While infantile hemangiomas are a very common lesion seen in infants and young children, congenital hemangiomas are much more rare and have been only recently described. Two types of congenital hemangiomas exist: rapidly involuting congenital hemangiomas and noninvoluting congenital hemangiomas. The goal of this article is to describe rapidly involuting and noninvoluting congenital hemangiomas as they differ from infantile hemangiomas in their presentation, natural history, histopathologic features, and treatment.

Infantile hemangioma (IH) occurs in 4% to 10% of white infants. Infantile hemangioma typically appears in the first few weeks of life, proliferates for weeks to several months, stabilizes between 6 and 12 months of age, and then begins to gradually involute over a period of several years. Infantile hemangioma presents either as focal, segmental, solitary or multiple lesions, which may be superficial, deep, or mixed (superficial and deep) lesions. Infantile hemangioma occurs 3 to 5 times more commonly in female infants and is seen most frequently in whites and less commonly in those of African or Asian descent.

Through organizations such as the International Society for the Study of Vascular Anomalies (ISSVA), a clear distinction of the classification and nosology of vascular tumors such as IH and vascular malformations has emerged. This classification system has improved communication between specialists and diagnostic accuracy. The classification system differentiates between “vascular tumors” (eg, hemangiomas, tufted angiomas, hemangioendotheliomas, and pyogenic granulomas) and “vascular malformations.” The importance in the distinction between the 2 categories of vascular lesions is that vascular tumors grow after birth, proliferate, and then involute. Vascular malformations are present at birth, never involute, and grow commensurate with the child.

PATHOGENESIS OF IH

The pathogenesis of IH is poorly understood. North et al have proposed that IH may originate from either invading angioblasts that differentiate toward a placental phenotype or from embolized placental cells. Glucose transporter isoform 1 (GLUT1), a glucose transporter enzyme, is uniquely expressed on the endothelial cells of hemangiomas but not in surrounding normal vascular endothelium. This positive expression persists throughout the life cycle of the hemangioma, making this histochemical marker a valuable tool in differentiating hemangiomas from vascular malformations and other vascular tumors. Recently, Dadras et al have shown that lymphatic endothelial hyaluronan receptor-1 (LYVE-1), a marker for both normal and tumor-associated lymphatic vessels, was strongly expressed in proliferating hemangiomas and that LYVE-1 staining diminished with the age of the hemangioma and was absent in involuted lesions, suggesting that vessels from IH are arrested at an early developmental stage of vascular development.

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PRESENTATION OF IH

Most IHs are not recognized at birth, and the skin at the site of the future hemangioma appears normal. However, approximately 30% of IHs present as nascent lesions at birth. Findings may include a blanched vasoconstricted area (Figure 1), a flat telangiectatic macule (Figure 2), a vascular stain (pseudo–port-wine stain), bruised area, or rarely cutaneous ulceration. These lesions later undergo postnatal proliferation typical of an IH (Figure 1B).

PRESENTATION OF CONGENITAL HEMANGIOMAS

Physicians caring for patients with IH had long noted that on occasion infants would present with congenital hemangiomas. Use of the term congenital implies a lesion fully grown at birth. Congenital hemangiomas are present at birth and do not exhibit accelerated or disproportionate postnatal growth. Some of these lesions appeared to involute much more rapidly compared with IH, while others seemed fully formed at birth, never proliferated, grew proportionally with the child’s growth, and never involuted.

Congenital hemangiomas are unique lesions, unlike precursor or nascent hemangiomas. They fall into 2 major types: rapidly involuting congenital hemangioma (RICH) and noninvoluting congenital hemangioma (NICH). They usually present as solitary lesions at birth and rarely coexist in the same patient with a typical IH. Based on negative GLUT1-staining results, these lesions are rare, with a combined incidence of less than 3% of all IHs.

Unlike IHs, congenital hemangiomas are mature at birth, having undergone their proliferative phase in utero. Mulliken and Enjolras have recently characterized the growth curves of RICH and NICH compared with IH (Figure 3).

Both RICH and NICH may be bossed plaques or tumors. Three morphologic variants of RICH exist: (1) a lesion with a characteristic red-purple color, often with coarse telangiectasia present on a portion of its surface or at the periphery of the tumor (Figure 4 and Figure 5); (2) a flat infiltrative tumor with violaceous overlying skin (Figure 6); and (3) a raised grayish tumor with multiple tiny telangiectasias, surrounded by a pale halo (Figure 7). Peripheral blanching or halolike effect may be seen in both RICH and NICH but is more characteristic of NICH.

RAPIDLY INVOLUTING CONGENITAL HEMANGIOMA

The most common location for RICH is on the limb, head, or neck. In the posterior nuchal area, RICH lesions may be seen on second-trimester ultrasound and be suggestive of lymphatic malformation, encephalocele, or other forms of cranial dysraphism. Both NICH and RICH exhibit fast flow by ultrasonography and may show flow voids on magnetic resonance imaging. Angiography of RICH shows inhomogeneous parenchymal staining, large and irregular feeding arteries in a disorganized pattern,
arterial aneurysms, arteriovenous shunts, and intravascular thrombi. Two infants with RICH have been reported with associated high output failure and intraleral bleeding. In the center of the lesion, RICH may involute rapidly shortly after birth much like a collapsed soufflé, resulting in central fissuring and ulceration. Such lesions may be difficult to heal, necessitating early surgical excision.

