

Diagnostic Imaging of Vascular Anomalies

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KEYWORDS

- Vascular anomaly • Vascular malformation • Imaging
- Radiology

Medical imaging has become critically important in the diagnosis and treatment planning of vascular anomalies. The classification of lesions into fast-flow and slow-flow categories, the identification of a soft tissue mass, and the determination of the extent of the lesions are all facilitated by the use of magnetic resonance imaging (MRI), ultrasonography, catheter angiography, and other imaging studies.^{1–3} Ultrasonography is typically the first-line imaging study for the evaluation of vascular anomalies in children because sedation is not required. MRI may be indicated for diagnostic confirmation or to better define the anatomy of the lesion. Computed tomography (CT) gives superior resolution for osseous lesions.

VASCULAR TUMORS

Infantile Hemangioma

Infantile hemangiomas are benign tumors composed of endothelial cells. These lesions follow a predictable clinical course of proliferation in infancy followed by involution, usually within the first 5 to 7 years of life. Most cases do not require imaging. If clinical features are atypical or the anatomic extent of the lesion must be determined, ultrasonography and MRI can be of use.

Typical ultrasonographic appearance of an infantile hemangioma, both in the proliferating stage as well as the involuting stage, is a well-circumscribed hypervascular mass showing low-resistance arterial waveforms (**Fig. 1**). Most hemangiomas are hypoechoic, although up to

18% have been reported to be hyperechoic.⁴ Hemangioma can be differentiated from arteriovenous malformations (AVMs) by the presence of solid parenchymal tissue.

Proliferating infantile hemangiomas are lobulated hypervascular masses. On MRI studies, the lesions are isointense to muscle on T1-weighted sequences and hyperintense on T2-weighted sequences. High-flow central and peripheral vessels, seen as flow voids, are evident on T2-weighted sequences.⁵ After contrast administration, these masses enhance intensely and diffusely (**Fig. 2**). In contrast to AVMs, arteriovenous shunting is not typically seen in infantile hemangioma.⁶

During involution, infantile hemangiomas become more heterogeneous in appearance. MRI of involuting infantile hemangiomas demonstrates regions of fibrofatty deposition, manifested by areas of increased signal on T1-weighted sequences. Contrast enhancement diminishes and becomes inhomogeneous.⁶

Rapidly Involuting Congenital Hemangioma and Noninvoluting Congenital Hemangioma

Congenital hemangiomas are tumors that have reached their maximal size at birth. Two variant forms of congenital hemangioma have been described: rapidly involuting congenital hemangioma (RICH) and noninvoluting congenital hemangioma (NICH). These lesions are distinguishable from infantile hemangioma by their clinical course, as described by their names.

Disclosures: None.

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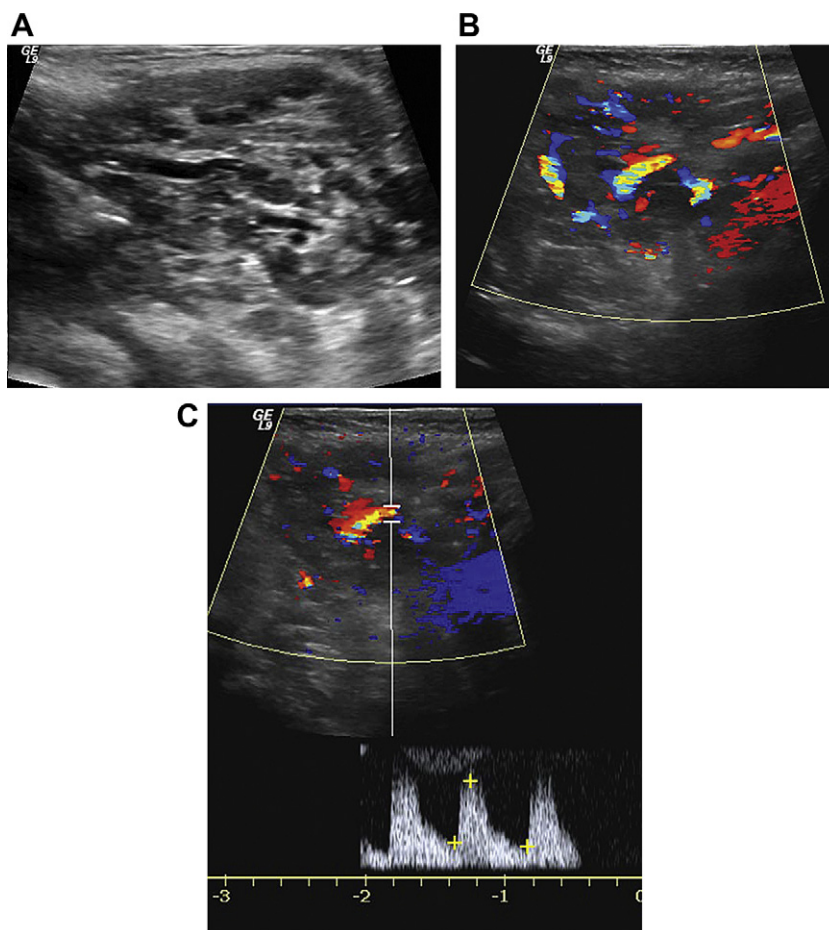


