Techniques in Vascular and Interventional Radiology

Brief Description of ISSVA Classification for Radiologists

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Ongoing discovery regarding the origin and treatment of vascular anomalies requires standardized nomenclature which itself must undergo iterative updating. This article introduces the 2018 International Society for the Study of Vascular Anomalies (ISSVA) classification, emphasizing the biologic basis of vascular anomalies, summarizing the key features of commonly encountered entities, and serving as a foundation for subsequent articles presented herein. Vascular tumors are discussed to highlight their distinction from vascular malformations which will receive greater attention with respect to management and technical considerations within the issue.
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KEYWORDS vascular, tumor, malformation, anomaly

Introduction

Clarity in nomenclature begets effective study and treatment of any condition. The field of vascular anomalies exemplifies the principle particularly well. In their 1982 papers, Mulliken and Glowaki set forth a binary classification system by histologic composition with the foremost distinction of a vascular anomaly into a mass or malformation on the presence or absence of increased mitotic activity and endothelial cell turnover.[1,2] This original binary classification based on histopathogenesis was adopted by the International Society for the Study of Vascular Anomalies (ISSVA) and receives periodic revision during biennial international workshops.[3] The use of the ISSVA classification among clinicians managing vascular anomalies continues to grow. Despite advances in consensus nomenclature, widespread use of incorrect terminology persists. Hassenein et al. published results of a PubMed literature search for instances of the term "hemangioma" appearing in the manuscript titles of abstracts, identifying incorrect use in 71% of these publications. Most importantly, erroneous classification led to a significantly increased likelihood of improper treatment, 20.6% compared to 0% for correctly designated lesions.[4] Prior to the discussion of management, therefor, a brief overview of the widely accepted ISSVA classification is warranted.

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Foremost Distinction—Mass or Malformation

Not everything with vessels and volume should be referred to as a "mass" and the term "vascular mass" is reserved specifically for vasoproliferative lesions with increased endothelial cell turnover. Relatedly, the suffix "-oma" implies neoplastic behavior. Terms ending in "-oma" and referring to lesions without increased mitotic activity or endothelial cell turnover, such as "cavernoma" "lymphangioma" or "cystic hygroma," add confusion and should largely be abandoned. The term "hemangioma" maintains its place in current terminology, but not as the indiscriminately inclusive descriptor of bygone days. Vascular masses are subsequently broadly subdivided by their malignant potential: benign, locally aggressive or borderline, and malignant (Table).

Unlike vascular masses, vascular malformations are non-neoplastic and represent focal, defective morphogenesis with quiescent endothelium. Simply put, they are badly ("mal-") formed vessels. Vascular malformations are present at birth and generally grow in proportion to the child. Histologically, they are composed of irregular, dilated, and thickened channels displaying mature endothelial lining.[5] These comprise a large spectrum of entities, ranging from simple, combined, malformations of major named vessels, and syndrome-associated malformations. Simple and complex malformations derive their names from their constituent malformed vessel type(s): capillary, lymphatic, venous, arteriovenous, and many combinations thereof (Table). The broad distinction into "high-flow" or

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Table International Society for the Study of Vascular Anomalies (ISSVA) Classification. Adapted from issva.org/classification

Vascular Tumors	Vascular Anomalies			
	Vascular Malformations			
	Simple	Combined	Of Major Named Vessels	Associated With Other Anomalies
Benign Borderline Malignant	Capillary Lymphatic Venous Arteriovenous	CVM, CLM, LVM, CAVM, others		

"slow-flow" malformations developed by Jackson et al. based on the presence of absence of arterial components is conceptually useful for both diagnosis and treatment.[6]

Benign Vascular Tumors

Infantile Hemangioma/Hemangioma of Infancy

Benign infantile hemangiomas (IH), typically occurring in their sporadic form, represent by far the most common vascular tumor. They present postnatally, typically between 2 weeks and 2 months, first undergoing a rapidly proliferative phase in the first year of life and then regressing during childhood.[7] Lesions frequently involve the skin of the head, neck, trunk and extremities, but may arise in deeper structures such as the parotid gland (Fig. 1) and liver. Distribution may be focal, regional or diffuse. On histopathology, IHs uniquely express glucose transporter-1 (*GLUT1*) protein.

Simple IHs frequently present where they are readily apparent and display a typical appearance, often obviating the need for imaging or biopsy. Color Doppler ultrasound avoids ionizing radiation and need for sedation and is thus the preferred imaging modality. Computed tomography (CT) and magnetic resonance (MR) are reserved for the characterization of deep or extensive involvement. Regardless of modality, IHs demonstrates well-defined lobulated contours,

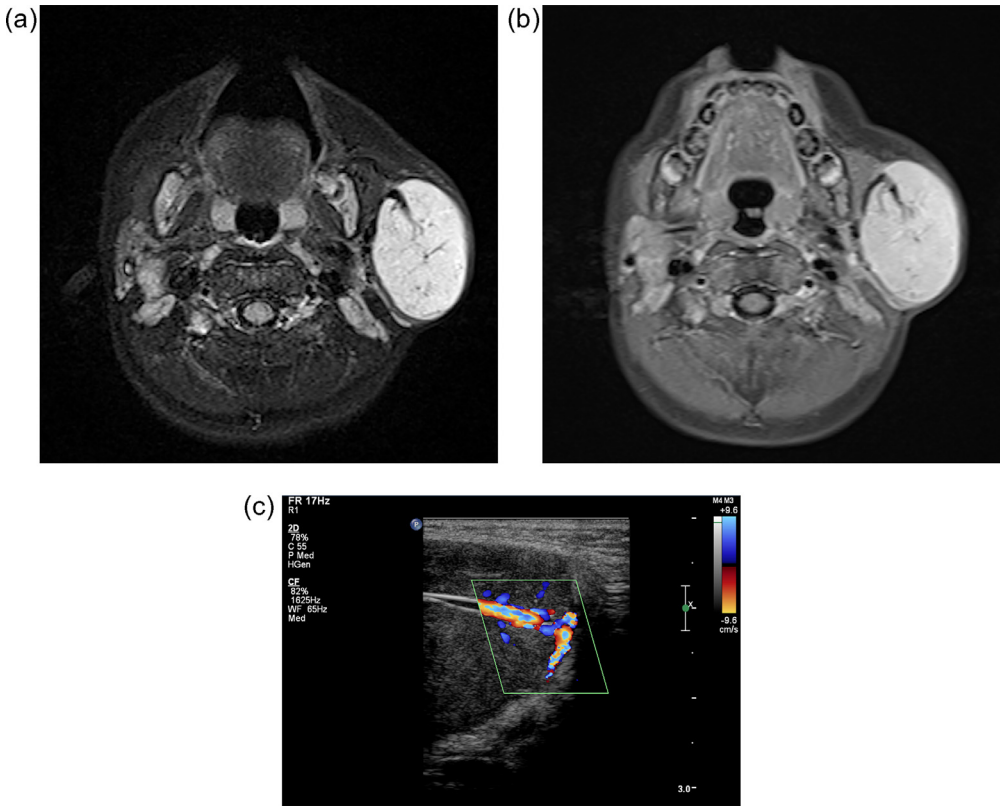

Figure 1 Infantile hemangioma. A 6-month-old female presents with progressive left facial swelling, present since birth. Axial STIR MR image (**a**) demonstrates a well circumscribed hyperintense lesion within the left parotid gland containing branching internal flow voids emanating from a pedicle at the medial aspect of the mass. Axial T1-weighted fat-saturated postcontrast MR image (**b**) demonstrates avid homogeneous enhancement. Doppler ultrasound image (**c**) confirms a well-circumscribed mass with internal vascularity and a distinct vascular pedicle. Excision after the mass was found to be unresponsive to propranolol confirmed a *GLUT1* positive infantile hemangioma.

solid lesional parenchyma with both arterial and venous flow, and brisk contrast enhancement.[8] Fibrofatty tissue gradually replaces these features with progressive involution.

