Radiographic Findings Associated with Vascular Anomalies

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Abstract

Imaging of patients with vascular tumors and malformations has been sufficiently refined to answer pertinent questions when making treatment decisions in this challenging subgroup of pediatric patients. The imaging modalities at hand include conventional radiography, Doppler ultrasound, and magnetic resonance imaging with time-resolved, contrast-material enhanced magnetic resonance angiography. This review article will focus on the characteristic imaging features of some focal and diffuse vascular lesions, which have been classified by their clinical history and physical exam, and further labeled as a vascular tumor or slow-flow versus high-flow vascular malformation based on the updated classification system proposed by the International Society for the Study of Vascular Anomalies. The recent advances in knowledge regarding the biology of these vascular anomalies have led to increased awareness of the current nomenclature. Moreover, with better understanding of the imaging features, the radiologist has become a key player in the multidisciplinary approach offered at various institutions where appropriate treatment algorithms and interventional strategies are put together. This is crucial in avoiding misdiagnosis and improper management.

Keywords

- ▶ vascular tumors
- ▶ malformations
- ▶ magnetic resonance angiography
- ▶ pediatric patients

The diagnosis of soft-tissue vascular neoplasms and malformations is achieved with clinical history and physical examination. Imaging modalities can be used both to confirm the diagnosis as well as to delineate the lesions further, including determining their characteristics (i.e., slow flow vs. high flow) and size. Today, magnetic resonance imaging (MRI) and ultrasound (US), along with magnetic resonance angiography (MRA), are most commonly used imaging modalities for vascular anomalies.^{1,2}

Ultrasound is safe, portable, and does not involve radiation exposure. It can be performed at the bedside or in clinic and is excellent for follow-up monitoring. Furthermore, it helps guide interventional therapies. With US, one can assess the vascularity of the anomaly, the type of vessels feeding the lesion, and the presence of a high-flow component.³ It is, however, challenging to determine the exact extent of the malformation with US because its range is limited. The choice

of transducer will depend on the location of the lesion, especially as it pertains to depth of the lesion.

Magnetic resonance imaging is excellent in depicting the anatomical extent, and multicompartamental involvement. Computed tomography is rarely used today for this purpose due to radiation concerns, although it is fast and can be performed without sedation. With MRI, the intra-articular and osseous involvement can be easily visualized with improved spatial resolution of the sequences. Commonly used protocols typically include T1, T2, and some gradient recalled-echo sequences.^{4,5} Addition of time resolved contrast-enhanced MRA along with contrast-enhanced spin echo sequences shows the vascular-filling pattern of the lesion.^{4–7} The arterial feeding vessel and early draining veins can be easily seen, as well as any associated lymphatic component within the malformation. Robust three-dimensional (3D) reconstruction techniques like maximum

Fig. 1 An 11-month-old infant with rapidly involuting congenital hemangioma (RICH). Axial T2-weighted sequences (A,B) and axial gradient echo, fat-suppressed post-contrast T1-weighted sequences (C,D) show a right hepatic lobe lesion with interval decrease in size over 3 months.

intensity projection and volume rendering have become very useful in depicting the abnormality. In addition, the sensitivity and specificity of combined conventional and dynamic contrast-enhanced MRI (CE-MRI) in differentiating venous and nonvenous malformations are 83 and 95%, respectively.⁴ There are limitations in terms of high cost, need for sedation, and longer scan time. Also, a fair degree of expertise is required in the interpretation of these scans.

The role of conventional radiographs is limited, but may show calcifications and underlying osseous changes. Digital subtraction angiography (DSA) is usually performed during therapeutic intervention; its imaging features are not discussed in this article.

Spectrum of Imaging Findings

Vascular Tumors

These are broadly divided into congenital (CH) and infantile hemangioma (IH), hemangioendothelioma, and angiosarcoma. The congenital variety is fully developed at birth and further subdivided into rapidly involuting and noninvoluting (RICH and NICH, respectively).^{2,8-12} Some other vascular neoplasms like tufted angioma are clinically diagnosed and their imaging appearance is nonspecific and of little help.

