Magnetic Resonance (MR) Imaging of Vascular Malformations

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Summary

Vascular malformations pose a diagnostic and therapeutic challenge due to the broad differential diagnosis as well as common utilization of inadequate or inaccurate classification systems among healthcare providers. Therapeutic approaches to these lesions vary based on the type, size, and extent of the vascular anomaly, necessitating accurate diagnosis and classification. Magnetic resonance (MR) imaging (MRI) is an effective modality for classifying vascular anomalies due to its ability to delineate the extent and anatomic relationship of the malformation to adjacent structures. In addition to anatomical mapping, the complete evaluation of vascular anomalies includes hemodynamic characterization. Dynamic time-resolved contrast-enhanced MR angiography provides information regarding hemodynamics of vascular anomalies, differentiating high- and low-flow vascular malformations. Radiologists must identify the MRI features of vascular malformations for better diagnosis and classification.

Background

Vascular malformations (VMs) are developmental anomalies, which often become symptomatic in late adolescence or adulthood and thus usually remain undiagnosed during childhood [1]. Historically, numerous classification schemes have existed, causing considerable confusion among clinicians regarding appropriately classifying and treating them. Besides, nomenclature inconsistencies such as the use of the terms 'hemangioma' and 'vascular malformation' synonymously by many exacerbate this confusion [2]. The landmark work done by Mulliken and Glowacki, published in 1982, helped ameliorate much of the disparity, broadly classifying vascular lesions as vascular tumors (hemangiomas) and vascular malformations [3]. While the original work differentiates lesions based on histological components, i.e., arterial, venous, capillary, and lymphatic, a useful imaging strategy is to classify VMs as high-flow or low-flow lesions based upon the degree of shunting and speed of flow through the lesion [4].

This is an important distinction, as management strategies may differ for high- and low-flow lesions, with the former often requiring some means of flow control using sclerosant injection or embolization prior to ablation.

Contrast-enhanced computed tomography (CT) or CT angiography is useful in the assessment of vascular malformations. Multi-detector CT helps to evaluate for vascular anatomy, enhancement, calcification, thrombus, phleboliths, and involvement of adjacent structures. CT provides high temporal resolution and is relatively easy to interpret.

Table 1. Vascular anomalies classification adopted by ISSVA. Rome 1996 [ref].

Vascular tumours (vasoproliferative)	Vascular malformations

• Infantile hemangioma
• Congenital hemangiomas
  • RICH
  • NICH
• Kaposiform
  • Hemangioendothelioma
  • Tufted angiomas
• Spindle cell
  • Hemangioendothelioma
• Epithelioid
  • Hemangioendothelioma
• Other (rare)
  • Hemangioendothelioma (composite, retiform, other)
• Angiosarcoma
• Dermatologic acquired vascular tumours
	• Low-flow vascular malformations
	  • Capillary malformation
	  • Venous malformation
	  • Lymphatic malformation
	• High-flow vascular malformations
	  • Arterial malformation
	  • Arteriovenous malformation
	  • Arteriovenous fistula
	• Combined vascular malformation (combination of above lesion types)

RICH – rapidly involuting congenital hemangioma; NICH – noninvoluting congenital hemangioma.

Hepatic hemangiomas are typically diagnosed on contrast-enhanced CT as hypoattenuating lesions on non-contrast sequence, with peripheral globular enhancement on early arterial phase. There is a progressive centripetal enhancement on venous phase, causing uniform filling. The uniform enhancement persists on delayed images. However, CT has an overall limited utility, as it provides less information on the flow dynamics, and is associated with exposure to ionizing radiation. Hence, ultrasound (US) and MR are the primary noninvasive imaging modalities for the evaluation of vascular malformations [5,6].

Although ultrasound is used to differentiate high-flow from low-flow lesions, MR imaging has proven advantageous due to its reproducibility, ability to reliably delineate the extent of these lesions, and its ability in differentiating high-flow VMs from low-flow VMs using dynamic post-contrast sequences. Due to these benefits, coupled with the lack of ionizing radiation and multi-planar capabilities, MRI has become the first-line imaging modality in the assessment of vascular malformations in many institutions [7–10]. In this review, we present typical MR imaging appearances of VMs, including examples from different body regions, to illustrate their ubiquitous occurrence and aid the radiologist in correctly classifying and diagnosing these lesions.

Classification and Management

Multiple classification systems have been proposed for the characterization and categorization of vascular anomalies. The first major classification system was presented in 1982 by Mulliken and Glowacki, who classified vascular anomalies into two major categories based on their biological and pathological characteristics including cellular growth rate and turnover, histologic features, physical examination findings, and natural history of the lesion [3]. The classification separated the vascular anomalies into hemangiomas (vascular tumors) and vascular malformations, and was adopted by the International Society for the Study of the Vascular Anomalies (ISSVA) at the 1996 ISSVA workshop

(Table 1) [11]. The inclusion of radiologic findings within the classification system was proposed by Jackson et al. in 1993, and then vascular malformations were further divided into the high-flow and low-flow lesions based on their flow characteristics [12,13]. Low-flow malformations include venous, lymphatic, capillary, and any combination of the mentioned vessels. High-flow malformations include arteriovenous (AV) malformations and AV fistulas (AVF).

