Low-Flow Vascular Malformation Pitfalls: From Clinical Examination to Practical Imaging Evaluation—Part 2, Venous Malformation Mimickers

Brandon Olivieri, Candace L. White, Ricardo Restrepo, Brett McKeon, S. Pinar Karakas, Edward Y. Lee

OBJECTIVE. The purpose of this article is to review the unusual clinical and radiographic features of venous malformations that can give rise to diagnostic confusion. Entities that can have overlapping clinical and imaging features with venous malformations are also reviewed.

CONCLUSION. Venous malformations are congenital endothelial malformations secondary to errors in vascular morphogenesis and are usually diagnosed in the first 2 decades of life. The clinical and imaging features of venous malformations often overlap those of other pathologic entities, creating diagnostic confusion. Furthermore, the clinical presentation and imaging appearance of venous malformations can vary, making the diagnosis challenging. Thorough knowledge of the various clinical and imaging features not only of venous malformations but also of the major potential mimic lesions is crucial for clinicians caring for patients with these lesions.

Keywords: anomaly, imaging, malformation, pitfalls, slow flow, vascular, venous

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Venous malformations (VMs) are congenital endothelial malformations that result from errors in vascular morphogenesis [1]. VMs are composed of vascular channels sometimes containing intraluminal thrombi, are lined by thin endothelium, and, with capillary and lymphatic malformations, are part of the low-flow subclassification of vascular malformations [2–5]. They are usually present at birth but are not always apparent and grow in proportion to the child’s growth until puberty [2, 4]. The estimated incidence of VMs is 1 in 10,000, and VMs are the low-flow vascular malformation most frequently referred to specialized centers [6–8].

The diagnostic workup of VMs includes a detailed history, clinical examination, and imaging evaluation. It is important that the radiologist personally interview the parents and directly examine the patient at the time of the imaging study. The goal of this article is to review the varied clinical and imaging appearance of VMs in infants and children. A variety of pediatric pathologic entities that can mimic VMs are also reviewed, as are clues to minimizing the risk of misdiagnosis and delayed treatment.

Clinical Presentation and Imaging Appearance of Venous Malformations

VM is one of the major subcategories of low-flow vascular malformations, along with capillary and lymphatic malformations. Most often located superficially within the head and neck (40%), trunk (20%), or limbs (40%), VMs can also be found in the viscera [3, 5, 9]. Although many pediatric patients with a VM initially undergo evaluation for cosmetic reasons, they may also seek medical care because of swelling from dependent stasis, pain from localized thrombosis, or limitation of activities. Like those with other vascular malformations, pediatric patients may present because of altered limb growth and gait abnormalities [4, 5, 10].

The typical appearance of a superficial VM at clinical examination is a nonpulsatile, compressible area of soft-tissue prominence or a discrete soft-tissue mass that causes no alteration in skin temperature, thrill, or bruit [4, 5, 9, 11]. The lesions usually increase in size and coloration during a Valsalva maneuver, with dependent positioning, and sometimes with the application of a tourniquet. If the skin is involved, a blue-purple hue or superficial veins can be seen [4, 5, 9] (Fig. 1). Sudden enlargement may occur after trauma and with intraleSIONAL thrombosis [4, 9, 12]. Enlargement has also been reported during the hormonal changes of puberty, pregnancy, and oral contraceptive use [13, 14]. The low-flow nature of VMs makes them inherently prone to repeated bouts of thrombosis [11]. Elevated levels of D-dimers are found to be
present in 42% of patients with VMs and are highly associated with pain due to thrombosis [8, 15]. There is considerable variation in lesion number, ranging from solitary to multiple, and in size, ranging from small circumscribed (masslike) to extensive infiltrative lesions crossing multiple tissue planes [4, 5, 9].

In addition to their variable clinical appearance and presentation, VMs also have substantial variability in imaging appearance. The three main venographic patterns—cavitary, spongiform, and dysmorphic—serve as the best lesion descriptors and can be translated into the interpretation of other imaging modalities [9]. Dubois et al. [16] and Puig et al. [17, 18] further subdivided these patterns into the following four types based on patterns of venous drainage (Fig. 2): type I, isolated malformations without venous drainage; type II, malformations that drain into normal veins; type III, malformations that drain into dysplastic veins; and type IV, malformations consisting primarily of venous ectasia [4, 16–18].

