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TOPIC HIGHLIGHT

RICH (Rapidly Involuting Congenital Hemangioma) with Fetal Involution

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ABSTRACT

A term newborn was evaluated for the presence of a lesion resembling an involuting hemangioma; dermoscopy showed some residual ectasic capillaries, consistent with the diagnosis. At 5 months, lesion appeared in further light regression, comparable to that of a classical capillary hemangioma at 2 years of life. Pediatric hemangiomas are classified into infantile and congenital hemangioma. Infantile hemangiomas typically are very pale at birth, proliferate in the first months of life and then gradually involute throughout childhood. Congenital hemangiomas are further subclassified into rapidly involuting (RICH) and non-involuting (NICH) congenital hemangioma. Recently, a new variant of RICH showing fetal involution has been described. The prominent part of the life cycle of this variant of RICH (ie, proliferation and rapid involution) seems to start prenatally, whereas regression in typical RICH usually occurs in early infancy.Our case seems to confirm the existence of such variant of RICH.

Key words: Congenital hemangioma; Pediatric dermatology; Perinatology; Neonatology

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Abbreviations
RICH: rapidly involuting congenital hemangioma; NICH: non-involuting congenital hemangioma

PATIENT PRESENTATION

A term newborn from a normal pregnancy came to our attention at 1 month of life for the presence of an atrophic lesion in the nucal region, clinically resembling an involuting hemangioma (Figure 1). This hypothesis was also strengthened by dermoscopy, which showed some residual ectasic capillaries (Figure 1, box).

The mother reported to have noticed posteriorly at the base of the neck, soon after birth, the presence of an ovalar area of atrophic skin lifted in folds.

No cutaneous markers suggestive of disraphism (ie, pits, fur, lipomas) were present. Therefore, our working diagnosis was that of an *in utero* rapidly involuting congenital hemangioma (RICH).

Given the absence of associated symptoms, we decided not to perform biopsy and opted for clinical follow up.

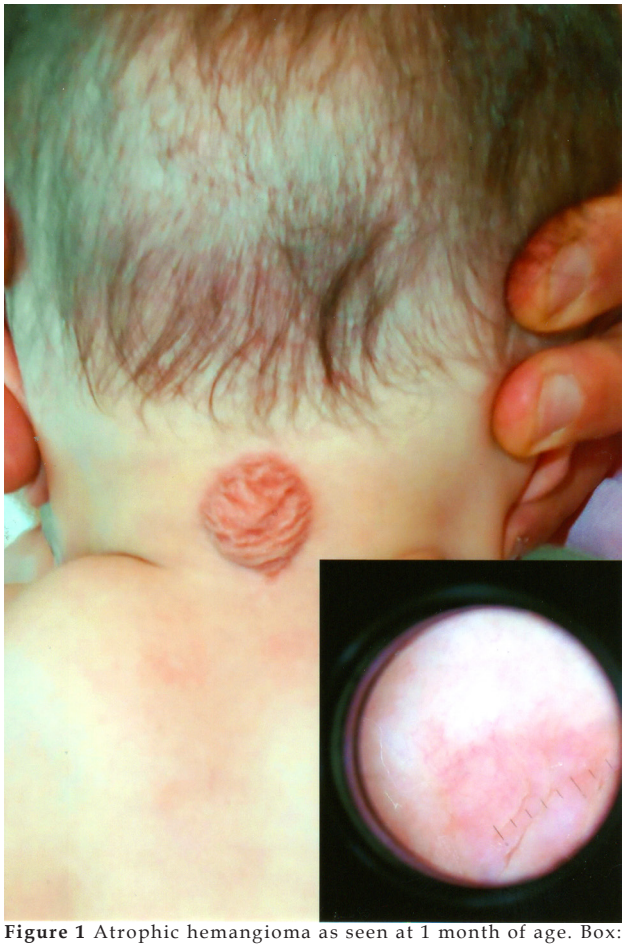

Figure 1 Atrophic hemangioma as seen at 1 month of age. Box: dermoscopy shows few ectasic capillaries, consistent with the diagnosis.

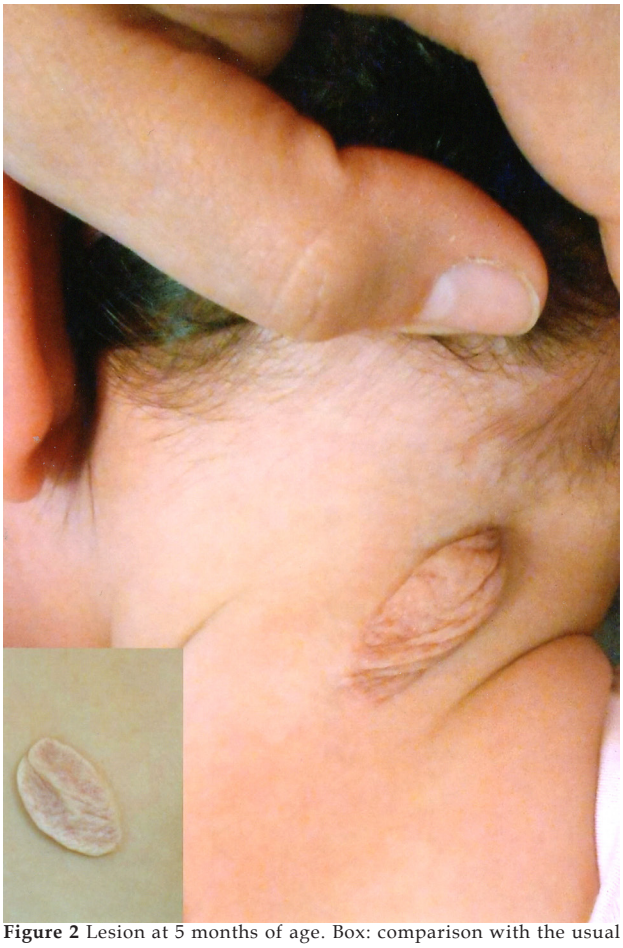

Figure 2 Lesion at 5 months of age. Box: comparison with the usual involution of an infantile capillary hemangioma at 2 years of life.

At 5-month follow up visit, lesion appeared in further light regression (Figure 2; in the box, a comparison with the usual involution of an infantile capillary hemangioma at 2 years of life).

DISCUSSION AND KEY CLINICAL MESSAGE

Hemangiomas, the most common vascular tumors in infancy, may be classified into infantile and congenital hemangioma. Infantile hemangiomas typically are either not detectable at birth or very pale and usually flat; they proliferate in the first months of life (lifting from the cutaneous plane and acquiring the typical reddish hue) and then gradually involute in a variable amount of months throughout childhood; up to 90% completely resolve by the age of 9 years[1]. On the contrary, congenital hemangiomas already present a mature aspect at birth [2].

Infantile capillary hemangiomas also differ from congenital hemangiomas in their histopathology and immunohistologic staining[3,4].

Congenital hemangiomas are further subclassified into rapidly involuting (RICH) and non-involuting (NICH) congenital hemangioma. RICH represent the more common variant of congenital hemangioma, typically showing complete involution by 12 months of age. Conversely, NICH do not show involution over time, usually demonstrating proportional growth and requiring eventual surgical excision[2].

Recently, Maguiness S et al[5]. reported 6 cases of RICH with fetal involution, a potentially new variant of congenital hemangioma sharing radiologic and histopathologic features of end-stage RICH. It is arguable that the prominent part of the life cycle of this variant of RICH (ie, proliferation and rapid involution) starts prenatally, whereas regression in typical RICH usually occurs in early infancy.

Our case seems to confirm the existence of such variant of RICH.

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