This flow characteristic-based categorization has implications for management: a suitably located slow-flow lesion may be amenable to sclerotherapy in one or multiple sessions, depending on the size of the malformation [4]. Treatment of high-flow vascular malformations usually requires flow control in the form of tourniquet application or embolization prior to sclerotherapy. It is important to note that among vascular malformations, high-flow and diffuse vascular malformations are difficult to treat and result in more frequent complications as well as early recurrence. Table 2 demonstrates treatment strategy based on histopathology and flow dynamics, as recommended by Jackson et al. [4]. Today, the most commonly used classification systems are the ISSVA classification and the Hamburg classification system, which utilizes embryologic characterization of vascular abnormalities in addition to anatomic and pathological features [14].

Epidemiology and Clinical Presentation

As part of the initial work-up and characterization of vascular malformations, the natural history, clinical presentation, and physical examination findings serve as a foundation for further diagnostic procedures including diagnostic imaging.

High-flow lesions

Symptoms of high-flow lesions correspond to the degree of AV shunting and the involved area of the body. The prevalence of AVMs in the general population has not been characterized, however, estimates range from 5 to 613 per

Table 2. Overview of classification and management of vascular malformations.

Type	Histopathology	Treatment
Slow flow vascular malformations	Capillary malformation (port-wine stain, telangiectasia, angiokeratoma)	Sclerosant injection (most commonly alchohol and Onyx) (1)
	Venous malformation Commonly sporadic, Bean syndrome, glomuvenous malformation, Mafucci syndrome	Sclerosant injection with embolization in a multi-disciplinary setting (2)
	Lymphatic malformation	Sclerosant injection
High flow vascular malformation	Arterial malformation Arteriovenous fistula (AVF) Arteriovenous malformation (AVM)	Embolisation/direct puncture sclerotherapy
Complex-combined vascular malformations	Capillary-venous, Capillary-lymphatic, Lymphatic-venous, Capillary-lymphatic-venous, AVM-lymphatic, Capillary malformation-AVM	Variable and depends upon relative proportion of low-flow and high-flow components.

100,000 people [15]. When fully developed, AVMs deepen in color and present with progressive erythema, local warmth, a palpable thrill, and a bruit. Patients with facial AVMs of the skin and/or facial bones may present with facial asymmetry, gingival hypertrophy, unstable teeth, periodontal bleeding, or skin/mucosal ulcers with secondary infection. Bony AVMs create osteolysis. Lower limb skin changes resembling brown-violaceous plaques may appear and are known histologically as pseudo-Kaposi sarcomas. Distal extremity AVMs may lead to ischemia of the tips of fingers or toes, which is associated with an arterial steal and venous hypertension.

Sequelae of expanding AVMs with AV shunting may include ischemic changes, indolent ulceration, intractable pain, acute life-threatening hemorrhage, or recurrent intermittent bleeding. Increased cardiac output with subsequent congestive heart failure occurs in less than 2% of cases [16]. Pain, overgrowth, and bleeding are the most common symptoms, while high-output cardiac failure is a relatively uncommon clinical presentation, seen mostly in patients with large AVMs. Following a localized trauma, AVMs may show rapid growth over a relatively short period. Schobinger developed a classification (Table 3) based on clinical signs and symptoms for determining the severity of AVMs and timing of intervention [17].

Low-flow lesions

Venous malformations are present in 1–4% of the population [18] and are non-tumorous developmental aberrations. Although present at birth, they are usually not detected until adolescence, when acute changes in lesion size or symptoms precipitated by growth, hormonal influence, infection or thrombosis may bring them to clinical attention. These lesions tend to occur more commonly in the head and neck (40%) and extremities (40%), and less commonly involve the trunk (20%) [19]. Superficially located venous malformations may cause bluish discoloration and appear as compressible non-pulsatile masses, whereas

Table 3. Schobinger classification of AVMs.

Stage	Clinical symptoms
I	(Quiescence) Skin warmth, discoloration
II	(Expansion) Enlargement, pulsation, bruit
III	(Destruction) Pain, ulceration, bleeding
IV	(Decompensation) Cardiac failure due to volume overload

lesions containing arterial components tend to appear as pulsatile masses with palpable thrill [20]. Deeper lesions may be extensive, and in the extremities, can be interspersed between muscular planes with atrophy of the surrounding muscles due to shunting. A high recurrence rate is noted due to the fact that these lesions contain immature cellular elements, which have a propensity to regrow after attempted ablation [21].