Ultrasound with color Doppler evaluation for perceptible flow is usually the first line of imaging in the evaluation of suspected VMs. At gray-scale sonographic evaluation, superficial VMs are compressible with heterogeneous echotexture (98%) and can be hypoechoic (82%), hyperechoic (10%), or isoechoic (8%) with respect to surrounding structures [19]. Although highly suggestive of VMs, calcified phleboliths are seen in only 16% of cases, whereas tubular anechoic structures indicative of vascular channels are seen in 4% [19]. According to the venographic classification, VMs can be seen as focal, well-defined, sponglike lobulated structures with varying echogenicity (Fig. 1) or as multiple tortuous and beaded varicosities arranged in a haphazard manner, violating multiple tissue planes [4, 16–18].

Color Doppler examination reveals waveforms with monophasic flow typical of venous structures in 78% of cases [19]. Sixteen percent of VMs have minimal or no flow, which may reflect very low flow below detectable limits or thrombosis and a possible source of diagnostic confusion [19] (Fig. 1). Biphasic flow is seen in 6% of lesions, possibly reflecting a mixed capillary component [19]. Arterial waveforms may be seen and likely represent the neighboring arteries traversing the VM (Fig. 1) or intravascular papillary endothelial hyperplasia (Masson tumor) (Fig. 3). When localized, spongiform, and in the presence of internal arteries, VMs can be confused with a neoplasm.

Given its superior soft-tissue contrast resolution, multiplanar capabilities, and lack of ionizing radiation, MRI is currently the preferred complementary imaging modality to ultrasound and is particularly useful in evaluating the full extent of lesions and for planning and assessing response to therapy. VMs typically are hypointense to isointense on T1-weighted MR images. Areas of high T1 signal intensity may be due to the presence of intraleSIONal fat or thrombi, and scattered punctate low-signal-intensity areas are likely due to phleboliths. Because of their low-flow nature, VMs are markedly hyperintense on fluid-sensitive sequences (Fig. 1). These sequences, especially with fat saturation, are therefore ideal for determining lesion extent. Areas of low signal intensity are usually related to thrombosis or phleboliths and can be confirmed on gradient-echo T2*-weighted MR images by identifying hemosiderin and calcification. VMs may have fluid-fluid levels that reflect layering hemorrhage [20]. On contrast-enhanced T1-weighted fat-saturated MR images, the circulatory portions of VMs have a wide range of contrast enhancement patterns: from homogeneous to heterogeneous, faint to vivid, and rapid to delayed [4, 9] (Fig. 1). The MRI protocol is shown in Table 1.

CT plays a limited role in the evaluation of VMs because of its lower soft-tissue contrast resolution, use of ionizing radiation, and erratic contrast enhancement, which is typical of these low-flow malformations often necessitating additional phases. Likewise, conventional radiography is not routinely used to evaluate VMs unless reactive bone changes are being assessed [4, 21]. The finding of phleboliths on radiographs is virtually but not always diagnostic of a VM [4, 5].

There are two specific situations in which VMs can pose diagnostic difficulties: when associated with intravascular papillary endothelial hyperplasia and when intramuscular in location. VMs are prone to stasis and intraluminal clotting secondary to their low-flow nature. When this occurs, exuberant intralesional intraluminal capillary ingrowth, also known as intravascular papillary endothelial hyperplasia or Masson tumor, can occur [3, 22]. This uncommon, locally occurring benign process is poorly understood but is thought to reflect an abnormal form of thrombus organization in which the clot serves as a scaffold for the ingrowth of endothelium-lined, papillary, frondlike projections that form intraluminal vascular channels [23–25]. Intravascular papillary endothelial hyperplasia may develop in a preexisting vascular malformation and therefore poses a diagnostic dilemma [25–28]. The lesion-to-lesion variation in proportions of internal solid components and septa and low-flow vascular channels result in variable amounts of vascular flow at Doppler imaging (Fig. 3) and no reliable intensity, homogeneity, or contrast-enhancement pattern at MRI [23, 25, 26]. Owing to