Hepatic hemangiomas (HH) arrive in focal, multifocal, or diffuse forms.[9] They are distinct from the incorrectly named "cavernous hemangiomas" encountered in the livers of adult patients, which histologically represent hepatic venous malformations.[10] Furthermore, as elucidated by the Liver Hemangioma Registry, mutifocal and diffuse forms parallel IHs in terms of clinical behavior, imaging, and histopathology while focal HHs represent a distinct entity more closely related to the subsequently discussed congenital hemangiomas (CHs).[10] Multifocal and diffuse forms are associated with cutaneous IHs and hypothyroidism. Children with multiple IHs should be screened for multifocal or diffuse HHs. Focal HHs, because they represent the hepatic variety of CH, may be detected on prenatal ultrasound (US). Extensive hepatic hemangiomas of any type may lead to heart failure or abdominal compartment syndrome (Fig. 2).

Congenital Hemangioma

As their name suggests and unlike IHs, CHs are present at birth. They appear as conspicuous exophytic masses or bossed plaques, occasionally with a pale halo, and most frequently occur on the head, neck, and extremities. CHs are classically divided into 2 subtypes based on their postnatal clinical behavior: rapidly involuting congenital hemangioma (RICH) and noninvoluting congenital hemangioma (NICH). RICH subtypes involute entirely within 1-2 years while NICH subtypes persist and grow proportionally with the child, occasionally necessitating surgical excision.[11,12] Both are *GLUT1* negative and display similar imaging features of heterogeneous parenchyma, visible vessels, and occasional calcification.[13] The 2018 ISSVA classification recognizes a CH with distinct behavior of partial involution (PICH). Lesions present in a similar fashion to the RICH variety, but arrest regression and subsequently exhibit behavior typical of the NICH variety. Imaging and histopathologic features overlap with both RICH and NICH, and these lesions are similarly *GLUT1* negative; distinction of this entity is therefor made primarily based on clinical behavior.[14] Some have theorized NICHs all represent lesions originally beginning as RICHs and subsequently aborting involution by a yet-to-be-determined mechanism.[15] Within this construct, the classic NICH would result from involution arrest in utero and a partially involuting congenital hemangioma (PICH) would result from postnatal involution arrest.[14]

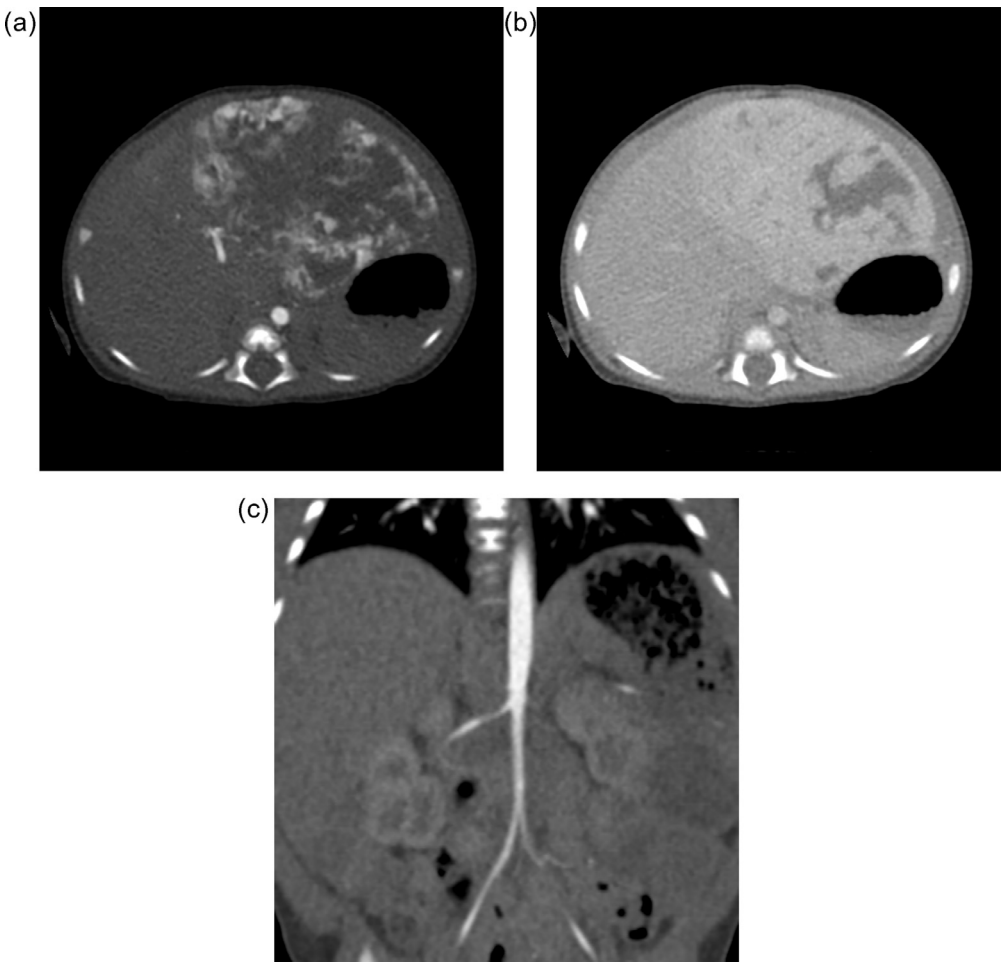

Figure 2 Hepatic hemangioma. A 4-week-old female found to have a large hepatic mass in utero presents with progressive heart failure. Axial arterial (**a**) and venous (**b**) phase CT images demonstrate a large left hepatic mass with centripetal enhancement. Coronal arterial phase image (**c**) demonstrates abrupt aortic tapering below the celiac axis.

Tufted Angioma

Tufted angiomas, occasionally referred to as progressive capillary hemangiomas, manifest as poorly defined, infiltrative lesions (Fig. 3). These rare vascular neoplasms typically affect the limbs followed by cervicofacial territories, particularly the mandibular area where they may be associated with hursuitism.[17] Cutaneous involvement often occurs without overt subcutaneous extension or bulk on examination. Clinical appearance and/or biopsy typically provide the diagnosis and imaging features are therefore infrequently described.[18] Tufted angiomas carry a risk of the Kasabach-Merritt phenomenon, or lesional sequestration of platelets, fibrinogen, and red blood cells with subsequent profound coagulopathy.[12] Management follows that of the potentially related and more aggressive kaposiform hemangioendothelioma.

