Abstract

A biological classification of vascular anomalies that correlated clinical features, natural history, and cellular characteristics was proposed by Mulliken and Glowacki in 1982 and in 1996 was accepted by the International Society for the Study of Vascular Anomalies (ISSVA). This classification clearly separating vascular tumors which result from active cell proliferation, from vascular malformations, which are inborn defects in vascular morphogenesis. These two types of lesions have different clinical behavior and require different diagnostic and therapeutic strategies. The most frequent vascular tumor is infantile hemangioma (IH). Vascular malformations can be classified according to the vessels types they are composed as simple and combined malformations. Simple vascular malformations are presented as: a) slow flow: capillary, venous, lymphatic and b) fast flow: arterial malformations. Combined vascular malformations are presented as a combination of simple vascular malformations and they can be slow flow, and fast flow. This classification system has facilitated diagnosis, communication, treatment, and research in this field.

INTRODUCTION

Vascular anomalies in children are divided into two types: 1) tumors (most being hemangiomas) and 2) vascular malformations(1-3). Infantile hemangiomas (IHs) have unique characteristics, consisting of a growth phase and an involuting phase(2-5). Unlike hemangiomas, vascular malformations which are structural anomalies derived from arteries, capillaries, veins, lymphatics or a combination, rarely involute and grow in proportion to the child(2,4,6).

VASCULAR TUMORS

a) Infantile hemangiomas

Infantile hemangiomas(IHs) occur in 4 and 12% of all Caucasian, with female predomination 3:1(1-4,7,8). Pathogenesis still remains unknown(8-10). Glucose transporter protein 1 (GLUT 1) is specific and useful immunohistochemical marker for IHs during all phases of these lesions(11). Most IHs involve the head and neck (up to 60%) and exist as solitary lesions, but up to 20% of infants may have two or more (Fig.1-A)(3,4). Clinical appearance allows differentiation between focal, indeterminate, segmental and diffuse IHs(8,10). The tumor has an initial growth phase during the first 12 months (proliferative phase), and then it involutes over approximately one to seven years (involuting and an involuted phase)(2-4,10). There are two types of congenital hemangioma that is present and fully grown at birth: noninvoluting congenital hemangioma (NICH) and rapidly involuting congenital hemangioma (RICH)(6,8,11). NICH do not go through a regression phase. RICH goes through a rapid regresion phase and may be completely gone by the time the child is 12 to 18 months old(6,8,11). Diffuse neonatal hemangiomatosis (DNH) is a rare disorder characterized by the presence of multiple cutaneous and visceral hemangiomas which appear at birth or shortly thereafter(12). The majority of IHs are uncomplicated and require no treatment (only clinical observation and serial clinic visits accompanied by photo documentation)(2-4,8,10). Intervention is typically required for
lesions that may interfere with a vital function such as airway or visual obstruction, ulcerated lesions, or lesions at increased risk for complications, including a poor cosmetic outcome (2-4,8,10,13). Current treatment options are conservative (corticosteroids, interferons, hemiotherapy), laser treatment, and surgical treatment (by lenticular excision, with a linear closure, or by circular excision and „purse-string” closure (2,4,8,10,13-16). Recently propranolol proved itself effective in inducing regression of growing hemangioma (17-19). The use of propranolol in the treatment of IHs was serendipitously discovered in 2008 by Léauté-Labrèze (17). A subgroup of children with IHs exhibits additional associated structural anomalies like in the syndrome called PHACES (Posterior Fossa malformations, Hemangioma, Arterial anomalies, Cardiovascular anomalies, abnormalities of the Eye, Sternal clefting (21).

b) Other tumors

Pyogenic granuloma is a benign, acquired vascular tumor of the skin and mucous membranes (Fig. 1-B). The treatment options are surgical excision with primary closure or shave excision at the base followed by electrocautery (3,20). Kaposiform hemangioendothelioma (KHE) is a vascular tumor associated with the Kasabach-Merritt phenomenon (severe coagulopathy due to platelet trapping and spontaneous bleeding) (Fig. 1-C) (2,3,22). Hemangiopericytoma, tufted angio- mas, spindle cell hemangioendothelioma are rarely seen vascular tumors (2,3).

VASCULAR MALFORMATION

Vascular malformation occurs as a result of errors in embryogenesis (1-4,6). They are histologically normal, but with abnormal architecture (1,6). Typically,
these anomalies are categorized according to the vessels types as simple (capillary, lymphatic, venous and arterial) and combined. Based on a flow-rate, they are classified as slow-flow and fast-flow (1-4,24,27).