Congenital Hemangioma

Congenital hemangiomas are present at birth and undergo involution by 2 years in most instances. When they fail to regress and persist beyond the first 2 years, the term NICH is preferred.^{9,13} Congenital hemangiomas affect both sexes equally and lack precursor lesions.^{9,14} On US and Doppler, CHs are heterogeneous, possess internal vascularity, and may contain calcifications.^{9,14} MRI shows T1 hypointense signal (skeletal muscle signal used as reference) and T2 hyperintense signal within a lobulated mass-like lesion (►Fig. 1). They may contain internal flow-voids on T2-weighted sequences representing central and peripheral vessel flow.¹⁵ Interspersed fat has T1 bright signal within the mass. During the involuting phase, central fibrofatty proliferation is evident. The postcontrast sequences may show diffuse enhancement and, on occasion, centripetal filling of contrast during the arterial phase of imaging. Delayed phases of the post-contrast dynamics demonstrate washout from the mass lesion. Late presentation of CH is more problematic, especially when birth history is not available, and the NICH lesion may need tissue sampling. Congenital hemangioma is GLUT-1 (glucose transporter protein-1) negative; this allows distinction from IH, which is GLUT-1 positive.^{2,9,16}

Infantile Hemangioma

A common tumor in infancy, IH is most commonly found in the head and neck region as well as the extremities.^{17,18} The diagnosis is made within the first few months, and after a period of proliferative growth in infancy, will involute by age of 7 years.¹⁸ There may be overlying skin discoloration. Infantile hemangioma is the only vascular tumor that is positive for GLUT-1 and thus helps to elucidate the diagnosis in atypical scenarios. Imaging is usually reserved for deep-seated or visceral lesions because most IHS are clinically apparent. Infantile hemangioma can be focal, multifocal, and diffuse.

Infantile hemangioma tends to be well circumscribed, echogenic, or heterogeneous on US with internal vascularity containing arterial as well as venous waveforms.^{3,19} Unlike high-flow lesions like arteriovenous malformations (AVMs), there is no arteriovenous shunting and the waveforms are low-resistance. On MRI, the findings are fairly similar to CHs, with T1 hypointense and T2 hyperintense signal within a lobulated mass (►Fig. 2). Central fibro-fatty change is sometimes seen, with flow voids (vessels) in the periphery. The enhancement is diffuse, starting in the early arterial phase, with variable washout. These features can help differentiate from other tumors, like sarcoma, which have a disorganized enhancement pattern due to tremendous arteriovenous shunting and randomly distributed vessels.^{6,7,20}

Visceral involvement can also be multifocal or diffuse. In the case of hemangioma of the liver, MRI can show multiple

contrast-enhancing masses for which the differential diagnosis can include metastatic neuroblastoma.² Urine catecholamines and concomitant adrenal mass can aid in this diagnosis. With visceral involvement, there are often signs of organ failure from excessive shunting, consumption coagulopathy, and associated hypothyroidism.

Kaposiform Hemangioendothelioma

Kaposiform hemangioendothelioma (KHE) is a rare vascular tumor that can be congenital, with 50% presenting at birth. It can be found in the trunk, extremities, retroperitoneum, and the head and neck regions.^{20–22} Kaposiform hemangioendothelioma grows rapidly and can be associated with Kasabach-Merritt phenomenon (consumptive coagulopathy).²² Ultrasound features are nonspecific with an ill-defined, variably echogenic lesion that may contain foci of calcification. On MRI, it presents as an infiltrative soft tissue mass involving multiple tissue planes. Kaposiform hemangioendothelioma has T1 hypointense and heterogeneously hyperintense T2 signal, with internal dark signal intensity channels due to vascular flow voids. Postcontrast sequences show diffuse, heterogeneous enhancement.²⁰ The soft tissue mass and calcifications may be evident on radiography.

Angiosarcoma

Angiosarcoma is a rare, aggressive, and malignant vascular tumor that carries a poor prognosis. Angiosarcoma is more common in girls and found throughout the body, including

Fig. 2 An 18-month-old child with infantile hemangioma. (A,B) Axial fat-suppressed T2-weighted and gradient echo sequence show predominantly hyperintense lesion in superficial perineum. (C) Coronal magnetic resonance angiography shows nodular peripheral enhancement on early dynamic phase. (D) Delayed spin echo postcontrast fat-suppressed T1-weighted sequence shows diffuse homogenous enhancement within the lesion.

the liver.²³ On US, it is large and heterogeneous with intense vascular flow. Prominent arterial vessels can be traced within the mass. On MRI, it is T1 hypointense and heterogeneously T2 hyperintense, and shows disorganized enhancement secondary to random vessel distribution on the arterial phase imaging. Typically, lesions show rapid enhancement with areas of contrast “pooling.”^{1,2} There is demonstrable washout secondary to arteriovenous shunting on subsequent phases. Internal foci of necrosis and hemorrhage can be seen. Local and regional spread can be determined with MRI as well.

