Vascular Malformations and Their Treatment in the Growing Patient

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KEYWORDS

• Vascular anomalies • Vascular tumors • Venous malformations

KEY POINTS

- Vascular anomalies, consistent of vascular tumors and malformations, frequently arise in the head and neck and often occur in the pediatric patient.
- Infantile hemangiomas are the most common tumors in infancy; 10% are found by 1 year of age, with white premature babies and girls affected more often.
- Vascular malformations represent an abnormal proliferation of mature vascular elements, are present at birth, and show equal distribution between the sexes. Vascular malformations grow with the child and are usually affected by hormonal changes with occasional accelerated growth during puberty.
- Capillary-venular and venous malformations represent the most common vascular malformations, approaching 1 in 10,000.
- Lymphangiomas or lymphatic malformations are areas of abnormal development of the lymphatic system of unknown etiology, commonly found in the head and neck region. Surgery, sclerotherapy, and laser treatments have all been used with various success rates in the management of lymphatic malformations.

INTRODUCTION

In 1982, Mulliken and Glowacki proposed a biological classification for these lesions based on their clinical and histologic findings. Based on this classification scheme, which has been supported by subsequent radiographic and biochemical studies, vascular anomalies are classified as hemangiomas (now known as *infantile hemangioma)* and vascular malformations (VMs). This classification was widely adopted and became the official classification of the International Society for the Study of Vascular Anomalies in 1996 with some updates to include infantile hemangioma variants, combined lesions, and other benign vascular origin tumors.[1–7]

Infantile hemangiomas are the most common tumors in infancy, found in as many as 10% of infants by 1 year of age, especially in white and premature babies weighing less than 1000 g, with girls affected 3 to 5 more often than boys. Whites are more prone to hemangioma development, whereas the incidence in African-Americans and Asians is low (1.4% and 0.8%, respectively). Other risk factors include multiparity, advanced maternal age, placental abnormalities (ie, placenta previa). Hemangiomas are proliferative lesions characterized by increased turnover of endothelial cells. Histologic landmarks are the hyperplasia of endothelial cells, which will stain positive for glucose transporter I (in infertile hemangiomas), and the large number of mast cells The lesions appear as

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solitary lesions in approximately 80% of children, and 60% of the tumors are found in the cervicofacial region. The lesions occur sporadically, although there are some tumors that may follow an autosomal dominant inheritance pattern in familial cases. Hemangiomas may be superficial, deep, or visceral, and, although the location does not alter the biologic behavior, it does affect the clinical manifestations.

The lesions typically appear during the first 2 years of life, if they are not present or evident at birth, as an erythematous patch, telangiectasia, or blanched area when they are located superficially. Deep lesions may appear bluish or a deep purple color, whereas the visceral ones will generally not be evident on clinical examination. Infantile hemangiomas follow a triphasic pattern of evolution during which there is classically a proliferative phase of rapid enlargement that follows and outpaces the child's body growth, which may last for 4 to 8 months before a plateau phase that is consistent with body habitus growth followed by involution. The involution phase starts by age 12 months and usually continues for the next 5 to 7 years. Roughly half of the lesions involute by age 5, and 70% by 7 years of age, but the process may continue until adolescence. Even with complete involution, some clinical signs of the tumor may still be evident on physical examination.[8–16]

VMs, represent an abnormal proliferation of mature vascular elements, are present at birth, and show equal distribution between the sexes. The lesion grows with the child and is usually affected by hormonal changes with occasional accelerated growth during puberty. Unlike hemangiomas, histopathologically there is no proliferation of endothelial cells, and the number of mast cells is normal. The lesions tend to infiltrate surrounding normal tissues, which makes their management challenging.

Further subclassification of VMs is based either on the type of vessels involved (capillary, venous, arterial, lymphatic, or combined forms) or the flow characteristics of the lesion (low- vs high-flow VMs). High-flow VMs are arteriovenous malformations (AVMs), whereas low-flow lesions are capillary-venular malformations or venous or lymphatic malformations (LMs). VMs are present at birth, as they represent errors in morphogenesis but, unlike hemangiomas, may go undetected until later in life, even into the teenage years; they do not regress and continue to expand with time.

The clinical appearance and characteristics vary according to the type of VM lesion. A palpable thrill or audible bruit, for example, may be evident in high-flow lesions, whereas diascopy can help distinguish venous malformations from subcutaneous or deep hemangiomas, as both may appear bluish on clinical examination. Diascopy is performed by pressing a glass slide over the lesion and observing for blanching of the bluish/deep purple color as blood is evacuated from the VM; whereas, in contrast, no color change is noted when diascopy is performed on a hemangioma.

HIGH-FLOW LESIONS

Arteriovenous Malformations

AVMs of the head and neck are among the most aggressive of VMs that often cause significant deformities and functional limitations. Although the exact etiology and pathogenesis of AVMs are currently unknown, defects of the transforming growth factor-beta signaling and a genetic 2-hit hypothesis seem to be the 2 most likely candidate theories. Although most of these lesions are present at birth, they are often misdiagnosed as other vascular lesions, or they lack classic presentation for appropriate diagnosis. Often cases are described as appearing in adults after trauma, or a rapid expansion is noted during puberty. Regardless of presentation, these lesions are characterized by direct connections between the arterial and venous system without intervening capillaries.[8,13,14,17–19]

Seventy percent of the AVMs in the head and neck involve the midface, with the cheek, nose, and forehead the most commonly involved sites. AVMs can invade the underlying tissues and the bone of the midface or the mandible, causing significant destruction and further complicating treatment. Interestingly, some of the intraosseous lesions diagnosed as hemangiomas do represent VMs (usually low flow) based on histologic examination and immunostaining. AVMs are staged based on presentation and clinical examination. Stage I AVMs are the quiescent lesions with only minimal clinical findings; stage II lesions show expansion, palpable thrills, or bruits or pulsations; stage III lesions are those that invade adjacent structures causing destruction, pain, ulceration, or bleeding; stage IV lesions cause congestive heart failure owing to decompensation from the rapid uncontrolled growth.[13,17,20,21]

Diagnosis of Arteriovenous Malformations

Clinical examination and imaging are crucial in the diagnosis of AVMs. Imaging, either contrast-enhanced computed tomography (CT) or MRI will distinguish between VMs and vascular tumors. The VM will lack a discrete soft tissue mass and will have enlarged feeding and draining vessels. Areas of thrombosis or intralesional hemorrhage may be found within the VM as well. Magnetic