Fig. 1. Ultrasonography in a 7-month-old girl with a parotid infantile hemangioma. (A) Two-dimensional image demonstrates a well-defined mass with multiple large internal vessels. (B) Color Doppler image shows the marked vascularity of the mass. (C) Spectral Doppler trace obtained from an intralesional vessel confirms fast flow.

Unfortunately, these lesions cannot be reliably differentiated from common infantile hemangiomas based on imaging alone. However, some imaging features may be suggestive of a specific lesion. On ultrasonography, the useful differentiating factors are the presence of more visible vessels in congenital hemangiomas in comparison to infantile hemangioma, as well as the presence of intravascular thrombi, calcifications, vascular aneurysms, and arteriovenous shunting (Fig. 3).^{7,8} RICH and NICH are less likely to be well defined than infantile hemangioma on MRI (Fig. 4).⁷

Kaposiform Hemangioendothelioma

Kaposiform hemangioendothelioma (KHE) is a rare vascular neoplasm with locally aggressive characteristics but without metastatic potential. MRI typically shows an ill-defined soft tissue mass that is hypo- or isointense on T1-weighted imaging and

hyperintense on T2-weighted imaging. On administration of contrast, there is intense but heterogeneous enhancement (Fig. 5). Subcutaneous fat stranding is an important feature that helps differentiate KHE from other benign fast-flow vascular masses. Prominent vascular channels, evidenced by flow voids, are usually present on MRI studies.^{9,10}

VASCULAR MALFORMATIONS ***Lymphatic Malformation***

Lymphatic malformations (LMs) are congenital malformations resulting from abnormal development of the lymphatic channels. The lesions may be classified as macrocystic, microcystic, or combined. On ultrasonography, macrocystic LM appears as a unilocular or multilocular cystic lesion, usually with thin septations. Doppler imaging often demonstrates vascular channels within the septations.⁸ MRI of macrocystic LM

