Management of Hemangiomas and Other Vascular Tumors

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KEYWORDS

• Hemangioma • Kaposiform hemangioendothelioma
• Pyogenic granuloma • Vascular anomaly • Vascular tumor

Vascular tumors of childhood are typically benign. The 4 most common types are infantile hemangioma (IH), congenital hemangioma (CH), kaposiform hemangioendothelioma (KHE), and pyogenic granuloma (PG). Vascular tumors *must* be differentiated from vascular malformations. Although tumors and malformations may appear as raised, blue, red, or purple lesions, their management differs significantly.

INFANTILE HEMANGIOMA

Clinical Features

IH is a benign tumor of the endothelium, and the most common tumor of infancy.[1,2] IH affects approximately 4% to 5% of Caucasian infants and is rare in dark-skinned individuals.[3–5] It is more frequent in premature children (23% of infants <1200 g) and females (3:1 to 5:1).[6,7] IH typically is single (80%) and involves the head and neck (60%), trunk (25%), or extremity (15%).[2] The median age of appearance is 2 weeks, although 30% to 50% of lesions are noted at birth as a telangiectatic stain or ecchymotic area.[8] IH grows faster than the rate of the child during the first 9 months of age (proliferating phase); 80% of its size is achieved by 3.2 (±1.7) months.[9] When IH involves the superficial dermis it appears red. A lesion beneath the skin may not be appreciated until 3 to 4 months of age when it has grown large enough to cause a visible deformity; the overlying skin may appear bluish. By 9 to 12 months of age the growth of IH plateaus to approximate that of the infant.[9] After 12 months of age the tumor begins to shrink (involuting phase); the color fades and the lesion flattens (**Fig. 1**). Involution stops in most children by 5 years of age (involuted phase).[10] After involution, one-half of children have an abnormality: residual telangiectasias, anetoderma from loss of elastic fibers, scarring, fibrofatty residuum, redundant skin, or destroyed anatomic structures.

Diagnosis

Ninety percent of IH are diagnosed by history and physical examination (**Fig. 2**). Deeper lesions may be more difficult to appreciate because they are noted later than superficial tumors, and may not have significant overlying skin changes. Diagnosis of subcutaneous IH is facilitated using a hand-held Doppler device showing fast flow. When history and physical examination are equivocal, ultrasonography is the first-line confirmatory study to differentiate IH from other lesions. IH appears as a soft-tissue mass with fast flow, decreased arterial resistance, and increased venous drainage.[11] Magnetic resonance imaging (MRI) can differentiate vascular anomalies, although young children require sedation.[12] During the proliferating phase, IH is isointense on T1 images, hyperintense on T2 sequences, and enhances with contrast. IH appears as a mass with dilated feeding and draining vessels; signal voids represent fast flow and shunting.[13] Involuting IH has increased lobularity and adipose tissue; the number of vessels and flow is reduced.[12]

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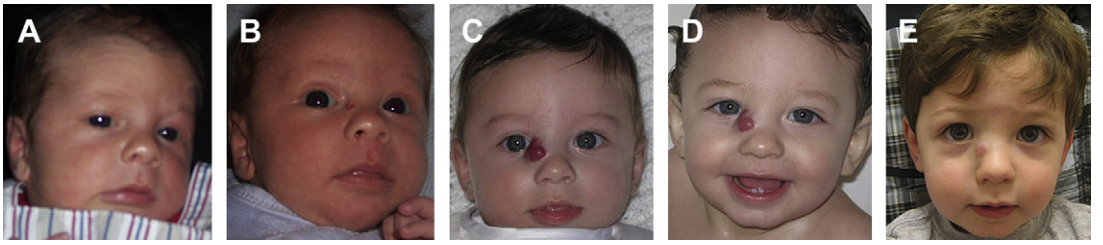

Fig. 1. Proliferation and involution of infantile hemangioma. (*A*) Age 2 weeks. (*B*) Age 5 weeks. (*C*) Age 6 months. (*D*) Age 12 months. (*E*) Age 18 months.

