary hypertension, and norepinephrine was initiated due to reported hypotension. A transfontanelar ultrasound reported grade II intraventricular hemorrhage, with preserved choroid plexus anatomy and no space-occupying masses, with the last report showing no alterations. A thyroid profile was requested due to its association with hypothyroidism, but thyroid hormones were reported within normal age parameters: TSH 7.3, T4L 2.45, total T4 14, T3L 2.7, total T3 1.1.

Before transfer to tertiary care, a thoracoabdominal CT scan was performed, reporting increased pulmonary hilum vascularity, without hyperdense masses suggestive of hemangiomas. A diagnosis of probable diffuse neonatal hemangiomatosis was made, and treatment with propranolol at a dose of 0.5 mg/kg/day and prednisone 2 mg/kg/day was initiated. However, upon admission to tertiary care, both treatments were suspended and only propranolol was resumed at 32 days of life, with the dose increased to 1 mg/kg/day, as indicated by the pediatric dermatology service.

Pediatric ophthalmology evaluation reported Zone 2, stage 0 in both eyes, and the cardiology service diagnosed a patent foramen ovale, without chamber dilation, with preserved biventricular function, no cardiovascular deterioration, and no need for any management. The approach to multiple diagnoses was initiated: disseminated hemangiomatosis (cutaneous and hepatic hemangiomatosis, thrombocytopenia under study, cholestatic syndrome, bronchopulmonary dysplasia, and grade II intraventricular hemorrhage).

After evaluation at the tertiary level, the patient was readmitted to the referral unit, and on physical examination, an increase in the number of hemangiomas was observed, affecting the face and back of the hands.

DISCUSSION

The prognosis for disseminated hemangiomatosis is severe, with a high mortality rate of 50% to 95%. The main cause of death is high-output heart failure due to hepatic hemangiomas causing high blood flow through arteriovenous shunts from left to right. Other complications can include liver failure, which includes consumption coagulopathy, as well as cutaneous, gastrointestinal, and cerebral bleeding. Due to the rapid progression, early intervention is aimed at stopping this progression or inducing lesion regression, reducing mortality to 27%.

The basic treatment usually focuses on corticosteroids, interferon alpha-2b, vincristine, propranolol, and non-pharmacological options like radiotherapy, embolization focused on focal lesions with shunts to improve refractory heart failure to treatment, or surgery with a high percentage of complications.

The patient presented with multiple cutaneous lesions and lesions in the lung, liver, and bile ducts, warranting timely treatment initiated with steroids and propranolol, later managed only with the latter. Upon referral to tertiary care, differential diagnoses considered included blueberry muffin baby syndrome and Kasabach-Merritt syndrome due to generalized hemangiomatosis associated with thrombocytopenia, prolonged coagulation times, and visceral involvement. However, these were ruled out as the type of lesions did not match the characteristics of both entities, and thrombocytopenia resolved after the antibiotic regimen, with no significant lesion growth or bleeding episodes. However, after returning to our unit, an increased presence of hemangiomas was observed, likely associated with the late initiation of propranolol and its suspension during the 8-day transfer to tertiary care, with more than twenty-five hemangiomas predominantly on the head, face, and trunk, although without growth of previously existing ones. We consider this highlights the importance of timely, adequate, and continuous treatment for this condition.

CONCLUSIONS

Diffuse neonatal hemangiomatosis highlights the complexity and the need for a multidisciplinary approach to address the multiple associated complications. The patient presented a series of clinical challenges, including cutaneous and hepatic hemangiomas, persistent thrombocytopenia, cholestatic syndrome, bronchopulmonary dysplasia, and grade II intraventricular hemorrhage. Despite initial interventions, such as the use of propranolol and the suspension of prednisone, management in a tertiary care unit allowed for reevaluation and adjustment of treatment, underscoring the importance of an individualized and coordinated approach. The stabilization of cardiac function and the absence of cardiovascular deterioration were significant achievements, although the increase in the number of hemangiomas highlights the progressive nature of the condition. This case illustrates the importance of early detection and timely treatment to minimize complications and improve prognosis in patients with diffuse neonatal hemangiomatosis.

