Segmental Beard Hemangioma

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ABSTRACT

Infantile hemangiomas arise from the proliferation of vascular endothelial cells and represent the most common benign tumors in infancy, with an estimated incidence of 4-10% in Caucasian infants. They vary according to their number, depth, and distribution. Within the latter classification are the so-called segmental ones, which feature an extensive distribution in areas of embryonic mesodermal extensions. We report the case of a patient evaluated at one and a half months of life with an extensive hemangioma of the mandibular area and anterior neck (segmental hemangioma of the beard). We describe the importance of complementary studies for evaluating the involvement of underlying organs, detecting associated syndromes, and defining the treatment based on these findings.

Key words: infantile hemangiomas, segmental beard hemangioma, ulceration, propranolol, PHACES.

INTRODUCTION

Infantile hemangiomas (IH) represent the most common benign vascular tumor in childhood, with an incidence of 4 to 10% in the first year of life. They are more frequent in the Caucasian population and female patients. Advanced maternal age, prematurity, low birth weight, and factors related to placental insufficiency, among others, have been described as risk factors[1,2].

IHs can be classified by number into single or multiple; by depth, in superficial, deep, or combined; and by distribution, in focal, segmental, or mutifocal[3]. Although they can appear anywhere on the tegument, they have predominance on the head and neck[3,2]. Segmental hemangiomas are those that occupy a large surface and involve areas of embryonal mesodermal extensions, have a worse prognosis, and are more prone to complications (the most frequent being ulceration), thus requiring more intensive and prolonged therapy, with a worse overall outcome[4]. On the other hand, they are more likely to be associated with extracutaneous anomalies. Such is the case of segmental hemangiomas of the beard, which are extensive hemangiomas of the mandibular area and anterior neck, which present more risk of concomitant airway hemangioma and PHACES syndrome[5].

CLINICAL CASE

Female patient first evaluated at one month and 28 days, born at full term of an uncomplicated pregnancy, with adequate weight for gestational age. She had no skin lesions at birth. At one week of life, she began to develop a fast-growing violaceous tumor lesion on the lower lip, with ulceration after the fifth week (Fig. 1 A, B, C). Concomitantly, reddishstrawberry-like tumors appeared on the anterior face of the neck and presternal region (Fig. 2). The patient had an electrocardiogram, an echocardiogram, and a cerebral and abdominal ultrasound within normal limits, requested by her

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Received: 02/13/23 Accepted: 06/01/23 Online: 06/30/2023

DOI: http://doi.org/10.51987/revhospitalbaires.v43i2.283

How to cite: Ruf M, Di Prinzio A, Martínez MF, Angles MV y Mazzuoccolo LD. Segmental Beard Hemangioma. Rev. Hosp. Ital. B.Aires. 2023;43(2):98-101.

ISSN (Online) 2314-3312

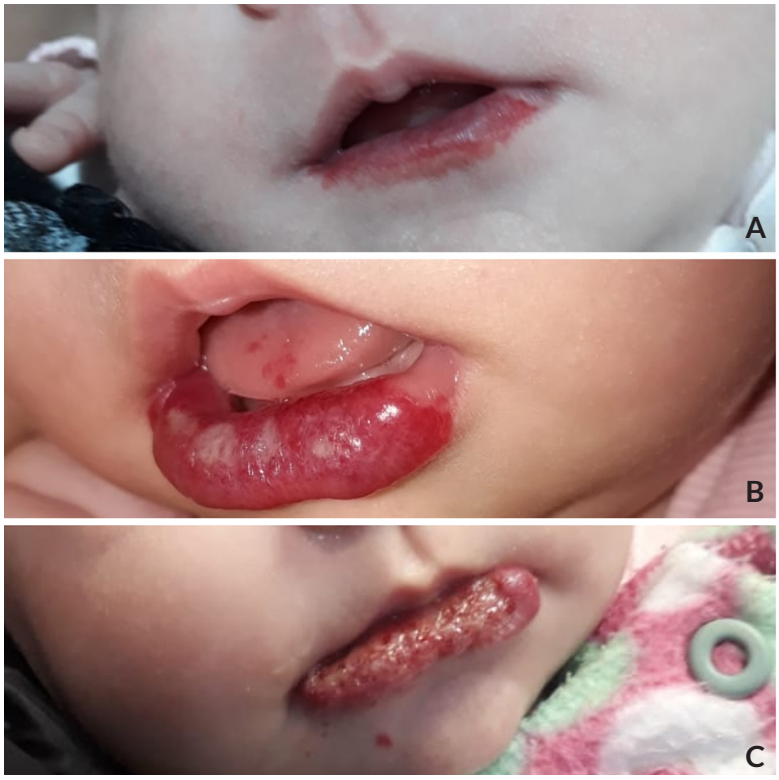

Figure 1 A. First week of life. Red-violet macula involving the vermilion border of the lower lip, slightly overlapping it. **B.** Fourth week of life. Violaceous tumor with a pale center, indicative of imminent ulceration. An extension of the lesion on the tip of the tongue is visible. **C.** Sixth week of life. The ulcerated violaceous tumor.

