Answer to Dermacase *continued from page 379*

2. Infantile hemangioma

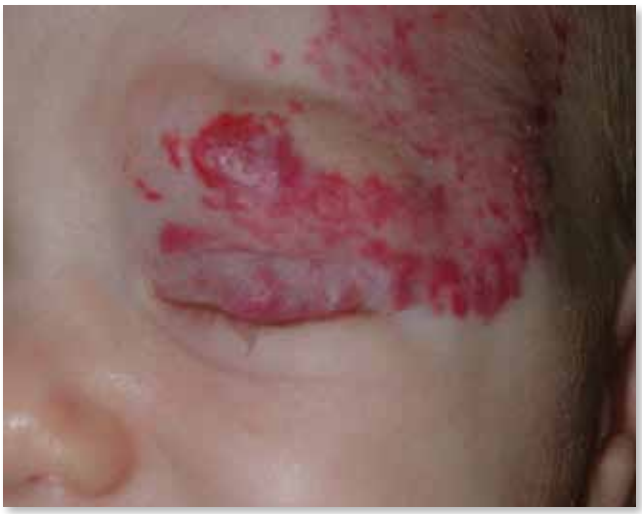
Infantile hemangioma (IH), previously referred to as *strawberry* or *capillary hemangioma*, is the most common benign vascular tumour of infancy. These tumours consist of endothelial cells that rapidly proliferate during the first few months of life and are characterized by slow involution, which can take years to complete.[1] They are thought to occur in as many as 10% of children, but this estimate is likely higher in theory than in practice.[2] Infantile hemangiomas are seen in children of all races, although less often in those of African or Asian descent. Female infants are 3 to 5 times more likely to be affected.[3] The prevalence of IH increases with both decreasing gestational age and lower birth weights.[4] Most cases occur sporadically, although some are presumed to be inherited in an autosomal dominant fashion.[5]

Most IHs appear in the first few weeks of life; however, one-third of infants exhibit a premonitory mark at birth.[3] Once they appear, IHs undergo rapid progressive enlargement for 8 to 18 months (the proliferative phase), followed by spontaneous regression (the involution phase).[6] Most IHs reach their maximum size by the time the infant is 9 to 12 months of age. They are classified as superficial, deep, or combined. Superficial IHs appear red, raised, and lobulated. Deep IHs have normal overlying skin and appear as raised, soft masses, often with a bluish cast.[4]

Diagnosis

Patient history and physical examination yield accurate diagnoses in 90% of infants.[3] It is important to differentiate between IHs and other vascular anomalies, such as vascular malformations, because of differences in treatment, associated symptoms, and outcomes. The history of the lesion is of great importance—IHs might not be present at birth and grow disproportionately and rapidly in a short period of time, while vascular malformations are present at birth and grow in proportion to the child. Ultrasonography or magnetic resonance imaging can be used to diagnose a difficult lesion. Biopsy should only be considered when history, physical examination, and imaging studies fail to clearly define the pathology.[4]

It is also important to differentiate IHs from other tumours that might carry malignant potential or be harbingers of systemic disease. Unlike IHs, port-wine stains are flat, dark, and do not blanch with pressure.[7] Rhabdomyosarcoma can present as a soft-tissue mass, but proptosis, eyelid edema, and extraocular motility impairment are the characteristic features.[8] Cutaneous lymphangioma is a rare vascular hamartoma of lymphatic channels, which presents as multiple cystic lesions of different sizes that are pink or dark red



in colour.[9] Plexiform neurofibroma occurs in patients with neurofibromatosis type 1; its lesion produces an S-shaped curvature to the upper eyelid and, on palpation, has been classically described as feeling "like a bag of worms."[9]

Progression

Most IHs spontaneously involute by the time the infant is 12 to 18 months of age. Superficial hemangiomas often change colour from bright red to dull red to gray, indicating involution of the lesion. The process usually begins centrally and spreads peripherally. Deep lesions become less blue and less warm.[4] Complete involution of IHs occurs at a rate of 10% per year per affected population, such that lesions in 50% of affected children will undergo spontaneous involution by age 5, and 90% by age 9.[4] Up to 50% of patients might be left with mild residual changes after full involution, including telangiectasia, atrophic wrinkling, yellowish discolouration, redundant skin, fibrofatty infiltration, or postulceration scarring.