Pyogenic Granuloma

Also known as lobular capillary hemangioma, pyogenic granulomas are typically superficial and small (<1 cm) but highly vascular lesions. They typically occur on the face, neck, and mucosal surfaces as a result of local trauma or inflammation, or in association with other vascular anomalies.[18] Additional benign vascular tumors beyond the scope of this discussion include intravascular papillary epithelial hyperplasia, cutaneous epithelioid angiomatous nodule, bacillary angiomatosis, eccrine angiomatous hamartoma as well as spindle-cell, epithelioid, hobnail, microvenular, anastomosing, glomeruloid, papillary, acquired elastotic, and splenic littoral cell type hemangiomas.

Locally Aggressive or Borderline Vascular Tumors

Kaposiform Hemangioendothelioma

Kaposiform hemangioendotheliomas are rare vasoproliferative lesions which present shortly after birth as enlarging cutaneous/subcutaneous masses. Lesions typically arise on the trunk, extremities, head, neck, and retroperitoneum. They are locally aggressive, frequently extending into adjacent soft tissues and lymphatics but do not commonly metastasize.[19] MR demonstrates an infiltrating T2-heterogenous lesion with avid

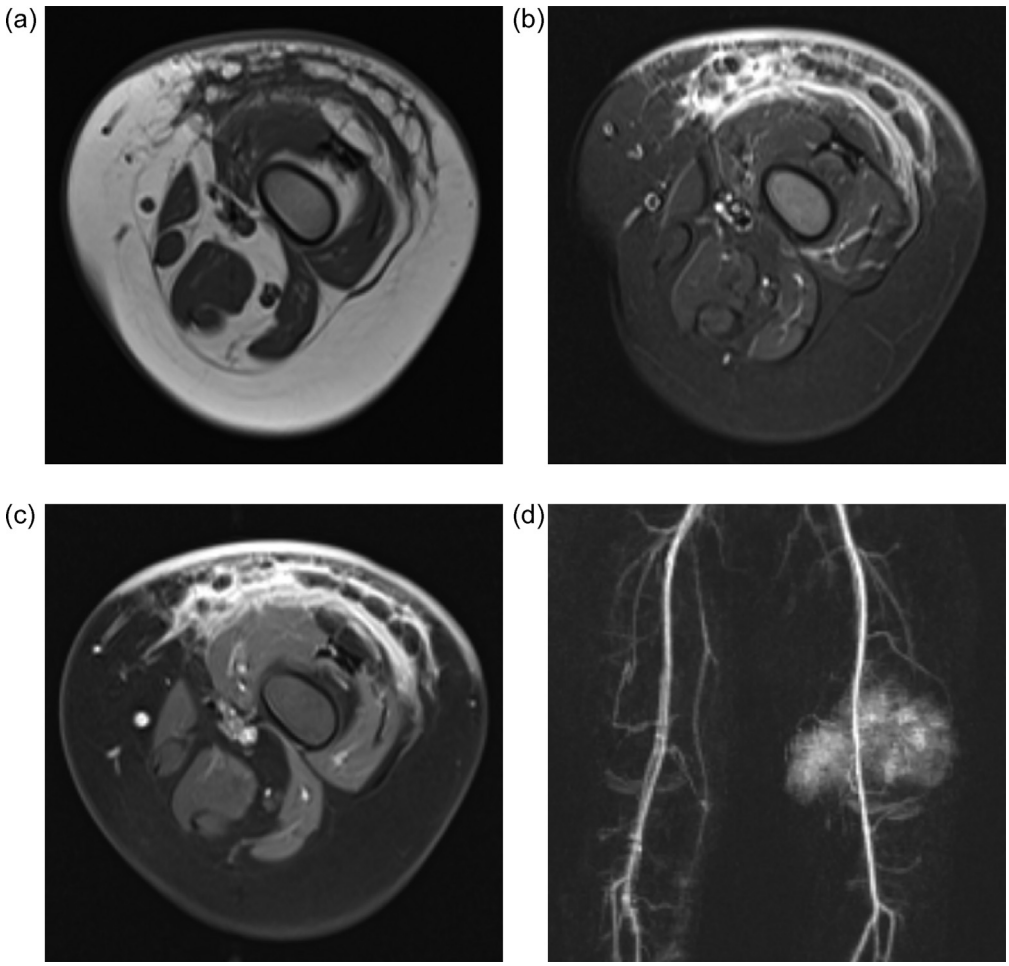

Figure 3 Tufted angioma. A 6-month-old male presents with progressive focal discoloration and swelling of the anterior left thigh and thrombocytopenia. Axial T1-weighted MR image (**a**) demonstrates ill-defined hypointensity within the subcutaenous fat deep to the cutaneous lesion. Axial STIR MR image (**b**) demonstrates corresponding hyperintensity. Axial T1-weighted fat-saturated post-contrast MR image (**c**) and coronal time resolved MR angiogram image (**d**) demonstrate avid enhancement.

enhancement and conspicuous vessels leading up to but not within the lesion.[16,20-22] Histologic similarities to tufted angioma have caused some to speculate kaposiform hemangioendothelioma to be the more aggressive variant on a single entity (Enjolras). They are similarly associated with the development of potentially fatal Kasabach-Merritt phenomenon. While medical management, including corticosteroids or chemotherapy (vincristine, sirolimus) is the mainstay of treatment, adjunctive transarterial chemoembolization has been reported for lesions complicated by Kasabach-Merritt phenomenon.[23] Additional borderline vascular tumors beyond the scope of this discussion include a variety of named hemangioendothelioma subtypes, papillary intralymphatic angioendothelioma ("Dabska tumor") and Kaposi sarcoma.

Malignant Vascular Tumors

Angiosarcoma

Angiosarcomas are uncommon vascular tumors in both pediatric and adult patients. While rare, prognosis is dismal. The arise spontaneously or in relation to pre-existing vascular anomalies, chronic lymphedema, or exposure to anabolic steroid and toxins such as arsenic, thorotrast, and vinyl chloride.[24,25] Imaging features are nonspecific but may include intralesional hemorrhage or contrast pooling and occasional fluid-fluid levels from layering blood products.[26]

Epithelioid Hemangioendothelioma

Epithelioid hemangioendotheliomas may be encountered in a variety of locations such as the bone and brain but most frequently arise within the liver. A hypoenhancing and T2 hypointense rim resulting in a targetoid appearance is classic, occasionally accompanied by capsular retraction.[26,27] These rare malignancies are not to be confused with multifocal hepatic hemangiomas, which have previously been erroneously referred to as epithelioid hemangioendothelioma.