Vascular Malformations

Vascular malformations (VMs) can be divided into high-flow or low-flow lesions according to the ISSVA classification system.^{24,25} High-flow malformations always have an arterial component with variable combinations of slow-flow elements.²⁶ Low-flow malformations involve a combination of venous, capillary, and lymphatic elements. Both these subsets will grow with the child without evidence of regression. Mixed vascular malformations may be seen more commonly in the setting of overgrowth syndromes like Klippel-Trenaunay (KTS) (►Fig. 3) or Parkes-Weber (►Fig. 4). Sudden presentations may be secondary to superimposed infection

or intra-lesional hemorrhage.¹ Malformations are differentiated from tumors by the lack of mitoses and GLUT-1 negativity.²⁷ These lesions can affect the face, trunk, extremities, internal organs, bones, and skeletal muscle. For the purpose of imaging description, the low-flow lesions are subdivided into lymphatic (LM), venous (VVM) and mixed venolymphatic (VLM) malformations, and the high-flow lesions into arteriovenous fistula (AVF) and AVM.

Low-Flow Malformation

Lymphatic Malformations

Lymphatic malformations (LMs) are the second most common type of vascular malformations after the VVMs.²⁸ Ninety percent are diagnosed by 2 years of age and almost 70 to 80% occur in the head-neck region.^{29,30} Lymphatic malformations can lead to bony and soft tissue overgrowth and may be a component, along with VVMs, in overgrowth syndromes. Lymphatic malformations are classified as macrocystic, when the individual cysts are larger than 2 cm, or microcystic, when the smaller locules in the LMs are 1 to 2 cm in size (►Fig. 5).^{19,29}

On US, LMs present as multiseptated cystic masses with no internal vascularity, mural nodule, or associated soft tissue mass. Perilesional hyperemia suggests superimposed infection. Low-level internal echoes may be present within the cysts in this setting. Pure microcystic LMs are ill-defined and diffusely echogenic in appearance due to the numerous wall interfaces.³ On MRI, the cysts are hyperintense on the fluid-sensitive T2-weighted sequences with corresponding T1 hypointense signal. If there is associated hemorrhage or infection, the cysts may show hyperintense signal on the T1 sequences related to the proteinaceous nature of the fluid. Mild septal and rim enhancement is seen on the postcontrast sequence.¹⁴ Fluid-fluid levels may be observed within the LM secondary to the layering debris from hemorrhage (►Fig. 5). Microcystic LM will appear as a region of diffuse hypointense signal on T1-weighted and hyperintense on T2-weighted sequences. They may have mild diffuse enhancement, sometimes mimicking a solid mass, especially if the cysts making up the LM are too small to discern.²⁰

The purpose of MRI is to determine the location, the exact extent (intra-articular or intraorgan extension) and the relationship of the LM to the adjacent structures (nerve or tendon involvement).¹⁴

Venous Vascular Malformations

Venous vascular malformations (VVMs) are classified into sporadic VVM, dominantly inherited cutaneomucosal VVMs and dominantly inherited glomuvenous VMs. Ninety-four percent fall under the sporadic category.^{27,31} VVMs are congenital malformations made up of dilated venous channels that are dysplastic and lack smooth muscle in the wall.³² VVMs typically enlarge in size as the child grows, with sudden increase during pregnancy and puberty due to hormonal influences. VVMs are commonly located in the head and neck (40%), trunk (20%), and extremities (40%).¹² VVMs can present as a focal mass or a diffuse abnormality. The venous

Fig. 3 A 16-year-old with Klippel-Trenaunay syndrome. (A,B) Coronal short tau inversion recovery sequences (STIR) demonstrate hyperintense serpiginous vessels within the superficial and deep soft tissues of the right lower extremity representing the low-flow venous malformation. Note also the extensive network of hyperintense dysplastic veins involving the pelvis and T2 hyperintense cystic LM within the right abdomen. (C) Coronal T1-weighted sequence depicts fatty atrophy of some of the right lower-extremity muscles.

Fig. 4 A 17-year-old with Parkes-Weber syndrome. (A,B) Coronal and sagittal STIR sequences show tortuous hyperintense vascular channels within the soft tissues around the right knee as well as the infiltration of the knee joint. (C-E) Maximum-intensity projection images generated from a time-resolved magnetic resonance angiography shows tortuous arterial branches arising from the distal superficial femoral and popliteal arteries. There is almost immediate opacification of serpiginous venous channels, consistent with multiple small arteriovenous fistulae as part of the high-flow vascular malformation.

varicosities and ectasia involving multiple tissue planes are a feature of diffuse VVMs, which may be seen in KTS and Parkes-Weber syndrome.