Lymphatic malformations are subdivided into macrocystic (also known as cystic hygroma), microcystic (also known as cavernous lymphangioma), and mixed lesions based on the size of the cystic component [22,23]. Microcystic lesions contain endothelium-lined small cavities and cysts measuring less than 2 cm^3 with a background of solid matrix of fibrous tissue and smooth muscles. Microcystic lesions are the most common type and usually involve the neck, shoulders, proximal limbs, and perineum. They present later in the childhood. Macrocystic lesions contain large, >2 cm^3, cystic spaces filled with proteinaceous fluid and some lymphocytes. These lesions often contain thick intralesional septae. Macrocystic lesions commonly involve the head and neck (70%) and are usually diagnosed within the first 2 years of life (Figure 1) [23,24]. They can also involve the trunk and limbs. However, the involvement of the mediastinum or abdomen is rare. Macrocystic lesions are typically subcutaneous with normal overlying skin and appear
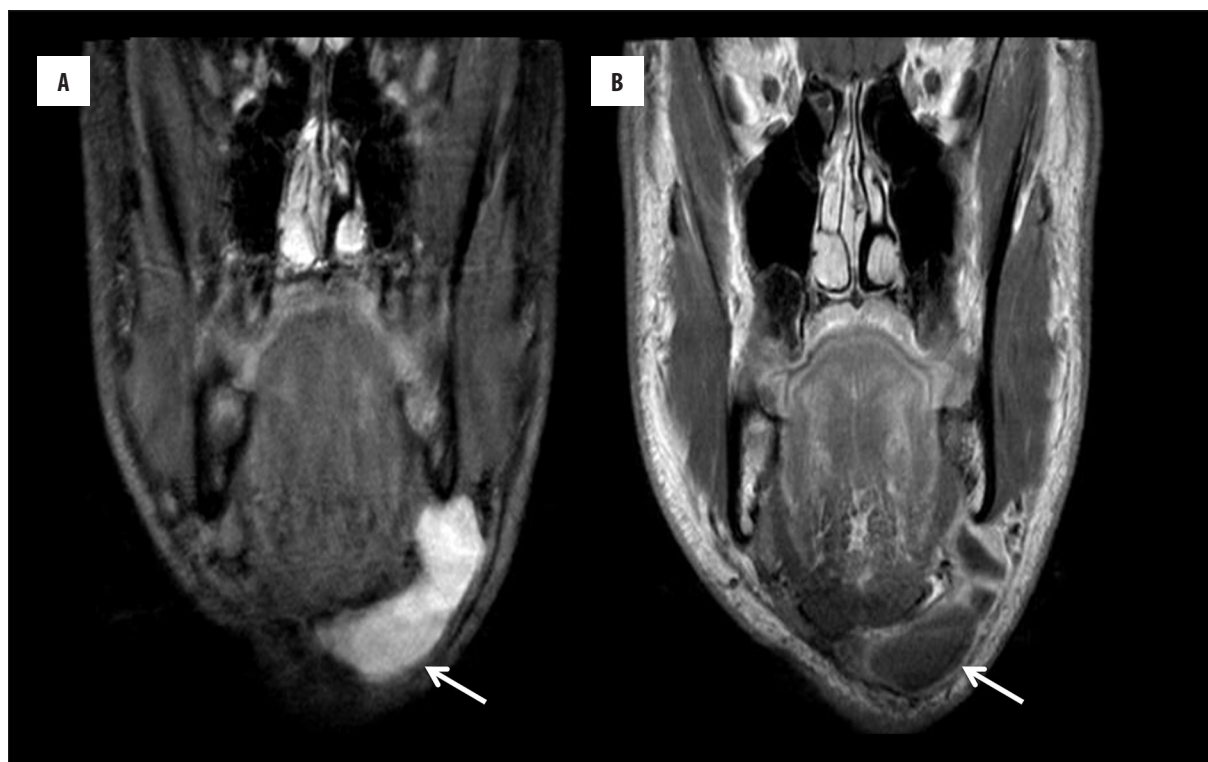
Figure 1. Coronal T2 with fat suppression (**A**) and coronal T1 post contrast (**B**) demonstrate left submandibular, fluid signal intensity lesion with thin peripheral enhancement after contrast administration compatible with a lymphatic malformation (arrows).

Table 4. MR features of vascular malformation.

Modality	Arteriovenous malformation	Venous malformation	Arteriovenous fistula	Pure lymphatic malformation
MR features	Dark T1 and T2, flow voids, enhances early with contrast, localized or infiltrative, enlarged feeding arteries and draining veins	Intermediate T1, bright T2, absent flow voids, phleboliths/areas of thrombosis, localized or infiltrative, enhances gradually with contrast, normal feeding arteries and draining veins	Dark T1 and T2, flow void, enlarged feeding artery and draining vein. Early venous filling on dynamic contrast studies.	Dark T1, bright T2, localized or infiltrative, minimal to no enhancement, no feeding arteries and draining veins

translucent on clinical examination, while microcystic lesions involve the skin and mucosal surfaces. Both macrocystic and microcystic lesions often present with acute exacerbation of their painful symptoms and rapid expansion of the lesions secondary to superimposed infection or intralesional hemorrhage. Since venous and lymphatic malformations are embryologically related, there is a considerable overlap in imaging characteristics [17].

MR imaging techniques

The standard sequences for characterization and diagnosis of suspected vascular malformations include spin echo (SE) or fast SE T1-weighted imaging (T1WI) for regional anatomy, and SE T2-weighted imaging (T2WI) with and without fat saturation to delineate the extent of the abnormality (Figure 1A). Gradient recalled echo (GRE) T2* WI is invaluable in differentiating areas of high-flow or hemosiderin deposition from bleeding.

The dynamic contrast-enhanced techniques like time-resolved MRI sequence of the lesion using three-dimensional (3D) fast gradient recalled echo (GRE) technique, which involves image acquisition at intervals as short as 2 seconds, has high specificity for differentiating venous from non-venous malformations [25,26]. This yields a volumetric dataset with high spatial and temporal resolution, which can also facilitate digital subtraction (using pre-contrast mask image), volume rendering, multi-planar reformats (MPR), and maximum intensity projection (MIP). Time-resolved angiography techniques allow assessment of flow direction, feeder arteries and drainage veins, and differentiation of high-flow from low-flow lesions [20,26].

Key Imaging Features

An important role of imaging is to classify vascular malformations into low-flow or high-flow lesions based on their hemodynamics. The distinction of low-flow from high-flow lesions has therapeutic and prognostic implications and is

more relevant to management than the prediction of individual vascular components [26,27]. Table 4 gives an overview of pertinent conventional and dynamic imaging findings of low-flow and high-flow lesions.