**TABLE 1: Suggested MRI Protocol for Suspected Vascular Malformation**

<table>
<thead>
<tr>
<th>Sequence</th>
<th>Feature Evaluated</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coronal (less frequently sagittal) STIR or T2-weighted fat-saturated</td>
<td>Extent of the lesion</td>
</tr>
<tr>
<td>Axial T1-weighted</td>
<td>Contents: blood, proteinaceous material, fat, and involvement of adjacent musculature</td>
</tr>
<tr>
<td>Axial STIR or T2-weighted fat-saturated</td>
<td>Relation between malformation and adjacent structures: skin, neurovascular bundle, bone</td>
</tr>
<tr>
<td>Axial gradient T1-weighted</td>
<td>Presence of phleboliths, recent bleeding, or flow voids in hypervascular lesions</td>
</tr>
<tr>
<td>Axial T1-weighted fat-saturated before gadolinium enhancement</td>
<td>Enhancement pattern (sequence required for this evaluation)</td>
</tr>
<tr>
<td>2D Time-of-flight MR angiography and MR venography⁴</td>
<td>Vascular pattern, deep venous system, and flow characteristics</td>
</tr>
<tr>
<td>Multiple-phase gadolinium-enhanced MR angiography⁴</td>
<td>Flow dynamics</td>
</tr>
<tr>
<td>Axial T1-weighted fat-saturated after gadolinium enhancement</td>
<td>Enhancement pattern: dynamics and possible solid component</td>
</tr>
<tr>
<td>Second-plane T1-weighted fat-saturated after gadolinium enhancement</td>
<td>Delayed enhancement; very useful in venous malformations</td>
</tr>
</tbody>
</table>

⁴Optional sequence for evaluation of definite lymphatic malformation or for follow-up of venous malformation.
the presence of solid component and internal arterial and capillary flow, these lesions are often confused with neoplasms [25, 26, 28].

Purely intramuscular vascular malformations are another clinical and imaging diagnostic dilemma. Unlike nonintramuscular VMs, intramuscular VMs tend to present later in life, probably because of a deeper location and delayed progression to a symptomatic state secondary to muscle contraction-related decreased intralesional venous stasis [22]. Furthermore, because of increased risk of local intravascular coagulation and thrombosis, intramuscular VMs are associated with higher rates of morbidity than their nonintramuscular counterparts. Intramuscular VMs also have a higher rate of pain as the presenting symptom (30% vs 15%) and cause greater limitation of physical activity (35% vs 16%) [22, 29]. Because of this greater propensity toward a painful, symptomatic presentation at later ages, intramuscular VMs can be confused clinically with soft-tissue neoplasms [3].

Purely intramuscular VMs tend to be well defined, oval, or round and often are adherent to neurovascular bundles, a trait that can mimic peripheral nerve sheath tumors and soft-tissue sarcomas. At ultrasound and MRI examinations, the venous channels are confined to and follow the long axis of the muscle [30] (Fig. 4). Intramuscular VMs also tend to have higher rates of calcification than their nonintramuscular counterparts, further clouding the diagnostic picture [22, 31, 32].

**Venous Malformation Mimickers**

The following soft-tissue lesions may have clinical and imaging appearances similar to those of VMs, especially the localized spongiform type (Fig. 1).

**Peripheral Nerve Sheath Tumors**

Neurofibromas and schwannomas are benign peripheral nerve sheath tumors (PNSTs) [33]. Approximately 90% of neurofibromas are localized and can be a superficial plaque-like or a deep fusiform mass [34]. Plexiform neurofibromas are multinodular or fascicular-tubular masses along a specific nerve distribution, ranging from a rosary-bead to a bag-of-worms appearance [33] (Fig. 5).

Schwannomas commonly occur along the spinal and sympathetic nerve roots and along the nerves of the flexor aspects of the limbs as encapsulated, eccentric, and fusiform masses [33, 35]. Schwannomas can resemble localized neurofibromas clinically and on images [33, 35].

