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Vascular cutaneous anomalies in children: malformations and hemangiomas

Abstract The vast majority of cutaneous vascular anomalies in infants and children are either malformations or hemangiomas. Vascular malformations are subgrouped, based on channel morphology and rheology: slow-flow (capillary, lymphatic, venous, or combined-complex types) and fast-flow malformations (ectasia, aneurysm, fistula, or arteriovenous anomalies). Noninvasive radiologic techniques, especially ultrasonography with Doppler flow studies and magnetic resonance imaging, serve to document the extent and flow characteristics. Management depends on the type of malformation: laser for capillary malformations; surgical excision for lymphatic malformations; compression, sclerotherapy, and resection for venous malformations; and embolization and/or surgical resection for arteriovenous fistulae/malformations. Hemangiomas are the most common tumors of infancy. The life cycle is divided into three phases: proliferating, involuting, and involuted. Most hemangiomas do not require treatment, although drug therapy is indicated for endangering or life-threatening hemangiomas. Corticosteroids (either systemic or local) and alpha-2a interferon are currently the most effective agents. Surgical resection of problematic hemangiomas can be undertaken during infancy, the preschool years, or childhood.

Key words Vascular malformation • Hemangioma • MRI • Embolization • Sclerotherapy

Introduction

Based on clinical features, hemodynamic characteristics, natural behavior, and histologic differences, there are two major categories of superficial vascular anomalies (VA): vascular malformations (VM) and hemangiomas [1–3]. VMs are composed of dysplastic vessels exhibiting a normal endothelial turnover. Most of these anomalies are visible at birth, although some are subtle cutaneous birthmarks that may go unnoticed in early childhood. VMs grow commensurately with the patient, however, some can enlarge suddenly or progressively worsen over a lifetime.

Hemangiomas are exclusively a pediatric problem. These benign tumors of the blood capillary networks are characterized by rapid postnatal growth of endothelial cells, pericytic nests, and capillaries (the proliferating phase), followed by slow, spontaneous, invariable regression (the involuting phase). Hemangiomas are the most common tumors of infancy and childhood; they always regress, and never appear in adolescents or adults.

Although VMs and hemangiomas are totally different pathologic entities, they are often discussed adjacent to one another in the literature. The aim of this paper is to review the evolution, differential diagnosis, and therapy of these two distinct major categories of VAs.

Vascular malformations

Depending on the type of malformed vascular channels and their flow characteristics, superficial VMs can be separated clinically into capillary malformations (CM) (port-wine stains and telangiectases), venous malformations (VeM), lymphatic malformations (LM), arterial malformations (AM) (aneurysms, ectasias, stenoses, rare in the skin area), and various complex combined slow-flow and fast-flow forms. In the latter category, arteriovenous malformations (AVM) (with AV fistulae) deserve special commentary.

Capillary malformations

Port-wine stains are the most common CM. These are obvious at birth as red, clearly delineated skin or mucosal
VeMs are present at birth, sometimes as an obvious spongy blue mass, but more often as bluish patches in the skin. They progressively enlarge and slowly worsen throughout childhood and adolescence, and to a lesser degree in adulthood. Their characteristic blue color is due to ectatic venous channels in the dermis. There is no increased local heat, bruit, or thrill on palpation. VeMs are easily compressed manually. Phleboliths are pathognomonic; they are palpated or seen on plain radiographs.

Specific problems occur depending on the location of the VeM. In the facial area the lesion swells in dependent areas, with the Valsalva maneuver, and when the patient cries or during physical exertion. With time, this swelling becomes conspicuous. There can be distortion of facial structures, e.g. periocular VeMs cause progressive orbital enlargement and proptosis. Enophthalmos can occur when the patient stands up. A venous lesion can become painful when the patient is exercising. Localized thrombosis is a common occurrence.

Pharyngeal, laryngeal and palatal VeMs can cause obstructive sleep apnea syndrome, easily demonstrated by polysomnographic investigation. Pelvic VeMs enlarge during pregnancy, causing pain and large vulvar varicose veins, yet rarely impede vaginal delivery. In the limbs, pure VeMs are less common than complex combined VMs with overgrowth (an older term is Klippel-Trenaunay syndrome).