Vascular Malformations

Simple Vascular Malformations

The 2018 ISSVA classification distinguishes simple vascular malformations from combined forms, malformations of major named vessels, and malformations associated with other anomalies (ie, syndromes) (Table). Simple and combined vascular malformations derive their name from constituent vessel type(s). Slow-flow malformations include dysplastic capillary, lymphatic, venous vessels or a combination thereof. Capillary malformations, frequently referred to as "port-wine stains," affect cutaneous and mucosal surfaces without a deep component. Unless suspected to represent a portion of a combined vascular malformation or part of a syndrome, capillary malformations are infrequently imaged. The common, simple variety, occasionally referred to as nevus simplex, will frequently spontaneously resolve.

Lymphatic malformations consist of dilated, ectatic lymphatic channels, and cystic spaces. They are subdivided into microcystic, macrocytic, and mixed subtypes based on the prevailing size of the cystic spaces. Distribution varies from discrete muticystic to ill-defined, infiltrating lesions. Although lymphatic malformations may occur anywhere, anterior cervical involvement predominates, where occasionally extensive and multicompartmental extension threatens the airway (Fig. 4). Cervicofacial lymphatic malformations of this variety are typically obvious on prenatal screening or immediately after birth, and bilateral involvement helps to differentiate from cervical teratomas. While occasionally referred to as "cystic hygromas" when presenting in this manner, the term is archaic and should be abandoned. Smaller and/or deeper malformations may remain clinically silent before coming to clinical attention as a result of intralesional hemorrhage or expansion during viral illness (Fig. 5). US imaging demonstrates thin walled, largely anechoic spaces with occasional echogenic debris. MR demonstrates thin septa with minimal or no enhancement. Cyst contents are generally T2 hyperintense with variable intrinsic T1 or T2 signal on the basis of internal proteinaceous content or

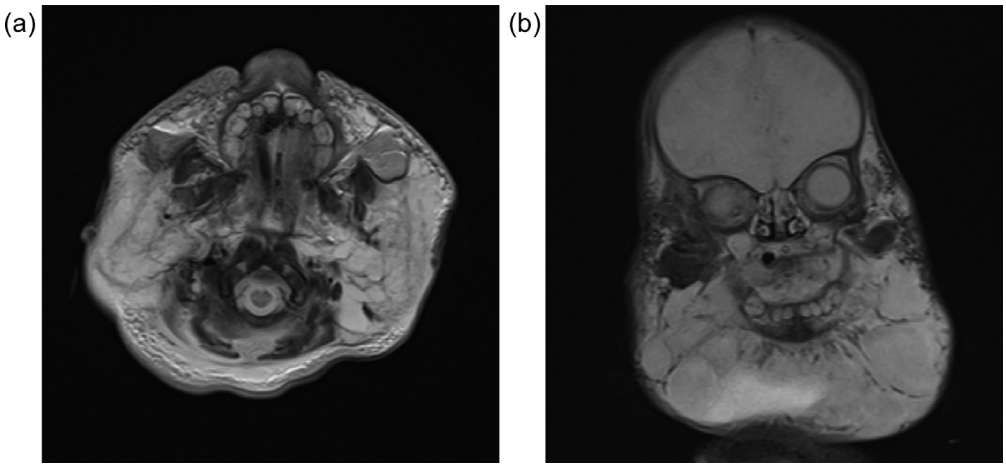

Figure 4 Cervicofacial mixed type lymphatic malformation. A 2-day-old boy diagnosed in utero with a cervicofacial cystic lesion. Axial T2-weighted (**a**) and coronal short tau inversion recovery (STIR) (**b**) MR images demonstrate bilateral extensive, multicompartment mixed type lymphatic malformation.

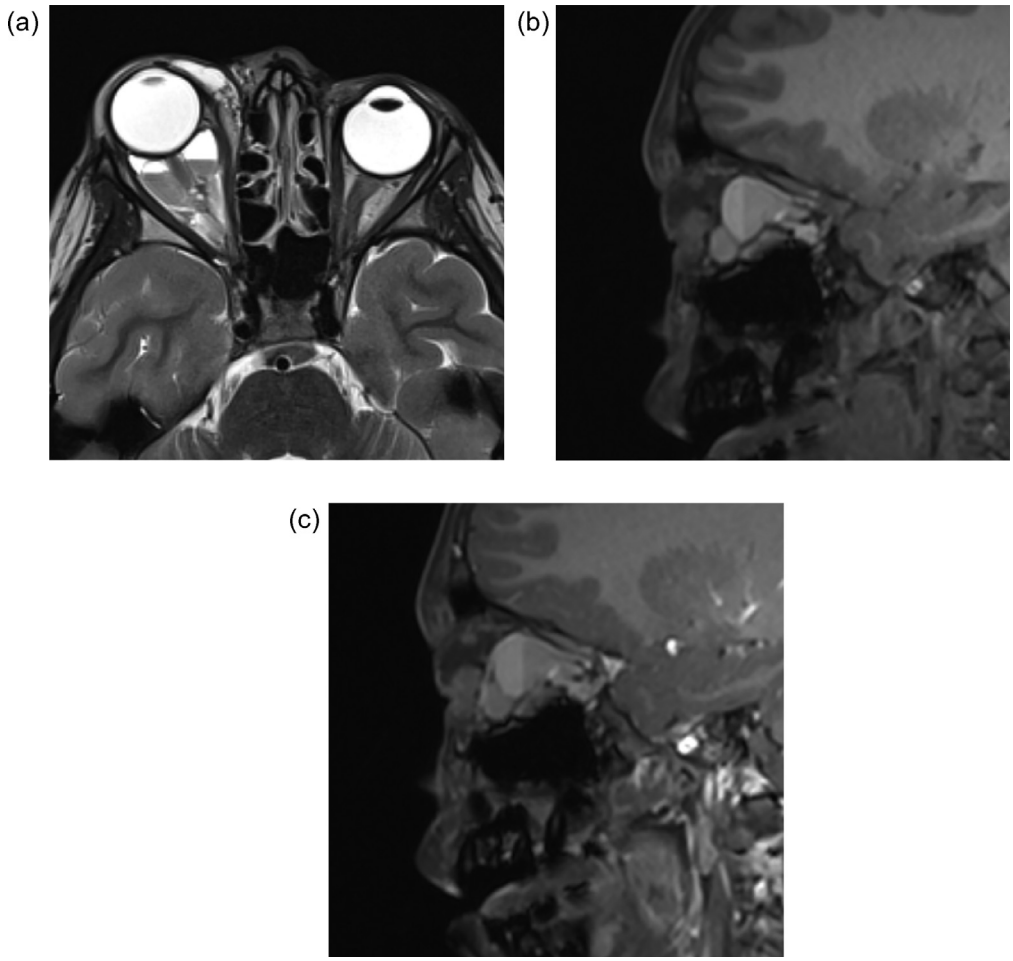

Figure 5 Orbital lymphatic malformation. An 8-year-old girl presents with recurrent right eye pain and proptosis exacerbated by upper respiratory tract infections. Axial T2-weighted MR image (**a**) demonstrates a multiloculated, macrocystic intraconal lesion with fluid-fluid levels. Sagittal T1-weighted, fat-saturated MR images before (**b**) and after (**c**) administration of IV contrast demonstrate layering fluid-fluid levels of variable intrinsic T1 hyperintensity of the orbital lesion with no enhancement.

intralesional hemorrhage and do not enhance following administration of intravenous contrast. Microcystic lesions, due to the high relative volumetric composition of septa, may appear solid and enhancing. Macrocystic lesions, while often large at presentation, typically respond impressively to drainage and sclerotherapy (Fig. 6). Microcystic varieties present greater therapeutic challenges, and may require multiple modes of treatment ranging from sclerotherapy to medical management and surgical debulking.