Low-Flow Vascular Malformations

Pure capillary malformations are present at birth in 0.3% of children [28] and are mostly diagnosed clinically. When imaged, the only feature may be of mild skin thickening. The MRI appearance of slow-flow lesions depends on their components. Pure venous malformations appear as lobulated and multi-septated masses with internal serpentine architecture. They exhibit intermediate to low signal intensity on T1- and high signal on T2-WI. The presence of internal hemorrhage or proteinaceous content may lend higher signal on T1WI or fluid-fluid levels. When present, phleboliths are diagnostic of venous elements [29]. Following IV contrast administration, there is gradual and complete enhancement, though this tends to be earlier in the case of mixed capillary venous malformations compared to pure VMs. Pure lymphatic malformations, on the other hand, do not show internal enhancement following contrast administration, being composed entirely of cystic spaces non-communicating with the venous system, although peripheral and septal enhancement may be occasionally seen (Figure 1B).

High-Flow Vascular Malformations

AVMs comprise approximately 10% of peripheral vascular malformations [28,30]. Their hallmark feature is the nidus, which is the confluence of serpentine vascular structures, e.g., feeder arteries and draining veins without an intervening capillary bed. Identifying the nidus has management implications, as it is frequently the target of sclerosant injection [7,29]. Conversely, AVFs are composed of a single channel between a vein and an artery. Following contrast administration, both types of lesions show rapid filling in 5–10 seconds, typically with contrast reaching the venous side of the lesion during the arterial phase, however, AVMs will demonstrate a confluence of many feeder arteries and draining veins, while AVFs will demonstrate a single large channel bridging the artery and vein.

The distinction between low-flow and high-flow vascular malformations relies on the late filling of the former and early filling of the latter in the arterial phase on dynamic contrast-enhanced MRI (DCE-MRI) (Figure 2). Other distinguishing features include the presence of dilated tortuous vessels and flow voids on spin echo images, which indicate high-flow components of a vascular malformation.

Pitfalls in MRI Interpretation

Calcification and phleboliths may also appear as flow-voids and can lead to erroneous labelling of low-flow lesions and high-flow lesions [31]. Since calcifications and phleboliths are better visualized on CT, a non-contrast CT may be obtained for better identification [5]. Similarly, septations have a low signal on T2WI. These potential errors are reduced utilizing high signal on GRE and contrast enhancement to differentiate true high-flow lesions from other causes of low signal on standard MRI [29].
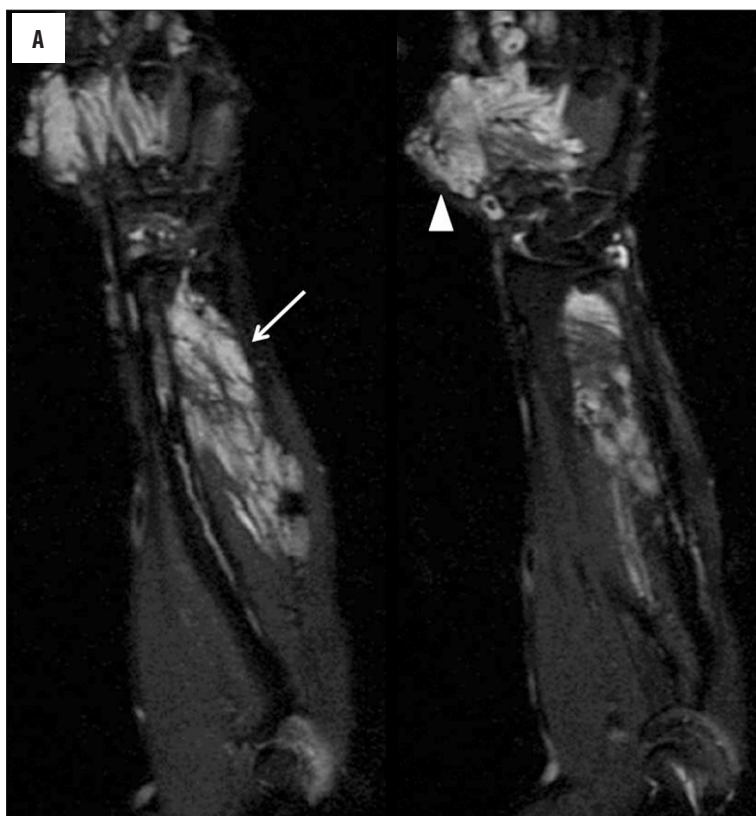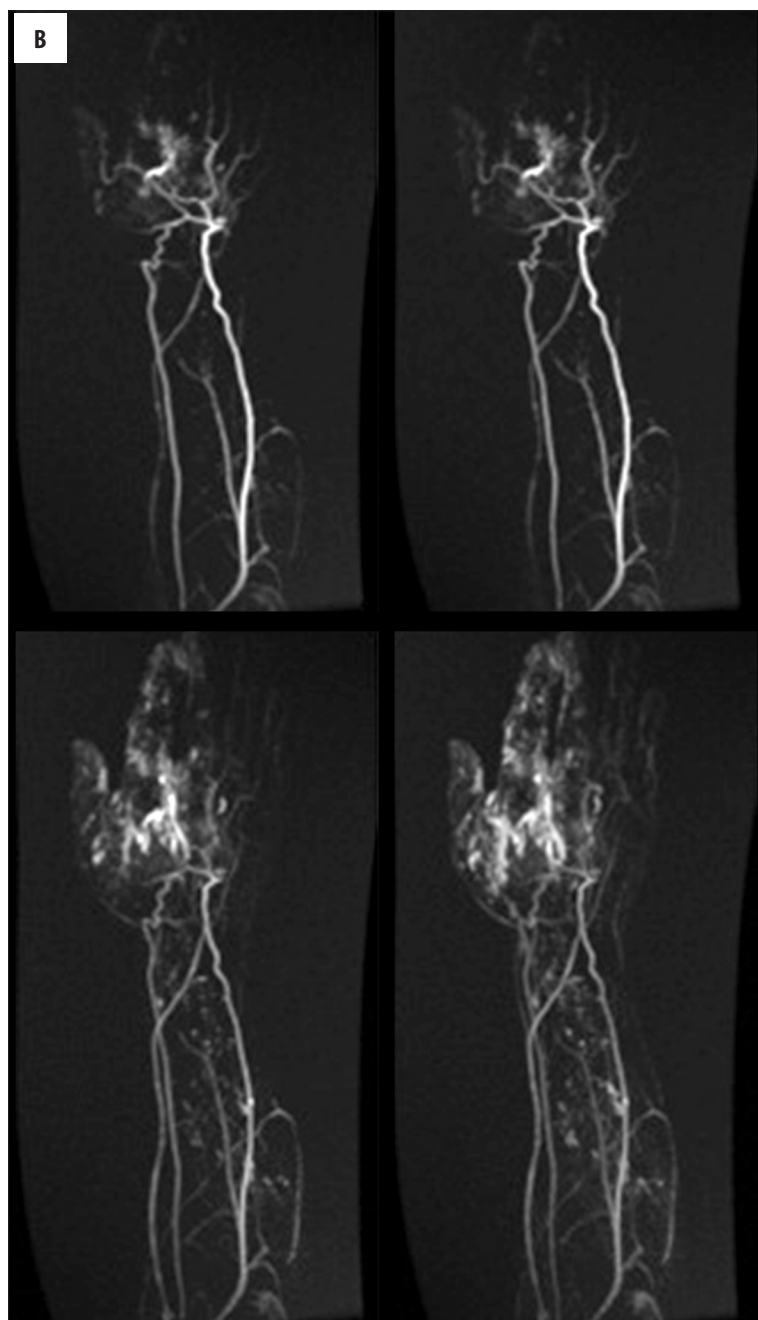
Figure 2. (**A**) Coronal T2 fat saturation images demonstrate a diffuse soft tissue lesion in the forearm (arrow) and hand (arrowhead). (**B**) Time resolved contrast images of the same lesion demonstrate early filling of the lesion consistent with arteriovenous malformation.