**Venous Malformation Mimickers**

On MR images PNSTs are isointense to hypointense to muscle on T1-weighted images and hyperintense on fluid-sensitive images, usually with vivid and homogeneous contrast enhancement, as can be seen with VMs. PNSTs can develop cystic areas due to hemorrhage or necrosis and therefore display fluid-fluid levels, a feature shared with VMs [36]. The localized and plexiform neurofibromas and schwannomas can exhibit a characteristic target sign with central low signal intensity on fluid-sensitive images (Fig. 5). After IV contrast administration, however, the center becomes more enhanced than the periphery, reversing the target sign [33]. Diffuse-type neurofibromas are indistinct, infiltrative masses with small fascicles or nodules without a target sign and extend to the skin surface [37, 38] (Fig. 5). They are isointense or hyperintense to muscle on T1- and T2-weighted MR images, exhibit diffuse and avid enhancement, and can be associated with ectatic low-flow vessels and fatty hypertrophy [37–39]. Additional helpful imaging findings of PNST include a fusiform mass with entering and exiting nerve roots at both ends of the mass (string sign); fascicular bundles inside the mass (fascicular sign); and splayed fat tissue around the mass (split-fat sign) [33–35]. When these classic signs are not present, however, neurofibromas can mimic VMs, especially when the patient does not have a history of neurofibromatosis type 1 [40, 41].

The bag-of-worms appearance of plexiform neurofibromas can mimic the dilated, tubular venous channels of dysplastic VMs on T1-weighted and fluid-sensitive MR images (Fig. 5). The different contrast enhancement patterns therefore are critical in differentiating the two entities. Both lesions can clinically present as spongy, poorly defined masses that grow with the patient until puberty. PNSTs are rubbery and firmer than VMs [40, 41]. Brownish skin discoloration and a hairy patch can be associated with subdermal neurofibroma [39]. A target appearance suggests but is not pathognomonic of PNST (Fig. 5). On fluid-sensitive images, a targetlike sign can be seen in VMs owing to the central hypointense thrombus and the peripheral hyperintensity of the flowing blood; however, the central hypointense thrombus displays no contrast enhancement (Fig. 6), as opposed to the central enhancement of PNST [41]. Identification of phleboliths at imaging or clinical examinations supports the diagnosis of VM. Ultrasound can be a problem-solving tool because the dilated and tortuous venous channels of VM visualized with gray-scale ultrasound exhibit venous flow or no flow and augmentation at color Doppler imaging [4, 19]. Applying a tourniquet proximally with subsequent enlargement of the venous channels, not seen with PNST, is also helpful.

**PTEN Hamartoma of Soft Tissue**

*PTEN* hamartoma syndrome is a spectrum of disorders resulting from mutations in the tumor suppressor gene *PTEN* on chromosome 10q23.3. These disorders include Cowden syndrome, Bannayan-Riley-Ruvalcaba syndrome, Proteus syndrome, and Proteus-like syndrome [42]. *PTEN* hamartoma syndrome entails proliferation of mesenchymal elements resulting in formation of multiple hamartomas in different locations, including the soft tissues [42, 43]. *PTEN* hamartomas of soft tissue are most commonly located on the lower extremity, followed by the upper extremity, trunk, head, and neck [43]. Size can vary; the range was 3–25 cm in one study, and 20–57% of lesions were multifocal [43, 44]. As in the clinical presentation of a VM, *PTEN* hamartoma of soft tissue usually presents during childhood or early adolescence with skin discoloration, pain, swelling, stiffness, prominent veins over the area of the tumor, or a combination of these findings [43, 44].

Commonly confused with VM or arteriovenous malformation, *PTEN* hamartomas of soft tissue are tumors composed of nonencapsulated clusters of high-flow feeding vessels and abnormally dilated draining veins nested within varying amounts of adipose and fibrous tissue. Thus more than 90% of *PTEN* hamartomas of soft tissue have CT or MRI evidence of disorganized ectopic fat [43, 44]. Although the tumor is usually centered intramuscularly (85%), the concomitant involvement of subcutaneous fat, dermis, and occasionally subjacent bone is often present without regard to fascial planes [43]. *PTEN* hamartomas of soft tissue have a large vascular component mainly in the form of arteriovenous fistulas with a draining vein that have characteristic areas of focal dilatation called disproportionate venous ectasia [44].