Venous malformations (VM), the most common vascular malformation type encountered, represent focally ectatic venous channels frequently deficient in elastic lamina and smooth muscle.[5] Most arise within the face and neck with the limbs, trunk, skeletal muscle, viscera, and bones less commonly affected.[28] Distribution ranges from focal to diffuse and infiltrative across tissue planes. As a result, symptoms range from total quiescence and incidental discovery to cosmetic disfigurement to debilitating pain and disability. Discomfort may result from mass effect on adjacent structures, venous engorgement, restricted range of motion, local hemorrhage, or thrombophlebitis.[29] Localized intravascular coagulopathy is common, particularly within large or extensive lesions.[30,31] Cutaneous, mucosal or otherwise superficial lesions may demonstrate a characteristic blue tinged compressible mass which distends with Valsalva or dependent positioning. While longstanding intralesional thrombi may result in phleboliths apparent on radiographs (Fig. 7), suspected VMs should first be evaluated with Doppler US.[32] Serpigenous, dilated and anechoic channels with monophasic Doppler flow is characteristic. Less commonly, VMs will demonstrate no flow, or flow velocity that is below the sensitivity of Doppler. Furthermore, very slow flow or intralesional thrombus produces variable channel echogenicity.[33] MR demonstrates lobulated T2 or short tau inversion recovery (STIR) hyperintense channels with late homogeneous or heterogeneous enhancement.[34,35] A wide variety of lesion channel and drainage morphologies are encountered with important therapeutic implications.[32]

Arteriovenous malformations (AVM) are high-flow lesions containing abnormal connections between primitive arteries and veins. They occur spontaneously or in syndromic forms such as hereditary hemorrhagic telangectasia (Fig. 8) or Parks

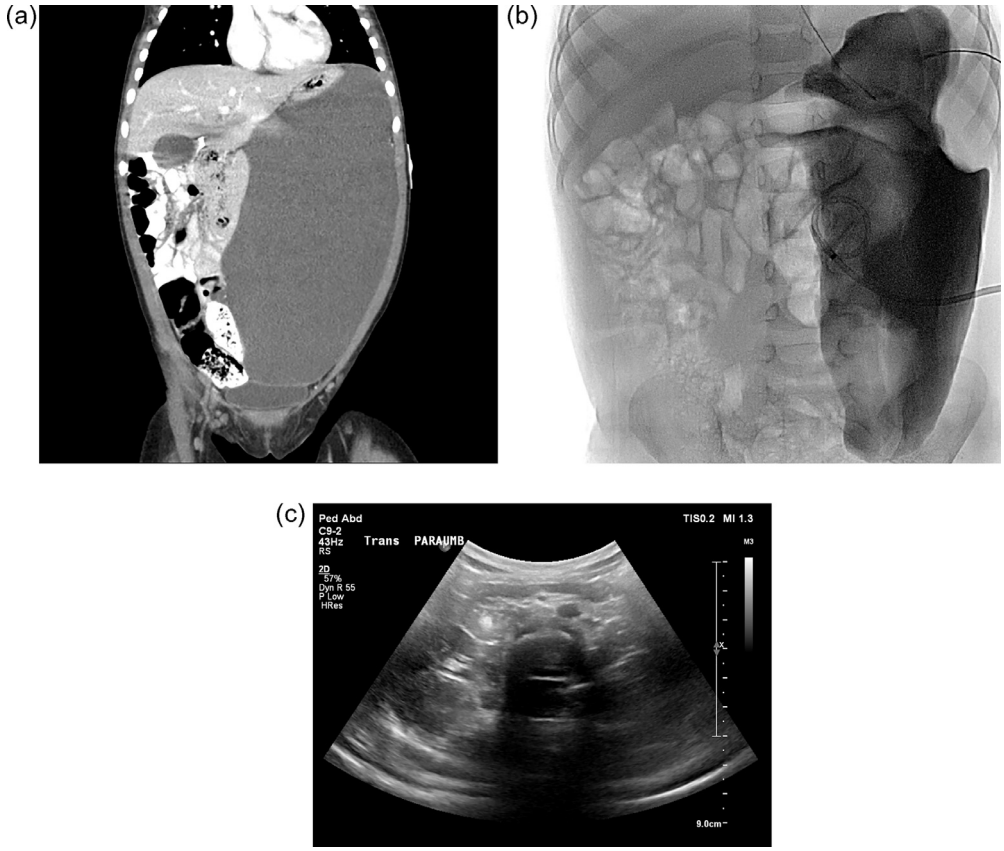

Figure 6 Abdominal macrocystic lymphatic malformation. A 2-year-old girl presented due to several months of progressive abdominal distension and early satiety. Coronal CT of the abdomen (**a**) demonstrates a large unilocular lymphatic malformation within the left aspect of the abdomen. Fluoroscopic image (**b**) following drain placement, evacuation of cyst contents and instillation of a contrast-opacified doxycycline sclerosant mixture. Transverse sonographic image of the abdomen 6-months post sclerotherapy (**c**) demonstrates no recurrence.

Weber syndrome.[36,37] The intervening dysplastic microvascular bed, otherwise known as the nidus, results in shunting of blood away from the normally supplied tissue; patients may present with ischemic pain (Fig. 9), ulceration, or hemorrhage.[37,38] Physical exam reveals a pulsatile mass occasionally with a palpable thrill or audible bruit. When a large volume of blood is being shunted, patients may present with high-output cardiac failure. US demonstrate engorged anechoic channels with turbulent, arterialized venous outflow. Waveform analysis of supplying arteries reveals high diastolic flow, reflecting loss of normal vascular impedance due to shunting. CT and MR demonstrate a cluster of enlarged arteries and veins with little to no solid component. Fast flow through large draining veins results in T2-weighted imaging flow voids and arterial phase venous enhancement on angiographic sequences. CT arteriography outperforms MR for the characterization of AVMs involving osseous structures.[39] Arteriovenous fistula, present in a similar fashion, but represent a focal connection between artery and vein without an intervening nidus. These occur in spontaneous forms (Fig. 10) but more frequently result from trauma (Fig. 11) or iatrogenic injury.

Combined Vascular Malformations

Combined vascular malformations are defined as 2 of more vascular malformations found in one lesion. They may occur in sporadic or syndrome forms and are named simply by hyphenation of the tissue types encountered.