HISTOPATHOLOGIC CHARACTERISTICS OF RICH

The histopathologic characteristics of RICH include small to large lobules of capillaries with moderately plump endothelial cells and pericytes. The lobules are surrounded by fibrous tissue with large, abnormal draining channels and may contain hemosiderin, thrombosis, cyst formation, focal calcification, and extramedullary hematopoiesis. Rapidly involuting congenital hemangioma lesions stain negative with GLUT1.

NATURAL COURSE OF RICH

In most infants with RICH, involution is complete, leaving an empty bag of anetodermic excess skin, within the first 6 to 14 months of life (Figure 4B and Figure 5B). In a smaller proportion of patients with RICH, involution may be rapid at first, but incomplete, leaving a vascular plaque with coarse telangiectasia on the surface and a peripheral bluish white border, resulting in a clinical lesion that is indistinguishable from NICH (Figure 8). Mulliken and Enjolras have proposed that NICH could be a later stage of RICH because of these similarities in appearance, persisting fast flow on ultrasound, and their similar histopathologic features. In patients with NICH, this would suggest that any preceding involution had occurred in utero.

MANAGEMENT OF RICH

Management of RICH lesions is straightforward in a patient with a typical clinical presentation whose lesion has...
characteristic features on ultrasound or magnetic resonance imaging. Observation during the rapid involution phase is usually the first line of treatment. In many patients, involution may begin within the first week of life with crusting and scaling on the surface of the lesion. Prevention of ulceration in these infants may be accomplished by applying petrolatum ointment to the surface of the lesion several times daily. Lesions that are firm or show no tendency to involute may be difficult or impossible to differentiate, even following imaging, from more serious tumors such as congenital fibrosarcoma or rhabdomyosarcoma. Such lesions require early biopsy and GLUT1 staining and/or surgical excision.

Patients with RICH may not always have an uncomplicated course. When ulceration occurs, adherence to good wound care practices, pain control, and treatment of secondary infection is essential. Surgical excision is indicated for persistent ulceration or development of Kasabach Merritt phenomenon in lesions that do not respond to medical therapy. Once involution is complete, any residual anetodermic skin may be excised surgically if it does not return to normal by the time the child is self-aware (usually by age 3-4 years).

NONINVOLUTING CONGENITAL HEMANGIOMA

Noninvoluting congenital hemangioma lesions are much less common lesion compared with RICH lesions. Only 53 cases were described over a 10-year period in 3 large subspecialty vascular anomalies clinics. There was a slight preponderance of males to females of 3:2. All lesions were congenital and single. Most lesions were in the head and neck, trunk, or limbs. Noninvoluting congenital hemangioma lesions tend to be plaquelike or bossed, with a pink to purple color with prominent coarse telangiectasia on the surface (Figure 9). Lesions are warmer to palpation than the surrounding normal skin.
HISTOPATHOLOGIC CHARACTERISTICS OF NICH

The histologic characteristics of resected specimens include lobular collections of small thin-walled vessels with large, stellate-shaped central vessels. Interlobular areas often contain predominantly dilated, often dysplastic veins. Small arteries, which are increased in number, are seen in close proximity with lobular vessels or abnormal extralobular veins. The endothelial cells in many areas have a hobnailed appearance. Increased mast cells were noted in all specimens. Noninvoluting congenital hemangioma lesions stain negative with GLUT1.

NATURAL COURSE OF NICH

Noninvoluting congenital hemangioma will never disappear or involute. They remain unchanged except for proportional growth and appearance of increased draining veins in the periphery of the lesion. Excision is the recommended treatment, with preoperative embolization to control bleeding in larger lesions. Imaging will confirm fast flow, and magnetic resonance imaging will show hyperintensity on T2-weighted sequences with flow voids similar to IH. Arteriographic examination reveals arterial-like feeders, a tumorlike capillary blush with small arterial channels. Early venous draining is not seen, which differentiates these lesions from arteriovenous malformations or arteriovenous fistulas on angiography.  

CONCLUSIONS

Congenital hemangiomas are distinct lesions, differing from nascent or precursor lesions of typical IH. Unlike IH, negative GLUT1 results on histologic examination are characteristic. Recognition of their unique clinical presentation, natural course, imaging, and histologic characteristics is essential for those who care for patients with vascular birthmarks. Congenital hemangiomas (RICH and NICH) may require a more aggressive workup and treatment course, including biopsy with immunohistochemical staining (GLUT1) and/or excision for diagnostic and therapeutic reasons. Further awareness and familiarity with these uncommon congenital lesions will facilitate accurate diagnosis and management.

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