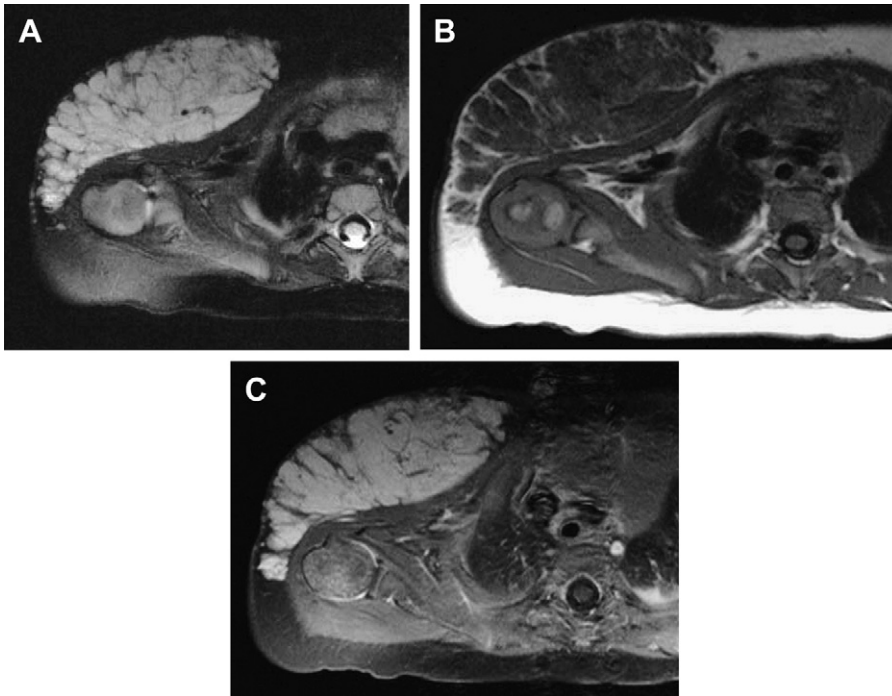


Fig. 2. MRI in a 6-month-old girl with infantile hemangioma of the right anterior chest wall and supraclavicular region. (A) T1 axial image shows well-defined low-signal mass with intralesional flow voids. (B) T2 axial image. The lesion is uniformly hyperintense on fluid sensitive sequences. (C) T1 axial postcontrast image. There is avid homogeneous enhancement of the entire lesion.

shows clearly defined cysts that are hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging. Fluid-fluid levels within the cysts may be present. The septa may enhance, creating a “rings and arcs” appearance (**Fig. 6**). No flow voids or phleboliths are expected within the cysts.¹¹

Depending on the size of the cysts, microcystic lesions may appear as ill-defined hyperechoic masses on ultrasonography.⁸ Likewise, on MRI, microcystic LMs can appear as solid lesions that are generally hypointense on T1 sequences and hyperintense on T2 sequences. There is minimal enhancement on administration of contrast (**Fig. 7**). Differentiation from soft tissue masses can be difficult.¹¹ Categorization of LMs as slow-flow lesions and analysis of the extent of the lesions are two important tasks when imaging LMs.¹²

Venous Malformation

Venous malformations (VMs) are congenital slow-flow malformations that are present at birth and typically grow proportionally with the child. VM may take 2 forms: the most common form

(cavitary) is a spongy mass of abnormal venous channels containing stagnant blood and the ectatic or dysplastic form presents as multiple irregular varicose veins.^{6,8} A combination of the 2 forms can be seen. When VM occurs in an extremity, undergrowth or overgrowth of the affected limb is possible.

Ultrasonography of VM typically shows a well-defined spongelike collection of vessels. Blood can be seen flowing into the cavities, especially after applying and releasing manual compression. Other features include phlebectasia, thickening of subcutaneous tissues, and the presence of phleboliths.^{4,8}

MRI studies of VM show multilocular, lobulated, septated masses that are hypo- or isointense to muscle on T1 sequences and hyperintense on T2 sequences. These lesions often infiltrate into adjacent organs, nerves, tendons, muscles, and joints.⁶ Slow-flow lesions can be confirmed with gradient-recalled echo imaging, and delayed postcontrast imaging may be used to show central enhancement (**Fig. 8**).¹³ Other important characteristics include the presence of phleboliths, best appreciated as focal low-signal areas on gradient-recalled echo