Malformations of Major Named Vessels

The 2018 classification includes anomalies of major named vessels, otherwise known as "channel type" or "truncal" vascular malformations, broadly including anomalies of origin, course, number, and diameter as well as residual embryonal structures. This category captures a wide range of entities such as "megacava" or a persistent sciatic artery.

Diffuse Lesions/Syndromes

PIK3CA-Related Overgrowth Spectrum

PIK3CA-related overgrowth spectrum encompasses a group of segmental overgrowth disorders driven by activating mutations in the *PIK3CA* gene on chromosome 3,[40,41] several of which are accompanied by vascular malformations. Klippel-Trénaunay syndrome classically comprises a triad of capillary malformations, VMs, and soft tissue and bone overgrowth of one or more (typically lower) extremities.[42] Capillary malformations typically affect the hypertrophied extremity but may occur remotely. Hypertrophy manifests as

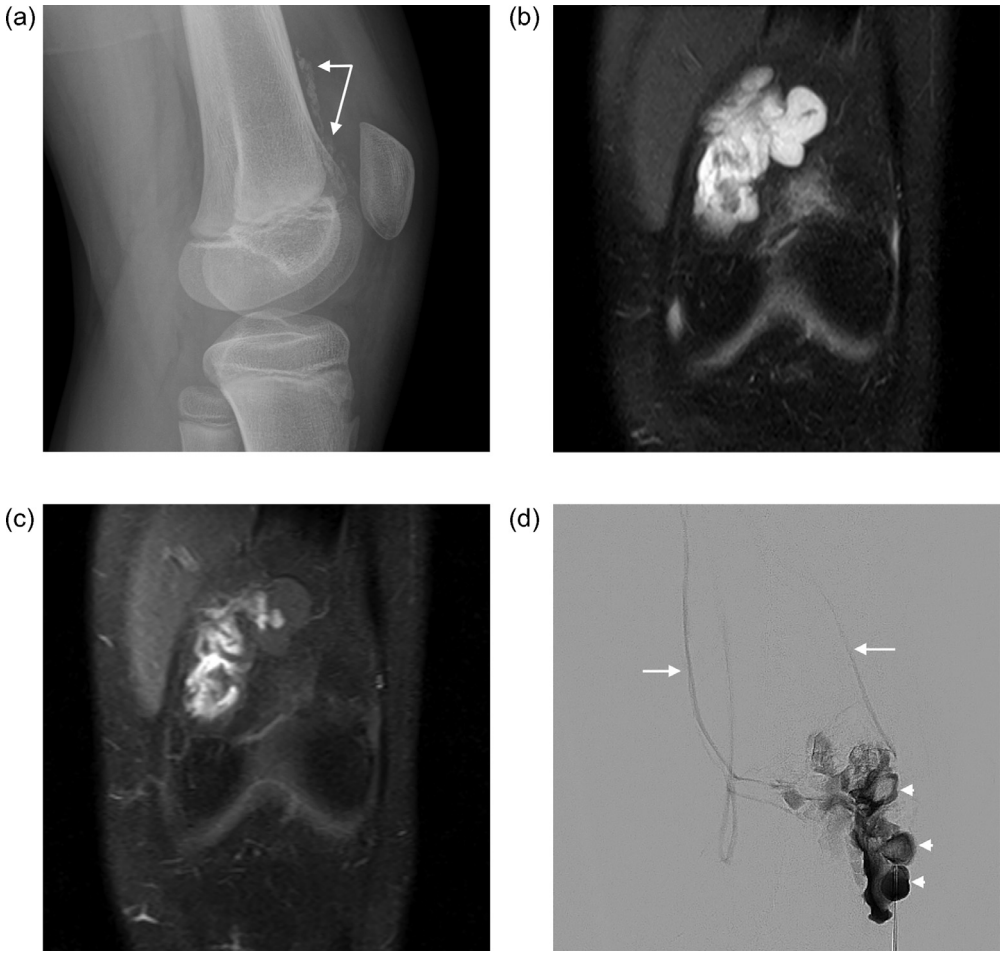

Figure 7 Intra-articular venous malformation. A 14-year-old girl presents with recurrent pain and swelling of the left knee. Lateral radiograph (**a**) demonstrates soft tissue thickening and phleboliths (arrows) within the suprapatellar bursa. Coronal proton density MR image (**b**) demonstrates a circumscribed, lobulated hyperintense lesion. Coronal T1-weighted fat-saturated postcontrast MR image (**c**) demonstrates channel enhancement and partial thrombosis. Lateral digital subtraction venogram following percutaneous lesion access (**d**) demonstrates slow-flowing ectatic channels containing phleboliths (arrowheads) draining to normal appearing superficial and deep veins (arrows).

bone elongation with leg length discrepancy and circumferential soft tissue overgrowth. A wide spectrum of venous anomalies may be encountered, including venous ectasia, persistence of embryologic veins (typically the lateral marginal vein of Servelle), hypoplasia or aplasia of normal deep veins, venous incompetence, and venous malformations of both deep and superficial systems (Fig. 12). Congenital lipomatous overgrowth, vascular malformations, epidermal nevi and skeletal abnormalities (CLOVES) syndrome is a rare condition manifesting with truncal lipomatous masses, spinal and acral anomalies, and a spectrum of vascular malformations ranging lymphatic malformations and ectatic thoracic veins to high-flow spinal/paraspinal lesions.[43,44] Both CLOVES and Klippel-Trénaunay syndrome place patients at increased risk of venous thromboembolism.[44,45] CLAPO syndrome is characterized by lower lip CM, cervicofacial LM, asymmetry and partial or generalized overgrowth. Additional entities within PIK3CA-related overgrowth spectrum include fibroadipose hyperplasia/overgrowth, hemihyperplasia multiple lipomatosis, macrodactyly, megalencephaly-capillary malformation and dysplastic megalencephaly.

RASA1-Related Disorders

Autosomal dominant RASA1-related disorders manifest with overlapping phenotypes of capillary malformation-arteriovenous malformations (CM-AVM) syndrome and Parkes Weber Syndrome (PKWS). RASA1-related CMs are characteristically small (1-2 cm diameter), round or oval, and randomly distributed primarily on the face and limbs.[46] Classic PKWS is characterized by multiple small arteriovenous fistula, associated CMs and overgrowth of an affected limb. While once considered to be sporadic and nongenetic, a subset of patients with PKWS, particularly those with multifocal CMs, demonstrated mutations in the RASA1 gene.[47] Individuals with RASA1-related disorders are at risk for potentially lethal high-flow intracranial malformations and Vein of Galen malformations.[47]

Hereditary Hemorrhagic Telangiectasia

Telangectasias represent bypasses the capillary vessels, or direct fusion of an arteriole and a dilated postcapillary