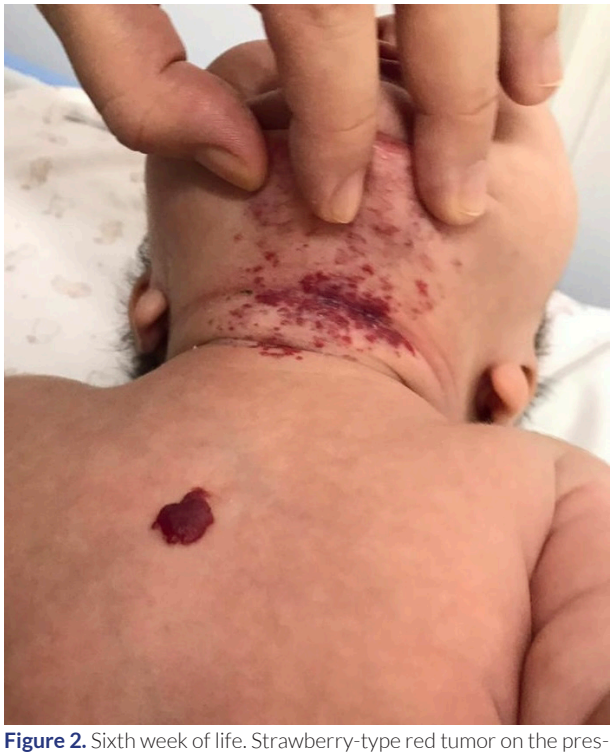

Figure 2. Sixth week of life. Strawberry-type red tumor on the presternal region and plaque-type red tumor on the anterior aspect of the neck.

attending pediatrician.Due to the location of the lesion, we ordered an ENT, and as there were no respiratory symptoms, it ruled out airway compromise for the time being.In addition, we requested complementary studies to rule out the PHACES syndrome. MRI of the brain with contrast, a magnetic resonance angiography (MRA) of the brain and neck vessels, and a nuclear magnetic resonance of the thorax showed no alterations. We decided to start propranolol at 1 mg/kg/day.The first dose was administered in the consulting room with heart rate and blood pressure monitoring before and one hour after administration. Due to good tolerance, at 24 hours, the dose was increased to 2 mg/kg/day. Improvement of the lesions became evident after 48 hours, with significant reduction and complete epithelialization of the lip ulceration after one month of treatment (Fig. 3 A and B).We maintained the dosage as the child gained weight until reaching a propranolol dose of 1 mg/kg/day at one year of age. Four months later, we decided to suspend it, continuing with topical timolol to avoid relapse. However, two months after discontinuation, there was evidence of increased erythema in the affected area but no ulceration; we interpreted this as a relapse of the hemangioma due to a rebound effect, so propranolol was started again at 1 mg/kg/day. At 33 months, we discontinued the medication; currently the patient is 43 months old and continues without relapse.

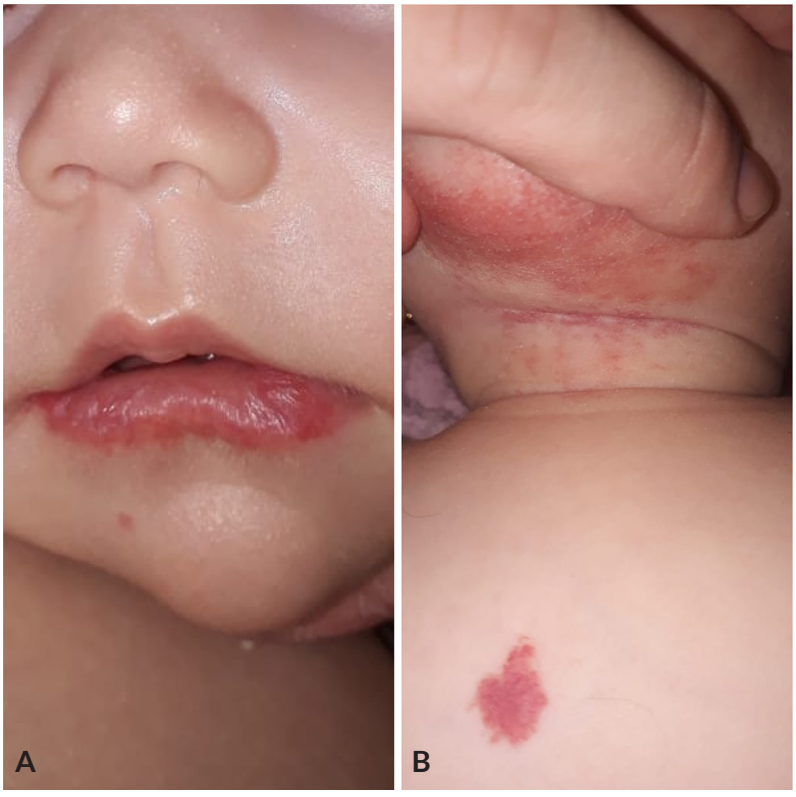

Figure 3. A. Control one month after initiation of propranolol. Resolution of ulceration, clearing, flattening, and decrease of the size of the hemangioma. **B.** Control at one month after initiation of propranolol. Clearing and flattening of the hemangioma.