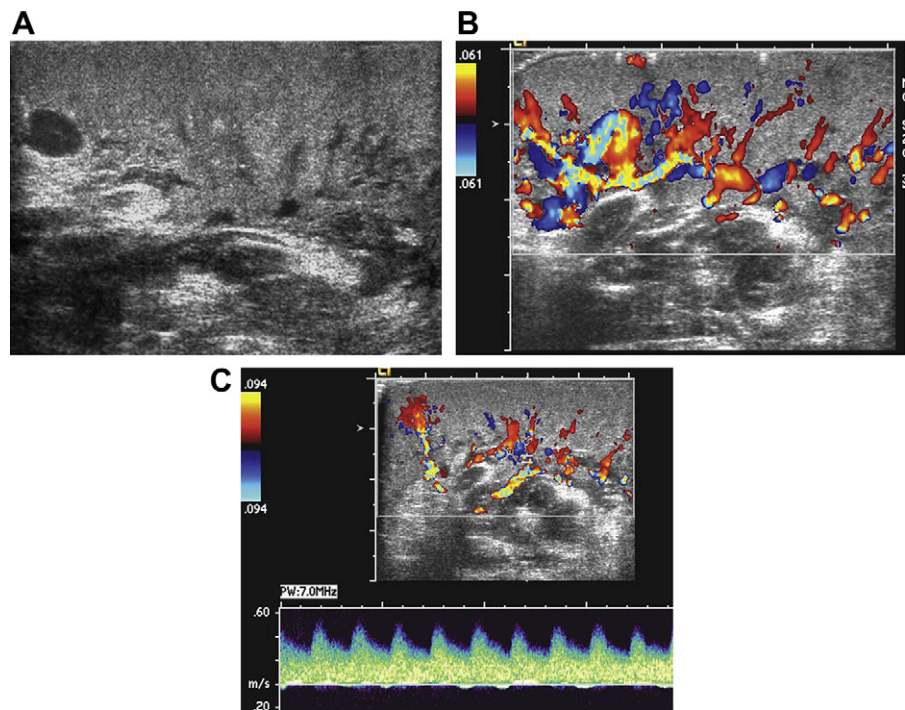


Fig. 3. Ultrasonography of a RICH of the right thigh in a 5-day-old boy. (A) Two-dimensional image shows echogenic subcutaneous lesion with prominent vessels and poorly defined borders. (B) Color Doppler image. The entire mass is markedly vascular with tortuosity of some intralesional vessels. (C) Spectral Doppler trace confirming pulsatile fast flow.

sequences, and the presence of an anomalous venous drainage system.¹¹ VMs are also more likely than LMs to primarily involve muscle.

Arteriovenous Malformation

AVMs are rare fast-flow lesions that result from dysplastic arterial and venous development, with the absence of a normal intervening capillary

bed. Although growth of the lesion is generally proportional to that of the child, AVMs may rapidly enlarge during puberty or after trauma.

On ultrasonography, AVM is a poorly defined hypervascular lesion without a soft tissue mass. Gray-scale imaging is often normal. Multiple tortuous feeding arteries showing increased diastolic flow are best seen with Doppler, power, and spectral imaging. The draining veins are

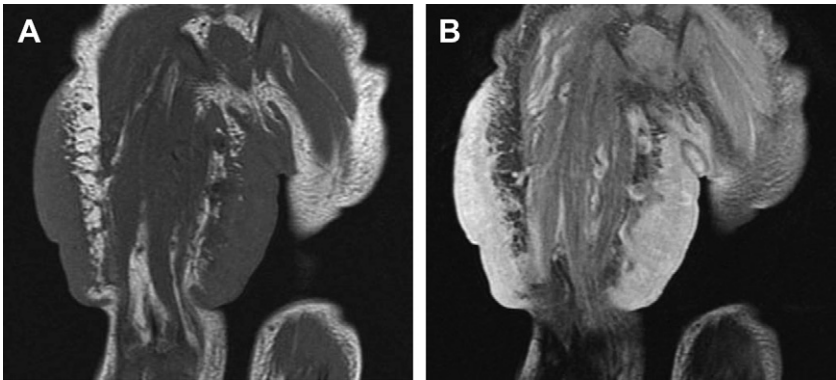


Fig. 4. MRI of a RICH. (A) Coronal T1 image. Low-signal subcutaneous lesion is seen with poorly defined borders. (B) Coronal T1 postcontrast image. Intense uniform enhancement is seen with stranding of the surrounding fat.

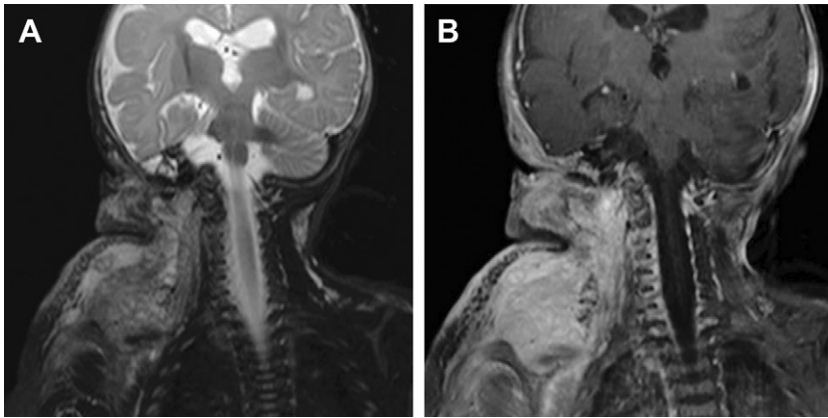


Fig. 5. MRI of a KHE involving the neck, right shoulder, and anterior chest wall in a 4-month-old girl. (A) Coronal T2 image. Heterogeneous hyperintensity is seen with involvement of multiple tissue planes. (B) Coronal T1 post-contrast image. Avid but heterogeneous enhancement is seen, with evidence of peripheral fat stranding.