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IMAGES

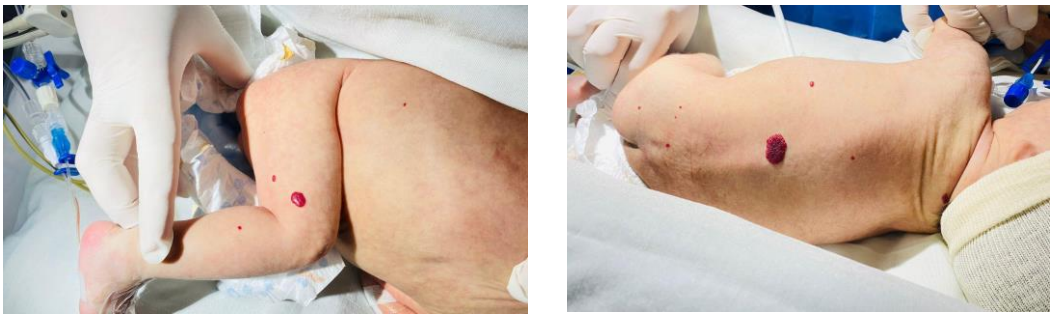

Figure 1. Presence of hemangiomas on the back and right leg upon admission to the NICU.

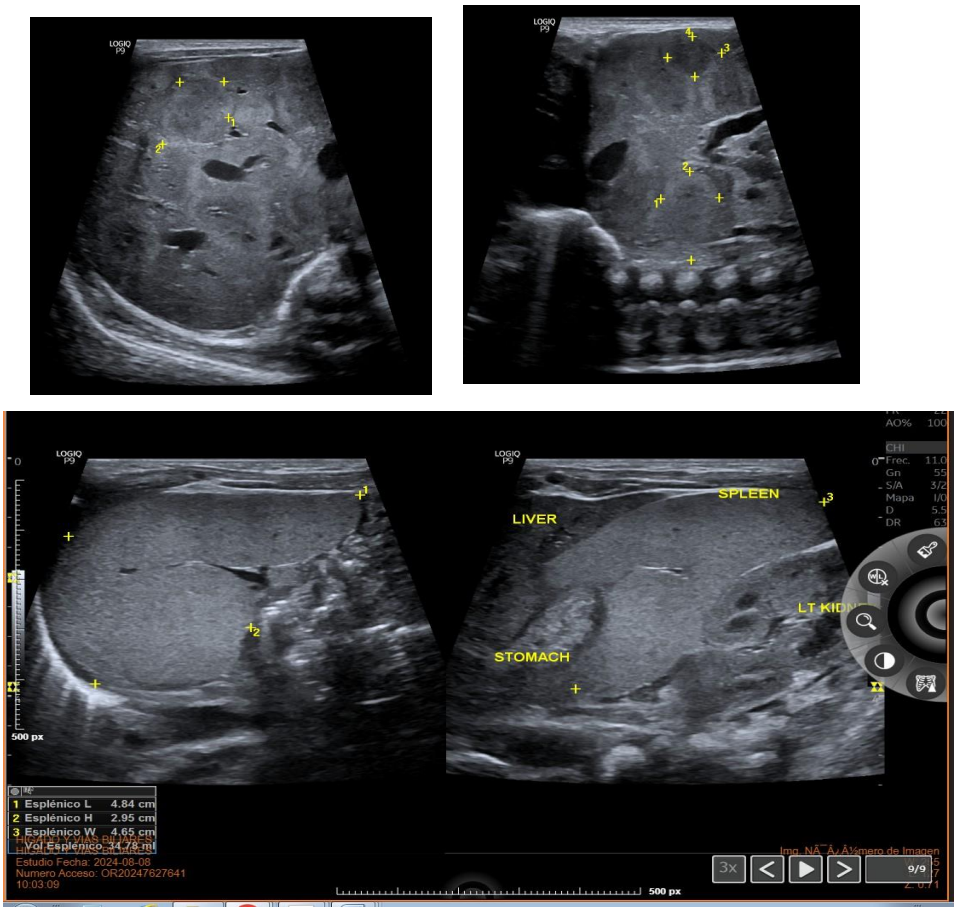

Figure 2. Liver: Heterogeneous parenchyma with countless iso-hypoechoic lesions with lobulated hyperechoic borders, the largest in the caudate lobe measuring 20 x 31 mm, consistent with transmission diameter; hepatomegaly with a longitudinal axis of 61 mm and extension to the left hypochondrium with mass effect on the spleen and stomach.

