

Vascular Anomalies (Birthmarks) of the Foot and Ankle

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Vascular anomalies (birthmarks) commonly involve the feet and ankles. Little is known about these anomalies among practicing physicians. In this article, vascular anomalies are described, and detailed information is presented regarding appropriate diagnostic work-up and treatment strategies. (J Am Podiatr Med Assoc 94(5): 477-482, 2004)

Vascular anomalies (birthmarks) are commonly encountered lesions and vary from tiny skin discolorations to large, devastating lesions requiring invasive interventions (Table 1).¹ These lesions can involve any part of the body, but the lower extremities, particularly the foot and ankle regions, are commonly affected. A vascular anomaly in the foot or ankle usually requires treatment because of significant pain and discomfort regardless of the size of the lesion, whereas the same lesion in a different location (eg, the trunk) may be managed simply by observation and may not require treatment.

It is common to see incorrect terminology used in the diagnostic work-up of vascular anomalies. A typical example is the term *hemangioma* or *cavernous hemangioma*, which is commonly used for *venous malformation* (VM). Also, it is common for practitioners, even pathologists, to refer to any non-neoplastic vascular lesion as an *arteriovenous malformation* (AVM), which causes confusion, erroneous staging, and mistreatment in some cases.

Limited experience with vascular anomalies among diagnosticians also leads to less-than-ideal testing being performed during the diagnostic work-up of these patients. For example, computed tomography (CT) and noncontrast magnetic resonance imaging (MRI) do not provide any useful information in most cases. Also, using advanced imaging techniques, such as MR angiography and CT angiography, requires solid knowledge not only of vascular anomalies but also of the new technologies and imaging modalities. Currently, the rate of misdiagnosis may be higher

than 50% of all patients referred to a dedicated tertiary-care medical center.²

This article aims to guide practitioners who occasionally encounter vascular anomalies in the process of diagnosing these anomalies and to briefly describe the currently available treatment options. Various vascular anomalies are described in descending order of prevalence in daily practice. More information on vascular anomalies can be found at my Web site (<http://www.birthmarks.us>).

Venous Malformations

Venous malformations are vascular lesions that are commonly (erroneously) called hemangiomas or cavernous hemangiomas. These slow-flow vascular anomalies are congenital, like any other birthmark, but may not be obvious at birth and may appear later in life. Venous malformations are probably the most common symptomatic vascular anomalies, and may be much more common than reported in the literature. Bluish skin discoloration, local swelling, and pain are usually present with VMs (Fig. 1). Pain is the most common reason for seeking medical help for foot and ankle VMs. On physical examination, they are soft and easily compressible lesions that usually engorge with dependency (less prominent early in the morning and more swollen and bothersome at the end of the day). The lesion may be focal and small, or it may involve the entire foot and ankle diffusely or in a scattered fashion (Figs. 2 and 3). Foot and ankle VMs are usually painful because most involve one or more muscle groups rather than just the subcutaneous fatty tissue. When the lesion involves the subcutaneous tissue as well as the foot muscles,

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Table 1. Biological Classification of Vascular Anomalies by Mulliken and Glowacki¹

I. Hemangiomas and congenital vascular tumors	Infantile, congenital, noninvoluting, intramuscular, kaposiform hemangioendothelioma
II. Vascular malformations	
A. High flow	Arteriovenous malformation Arteriovenous fistula
B. Low flow	Capillary malformation (port-wine stain) Venous malformation Lymphatic malformation
C. Combined	
1. High flow	The Parkes-Weber syndrome (CLAVM with limb overgrowth)
2. Low flow	The Klippel-Trénaunay syndrome (CLVM with limb overgrowth) The Maffucci syndrome (venous malformation–like lesions with enchondromatosis)

Abbreviations: CLAVM, capillary lymphatic arteriovenous malformation; CLVM, capillary lymphatic venous malformation.