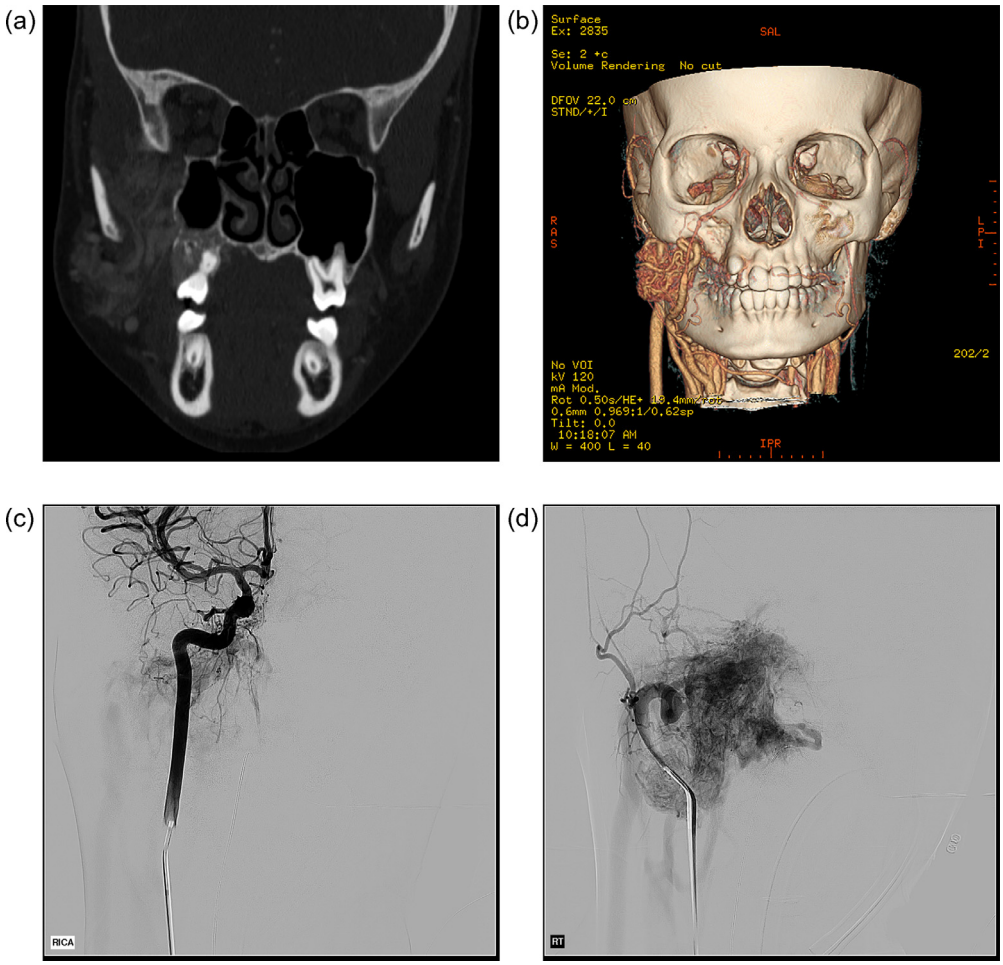

Figure 8 Arteriovenous malformation in hereditary hemorrhagic telangectasia. A 13-year-old with a history of recurrent epistaxis presents with brisk oral bleeding. Coronal (**a**) and three-dimensional surface rendering (**b**) CT images demonstrate an extensive AVM of the right face and skull base. Angiography prior to embolization confirms supply from internal (**c**) and external (**d**) carotid arteries.

venule. The current classification places telangectasias as a subtype of CM while acknowledging a source of debate which may be clarified in subsequent iterations.[3] Telangectasias are the hallmark of hereditary hemorrhagic telangectasia, formerly known as Ossler-Weber-Rendu disease, where they may be encountered along with a spectrum of high-flow vascular malformations affecting the central nervous system (Fig. 8), lungs (Fig. 13) and abdominal viscera. Five subtypes are currently recognized, with phenotypes largely influenced by the specific genetic mutations. Diagnosis is facilitated by the Curaçao criteria.[48]

Sturge-Weber Syndrome

Sturge-Weber syndrome is characterized by trigeminal nerve ophthalmic (V1) division and (typically) ipsilateral pial CM with occasional soft tissue overgrowth. Somatic mutations in GNAQ have recently been implicated.[49] Pial CM results in gyriform calcifications of the cortical and subcortical white matter, producing the classic tram-track sign. Ipsilateral calvarial expansion and choroid plexus enlargement are typical.

Additional syndromic associations included in the 2018 ISSVA classification include Sevelle-Martorell syndrome (limb VMs with bone undergrowth), limb CMs with congenital nonprogressive limb overgrowth syndrome, Maffucci syndrome (VMs with spindle-cell hemangioma and enchondroma), Proteus syndrome (CM, VM, and asymmetrical somatic overgrowth), Bannayan-Riley-Ruvalcaba syndrome (AVM, VM, macrocephaly, and lipomatous overgrowth).

Provisionally Unclassified

Lastly, the 2018 ISSVA update captures an expanded list of provisionally unclassified entities. Fibroadipose vascular anomalies (FAVAs) are composed of dense fibrous and adipose tissue with admixed slow-flow (venous or venolymphatic) malformations, typically involving the