Slow-flow venous malformations might be indistinguishable from lymphoid malformations due to cystic appearance and sedimentation of blood products. Post-contrast imaging with or without fat-suppressed T1 sequence is imperative, as lymphoid malformations enhance peripherally, while venous malformations enhance homogeneously throughout the lesion [17].

Central Nervous System Vascular Malformations

Central nervous system (CNS) vascular malformations have been extensively studied owing to their profound sequelae and related complications [32–36]. The most common presenting symptoms for lesions in the brain are seizures, headache, and focal neurological deficits. Similar to

vascular malformations located in other organ systems, identifying the nature of vascular supply and differentiating a slow-flow malformation from a high-flow malformation are important determinations to be made on imaging. Generally, superficially located brain vascular malformations are supplied by pial arteries and drain into superficial cortical veins (Figure 3), whereas deeper vascular malformations are supplied by perforators e.g., the lenticulostriate arteries and choroidal arteries and drain into the deep venous system [1].

In addition to the general characterization outlined above, intracranial vascular malformations have specific features related to sensitive functional regions, i.e., the eloquent cortex and involvement of adjacent brain parenchyma in
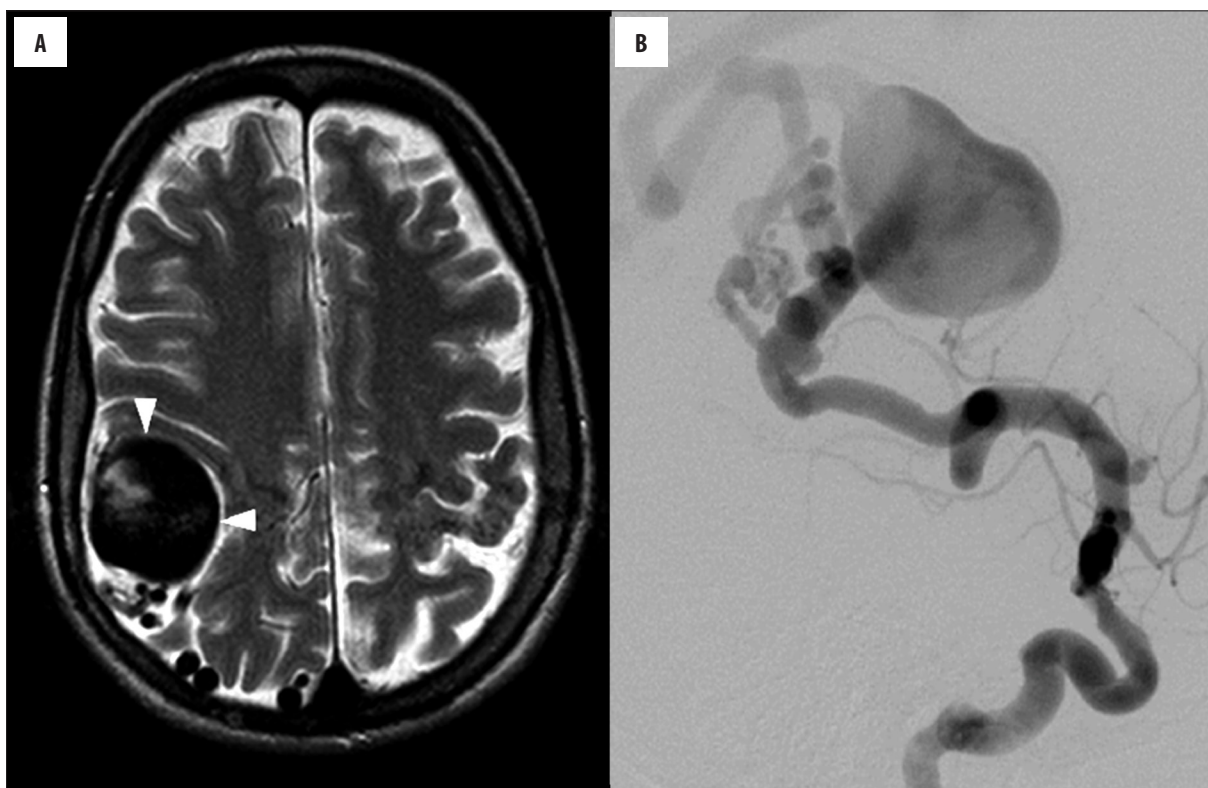
Figure 3. (**A**) Axial T2 image (1) demonstrates a dural AVF with a large venous varix (arrowhead), which enhances on a T1 contrast-enhanced axial image (2). There is early enhancement of dilated veins (arrow). (**B**) Conventional angiogram demonstrating large dural AVF with distended draining veins and early filling of the sagittal sinus (arrow).
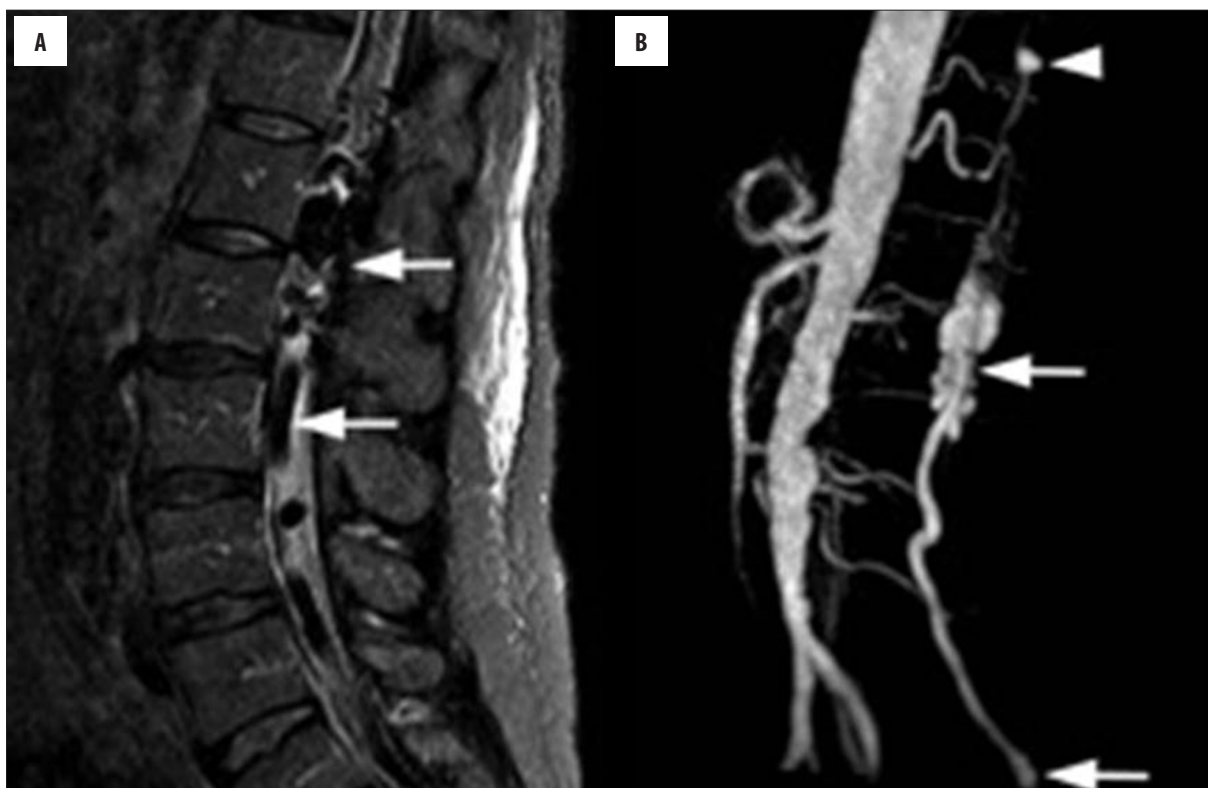

Figure 4. Sagittal MRI (T2WI) (**A**) and MRA (MIP images) (**B**) demonstrate a spinal cord AVM with an entanglement of blood vessels on the surface of the cord and cauda equine. AVM is demonstrated with signal void (arrow) on T2WI (**A**) and hyperintense signal (arrow) on MRA (**B**). The nidus (arrowhead in b) is present at the cranial end of the lesion.

