Endovascular Methods for the Treatment of Vascular Anomalies

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KEYWORDS

• Vascular malformations • Vascular anomalies • Venous malformation • Lymphatic malformation • Capillary malformation • Hemangioma • Arteriovenous malformation

KEY POINTS

- Venous malformations can be treated percutaneously by injecting a sclerosing agent. The most commonly used sclerosing agents include alcohol, sodium tetradecol, Ethibloc, polidocanol, and bleomycin.
- Multiple sclerosing agents have been used effectively for the treatment of lymphatic malformations, including absolute alcohol, sodium tetradecyl sulfate, doxycycline, Ethibloc, OK 432, and bleomycin.
- Approximately 8% of capillary malformations (CMs) are associated with Sturge-Weber syndrome and unilateral glaucoma. CMs are also associated with Klippel-Trenaunay, Parkes-Weber, macrocephaly CM, and CM-arteriovenous malformation (AVM) syndromes.
- Treatment of AVMs is difficult. Endovascular embolization is often the first treatment option with or without surgical resection. The main goal of any form of treatment is to eradicate the nidus.
- Endovascular treatment of an AVM can be performed from a transarterial approach, direct percutaneous puncture of the nidus, or a retrograde transvenous approach with alcohol, *N*-butyl-2-cyanoacrylate (n-BCA), or Onyx.
- Congenital hemangioma is a vascular lesion that completes the proliferative phase before birth. Two forms exist: the rapidly involuting congenital hemangioma (RICH) and the noninvoluting congenital hemangioma (NICH). RICHs involute more rapidly than hemangiomas of infancy, usually within the first 14 months of life. NICHs do not involute but grow in proportion to the child.

INTRODUCTION

In 1982, John Mulliken and Julie Glowacki proposed a classification of vascular anomalies based on clinical behavior, histology, and histochemistry.[1] The International Society for the Study of Vascular Anomalies (ISSVA) accepted this classification in 1992.[2–4] The classification divides vascular anomalies into 2 groups: tumors (eg, hemangiomas), the cause of which is endothelial cell proliferation, and vascular malformations, in which developmental error has resulted in abnormally formed vascular channels. In the tumor group, this article

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focuses on hemangiomas; the other tumor subtypes are beyond the scope of this review. Vascular malformations can be further subdivided into lesions consisting of arterial, capillary, lymphatic, venous, and fistulous networks. Furthermore, they can be further subdivided functionally based on the flow characteristics (ie, high-flow vs low-flow lesions).[5] This article discusses the clinical features, natural history and epidemiology, and presents the diagnostic imaging features of vascular anomalies of the head and neck. The percutaneous/endovascular treatment of each of the vascular anomalies are presented. Additional treatment options, such as surgery are discussed briefly. Finally, the clinical outcomes of the main forms of treatment and level of evidence are presented.

VENOUS MALFORMATIONS

Clinical Features

Venous malformations (VMs) can occur anywhere in the body, but are most frequently located in the head and neck. They can be solitary, small, well circumscribed, large, superficial, or infiltrative involving multiple tissue planes. The lesion is nonpulsatile, may have a light blue to deep purple color, and can be associated with telangiectasias, varicosities, or ecchymosis. The mass may increase in size in a dependent position, with a tourniquet, or during a Valsalva maneuver. Superficial lesions are soft and compressible and can usually be emptied of blood. Patients usually present with pain associated with compression on the surrounding nerves or thrombosis of a portion of the mass. An increased level of D-dimer has been determined to be highly specific for a VM and can help to distinguish VMs from lymphatic malformations and slow-flow Klippel-Trenaunay syndrome from high-flow Parks-Weber syndrome.[6]

VMs are usually isolated findings; however, they may be associated with the following syndromes (**Table 1**):

Klippel-Trenaunay syndrome
Blue rubber bleb nevus (BRBN) syndrome
Mucocutaneous familial VMs
Glomuvenous malformation
Maffucci syndrome
Proteus syndrome
Bannayan-Riley-Ruvalcaba syndrome
CLOVES/S syndrome

Natural History/Epidemiology

VMs are present at birth. They are not always clinically apparent and tend to grow in proportion to the growth of the child. The growth is most pronounced during puberty and pregnancy. These are congenital lesions that affect boys and girls with equal frequency with a reported incidence of 1 to 2 per 100,000 births and a prevalence of 1%.[7]

Most VMs (95%) are sporadic, but can be seen in several heritable conditions. The molecular basis for sporadic occurrence has yet to be discovered, however there are familial cases where the genetic defect has been localized on a specific chromosome. In 1994/1995, 2 families were identified to have autosomal dominant inherited cutaneous and mucosal VMs. Genetic analysis mapped a locus for both of these families to chromosome 9p21.[8,9] These families shared a mutation resulting in an arginine to tryptophan substitution R849W in the gene that encodes for the kinase domain of the endothelial cell receptor Tie2.[10] In 1999, 4 more families with autosomal dominant inherited VMs were identified.[11] Only 1 of those families shared the same mutation as the previous 2 reports. The second family had a novel hyperphosphorylating Y897S mutation in the *TIE2* gene. The other families showed no evidence of linkage to 9p21, which suggests genetic heterogeneity. Multifocal VMs are most commonly seen in the familial forms of VM, including the following: BRBN syndrome, mucocutaneous familial VMs, glomuvenous malformation, and Maffucci syndrome.[12]

Diagnostic Imaging

Ultrasonography

On grayscale imaging, VMs are usually hypoechoic to anechoic with the shape of tubular structures.[13] Some lesions can have a heterogeneous echotexture if phleboliths or different forms of thrombus are present within the lesion. Doppler flow is usually a monophasic low-velocity flow. Sometimes flow can only be seen with compression and release of the lesion.[14]

Computed tomography

On noncontrast computed tomography (CT), VMs are usually hypoattentuating, however they can be heterogeneous depending on the amount of fatty tissue within the lesion. Phleboliths or dystrophic calcifications can be seen within the lesion. After the administration of contrast, the lesion usually enhances on the periphery and then fills in centrally on the delay images.[14] CT is excellent at looking for boney involvement from the lesion. Magnetic resonance (MR) imaging is better at characterizing the relationship of the lesion with surrounding soft tissue structures.

MR imaging

VMs usually appear as hypointense to isointense on T1-weighted imaging. They can, however,