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CASE REPORT

Clinical diagnosis of disseminated neonatal hemangiomatosis: a case report.

Wallace William da Silva Meireles[1,2], Janinne Barboza Rangel[1,2], Graziela Paronetto Machado Antonialli[1,2], Cristina Touguinha Neves Medina[1,2], Romina Soledad Heredia Garcia Silva[1,2]

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Abstract

Hemangioma is the most common vascular tumor of childhood. It may present as multiple cutaneous hemangiomas on rare occasions, with or without visceral involvement. The condition of lesions restricted to the skin is called benign neonatal hemangiomatosis, and in the case of visceral lesions associated with cutaneous lesions, the denomination is disseminated or diffuse neonatal hemangiomatosis (DNH). For the diagnosis of DNH, three criteria must be met: onset in the neonatal period, involvement of three or more organs, and absence of malignancy in hemangiomas. ASAS, a full-term, vaginally delivered newborn, weight: 3560g, length: 52cm, head circumference: 35cm, APGAR: 9/9. Maternal history of syphilis during pregnancy - mother adequately treated in pregnancy. At birth, she presented with purpuric lesions of variable size on the chest, limbs, feet, and hands, which discolored under digital pressure; larger lesions with relief, not scaly. He developed severe heart failure and was admitted to the Neonatal Intensive Care Unit (NICU) in a secondary hospital in the Federal District. Imaging tests were performed: Computed tomography of the skull, thorax, and total abdomen, which revealed the presence of a possible hemangioma in the left frontal lobe, telangiectasias in the lungs, and multiple hepatic hemangiomas. He had a clinical diagnosis of DNH. The case described corroborates the fact that knowledge about the disease is relevant, as early diagnosis can lead to more accurate approaches that may reduce morbidity and mortality from the disease.

[1] Escola Superior de Ciências da Saúde (ESCS), Residência em Genética Médica - Brasília - Distrito Federal – Brazil.
[2] Hospital Materno-Infantil de Brasília (HMIB), Unidade de Genética Médica - Brasília - Distrito Federal - Brazil.

Endereço para correspondência:
Wallace William da Silva Meireles.
Escola Superior de Ciências da Saúde. ESMHN Quadra 3 Conjunto A Bloco 01 Edifício Fepecs, SMHN Conjunto A Bloco 01 Edifício Fepecs Asa Norte, Brasília/DF, 70710-907
E-mail: wallacemeireles@hotmail.com

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INTRODUCTION

Hemangioma is the most common vascular tumor and benign soft tissue tumor in children[1 2 3]. On rare occasions, it may present as multiple cutaneous hemangiomas, with or without visceral involvement. The condition in which lesions appear solely on the skin is called benign neonatal hemangiomatosis, whereas the one in which visceral lesions appear in association with skin lesions is called diffuse neonatal hemangiomatosis (DNH)[4]. Patients with the benign form of hemangiomatosis usually present rapid involution of the tumors and with a good prognosis, while individuals with DNH may have poorer outcomes[1].

Three criteria must be met for a patient to be diagnosed with DNH, namely: onset in the neonatal period; involvement of three or more organs; and absence of malignancy in the hemangiomas[1]. Investigation of visceral lesions is warranted in children with five or more skin hemangiomas suspected of DNH.

The etiology of DNH is still undefined. Some back up the idea of multifactorial inheritance, while the possibility of autosomal dominant inheritance is also considered. Regardless of the type of inheritance, it is currently believed that hemangiomas are the result of an imbalance in angiogenesis that causes the uncontrolled proliferation of vascular elements[1].

The treatments proposed for the condition include systemic beta-blockers (especially propranolol), topical beta-blockers (such as timolol), corticosteroids, sirolimus (a mTor pathway inhibitor) and vascular embolization[1 6 7 8].

