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Photo Vignette

Rapidly involuting congenital hemangioma (RICH): a brief case report

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Abstract

Congenital hemangiomas (CH) are benign vascular neoplasms that proliferate in utero and have completed development by birth. Two subtypes of CH are recognized: rapidly involuting congenital hemangiomas (RICH) and non-involuting congenital hemangiomas (NICH). Involution of the RICH subtype often begins in the first weeks of life. NICH does not involute, allowing the distinction between RICH and NICH. We report a case of an infant with RICH occurring on the scalp, examined at birth and followed for 26 weeks.

Case synopsis

We report the case of an infant diagnosed with rapidly-involuting congenital hemangioma (RICH). Birth was uncomplicated and appropriate APGARS were noted. Exam 4 hours postpartum revealed a nodule at the temporoparietal scalp, 2.5 by 2.0 centimeters, raised, ovaloid, and purple-red in color.

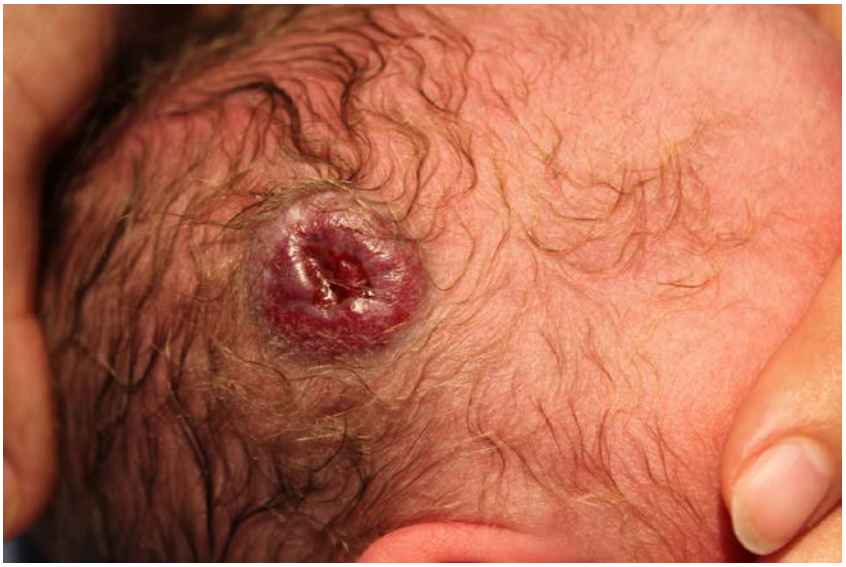

Figure 1. 4 hours post-partum

The nodule was centrally ulcerated without active bleeding or apparent pain. Alopecia over the nodule was observed without a corresponding “hair collar” sign. A complete blood count was normal. An infectious source was considered unlikely and there were no concerning maternal factors. The initial differential diagnosis included RICH, NICH, arteriovenous malformation, tufted angioma (TA) congenital kaposiform hemangioendothelioma (KHE), primary versus metastatic tumor, and aplasia cutis congenita (ACC). Perinatal scalp trauma causing hematoma was also considered initially but excluded by review of the birth history. Investigation with ultrasound and skull films revealed an intact cranium with a superficial vascular mass, directing the diagnosis away from congenital malformations and solid or malignant lesions. Biopsy was considered but ultimately deferred by parents for clinical monitoring. Conservative wound care with protection of the site was recommended.