Simple vascular malformation  
Capillary malformations (CM) (known as “port-wine stain”), occur in 0.1-0.3% of newborns, and can occur in any cutaneous location (Fig. 1-D)2-4. The latter occurs in 50% of white newborns and is popularly known as “angel kiss” (on the forehead, eyelids, nose, and upper lip) and “stork bite” (on the nuchal area). These irregular macular stains predictably fade and thus represent a minor transient dilatation of dermal vessels. When a CM afflicts the face, the Sturge-Weber syndrome must be suspected. Ipsilateral leptomeningal and ocular vascular abnormalities may occur23. Pulsed dye laser is a first treatment option for CMs. Partial excision is the primary treatment for thick, nodular lesions2-4.

Venous malformations (VM) are present at birth, but they are not always evident. These slow-flow anomalies manifest in many forms (Fig. 1- E). They are bluish, soft, and compressible. They can be localized or extensivewithin an anatomic region.3,4. Histologically, VM is composed of thin-walled, dilated, sponge-like abnormal channels. Unlike hemangiomas, VMs grow in proportion to the child and do not involute4. MRI is the most informative imaging modality, but venography may be required preoperatively 26,27. Treatment of VMs is indicated for appearance, pain, or functional problems. The therapeutic mainstays are sclerotherapy and surgical resection. Treatment is multimodal: sclerotherapy or complete surgical extirpation 2-4,26,27. Elastic compression devices aid in pain control, as will aspirin to prevent thrombosis4,27.

Lymphatic malformations (LM) (known as lymphangioma) are present at birth, and larger lesions are sometimes detected on prenatal ultrasound2-4,25,26. Common sites of occurrence are the face, neck, axilla, chest, buttock, perineum and retroperitoneum/mediastinum (Fig.1-F)25,26. LMs are categorized as microcystic, macrocystic or a combination25. In the head and neck, such bulky lesions may compromise the aerodigestive tract, which may necessitate tracheostomy or feeding devices3,25,26. Therapy is usually multimodal25. Surgical resection is infrequently realized due to lesion extent and inevitable incorporation of vital structures3,26. Sclerotherapy with OK-432 substance (a killed strain of groupA Streptococcus pyogenes), is only possible with the macrocystic subtype25. Stocking compression is the mainstay of therapy for extremity lesions2-4.

Arteriovenous malformations (AVM) is distinctive from other vascular malformations due to its high flow, expressed outwardly by a palpable thrill, audible bruit and general warmth and redness (Fig.1-G)3,4,27. Schobinger proposed a four stagesclinical system in 1990, according to clinical manifestations and present complications4,27. Ideal initial imaging is via ultrasonography and color Doppler, with MRI to evaluate extent of malformation26. Treatment options are embolization and surgical resection3,27.

COMBINED VASCULAR MALFORMATIONS  
Slow flow combined vascular malformations  
They can be different according to vessel type combination (capillary, lymphatic, venous).

Klippel-Trenaunay syndrome is a well-worn eponym for capillary-lymphatic-venous malformation (CLVM) that is associated with soft tissue/skeletal hypertrophy, usually of one or more limbs (Fig. 1-H)2,3. The disorder is thought to be sporadic; it is obvious at birth3. Proteus syndrome is a sporadic and progressive vascular, skeletal, and soft tissue condition2. The major diagnostic features include verrucous (linear) nevus, lipomas and lipomatosis, macrocephaly, calvarial hyperostosis, asymmetric limbs with partial gigantism of the hands and feet or both, and aneurysmic cerebriform plantar thickening (“moccasin” feet) 2,3. Maffucci syndrome denotes the coexistence of exophytic vascular anomalies with bony exostoses and enchondromas2-4.

Fast flow combined vascular malformations  
These anomalies are less common than slow-flow combined disorders. The abbreviations for capillary-arteriovenous fistulas (CAVFs) and capillary-arteriovenous malformation (CAVM) correspond to the old eponym Parkes-Weber syndrome2-4. The vascular anomaly is obvious at birth. The involved limb is covered by a geographic pink, macular stain and is enlarged symmetrically2,3.

CONCLUSION  
This new classification system has facilitated diagnosis, communication, treatment, and research in this field. Most large pediatric centers now have a vascular anomalies team which includes plastic or pediatric surgeons, pediatric dermatologists, radiologist, anesthesiologist, ENT specialist, ophthalmologists, physical therapy specialist etc. These multifaceted teams can help provide all the essential medical and surgical options.
Apstrakt


REFERENCES