Note: It is important to categorize the lesions as high- or low-flow abnormalities. In general, high-flow anomalies are treated by transcatheter embolization, whereas percutaneous sclerotherapy is the treatment of choice for low-flow anomalies.

usually the portion of the lesion involving the muscle groups causes pain. Venous malformations are known to be a “benign” form of vascular anomaly, usually stable in size, but they typically grow or expand commensurately with the patient. On rare occasions, a foot or ankle VM may occur as part of a complex vascular anomalies syndrome (the Klippel-Trénaunay syndrome, the Maffucci syndrome, or the blue rubber bleb nevus syndrome).

Because they are slow-flow anomalies, the lesions fill with blood slowly during the venous phase (in contrast, lesions fill rapidly in high-flow anomalies, such as AVMs). In most patients, the diagnosis of VM can be made on the basis of clinical findings alone, although imaging (typically MRI) is indicated to confirm the diagnosis and, more importantly, to evaluate the extent of the malformation for treatment planning. On plain film radiographs, VMs may demonstrate single or multiple rounded calcifications (phleboliths) in the involved soft tissues, which may or may not be associated with adjacent bony changes (Fig. 4). Radiographic bony changes include lacelike lucencies, thinning of the osseous shaft, erosions, and bony expansions. The presence of osseous change, however, does not correlate with patient symptoms.³



Figure 1. An extensive venous malformation in the foot and ankle. Bluish skin changes with easily compressible swelling are characteristic of this malformation. This lesion involves almost the entire foot and extends into the ankle region. Surgical excision of this lesion is not feasible because several muscle groups and bony structures are also involved. However, this lesion can be treated with several sessions of direct, intralésional sclerosant injections (sclerotherapy).

Magnetic resonance imaging needs to be performed using the vascular anomalies protocol, which specifically requires postcontrast fat-saturated scanning and gradient echo imaging (Figs. 5 and 6). The extent of the lesion is best evaluated on T2-weighted images, whereas postcontrast images and gradient echo images are usually helpful to rule out high-flow anomalies, such as AVMs.²



Figure 2. A relatively small venous malformation in the lateral dorsal aspect of the foot involving the fourth and fifth toes. This foot deformity is easily compressible and is not tender on palpation. Magnetic resonance imaging is indicated to assess the extent of the lesion. Surgical excision can be attempted for this relatively focal lesion; however, the portion of the lesion that extends into the musculature is the area that causes pain, and it cannot be removed surgically without causing sequelae. Therefore, this lesion also needs to be treated with sclerotherapy.



Figure 3. Another venous malformation in the medial plantar aspect of the midfoot. The surgical scar is from a failed attempt to remove the lesion. This lesion was treated with sclerotherapy with good results.



Figure 4. A plain film radiograph of the foot shows multiple phleboliths and lacy osseous change in the first metatarsal and proximal phalanx of the first toe and thinning of the second metatarsal. These radiographic findings are characteristic of venous malformations.

Treatment of symptomatic foot and ankle VMs varies depending on the size and type of the lesion, which ranges from a focal abnormality to multiple small, scattered lesions with or without varicoid venous abnormalities. Small subcutaneous VMs can be removed surgically; however, subcutaneous lesions are usually asymptomatic or are the asymptomatic portion of a larger lesion. More problematic VMs involve the muscles of the foot and ankle, and these lesions cannot safely be surgically removed. Sclerotherapy (percutaneous sclerosant injections into the lesion) is the only treatment option for these patients. To obtain the best possible results, sclerotherapy requires solid knowledge and technical skills and should

be performed by an interventional radiologist who has experience with vascular anomalies. Of the different sclerosants available (eg, absolute alcohol, sodium tetradecyl sulfate, and ethanolamine oleate), absolute alcohol is the most effective; however, using it requires precise and careful infusion of the agent into the malformation under real-time imaging so that complications related to extravasation of the sclerosing agent can be minimized.⁴ Other sclerosants, particularly sodium tetradecyl sulfate and ethanolamine oleate, are generally preferred when the lesion involves important nerve bundles directly or when the skin is significantly involved to reduce alcohol-related nerve or skin damage. The patient should also