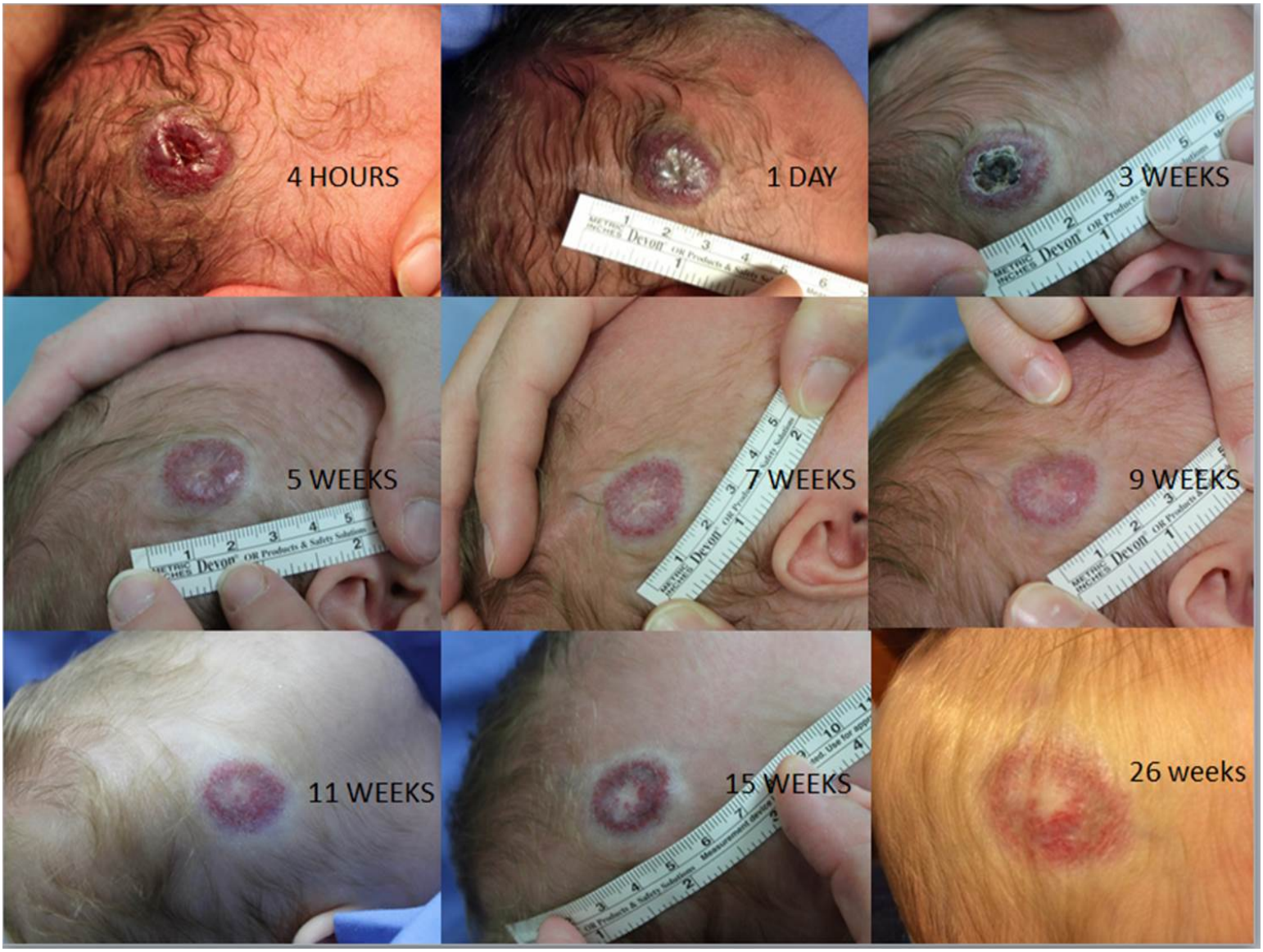

Figure 2. Photo progression from 4 hours post-partum to 26 weeks of life

In the following weeks, close observation showed the lesion to be involuting centrally and peripherally. Based on congenital presentation, physical exam, laboratory values, imaging, and clinical progression, a diagnosis of RICH was rendered. Follow up exam at 26 weeks showed hair growing throughout the site with the exception of the central scarred area corresponding to the ulceration present at birth.

Discussion

Hemangiomas are the most common tumor of infancy and are classified as infantile or congenital. Infantile hemangiomas are not present at birth unlike congenital hemangiomas, which are further characterized as rapidly involuting (RICH) or non-involuting (NICH). In a prospective study, RICH occurred in 2 of 594 newborns (0.3 percent) [1]. The female to male ratio is 3:1 and females present more often with problematic tumors [2]. In most cases, RICH completes involution in the first 16 months of life and in rare instances, involution may occur in utero [3]. Diagnosis is based upon history and physical examination in most cases [3]. RICH typically presents as a solitary, raised, plaque-like nodule that is pink to purple in color, often with telangiectasias [4].

These lesions vary in size from a few to several centimeters [4]. RICH appears on the head and neck (60%), trunk (25%), and extremities (15%) [2]. Ultrasonography, MRI, or biopsy may be helpful when there is a clinical suspicion of malignancy such as firmness on palpation, rapid growth, ulceration, and fixation to the fascia [4]. Because RICH is self-resolving, close monitoring is the standard of care for uncomplicated cases. Cutaneous ulceration is the most frequent complication [3]. When ulceration occurs, the application of petrolatum under occlusion may help prevent local infection [4]. Rarely, larger lesions may present with transient thrombocytopenia and cardiac overload [3, 5]. Post-involution skin may appear with textural changes, discoloration, persistent telangiectasias, and redundant skin [3]. Surgery can often correct post-involution redundancy. RICH and NICH stain negative for glucose transporter 1 (GLUT1), unlike infantile hemangioma [5]. The underlying cause for RICH is not clearly understood.

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