enlarged and demonstrate pulsatile high-velocity blood flow.^{4,8}

MRI can show the feeding and draining vessels connected by enlarged central channels. Signal voids can be identified on most standard

sequences (**Fig. 9A**). Although no soft tissue mass is seen, edema and abnormal enhancement may be present in the surrounding tissues. AVM may result in overlying skin thickening and underlying bony lytic changes.^{6,11} Magnetic resonance

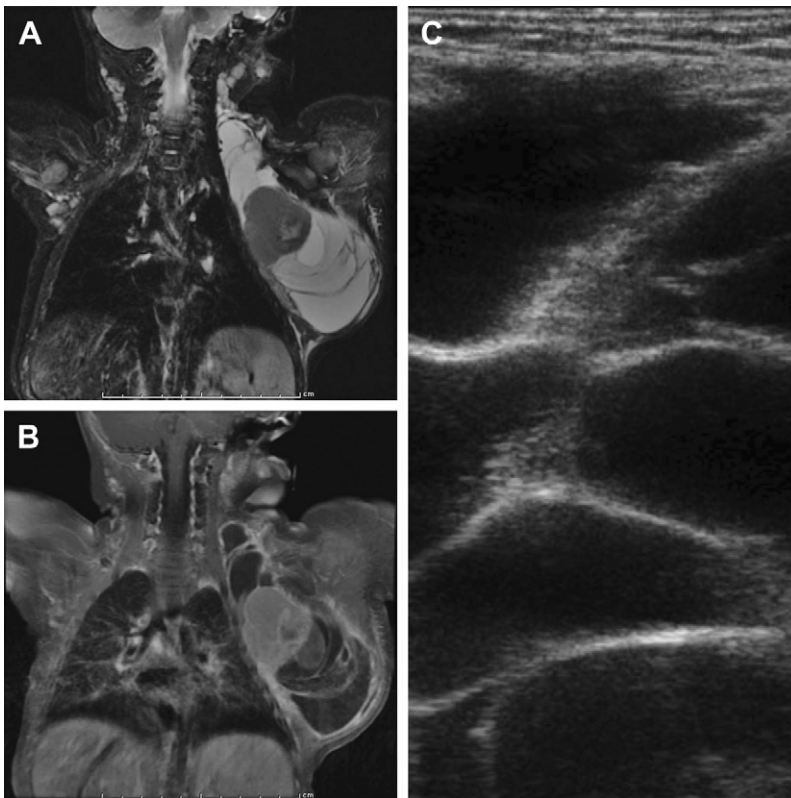


Fig. 6. A 4-month-old girl with a macrocystic LM of the left axilla, lateral chest wall, and neck. (A) Coronal T2 MRI. An almost entirely macrocystic lesion is seen. The focal area of low signal corresponds to prior hemorrhage within a macrocyst. (B) Coronal T1 postcontrast MRI. Peripheral and septal enhancement is noted. (C) Ultrasonography demonstrates the anechoic macrocysts with echogenic intervening septa.

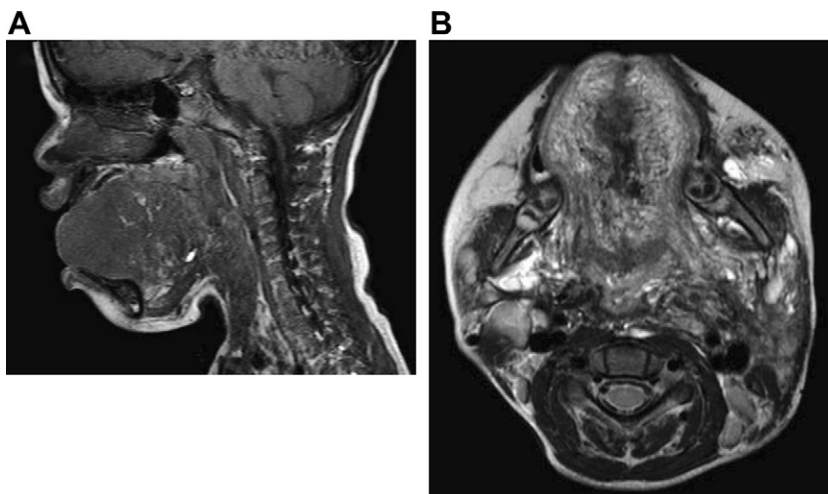


Fig. 7. MRI in a 3-year-old boy with microcystic LM of the tongue and floor of the mouth. (A) T1 sagittal image. A grossly enlarged tongue is seen occupying the oral cavity and oropharynx. (B) T2 axial image. Heterogeneous high signal is seen throughout the tongue, consistent with microcystic disease.