Ultrasound demonstrates multiple anechoic, tubular venous channels that infiltrate subcutaneous fat, muscles, fascia, and tendons (► **Fig. 6**).^{1,14,19} There is flow augmentation on performing the Valsalva maneuver. Most VVMs show low-velocity monophasic flow, but there may not be detectable flow in a small number of patients.²⁶ Shadowing echogenic foci represent calcified phleboliths, which are hallmark of VVMs. Radiographs can demonstrate phleboliths effectively as well (► **Fig. 6**).

Magnetic resonance imaging is the mainstay in determining the extent of VVMs, especially with regards to intra-articular and intraosseous extension that are challenging to visualize with other imaging modalities (► **Fig. 7**). Magnetic resonance imaging depicts the relationship of the malformation with adjacent organs, nerves, tendons, and bones. The initial sequences should cover a larger field of view to showcase the entire extent of the malformation.

In diffuse VVM, the dysplastic veins are hypointense on T1 and hyperintense on T2 sequences. Associated findings such as subcutaneous fat hypertrophy, muscle atrophy, and fatty replacement are commonly seen. Magnetic resonance angiography should be performed for all cases because the gradual filling seen in VVM is the key distinguishing feature from the high-flow lesions. There is also a lack of hypertrophied feeding arteries or early arteriovenous shunting across the lesion. The signal voids occasionally seen within the VVM are secondary to thrombosed vessel, phleboliths, or fibrous striations.³³ Phleboliths have dark signal on T1 and T2 sequences and old blood products (hemosiderin) are seen on gradient echo sequences. Fluid-fluid levels can be seen in VVMs, although less frequently when compared with LMs.¹⁴ The dynamic postcontrast enhanced MRA images should be postprocessed using maximum intensity projection and volume rendering on a dedicated 3D-enabled workstation. This allows adequate mapping of the superficial and deep venous anatomy, which is critical for treatment planning.

The focal VVMs can be superficial or deep and can even be intramuscular with involvement of any muscle group. They

Fig. 5 A 12-year-old with macrocystic lymphatic malformation (LM). (A,B) Coronal and axial STIR sequences depict a hyperintense multilocular cystic mass with septations and fluid levels, involving the right axillary soft tissues. (C) Axial precontrast T1 sequence depicts hyperintense signal within the LM secondary to proteinaceous material. (D) Axial postcontrast fat-suppressed T1 sequence depicts enhancement of the intervening septa and the periphery of the macrocystic LM with mild edema in the surrounding tissues.

present as lobulated masses, with T2 hyperintense signal and foci of dark signal intensity representing phleboliths. Central fibrosis has T1 and T2 hypointense signal and gradual fill-in of contrast is noted on the postgadolinium sequences.

Venolymphatic Malformation

Venolymphatic malformations have components of both venous and lymphatic malformation (► **Figs. 8 & 9**). They present as discrete soft tissue masses, commonly involving

Fig. 6 Phleboliths in forearm of a 5-year-old with VVM. (A) Radiograph demonstrates multiple calcified phleboliths within the soft tissues. (B) Ultrasound shows shadowing echogenic foci representing the phleboliths.

Fig. 7 Magnetic resonance imaging findings in a 5-year-old with forearm VVM. (A) Coronal STIR sequence shows predominant hyperintense signal, lobulated contours, and dark signal intensity flow voids due to phleboliths. (B) Coronal T1-weighted sequence shows iso- to mild hyperintense signal within the malformation with some fatty atrophy of the underlying muscle. (C,D) Axial pre-contrast and fat-suppressed post-contrast T1-weighted sequences show gradual moderate enhancement within the VVM.

the extremities. Magnetic resonance imaging demonstrates the fluid-fluid levels and T1 hyperintensity within the cysts representing the lymphatic component (► **Fig. 9**). The venous component is T2 hyperintense and may show phleboliths.

Hemosiderin deposits within the lesion are susceptible and “bloom” on gradient echo sequences. There is heterogeneous enhancement of the mass, with rim and septal enhancement of the LM component.

Fig. 8 A 6-year-old with mixed venolymphatic malformation in the elbow region. (A) Radiograph shows a soft tissue mass medial to the distal humerus with calcified phleboliths. (B) Gray-scale ultrasound (US) shows a heterogeneous mass within the soft tissues adjacent to the left humerus and elbow, containing tubular, anechoic channels and shadowing echogenic foci (phlebolith). (C) Normal US of same region shown as comparison.