CASE REPORT

The case described in this report involves an infant born from a vaginal delivery weighing 3560g, with a length of 52cm, a head circumference of 35cm, and an Apgar score of 9/9. His mother had syphilis during pregnancy, for which she was adequately treated. At birth, the infant had purpuric lesions of varying sizes on the chest, limbs, feet, and hands, which discolored upon digital pressure. Larger lesions were non-scaling and could be palpated (Figures 1 and 2). The patient developed severe congestive heart failure (CHF) and was admitted to the Neonatal Intensive Care Unit (NICU) of a secondary hospital in the Federal District of Brasília. Imaging tests performed included computed tomography (CT) of the head, chest, and total abdomen, which showed a potential hemangioma in the left frontal lobe of the brain, telangiectasias in the lungs, and multiple hepatic hemangiomas. The patient was diagnosed with DNH.

Twenty-seven days after birth, the infant developed respiratory failure. He was intubated and placed on a ventilator. The patient went into cardiorespiratory arrest twice, which resulted in focal lesions secondary to hypoxic-ischemic injury to the central nervous system. At the time, a Doppler echocardiogram showed left ventricular hypertrophy



Figure 1. Neonatal patient with multiple hemangiomas on the face, trunk, and limbs.



Figure 2. Close-up view of the patient's feet with hemangiomatous lesions, also involving the plantar region.

and dilation of cardiac chambers, elevated cardiac output, pulmonary hypertension, and a patent foramen ovale.

The patient was started on propranolol (2mg/kg/day) one month after birth and, two days later, prednisolone (3mg/kg/day) was introduced, causing a partial involution of the hemangiomatous lesions. At two months of age, he presented with generalized tonic and clonic seizures caused by refractory hypoglycemia, without evidence of an inborn error of metabolism or endocrinological disorder, which led to the consideration of hyperinsulinism secondary to the use of corticosteroids, low liver reserve (secondary to hepatic hemangioma) and low-calorie intake.

At three months of age, given the patient's favorable progression with propranolol, the medical team, after discussing the case with the vascular tumor team of a tertiary hospital in the state of São Paulo, decided to increase the dose of propranolol to 3mg/kg /day, gradually decrease the dose of prednisolone until it was completely removed (due to the risk of immunosuppression), and add vincristine (0.05 mg/kg/week) to the treatment.

The infant was discharged at four months of age and placed on outpatient follow-up with a multidisciplinary team from the cardiology, neurology, hematology, hepatology, and medical genetics departments.

At five months of age, an ultrasound examination to control for liver lesions found extensive venous thrombosis. At ten months of age, a CT scan of the upper abdomen revealed the disappearance of the hepatic nodules, the appearance of chronic thrombosis in the middle hepatic vein and the portal branch into segment VIII, the appearance of chronic thrombosis in the splenic vein with splenomegaly and densely packed lymph nodes in the root of the mesentery. At this time (Figures 3 and 4), he no longer had clinical signs suggestive of CHF, and no alterations were seen in the echocardiogram. The infant was on spironolactone, atenolol, ursodeoxycholic acid, furosemide, amlodipine, and phenobarbital.

The patient will be re-evaluated by a multidisciplinary team for the need for embolization of vascular lesions.

COMMENTS

Since the differential diagnosis of DNH includes conditions such as multifocal lymphangioendotheliomatosis with thrombocytopenia and kaposiform hemangioendothelioma, as described by Glick et al., and DNH can develop into a severe condition, cases of the disease must be described, evaluated and studied[9].

The patient met the clinical criteria for disseminated neonatal hemangiomatosis and was diagnosed at an early age, which facilitated the use of more assertive therapies. The case described corroborates the study by Santos et al., which demonstrates that knowledge about the disease is relevant, as early diagnosis can guide better approaches to decrease the morbidity and mortality of the condition[10].



Figure 3. Patient at 10 months of age, with involution of hemangiomatous lesions after treatment.



Figure 4. Close-up view of the patient's left arm at 10 months of age, with involution of hemangiomatous lesions after treatment.

Furthermore, the medications prescribed to manage the patient's symptoms (especially propranolol and prednisolone) are also used to treat the disease, as described by Bau et al[2]. In the case in question, in addition to propranolol and corticosteroids, the infant was also prescribed vincristine, unlike the case described by Iribarren et al., in which treatment was performed only with propranolol, with good clinical response and reductions in skin and visceral lesions[11].

The perception of the need and importance of multidisciplinary teamwork for the diagnosis and management of patients with DNH is equally relevant, as is also observed by Iribarren et al[11].

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