angiography demonstrates the feeding arterial branches and shows early enhancement of the draining veins.⁸

Catheter angiography of AVM typically shows one or more feeding arteries supplying a nidus formed by entangled vascular loops interconnected

by small venules. Several large draining veins are usually seen (see **Fig. 9B**). Although catheter angiography is superior to magnetic resonance angiography in demonstrating the vascular details of AVM, given the invasive nature, it should be reserved as an adjunct to endovascular embolotherapy.¹⁴

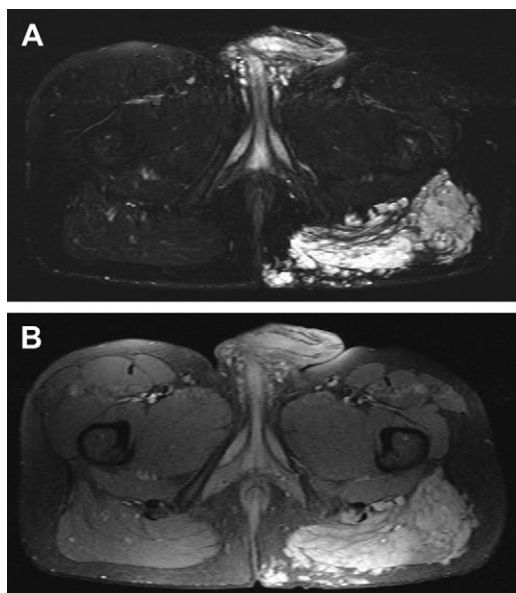


Fig. 8. MRI in a 24-year-old male with a VM of the left buttock, presenting with enlargement and pain on exertion. (A) Axial T2 image with fat saturation. Hyperintense lesion with clear lobulated margins is seen in the left gluteus maximus muscle, with subcutaneous extension noted medially. (B) Fat-saturated axial T1 postcontrast image showing uniform enhancement of the entire lesion.

COMBINED MALFORMATIONS AND VASCULAR ANOMALY SYNDROMES

Combined Malformations

Combinations of the various types of vascular anomalies are frequently seen. In particular, imaging characteristics of lymphaticovenous malformations have been described.^{15,16} These entities display characteristics of both of their individually occurring constituents. In such cases, cross-sectional imaging is useful in excluding fast-flow components and soft tissue masses.

Klippel-Trénaunay Syndrome

Klippel-Trénaunay syndrome (KTS) is characterized by a capillary-lymphaticovenous malformation affecting an extremity with associated overgrowth. Involvement of a lower extremity occurs 95% of the time.¹⁷ Imaging is used for initial diagnosis if the clinical presentation is atypical and it can also be used in the diagnosis and treatment of complications of the disease.

Ultrasonography, CT, and MRI demonstrate soft tissue hypertrophy in KTS and can define the numerous vascular malformations extending throughout the limb (**Fig. 10**). Involvement of vascular lesions in the pelvis and abdomen is