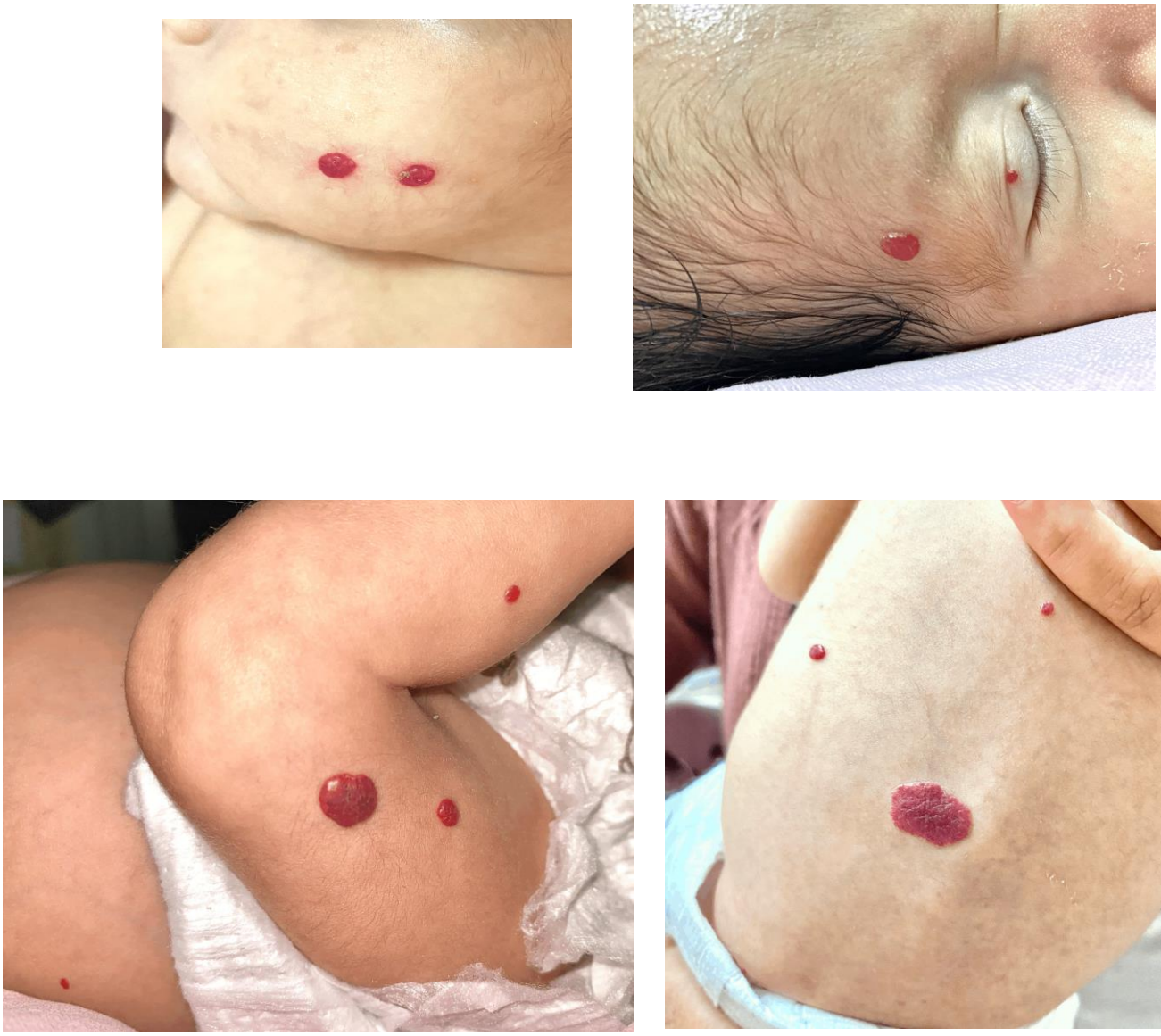

Figure 3. Lesions after evaluation at the tertiary level. A) Two superficial hemangiomas on the left cheek. B) Superficial hemangiomas in the supraciliary and right palpebral region. C) Superficial hemangioma on the right thigh and leg. D) Three hemangiomas on the back.