Figure 5. Coronal STIR (**A**) and coronal T1 (**B**) post-contrast image demonstrate a large AVM of the left hand, with auto-amputation of the distal phalanges of the ring and little finger (arrows).
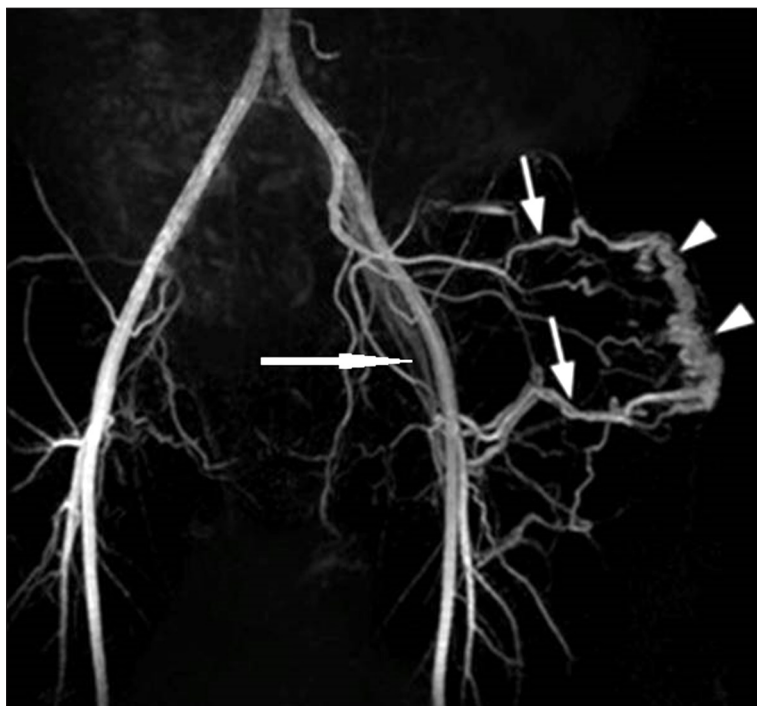

Figure 6. Coronal MRA demonstrates a gluteal AVM with a large tangle of vessels (arrow heads) supplied by branches of the internal iliac and the common femoral artery (arrows) and drainage via the common femoral vein (long arrow). This is a high-flow lesion.

vascular supply, which would naturally be affected, if the vascular supply to the lesion were reduced as part of treatment. The Spetzler-Martin grading system is frequently used to score the lesions and predict post-treatment outcome of the patient [37,38].

Spinal vascular malformations present with paresthesias, motor symptoms, or autonomic dysfunction such as incontinence or retention, depending on the spinal cord level involved as well as the degree of cord compression or ischemia due to the steal phenomenon [39]. Lesions may be intra-medullary or extra-medullary and similar to vascular malformations elsewhere, they can be high-flow, e.g. dural AVM (Figure 4) or low-flow lesions, with the former category constituting about 70% of all lesions [40,41]. Diagnosis is based on finding a long segment enhancing serpiginous
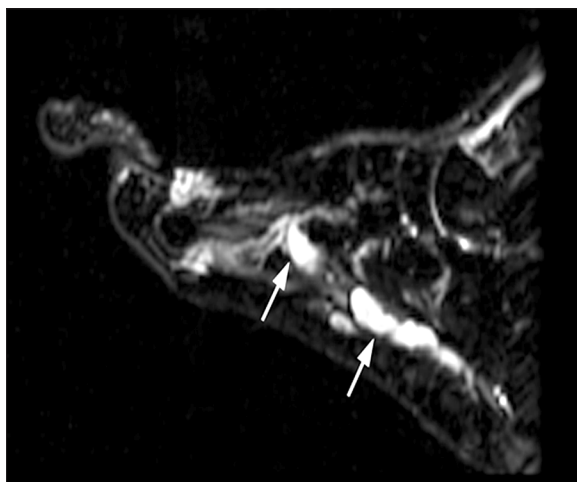
Figure 7. Sagittal MRA demonstrate an AVM along the plantar surface of the foot (arrows).
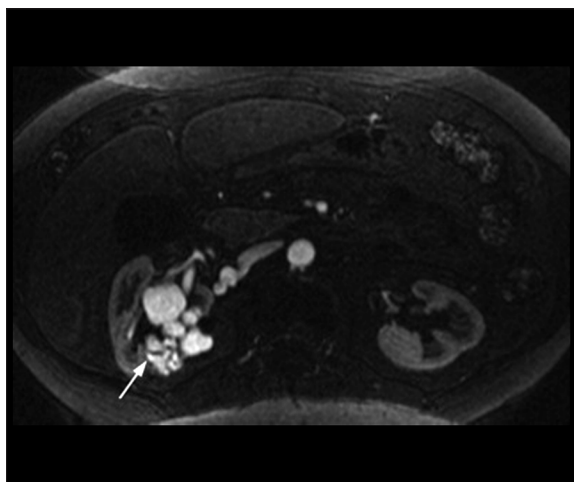