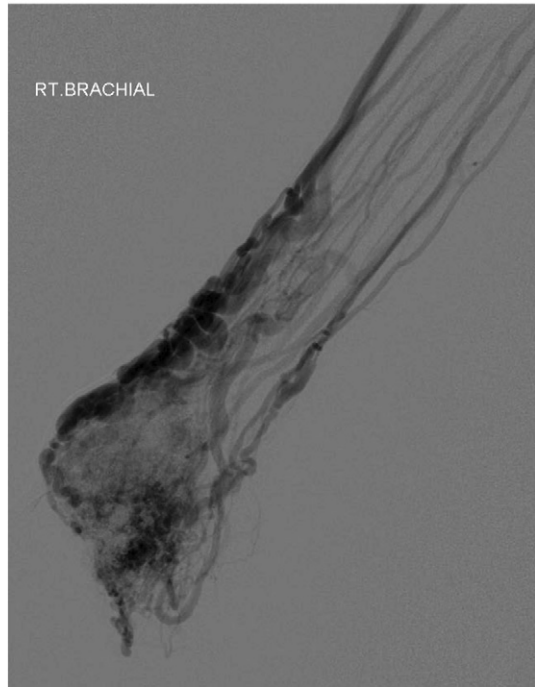
A**B**

Fig. 9. AVM of the right hand in a 16-year-old boy. (A) Coronal T1 MRI showing multiple flow voids without an associated mass. The malformation predominantly involves the thenar muscles, and the second and third rays have been amputated. (B) Angiogram with contrast injected into the right brachial artery. The malformation is largely supplied by the enlarged radial artery, with smaller feeding branches from the ulnar artery. There is marked dilatation and tortuosity of the draining veins, with evidence of early filling consistent with arteriovenous shunting.

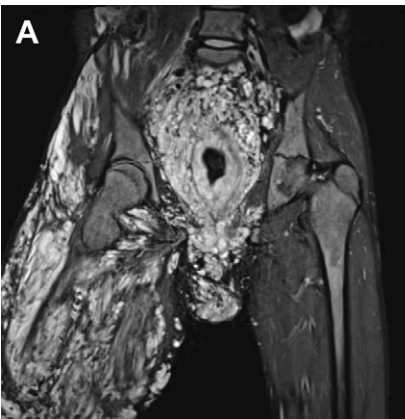


Fig. 10. MRI in a 16-year-old adolescent with KTS, who presented with rectal bleeding. (A) Coronal inversion recovery sequence. There is a diffuse high signal within the right thigh, perirectal area, and scrotum. (B) Coronal fat-saturated T1 postcontrast image. In the right thigh, there is heterogeneous enhancement of the subcutaneous and intramuscular venous components of the malformation, with minimal enhancement of the perirectal and scrotal LMs.

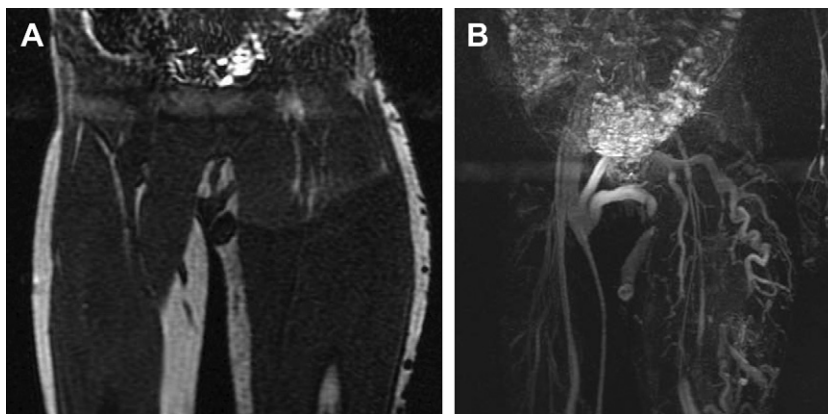


Fig. 11. MRI in a 7-year-old girl with PWS. (A) Coronal T1 image showing overgrowth of the affected left lower extremity with multiple subcutaneous flow voids. (B) Maximum intensity projection MRA/MRV. Multiple tortuous draining veins are seen on the left. These veins then drain into 2 grossly dilated anomalous veins that cross the midline to empty into the right femoral vein. MRA, magnetic resonance angiography; MRV, magnetic resonance venography.

common. A dilated superficial vein (lateral vein of Servelle) coursing along the subcutaneous fat of the lateral calf and thigh is often identified. Imaging of the vascular abnormalities can direct targeted treatments, such as sclerotherapy and resection. Complications of KTS that are often diagnosed with the aid of medical imaging include thrombophlebitis, deep venous thrombosis, pulmonary embolism, and congestive heart failure.^{8,17}

Parkes Weber Syndrome

A combination of arteriovenous fistulae and a cutaneous capillary-lymphatic malformation associated with limb hypertrophy has been termed Parkes Weber syndrome (PWS). This syndrome is often confused with KTS, but the distinction is important in predicting prognosis as well as in suggesting appropriate therapy. The arteriovenous fistulae of PWS are fast-flow lesions that can result in heart failure. These lesions can be identified with ultrasonography, CT, MRI, and catheter angiography (**Fig. 11**). Embolization to reduce shunting may be considered.¹⁸

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