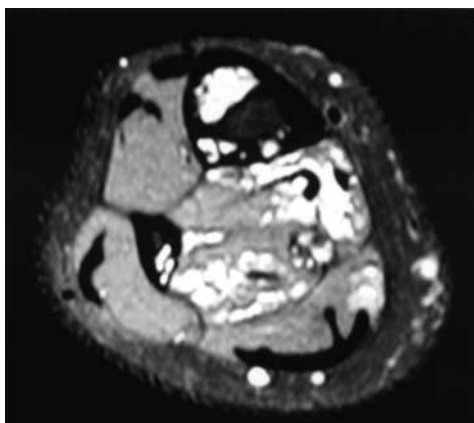


Figure 5. An axial T2-weighted magnetic resonance image of the ankle shows scattered bright lesions in the subcutaneous space, in the muscles, and in the distal tibia and fibula. T2-weighted imaging is the ideal method of evaluating the extent of venous malformations.

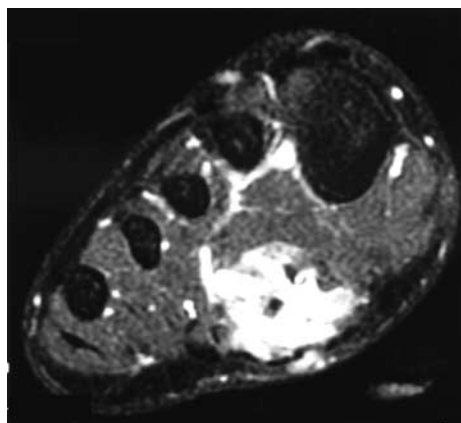


Figure 6. Axial fat-saturated postcontrast magnetic resonance image of the midfoot shows an intense contrast enhancement in the lesion, characteristic of venous malformations. Lymphatic malformations show no contrast enhancement or only a minimal degree of peripheral enhancement.

be observed closely for compartment syndrome after sclerotherapy. Sclerotherapy is particularly difficult in scattered multifocal small lesions owing to difficult access into the lesion.

Lymphatic Malformations

Lymphatic malformations (LMs), less common than VMs, are also slow-flow vascular anomalies, and the lower extremities are probably the most common sites for occurrence after the head and neck. Similar to VMs, LMs are congenital vascular malformations that are generally noticed at birth. Three morphological types have been described: microcystic, macrocystic, and mixed. Lymphatic cysts typically contain lymphatic fluid (which may be bloody owing to intralesional bleeding). If the lesion does not have any cysts (lesion appears totally solid) on MRI, it is considered microcystic; if the lesion contains a cyst or cysts without any solid-appearing abnormal tissue, it is considered macrocystic; and if the lesion contains both cystic and solid-appearing portions, the lesion is considered mixed.² This classification is particularly helpful in treatment planning (macrocystic and mixed-form lesions can be treated with sclerotherapy). An experienced practitioner can make the diagnosis on the basis of history and physical findings, without any imaging or testing (Fig. 7). Pain is less likely to be the reason for seeking treatment than is the appearance of the involved body part. The involved area usually shows an obvious soft-tissue mass-like deformity, usually associated with vesicular skin changes. If the



Figure 7. A focal lymphatic malformation is seen as an obvious soft-tissue mass-like lesion involving the second and third toes with characteristic vesicular skin changes. This lesion is not compressible or tender on palpation. Magnetic resonance imaging (not shown) revealed microcystic disease; therefore, this lesion cannot be treated with sclerotherapy. If a lymphatic malformation has a cystic morphology (macrocystic), sclerotherapy is an excellent alternative to surgery.

lesion consists mainly of large cysts, the lesion is soft and not painful on palpation; if the lesion is mostly microcystic, it generally appears as a more solid, non-compressible lesion. Enlargement of the lesion with infection and infection of the lesion are common.

