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Case Report

Persistent Congenital Hemangioma

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KEYWORDS

Vascular lesion,
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RICH,
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NICH

ABSTRACT

The author present a case of a 23-month-old girl with a vascular soft and violaceous neoformation located in the frontal region at the level of the glabella. Since the parents reported that the lesion had been present since birth, the diagnosis of infantile hemangioma was ruled out. The persistence of the lesion at 12 months still did not allow for a definitive distinction between Non Involuting Congenital Hemangioma (NICH) and Rapid Involuting Congenital Hemangiona (RICH). However, its persistence at 23 months confirmed the definitive diagnosis of NICH.

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Introduction

Congenital hemangioma is a rare benign vascular tumor, typically characterized by a violaceous coloration and medium to large size, that develops during intrauterine life. Based on its pattern of evolution, congenital hemangiomas are classified into three subtypes:
RICH (Rapidly Involuting Congenital Hemangioma)
NICH (Non-Involuting Congenital Hemangioma)
PICH (Partially Involuting Congenital Hemangioma)

A differential diagnosis among these subtypes can be made after the first 12 months of life. RICH lesions undergo complete involution by this time, whereas NICH remain stable in size. PICH lesions initially regress rapidly during the first year, but then involution halts and the lesion remains unchanged without disappearing, eventually becoming indistinguishable from NICH (1).

Case description

A 23-month-old girl was admitted to the clinic for evaluation of a neoformation measuring approximately 1.5 cm in diameter, located in the frontal region at the level of the glabella. The lesion was elastic in consistency, mobile over the underlying planes, and exhibited a violaceous coloration (Fig. 1). A previously performed ultrasound revealed an anechoic content.

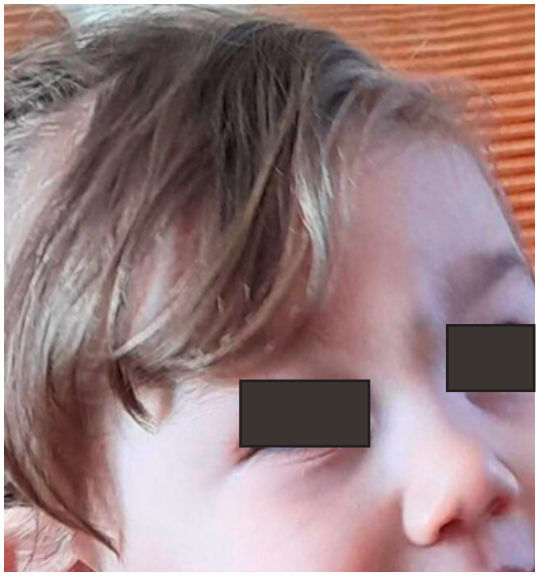

Fig. 1. *The patient at the age of 23 months.*

The initial suspected diagnosis, based primarily on the lesion's color and size, was subcutaneous infantile hemangioma. However, following a thorough medical history, this diagnosis was reconsidered, as the parents reported that the lesion had been present since birth.

Infantile Hemangiomas, whether superficial, subcutaneous, or mixed, are never present at birth. They usually appear in the first weeks of life, go through an initial proliferative phase in the first 4-5 months, followed by a maturation phase that extends up to 12 months of age. Subsequently, a regression phase ensues, usually concluding between 7 and 9 years of age. In this case, the parents were asked to provide photographic documentation to assess the lesion's evolution over time (Fig. 2, 3, 4).