Clinical history may seem crucial in differentiating *PTEN* hamartoma syndrome and VMs, yet the features of *PTEN* hamartoma syndrome may not always be clinically apparent. Therefore, certain key imaging characteristics may be diagnostic. First, *PTEN* hamartomas of soft tissue are histopathologi-
Fibroadipose Vascular Anomaly

Fibroadipose vascular anomaly is a distinct vascular disorder characterized by fibrofatty muscular infiltration, phlebectasia with pain, and contracture of the affected extremity [45, 46]. Although newly recognized, the condition itself has been documented in the past as gastrocnemius cavernous hemangioma [46–49]. It typically presents in girls as a severely painful intramuscular lesion with associated limited range of motion of the adjacent joint [45, 46]. Although it has been described in the forearm, wrist, and thigh, this anomaly typically involves the gastrocnemius muscle with limited ankle dorsiflexion [45, 46]. The skin is typically normal [46].

Histologic analysis of fibroadipose vascular anomaly reveals a combination of adipose tissue, dense fibrous tissue, large irregular muscularized venous channels, clusters of thin-walled adjacent venous channels, interspersed lymphatic channels, lymphoplasmacytic aggregates, atrophic patches of skeletal muscle, and trapped nerves [45, 46]. Sonographic evaluation shows a solid, heterogeneously echogenic, poorly defined structure with internal dilated vascular channels and thrombus without arterial waveforms [45, 46].

MRI typically shows an intramuscular lesion with muscle fibers replaced by serpiginous, ectatic T2-hyperintense channels (dilated veins or phlebectasia) with areas of high signal intensity on T1-weighted images. These hyperintense areas represent the fatty component and extend across fascial planes [45, 46, 48] (Fig. 7). Phleboliths have been reported in an isolated case [45]. Fibroadipose vascular anomaly exhibits moderate to avid gadolinium enhancement with large-dilated enhancing veins representing phlebectasia [45, 46].

A constant and differentiating criterion in fibroadipose vascular anomaly is its associated muscle contracture. Unlike VMs, fibroadipose vascular anomalies are constantly painful, and the overlying skin is normal [4, 5, 9, 45, 46].

Ultrasound findings should differentiate the solid fibroadipose vascular anomaly from the compressible, usually hypoechoic VM. MRI shows that the fibrofatty component of fibroadipose vascular anomaly comprises a dominant portion of the lesion. In VMs, however, the fibrofatty component is typically the lesser portion of the lesion. MR angiographic images show that the large dilated phlebectatic venous channels of fibroadipose vascular anomaly closely resemble those of a type IV VM [4, 9, 16, 17, 45, 46].

Spindle Cell Hemangioendothelioma

Spindle cell hemangioendothelioma (SCH) was first described as a low-grade angiosarcoma resembling cavernous hemangiomas (which in actuality were probably VMs) and Kaposi sarcoma [50]. Composed of areas of kaposiform spindle cells intermixed with dilated, partially collapsed, or thrombosed vascular channels containing phleboliths, SCH can have a variable histologic appearance ranging from that of a benign vascular malformation to that of a low-grade sarcoma [50–52].

SCH usually presents as a slow-growing solitary (59%) or multinodular (41%) mass, usually within the dermal and subcutaneous tissues of the distal lower (38%) or upper (36%) extremities [52]. Clinically, SCHs are superficially located nodules or masses usually causing mild bluish discoloration of the overlying skin [50, 52, 53] (Fig. 8). SCHs tend to be focal and appear as fixed or mobile, compressible or firm, and poorly defined or discrete nodules of variable size [50, 52, 53]. The lesions may have periods of growth and quiescence and can be painful to palpation or during growth spurts, as are VMs [51, 52]. Age at presentation and location can play a role in successfully differentiating VM from SCH. Unlike VMs, the superficially located SCH usually is not present in the first few years of life. Although approximately 40% of VMs involve the muscle only, approximately 6% of SCHs have been described in intramuscular locations [22, 54].