DISCUSSION

IHs are considered benign vascular tumors, according to the classification made by the ISSVA (International Society for the Study of Vascular Anomalies)[3]. They are not usually present at birth and appear during the first days or weeks of life. In their natural course, three phases can be distinguished: a proliferative phase, which reaches its maximum growth peak at three months of life; a plateau phase and a phase of spontaneous involution, which frequently begins after one year of life and can extend up to 10 years of age[1,2,6].

High-risk or complicated hemangiomas refer to those that cause dysfunction of a vital organ, bleeding, ulceration (with risk of superinfection), or risk of permanent disfigurement[1].

The importance of evaluating the pattern of involvement (focal, multifocal, or segmental) and the location (e.g., mandibular) lies in being able to detect those hemangiomas that may be part of an underlying syndrome[7].

Hemangiomas are diagnosed clinically and do not usually require the completion of complementary studies, except in specific cases that generate diagnostic doubt or in which systemic associations are suspected, as in our patient[1].

Segmental hemangiomas of the face have different patterns: those located in the frontotemporal (segment 1), maxillary (segment 2), mandibular (segment 3), and frontonasal (segment 4) areas. In the presence of a segmental hemangioma in the beard area (S3), as in the case presented, PHACES syndrome and association with airway hemangiomas should be ruled out[2,8]. The latter should be considered in any child who has a hemangioma in that location and develops respiratory symptoms, such as progressive hoarseness or stridor. Laryngoscopy is the study of choice for its detection1. PHACES syndrome is rare and named after its acronym: P: posterior fossa anomalies (Dandy-Walker complex and/or cerebellar hypoplasia or dysgenesis); H: segmental hemangiomas of the face A: intracranial or extracranial arterial anomalies; C: cardiac or aortic defects (coarctation of the aorta is the most frequent alteration); E: ocular anomalies; S: supraumbilical raphe or sternal agenesis[1,5].To rule out this syndrome, we suggest requesting brain magnetic resonance imaging with and without contrast, magnetic resonance angiography, echocardiography, Doppler ultrasound

of the neck vessels, ophthalmologic evaluation, and an abdominal ultrasound to evaluate the presence of hepatic hemangiomas[1].

In uncomplicated IH, the treatment of choice is watchful waiting or topical therapies (topical beta-blockers, corticosteroids or imiqui mod). In high-risk or complicated hemangiomas, a systemic treatment is usually indicated. Propranolol is currently considered the first-line treatment.Generally, it starts with a dose of 1 mg/kg/day or lower, and if well tolerated, it is increased to 2 or 3 mg/kg/day divided into 2 or 3 doses[1,7]. In cases of ulcerated IH or with PHACES syndrome, initiation of propranolol at a lower dose (≤ 1 mg/kg/d) should be considered; in the former scenario, we suggest such dosage as longer healing times have shown to occur with doses higher than 1 mg/kg/day compared to lower doses, and in PHACES syndrome lower doses minimize abrupt fluctuations in blood pressure that may increase the risk of stroke[9-11].

Depending on the patient's age, weight, and comorbidities, among other factors, doctors will determine whether the first administration will be on a supervised outpatient basis or will require hospitalization. In our case, since it is a segmental hemangioma of the beard, we chose to administer the medication in the consulting room under the monitoring of vital signs, with monitoring of blood pressure and heart rate[1].

Propranolol is an effective treatment in most cases; however, relapse after its suspension has an estimated frequency of 19-25%. The predictive factors for this are discontinuation of therapy before nine months of life, deep involvement, female sex, head and neck region localization, and segmental pattern[11]. Our patient fulfilled three of the predictive factors, which would explain the regrowth after the first discontinuation of the treatment.

Other therapeutic alternatives are systemic corticosteroids, vincristine, and interferon alpha. In specific situations, pulsed light laser, surgical excision, and embolization could be helpful[1,11].

CONCLUSIONS

We presented the case of a patient with segmental hemangioma of the beard, illustrating the importance of performing studies for detecting extracutaneous involvement in the presence of segmental hemangiomas in specific locations. In this case, it was a way to rule out PHACES syndrome and airway involvement, which may be present in hemangiomas with distribution in the mandibular area.

In addition, it's worth noting that the patient experienced the development of lower lip ulceration in association with the rapid growth of the hemangioma, a complication more frequent in segmental hemangiomas, especially those in that area. Therefore, the timely treatment helped the scarring, improving the symptomatology and probably avoiding worse esthetic sequelae.

Conflict of interest: the authors declare no conflict of interest.

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