Complications

The most common complication of IHs is ulceration, which occurs in 15% of cases. Large lesions, rapid proliferation, and facial or perineal location are all risk factors for ulceration.[10] Airway involvement, although rare, might require surgery or medical treatment. Hemangiomas distributed in the preauricular cheeks, chin, lower lip, and anterior neck are more likely to be associated with airway lesions.[11] Periorbital IHs can obstruct the visual axis, induce astigmatism, and increase intraocular pressure, resulting in amblyopia and permanent visual damage if left unaddressed. These lesions require early and active treatment.[12] Multiple hemangiomas (>5) warrant further imaging investigations, as lesions might also be present internally. Most of these internal lesions are found on the liver but can also develop in the lungs, brain, and intestines.[9]

Treatment

Most hemangiomas are asymptomatic and can be managed with close observation. Reassuring and educating parents is of utmost importance. Treatment is required for IHs that cause facial disfigurement, airway obstruction, high-output cardiac failure, bleeding, or lesion ulceration.[6] Periorbital lesions require referral to a pediatric ophthalmologist. Although some management strategies are controversial, the mainstay treatment is short-term oral corticosteroids (eg, 2 to 3 mg/kg prednisolone daily).[13,14] Intralesional corticosteroid injections are successful for orbital IHs but have not been well studied in other sites. Both vincristine and interferon alfa are considered second-line treatments for lesions that fail to respond to corticosteroid therapy. Laser treatment is not effective for proliferating hemangiomas—it is better suited for treating ulceration and residual telangiectasia.[1]

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Competing interests
None declared

References

1. Bruckner AL, Frieden IJ. Infantile hemangiomas. *J Am Acad Dermatol* 2006;55(4):671-82.
2. Kilcline C, Frieden IJ. Infantile hemangiomas: how common are they? A systematic review of the medical literature. *Pediatr Dermatol* 2008;25(2):168-73.
3. Mulliken JB, Fishman SJ, Burrows PE. Vascular anomalies. *Curr Probl Surg* 2000;37(8):517-84.
4. Bruckner AL, Frieden IJ. Hemangiomas of infancy. *J Am Acad Dermatol* 2003;48(4):477-93.
5. Blei F, Walter J, Orlow SJ, Marchuk DA. Familial segregation of hemangiomas and vascular malformations as an autosomal dominant trait. *Arch Dermatol* 1998;134(6):718-22. Erratum in: *Arch Dermatol* 1998;134(11):1425.
6. Lopriore E, Markhorst DG. Diffuse neonatal hemangiomatosis: new views on diagnostic criteria and prognosis. *Acta Paediatr* 1999;88(1):93-7.
7. Spring MA, Bentz ML. Cutaneous vascular lesions. *Clin Plast Surg* 2005;32(2):171-86.
8. De la Luz Orozco-Covarrubias M, Tamayo-Sanchez L, Duran-McKinster C, Ridaura C, Ruiz-Maldonado R. Malignant cutaneous tumors in children. Twenty years of experience at a large pediatric hospital. *J Am Acad Dermatol* 1994;30(2 Pt 1):243-9.
9. Rudolph CD, Rudolph AM, Hostetter MK, Lister GE, Siegel NJ. *Rudolph's pediatrics.* 21st ed. New York, NY: The McGraw Hill Companies; 2003.
10. Kim HJ, Colombo M, Frieden IJ. Ulcerated hemangiomas: clinical characteristics and response to therapy. *J Am Acad Dermatol* 2001;44(6):962-72.
11. Orlow SJ, Isakoff MS, Blei F. Increased risk of symptomatic hemangiomas of the airway in association with cutaneous hemangiomas in a "beard" distribution. *J Pediatr* 1997;131(4):643-6.
12. Ceisler EJ, Santos L, Blei F. Periocular hemangiomas: what every physician should know. *Pediatr Dermatol* 2004;21(1):1-9.
13. Bennett ML, Fleischer AB Jr, Chamlin SL, Frieden IJ. Oral corticosteroid use is effective for cutaneous hemangiomas. *Arch Dermatol* 2001;137(9):1208-13.
14. Pope E, Krafchik BR, Macarthur C, Stempak D, Stephens D, Weinstein M, et al. Oral versus high-dose pulse corticosteroids for problematic infantile hemangiomas: a randomized, controlled trial. *Pediatrics* 2007;119(6):e1239-47. Epub 2007 May 7.