Fig. 9 A 6-year-old with mixed venolymphatic malformation (VLM) in the elbow region. (A) Axial STIR sequence shows a discrete hyperintense lesion containing dark signal intensity foci representing phleboliths. Note the fluid-fluid levels within some areas of this lesion posteromedial to the elbow representing a mixed VLM. (B) Axial T1-weighted sequence shows hyperintense signal within the lesion secondary to proteinaceous content within the lymphatic component. (C,D) Axial and sagittal fat-suppressed post-contrast T1-weighted spin-echo sequences show patchy, heterogeneous enhancement within the venous component. Note that the lymphatic component only has rim enhancement.

High-Flow Malformation

Arteriovenous Malformation

Arteriovenous malformations constitute 10% of the peripheral vascular malformations.³⁴ They are composed of dysplastic vessels, which form the nidus, and connect hypertrophied feeding arteries and large draining veins without an intervening capillary bed.³⁵ Arteriovenous malformations most commonly involve the central nervous system, followed by extracranial head and neck, extremities, trunk, and visceral organs.³⁶ Patients can present with symptoms from shunting and heart failure, limb overgrowth or arterial steal, and ischemia of the affected limb. They can be congenital or acquired secondary to trauma. Most AVMs are solitary and sporadic, but can be multiple and even associated with syndromes.^{1,26}

On US, AVMs possess a mixed echogenic nidus with tortuous, hypertrophied arteries leading into the nidus (→Fig. 10). These arterial feeders have continuous diastolic flow, and the enlarged draining veins are arterialized, hence demonstrate pulsatile flow.^{3,19} High-flow vessels comprising the AVM appear as tortuous dark-signal intensity flow voids on the conventional T1 and T2 sequences. These serpentine high-flow vessels appear as high-signal-intensity foci on gradient sequences. T1 high signal intensity foci represent hemorrhage, intravascular thrombosis, or flow-related en-

Fig. 10 An 18-year-old with bruit along the right calf (secondary to arteriovenous malformation [AVM]). (A) Doppler ultrasound shows turbulent high velocity flow within the AVM nidus in the right calf. (B) Coronal STIR sequence shows hyperintense signal abnormality with a tortuous flow void within. (C) Time-resolved magnetic resonance angiography (MRA) depicts the feeding artery from the posterior tibial filling the AVM nidus. (D) Note the early draining vein opacifying almost immediately on the closely timed dynamic MRA.

hancement.^{14,37} Hypertrophy or atrophy of the soft tissues in the vicinity may be present. Dynamic MRA provides detailed mapping of the feeding artery and shows early filling of the enlarged draining vein (→Fig. 11). The contrast rise time is exceptionally short (< 20 seconds), as opposed to VVMs where the contrast rise time is typically 80 to 100 seconds.³⁸ Multiplanar reconstructions and 3D reconstruction techniques display the relevant information for the treating physician. The lack of associated soft tissue mass allows differentiation from soft tissue sarcomas.³³ Diffuse syndromes associated with high-flow malformations include hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu) and epidermal nevus syndrome.

Arteriovenous Fistulas

Arteriovenous fistulas do not contain a nidus, are often acquired, and are more commonly found in the brain.²⁶ In other words, there is more of a direct connection between the arterial and venous components. Ultrasound shows a cluster of serpentine vessels without intervening solid tissue.

Fig. 11 An 18-year-old with AVM in the right calf. (A-C) Digital subtraction angiography (DSA) confirms the filling of AVM nidus from the arterial feeders arising from the posterior tibial artery and opacification of the early draining vein (posterior tibial). (D) Posttreatment DSA image in the coronal plane showing complete obliteration of the AVM nidus.

Magnetic resonance imaging findings mimic those seen in AVMs, where the routine spin-echo T1 and T2 sequences as well as the gradient echo sequences depict multiple flow voids. Time-resolved MRA reveals the tortuous feeding artery and almost immediate filling of the draining veins.

Summary

The recent understanding of histology and the biologic behavior of vascular anomalies have led to increasing awareness and universal acceptance of the proposed nomenclature. Moreover, the multidisciplinary team approach to management of vascular anomalies has led to better patient care and outcomes. To this end, radiology is an essential component of the team approach. Of utmost importance is to be able to differentiate neoplasm from malformation. Among vascular malformations, the distinction between slow-flow and fast-flow lesions is critical. Today, MRI and US, along with MRA, are the most commonly used imaging modalities for vascular anomalies. The vastly improved MRA techniques allow for shorter dynamic scan times with improved spatial resolution, which in turn, help better delineate the type, location, and extent of the malformation.

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