Congenital Fibrosarcoma

Congenital fibrosarcoma is a rare malignancy composed of irregular fascicles of spindle-shaped cells (t12;15) [60]. This lesion usually presents in newborns and infants as palpable masses, most commonly in the extremities [60]. Although the overlying skin is usually intact, there have been case reports of violaceous skin discoloration or ulceration and associated thrombocytopenia and coagulopathies, leading to confusion with hemangiomas and VMs [22, 61, 62].

Because of their clinical propensity to occasionally present with overlying skin discoloration, propensity for internal hemorrhage, and similar imaging appearances, congenital fibrosarcomas have often been labeled as mimickers of hemangiomas and VMs [61–63]. They tend to be firmer and present as large, growing masses, which would be unusual for VMs, especially early in life [4, 61, 63]. At sonographic evaluation, congenital fibrosarcomas are solid masses with heterogeneous echogenicity and internal low-resistance arterial or venous flow [60]. At MRI, congenital fibrosarcomas appear as solid masses centered in the soft tissues [60]. They can be of varying size, and most...
have well-defined margins, but they can also be infiltrative [60]. They are T1-isointense or -hyperintense and T2-hyperintense compared with muscle and are heterogeneously enhancing after IV contrast administration (Fig. 9). There have been case reports of MRI and MR angiographic visualization of congenital fibrosarcomas as soft-tissue masses with peripheral followed by central contrast enhancement similar to that of VMs [63]. Approximately 50% of congenital fibrosarcomas exhibit internal hemorrhage but rarely have internal calcifications or cystic components [60].

**Conclusion**

VMs are congenital endothelial malformations secondary to errors in vascular morphogenesis and are usually diagnosed in the first 2 decades of life. The overlapping clinical and imaging features of VMs and other pathologic entities create a diagnostic dilemma. Furthermore, the clinical presentation and imaging appearance of VMs vary, sometimes making the diagnosis challenging. A thorough knowledge of the various clinical and imaging features of not only VMs but also of the major potential mimetic lesions is crucial for clinicians caring for these patients with VMs. As radiologists, we must be aware that the patient’s history and physical examination findings play an important role in elucidating the nature of vascular lesions and their pitfalls in children, emphasizing the importance of a vascular anomaly clinic.

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**Fig. 1**—5-year-old boy with focal spongiform venous malformation in hand. 
A, Clinical photograph shows focal mass in hand with bluish skin discoloration. 
B, Sagittal left palmar gray-scale ultrasound image shows well-defined oval mass (arrows) in soft tissues of hand with hypoechoic and hyperechoic channels. Hyperechoic channels are due to very slow venous flow (stars). (Fig. 1 continues on next page)
Venous Malformation Mimickers

Fig. 1 (continued)—5-year-old boy with focal spongiform venous malformation in hand.
C, Sagittal left palmar color Doppler ultrasound image shows venous malformation containing virtually no color flow, indicating extremely slow flow. Arterial waveform is likely caused by neighboring artery. Shadowing echogenic focus represents phlebolith (arrow).
D, Axial T2-weighted MR image shows venous malformation as hyperintense well-defined mass (calipers) in hypothenar region encasing two flexor tendons (white arrow). Two phleboliths (black arrows) are visible as round hypointense foci inside lesion. Remodeling of second and third metacarpals (M) is present.
E, Axial contrast-enhanced fat-saturated T1-weighted MR image shows mild patchy enhancement (thin arrows) of venous malformation (calipers) confirming low-flow nature of lesion. Thick arrow indicates flexor tendons. M = metacarpals.