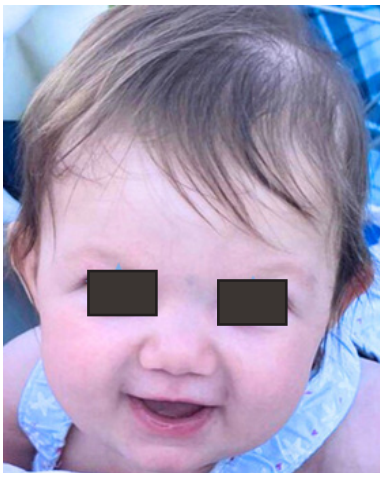

Fig. 2. *The patient at the age of 4 months.*

Fig. 3. *The patient at the age of 9 months.*

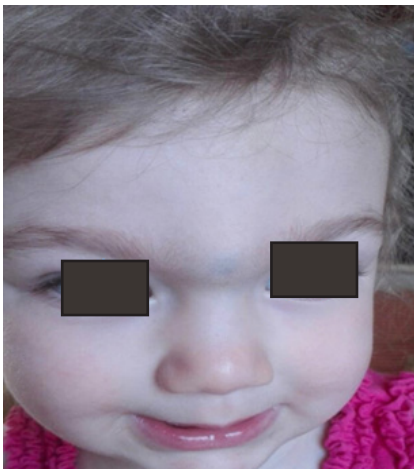

Fig. 4. *The patient at the age of 12 months.*

The earliest available images date back to the age of four months, when the lesion was evident but inconclusive for diagnosis. The persistence of the lesion at 12 months still did not allow for a definitive distinction between NICH and RICH. However, its persistence at 23 months confirmed the definitive diagnosis of NICH.

Discussion

Non-Involuting Congenital Hemangiomas (NICH) are characterized by their stability over time, showing neither expansion nor regression. In approximately 10% of cases, postnatal growth may occur, usually proportional to the child's somatic development.

Morphologically, two clinical variants of NICH have been described: one presents as a flat patch with a slightly atrophic surface and firm consistency; the other appears as a nodular or plaque-like lesion, dark red to violet in color, with telangiectasias and a pale peripheral halo. These lesions predominantly affect the head and neck region (43% of cases) and the limbs (38%).

The distinction between infantile hemangiomas and congenital hemangiomas lies not only in their presence at birth and evolutionary behavior, but also in their incidence, sex distribution, coloration, and size (2, 3) (Table I).

Table I. *Comparison between Infantile and Congenital Hemangioma.*

INFANTILE HEMANGIOMAS	CONGENITAL HEMANGIOMAS
Absent at birth	Fully formed and visible at birth
10% incidence	Rare
M/F: 1/4 Primarily located on head and neck	M/F: 1/1 Primarily located on limbs and trunk
Red or bluish nodule Glut-1 positive	Violaceous nodule often with telangiectasias Glut-1 negative
Variable volume Rapid growth phase during first 4-5 months and slow involution	Medium-to-large volume Distinct pattern of evolution: RICH (rapid), PICH (partial), NICH (none)

However, the most defining difference lies in immunohistochemistry. Infantile Hemangiomas are characterized by the presence of a glucose transporter protein (GLUT1) found on erythrocytes, which is absent in Congenital Hemangiomas (2).

This immunohistochemical marker regulates neoangiogenesis and acts as a capillary growth factor. It is normally found only in the placenta and the blood-brain barrier. During childbirth, microemboli containing GLUT-1 may detach from the placenta, enter the newborn's bloodstream, and settle in the skin, erroneously stimulating capillary proliferation for 4–5 months until, having exhausted its function, the marker disappears, causing the cessation of endothelial proliferation.

This hypothesis explains why Infantile Hemangiomas cannot be present at birth, since the microemboli detach from the placenta only during delivery.

Therefore, it can be deduced that the natural history of Infantile Hemangiomas differs from that of Congenital Hemangiomas, as the former are believed to be perinatal proliferative disorders of angioblastic tissue that occur during the final stage of differentiation of the capillary system, while the latter could be due to malformative disorders.

The exact cause is still unknown, though genetic (mutations in GNAQ and GNA11 genes have been described), epigenetic, and environmental factors might be implicated (4,5).

Diagnosis is clinical, and instrumental tests, including ultrasound, are not necessary and often misleading.

What should be communicated to the parents?

1) Non-Involuting Congenital Hemangiomas (NICH) are benign vascular tumors with a good prognosis.

2) They do not require further diagnostic testing, as the diagnosis is clinical.

3) Surgical excision is the only treatment option, considered only if the location of the lesion interferes with the child's relational life. Multidisciplinary specialist evaluation is necessary if the lesion's location may affect organ function.

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