Partially involuting congenital hemangiomas: A report of 8 cases and review of the literature

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Background: Congenital hemangiomas have been divided into 2 major subtypes based on clinical behavior: rapidly involuting congenital hemangioma (RICH) and noninvoluting congenital hemangioma (NICH).

Objective: We describe a clinical subtype of congenital hemangioma that begins as a RICH but fails to completely involute and persists as a NICH-like lesion. We propose the term "partially involuting congenital hemangioma" for this lesion with overlapping features.

Methods: A review of the medical charts, serial clinical photographs, imaging, and biopsies performed on children with a diagnosis of partially involuting congenital hemangioma between 2001 and 2012 at Centre Hospitalier Universitaire Sainte-Justine pediatric dermatology/vascular anomalies clinic was performed.

Results: Eight full-term, healthy infants presented at birth with vascular lesions typical of RICH. Affected locations included the head and neck, trunk, or extremities. Size varied from 2.0 × 1.5 cm to 13.0 × 8.5 cm. All had rapid involution during the first 12 to 30 months of life before stabilizing in size and appearance.

Limitations: Only a small number of cases were identified.

Conclusion: Partially involuting congenital hemangiomas are congenital hemangiomas with a distinct behavior, evolving from RICH to persistent NICH-like lesions. Their recognition and study will help us better understand whether RICH and NICH are indeed separate entities or simply part of a spectrum. (J Am Acad Dermatol 2014;70:75-9.)

Key words: infantile hemangioma; noninvoluting congenital hemangioma; partially involuting congenital hemangioma; rapidly involuting congenital hemangioma.

In contrast to infantile hemangiomas, which typically enlarge after birth and then involute, congenital hemangiomas arise and proliferate in utero.[1,2] Two major subtypes of congenital hemangiomas are defined: rapidly involuting congenital hemangioma (RICH)[3] and noninvoluting congenital hemangioma (NICH).[4] Although RICH involutes completely within the first 6 to 14 months of life, NICH grows proportionally with the child and does not regress.

We report 8 cases of congenital hemangioma that presented clinically and radiologically as RICH but underwent only partial involution (Table I). We propose the term "partially involuting congenital hemangioma" to characterize these lesions.

METHODS

Between 2001 and 2012, 116 children with a diagnosis of congenital hemangioma were seen at Centre Hospitalier Universitaire Sainte-Justine

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Funding sources: None.
Conflicts of interest: None declared.
Accepted for publication September 8, 2013.
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Published online October 31, 2013.
0190-9622/$36.00

http://dx.doi.org/10.1016/j.jaad.2013.09.018

pediatric dermatology/vascular anomalies clinic. We reviewed the medical charts, serial clinical photographs, imaging, and biopsy specimens of these children.

RESULTS

We identified 8 patients (6.9%) with cutaneous lesions that could be categorized as partially involuting congenital hemangioma. Three male and 5 female full-term, healthy infants presented at birth with a vascular tumor involving the head and neck (n = 2), trunk (n = 3), or extremity (n = 3). The size of the tumor ranged from 2.0 × 1.5 cm to 13.0 × 8.5 cm and the clinical appearance uniformly included a red-purple color, telangiectasia, and surrounding pallor (Fig 1). Two lesions displayed prominent surrounding subcutaneous veins and 1 developed central ulceration. All congenital hemangiomas began to involute shortly after birth.

CAPSULE SUMMARY

- Congenital hemangiomas have been described as either rapidly involuting or noninvoluting.
- We observed 8 congenital hemangiomas that presented as rapidly involuting congenital hemangioma but underwent only partial involution.
- Recognition of a partially involuting congenital hemangioma may contribute to our understanding of congenital hemangiomas.

An initial diagnosis of RICH was made on clinical grounds and all 8 patients were followed up for an average of 5 years (range 2.5-10 years) in our outpatient dermatology/vascular anomalies clinic. Four hemangiomas decreased in size and thickness within the first 12 months of life before stabilizing, whereas the other 4 continued to regress slowly and stabilized between 14 and 30 months of life. All patients had residual persistent lesions that grew proportionally with the child. The patches or plaques maintained their red-purple color, telangiectasia, prominent veins, and pale rim (Fig 2). Anetoderma or skin atrophy was noted in 3 patients. Thus, although all 8 cases initially resembled and behaved like RICH, they evolved into the typical clinical appearance and behavior of NICH.

Doppler ultrasound imaging performed at birth (n = 6) and of residual hemangioma (n = 8) consistently demonstrated hypervascular subcutaneous lesions with low-resistance, high-velocity arterial flow (Fig 3). Magnetic resonance imaging (n = 2) showed subcutaneous lesions with well-defined limits and patchy hyperintensity on T2-weighted sequences, flow voids representing microfistulae, and fat stranding. Histopathologic examination of lesional biopsy specimens (n = 2) showed large capillary lobules, hobnail endothelial cells, and interlobular fibrosis (Fig 4). Glucose transporter-1 staining was negative. In all instances, imaging studies and histopathologic findings were compatible with congenital hemangioma and indistinguishable from RICH (before complete involution) or NICH.

Treatment by partial surgical excision (n = 1), compression stocking (n = 1), and pulsed dye laser photocoagulation (n = 3) was attempted on residual lesions with moderate or no clinical improvement.

DISCUSSION

Clinically, RICH and NICH have overlapping features. They show an equal sex distribution and typically present as solitary bossed plaques or flat infiltrative tumors of red to violaceous color with overlying tiny or coarse telangiectasia. Blanching or a halo-like effect may be seen surrounding the lesion. The most common locations are the head, neck, and limbs. Involution of RICH begins shortly after birth, and may present with crusting and scaling of the skin along with central fissuring and ulceration. Eventually, there is a residual anetodermal patch of skin with normal blood flow.[3-5] Cutaneous RICH and NICH are rarely associated with high-output cardiac failure and hemorrhage.[6,7]

On Doppler ultrasound imaging, both RICH and NICH are usually confined to the subcutaneous fat and appear heterogeneous, with occasional calcifications and multiple arteries and veins exhibiting high-velocity blood flow. NICH contains vascular shunts more frequently than RICH.[8,9] On magnetic resonance imaging, both tumors demonstrate well-defined limits with homogeneous or patchy enhancement, hyperintensity on T2-weighted sequences, flow voids, and fat stranding.[6,9,10] Angiography shows inhomogeneous parenchymal staining, large and irregular feeding arteries in a disorganized pattern, multiple various-sized aneurysms, arteriovenous shunts, and intravascular thrombi. Early venous draining is not seen, which differentiates congenital hemangiomas from arteriovenous malformations.[6,9]

Histopathologically, RICH and NICH also demonstrate overlapping features, although certain findings may help distinguish them. RICH is characterized by well-defined, variably sized lobules containing small capillaries and prominent draining vessels in the dermis and subcutaneous tissue. There