Although LMs can be recognized clinically, imaging (typically MRI) is required to identify the morphological type that would eventually determine the treatment modality. If the lesion is macrocystic, sclerotherapy should be the initial approach, although some surgeons advocate a surgical approach. However, most are more complex than a simple loculated cyst or cysts and involve multiple tissue planes that make a surgical approach very difficult, if not impossible. In the foot and ankle, most lymphatic malformations are mixed, containing some cysts and some solid-appearing soft-tissue abnormalities on imaging. In the extremities, LMs can cause diffuse or localized swelling with soft-tissue and skeletal overgrowth. Rarely they are associated with progressive osteolysis (the Gorham-Stout syndrome).⁵

At The Cleveland Clinic Foundation, Cleveland, Ohio, the general therapeutic approach is conservative (observation and symptomatic measures, such as infection control and pain management) in most cases; we use sclerotherapy in symptomatic patients. If sclerotherapy is not feasible because of either the lack of treatable cysts within the lesion (microcystic or mostly microcystic mixed-form LM), a surgical approach is selected. Potential surgical complications include poor wound healing owing to lymph leak and scarring.

In sclerotherapy, the sclerosants used for VMs are generally accepted, with one major exception—doxycycline. This antibiotic can be effective as a sclerosing agent for LMs. Its only drawback is the potential risk of tooth discoloration (darkening) when it is used in children (<8 years of age). Although this concept is well established, the available data are based on systemic tetracycline use, and it is speculated that tooth discoloration should be less problematic with its semisynthetic derivatives (doxycycline).⁶ No data are available regarding whether intralesional doxycycline injections cause permanent tooth discoloration. The other sclerosant that can be used for LMs is OK432,⁷ a heat- and penicillin-treated lyophilized powder of the Su strain of *Streptococcus pyogenes* that is available in Japan.

During sclerotherapy, the cyst content is drained as much as possible, and then the lesion is infused with the selected sclerosant.⁴ Depending on the lesion size, sclerotherapy may need to be repeated. Significant swelling can be seen after sclerotherapy, which may take several days to resolve. Infection

control is important in LMs during and after the sclerotherapy procedure.

Arteriovenous Malformations

Arteriovenous malformations are less common than generally believed or speculated. Most patients diagnosed as having an AVM actually have a VM. The most common reason for this misconception or misnomer is lack of experience in the field of vascular anomalies and the use of the term AVM for any vascular malformation. Yet AVMs are completely different vascular lesions from VMs, not only in their presentation but also in their management options and expected lesion progression. Arteriovenous malformations are fast-flow lesions characterized by abnormal vascular connections (“nidus”) between arteries and veins. Therefore, there is significant arteriovenous shunting through the nidus, which may cause high-output cardiac failure. Generally, they are present at birth, but they usually become obvious because of various stimuli, such as trauma, pregnancy, or puberty. Pain is the leading complaint, but overgrowth, ischemia, and hemorrhage may also be present.⁸ Bleeding (spontaneous or due to trauma) from an AVM can be serious because high pressure (arterial pressure) is present in the lesion. On physical examination, increased warmth and redness and a pulsatile lesion are typically found (Fig. 8). If the lesion involves the entire extremity, the condition is known as the Parkes-Weber syndrome.

Although an AVM is rare in the foot and ankle, it can occur as a small focal lesion in the foot or ankle, or the foot and ankle may be involved in an extensive leg AVM. The high-flow nature of the malformation can be confirmed with a sonographic examination. Magnetic resonance imaging shows multiple enlarged vascular channels in the lesion without a “mass” lesion. However, a masslike signal abnormality can be seen in the area, which is generally due to edema.² Increased fatty tissue may also be found in the involved area. Occasionally, the bones are involved as well, which may cause lytic or lacy osseous changes or cortical thinning.