Fig. 2—Drawings show vascular malformation subtypes based on patterns of venous drainage. [Reprinted from Orthopedics Clinics of North America, Vol. 37, Legiehn GM, Heran MKS. Classification, diagnosis, and interventional radiologic management of vascular malformations, Pages 435–474, Copyright 2006, with permission from Elsevier]
A, Type I, consist of isolated malformations without venous drainage.
B, Type II, drain into normal veins.
C, Type III, drain into dysplastic veins.
D, Type IV, primarily consist of venous ectasia.
Fig. 4—6-month-old boy with intramuscular venous malformation presents with painless mass in posterior arm without skin discoloration. A, Color Doppler ultrasound image shows sharp oval mass (arrows) with slightly heterogeneous parenchyma embedded in triceps muscle (T). Mild internal vascularity is evident. H = humerus. B, Coronal T2-weighted MR image shows well-defined mass (white arrows) composed of multiple hyperintense venous channels simulating septations. Round hypointense foci represent thrombi and phleboliths (black arrows). H = humerus. C, Axial contrast-enhanced T1-weighted fat-saturated MR image shows enhancement of some venous channels inside mass (arrows), which gives heterogeneous appearance to venous malformation. B = biceps.

Fig. 3—6-year-old girl with painful lump in calf. Excisional biopsy was consistent with venous malformation with thrombosis and Masson tumor (intravascular papillary endothelial hyperplasia). A, Gray-scale ultrasound image shows solid well-defined nodule (arrows) abutting muscle fascia in subcutaneous soft tissues with minimal perilesional edema. B and C, Transverse color Doppler images of left medial calf show hypervascularity inside nodule with arterial waveform (B) and venous waveform (C).
Venous Malformation Mimickers

Fig. 5—11-year-old girl with neurofibromatosis. 
A, Coronal STIR MR image shows multiple neurofibromas as tubular hyperintensities (arrows) in muscles and subcutaneous soft tissues of forearm with thickening and increased signal intensity of overlying skin (arrowheads). 
B, Axial T2-weighted fat-saturated MR image shows target sign with central areas of hypointensity surrounded by hyperintensity within several neurofibromas (stars). Superficial diffuse neurofibromas appear as focal hyperintense plaques (arrows) involving superficial subcutaneous soft tissues and skin.
Fig. 6—9-year-old boy with extensive forearm venous malformation. Example of pseudotarget sign. 
A, Axial fat-saturated T2-weighted MR image shows several venous channels (arrows) in anterior and posterior forearm with hypointensity in center of most prominent varicosity (asterisk) corresponding to thrombus mimicking target sign seen in peripheral nerve sheath tumors. 
B, Axial contrast-enhanced fat-saturated TI-weighted MR image shows enhancement of venous channels (arrows) in anterior forearm. Absence of contrast enhancement of hypointensive center of most prominent varicosity (asterisk) confirms thrombus.

Fig. 7—13-year-old girl with fibroadipose vascular anomaly presents with constant chronic forearm pain and digital contraction. 
A, Axial T1-weighted MR image shows interspersed areas of fat and muscle atrophy (arrows) in flexor compartment. 
B, Coronal STIR MR image shows hyperintense lesion (arrows) composed of multiple vascular channels.
Venous Malformation Mimickers

Fig. 8—8-year-old boy with multiple painless soft nodules in both feet for several years, some with associated bluish skin discoloration. Excisional biopsy of nodule was consistent with spindle cell hemangioendothelioma. Diagnosis of Maffucci syndrome was made after enchondroma was removed from finger. Sagittal STIR MR image shows two well-defined lobulated hyperintense nodules (arrows) along dorsum of second metatarsal diaphysis and surrounding second phalanx. Lesion at toe is eroding phalanx (asterisk).

Fig. 9—7-month-old boy with congenital fibrosarcoma presenting as bluish discoloration on back for 3 months. Sagittal STIR MR image shows large well-defined elliptical mass (white arrow) involving subcutaneous soft tissues and muscles of back. Mass is predominantly hyperintense with small focal areas of intermediate signal intensity and prominent flow voids (black arrow). K = kidney, L = lung.

FOR YOUR INFORMATION
The reader’s attention is directed to part 1 accompanying this article, titled “Low-Flow Vascular Malformation Pitfalls: From Clinical Examination to Practical Imaging Evaluation—Part 1, Lymphatic Malformation Mimickers.”