Rarely, biopsy is indicated if malignancy is suspected or if the diagnosis remains unclear following imaging studies. Tumors or fast-flow lesions that may be confused with IH include arteriovenous malformation, CHs, cutaneous leukemia (chloroma), hemangioendotheliomas, infantile fibrosarcoma, infantile myofibromatosis, lymphoma, metastatic neuroblastoma, PTEN-associated vascular anomaly, and PG.[14–18] In the liver, the differential diagnosis of hemangioma includes arteriovenous malformation, hepatoblastoma, or metastatic neuroblastoma. Because an erythrocyte-type glucose transporter (GLUT1) is specifically expressed in IH, immunostaining for GLUT1 can differentiate IH from other tumors and malformations.[19]

Clinical Considerations

Head and neck hemangiomas

Ten percent of proliferating IH cause significant deformity or complications, usually when located on the head or neck.[20] Ulcerated lesions may destroy the eyelid, ear, nose, or lip. IH of the scalp or eyebrow can injure hair follicles, resulting in alopecia. Obstruction of the external auditory canal can cause otitis externa, but sensorineural hearing loss does not occur if the contralateral canal is patent. Periorbital hemangioma can block the visual axis or distort the cornea causing deprivation or astigmatic amblyopia, respectively. IH involving the upper eyelid is more likely to be problematic than a lesion involving the lower eyelid. Infants with periorbital IH are referred to an ophthalmologist; the noninvolved eye may be patched for a minimum of 2 hours per day to stimulate use of the affected eye. Subglottic hemangioma, associated with large cervicofacial lesions, may obstruct the airway; patients are referred to an otolaryngologist for evaluation. Although the patency of the airway is usually maintained with oral corticosteroid, laser treatment or tracheostomy may be necessary.[21,22]

Multiple hemangiomas

Although 20% of infants have more than one IH, occasionally a child will have 5 or more small (<5 mm), dome-like lesions termed disseminated hemangiomatosis.[8] These children are at increased risk for IH of internal organs, although the risk is low. The liver is most commonly affected; the brain, gut, or lung are rarely involved. Ultrasonography should be considered to rule out hepatic IH. However, because intervention is not necessary for an incidentally found liver lesion, families may not elect to pursue imaging. In addition, infants with problematic hepatic IH are usually symptomatic early in infancy, before they are referred for evaluation of the cutaneous lesions. If symptomatic

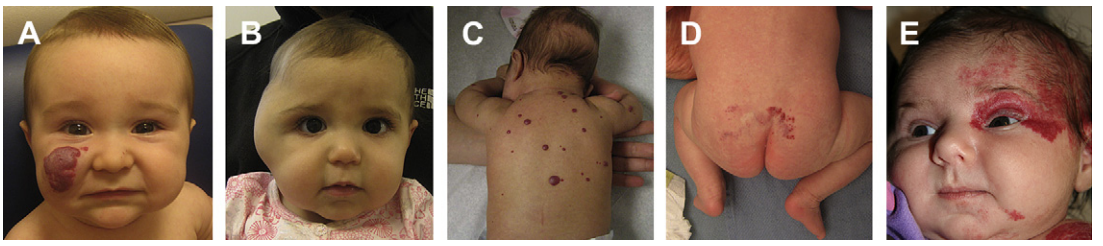

Fig. 2. Clinical features of infantile hemangioma (IH). (*A*) A 5-month-old male with a superficial tumor of the cheek. (*B*) A 12-month-old female with a deep IH of the cheek and temporal area without overlying skin changes. (*C*) A 2-month-old infant with hemangiomatosis, including hepatic hemangioma. (*D*) A 2-month-old female with midline lumbosacral IH and underlying lipomyelomeningocele. (*E*) An infant female with PHACE association (*P*osterior fossa brain malformation, *H*emangioma, *A*rterial cerebrovascular anomalies, *C*oarctation of the aorta and cardiac defects, *E*ye/*E*ndocrine abnormalities) and cerebrovascular anomalies.