Figure 8. Axial MRA (MIP) demonstrates an enhancing entangle of vessels (arrow) consistent with right renal hilum AVM.
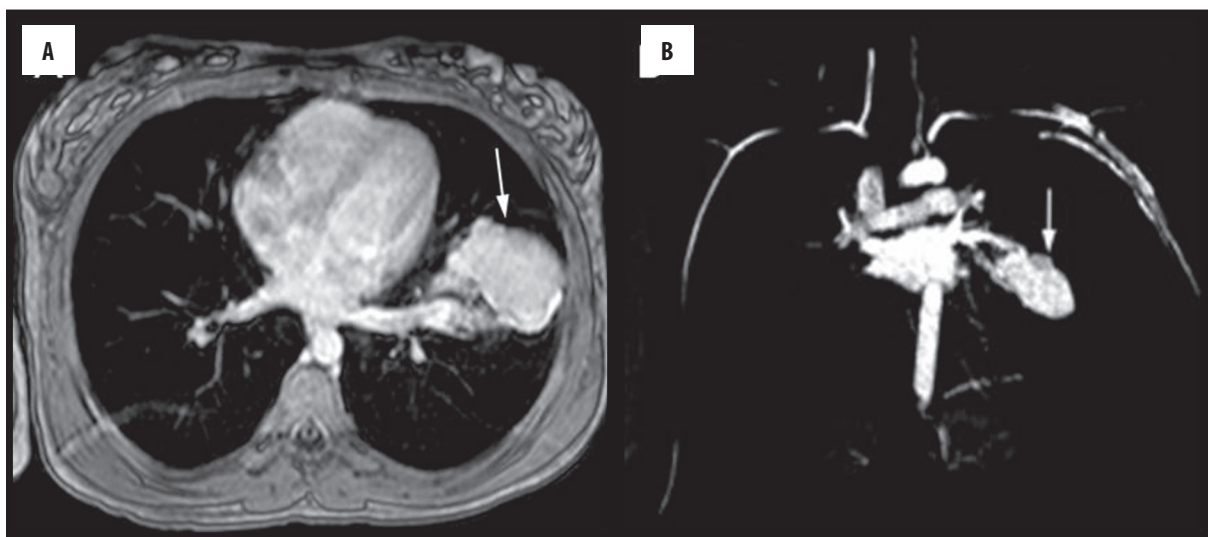

Figure 9. Axial contrast-enhanced MRI (**A**) and coronal MIP (**B**) images demonstrate a large left lung AVM (arrows).

structure within the spinal canal composed of abnormal vessels. Low-flow lesions demonstrating a paucity of flow-void on MR imaging can be confused for spinal neoplasm; conversely, cerebrospinal fluid flow artifact can be mistaken for an AVM of the cord by the unwary.

Peripheral Vascular and Visceral Malformations

The majority of the peripheral vascular malformations are low-flow venous malformations [7]. Within the trunk and limbs, lesions frequently spread across several muscles and tissue compartments owing to their congenital nature. Syndromic associations have been described, including Proteus, Mafucci, Klippel-Trenauny-Weber [42–44] and blue rubber bleb syndromes [45]. On MRI, peripheral vascular malformations appear as serpiginous structures with or without a nidus. Orientation along the long axis of the limb, multifocal involvement, and occasionally, the presence of atrophy of the surrounding structures may also be seen (Figure 5) [25,27,46]. Imaging evaluation, as a general principle, should take into account the presence or absence of a nidus, extent of lesion, and size of the affected area. As

outlined earlier, the distinction of the lesion into high-flow and low-flow hemodynamic categories is critical; this, in turn, depends on finding dilated vascular structures and early venous filling on dynamic T1WI – features indicative of high-flow lesions. Examples of peripheral and visceral malformations are seen in Figures 6–9.

Management of peripheral vascular malformations is generally conservative, unless the patient develops complications such as cosmetic disfiguration, functional impairment, pain, or high output cardiac failure [7]. Quite often, high-flow lesions are subjected to some manner of flow-control by tourniquet or embolization prior to sclerosant injection into the nidus for definitive closure. Management decisions are based on a multidisciplinary team-approach, as these lesions are difficult to treat and recur frequently. As a result, multi-staged, multi-disciplinary treatment approaches involving a cosmetic surgeon, a vascular surgeon, and an interventional radiologist may be necessary for the larger high-flow lesions [7,47]. In contrast, smaller low-flow lesions can usually be treated in a single session of sclerosant injection. It is imperative that radiologists

are aware of the specific challenges faced in the management of these lesions and structure their report to facilitate decision-making.

Post Therapy Changes

Percutaneous sclerotherapy is the treatment of choice for low-flow vascular lesions, while trans-arterial embolization followed by surgical resection is the treatment of choice for high-flow lesions. MR is the imaging modality of choice for post-therapy evaluation of lesions [48].

Venous malformations: Immediate post therapy, T2WI shows hyperintense signal which persists up to 3 months. On MR angiography, there is an absence of enhancement in the central portion of the treated lesion. However, there is peripheral, reactive arterial enhancement up to 3 months post therapy. After 3 months, there is a loss of peripheral enhancement and a central scar may be seen which is dark on T1 and STIR images. Post-contrast images are imperative in assessing for any residual lesions and planning subsequent therapy if needed [48].

Arterial malformations and AVMs: After trans-arterial embolization, there is thrombosis of the lesion resulting in absent shunting and reduced or absent early opacification of draining vein. Early post-therapy imaging is recommended for early identification of residual malformations for planning the second stage of treatment [48].

Conclusions

Vascular malformations are developmental abnormalities, which usually do not present before puberty. MRI with dynamic contrast-enhanced sequences is the mainstay of diagnosis and characterization of these lesions. The identification of a nidus and the classification into high-flow or low-flow categories are important determinants of treatment modality and should be sought out during the evaluation of these lesions. It is important to note that despite optimal therapeutic intervention, many these lesions recur and require multiple treatment sessions and complex management decisions.

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