Treatment depends on the extent of the malformation. Most AVMs can be controlled, if not cured, with a series of careful transcatheter embolizations, although there is considerable risk of digit necrosis after embolization, which may require amputation. Therefore, embolization should be performed by a dedicated interventional radiologist who has adequate skills, training, and experience in vascular anomalies. A commonly used embolization agent is absolute alcohol or glue. Particles (polyvinyl alcohol



Figure 8. An extensive arteriovenous malformation in the leg; the foot and ankle are greatly involved. There are multiple various-sized varicoid venous abnormalities due to rapid arteriovenous shunting. Scattered skin color changes are seen in the medial aspect of the distal calf and ankle. Scars seen in the upper medial calf are from failed surgical procedures.

or microspheres) can also be used in selected cases. Coils can be used only in the venous side of the AVM in selected cases. In some cases, a percutaneous approach can also be used effectively. Coiling of the feeding arteries is contraindicated because this results in more arterial small feeders with no easy arterial access remaining into the AVM nidus.

The Klippel-Trénaunay Syndrome

The Klippel-Trénaunay syndrome is a capillary-lymphatic-venous malformation that is typically associated with an overgrowth of the leg. It may be associated with muscle atrophy.⁸ There is generally a capillary stain in a geographic pattern. Abnormally dilated lateral veins are typically seen (Fig. 9). Also, the pathognomonic marginal vein of Servelle is frequently identified in the lateral calf and thigh. There may be incompetent venous valves or an absent deep venous system. Toe abnormalities are common, including macrodactyly, syndactyly, metatarsus primus varus, clindactyly, polydactyly, and camptodactyly.⁹ Soft-tissue signal abnormalities characteristic of slow-flow vascular anomalies are generally seen on MRI. Lymphatic abnormalities are common in the involved leg.



Figure 9. An example of Klippel-Trénaunay syndrome; some skin color changes are seen around the knee. Abnormal lateral veins are seen in the calf, which are usually associated with low-flow vascular anomalies (such as venous and lymphatic malformations) of the leg. The deep venous system must be evaluated in detail before any attempts are made to remove the venous abnormalities surgically or by using interventional techniques (eg, sclerotherapy).

Treatment is conservative in most patients. If there are isolated slow-flow anomalies or large, painful varicoid veins, sclerotherapy can be performed. However, it is important to confirm the patency of the deep veins before any intervention.

Other Vascular Anomalies That May Involve the Foot and Ankle

The remaining vascular anomalies are rarely seen in the foot and ankle and are described very briefly. Capillary malformation (port-wine stain) is actually very common but rarely requires treatment because the malformation involves only the skin and causes minimal or no symptoms. Capillary malformations are typically seen in the head and neck or trunk. Hemangiomas are benign tumors that commonly develop in the first few months of life; typically, they grow within the first year and involute within the first few years. Hemangiomas also occur commonly in the head and neck. An arteriovenous fistula is a single connection between an artery and a vein, causing arteriove-

nous shunting similar to an AVM. Arteriovenous fistulas are much more benign clinically than AVMs and can be cured much more easily by either embolization or surgical ligation of the vascular connection. Proteus syndrome is characterized by verrucous nevus, lipomas/lipomatosis, macrocephaly, and asymmetrical limbs with partial gigantism of the hands and feet. Abundant fat deposition is characteristic on imaging studies.¹⁰ The Maffucci syndrome is characterized by slow-flow vascular anomalies (similar to VMs) associated with bony exostoses and enchondromas. Malignant transformation, most commonly chondrosarcoma, occurs in some patients.¹¹

Conclusion

Vascular anomalies are common vascular birthmarks of the foot and ankle. These abnormalities require a careful diagnostic work-up to select an appropriate therapeutic modality. Most vascular birthmarks can be treated using percutaneous means, transcatheter embolization, or sclerotherapy.

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