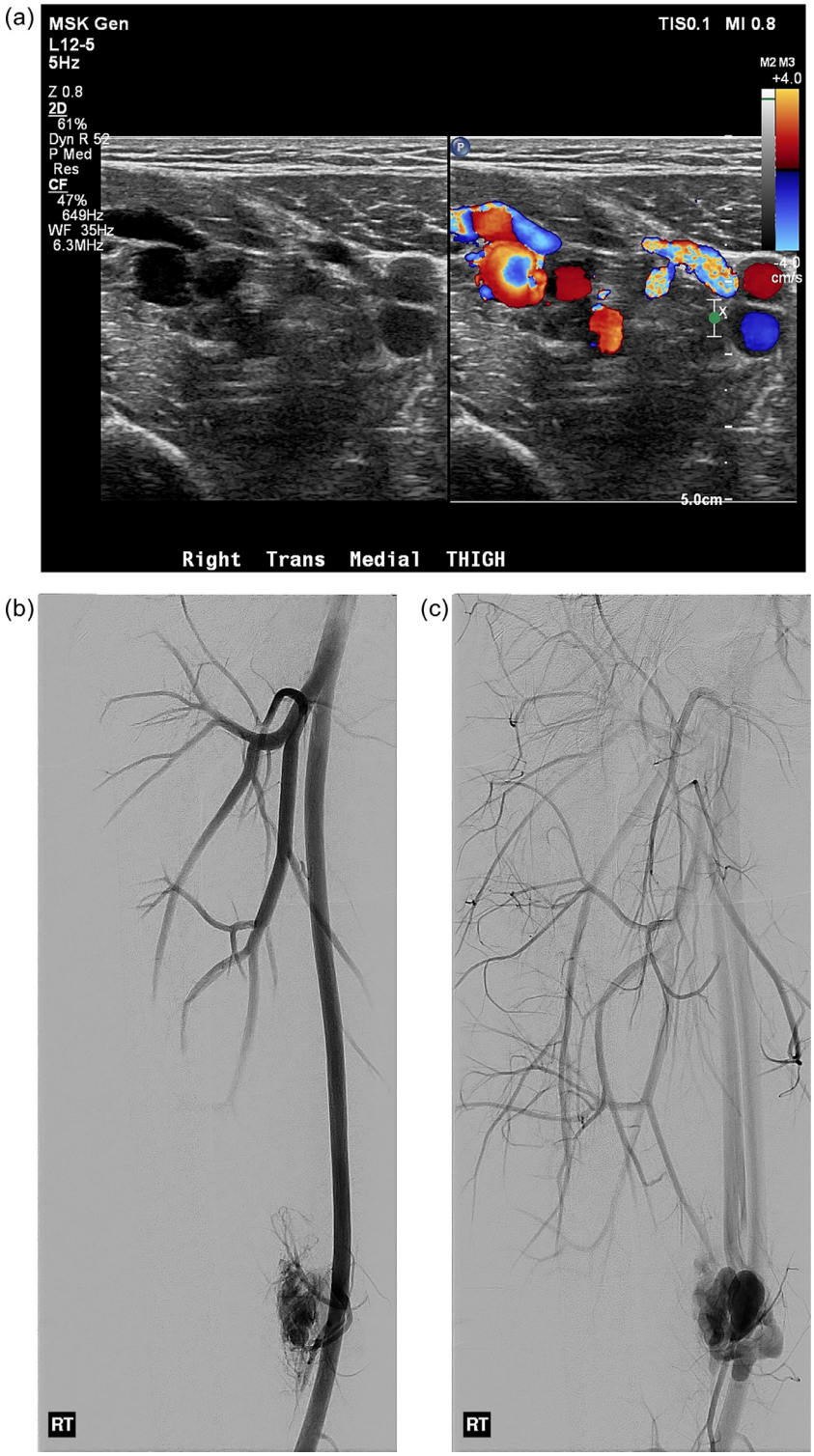

Figure 9 Arteriovenous malformation (sporadic). A 17-year-old male athlete presents with right lower extremity claudication. Ultrasound (**a**) demonstrates ectatic vascular channels with arterialized flow and no appreciable parenchymal component. Early (**b**) and late (**c**) arterial phase digital subtraction images demonstrate an intramuscular nidus supplied by multiple branches of the superficial femoral artery and brisk arteriovenous shunting.

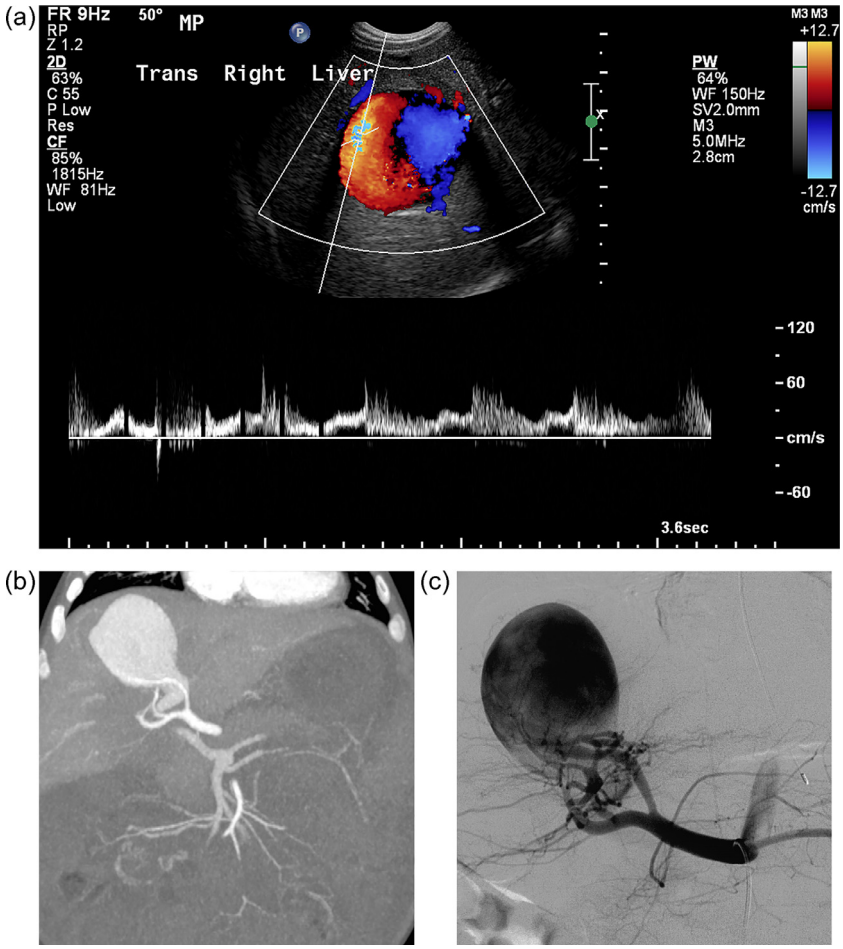

Figure 10 Arterioportal fistula. A 4-month-old girl presents with abdominal distension and failure to thrive. Doppler ultrasound (**a**) demonstrates a large intrahepatic aneurysm within arterialized flow. Coronal maximum intensity projection CT angiogram image (**b**) demonstrates an arterioportal shunting with an associated aneurysm and early mesenteric venous opacification. Digital subtraction celiac arteriogram image (**c**) in preparation for embolization confirms the previous findings.

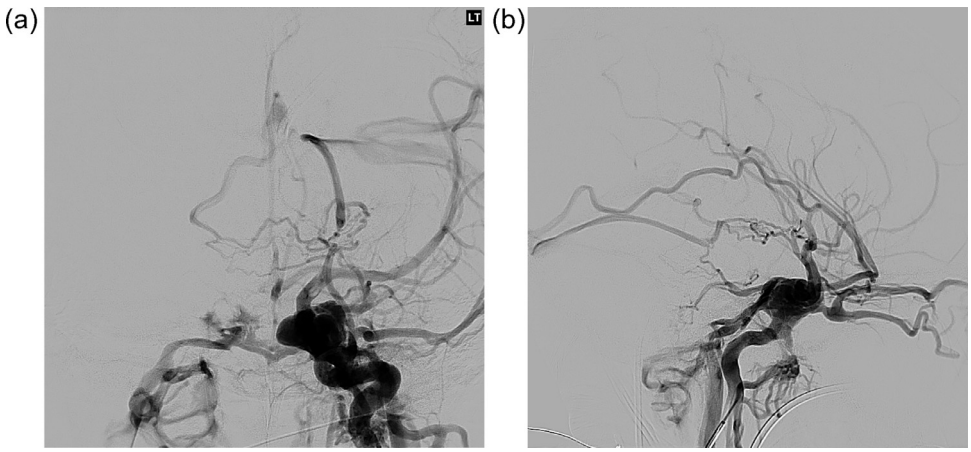

Figure 11 Carotid-cavernous arteriovenous fistula. A 6-year-old boy presents with progressive left eye proptosis 5 months post motor vehicle collision. Arteroposterior (**a**) and lateral (**b**) images from a left internal carotid angiogram demonstrate arterial phase filling of engorged intracranial and orbital veins with poor antegrade filling of the internal carotid artery branches.

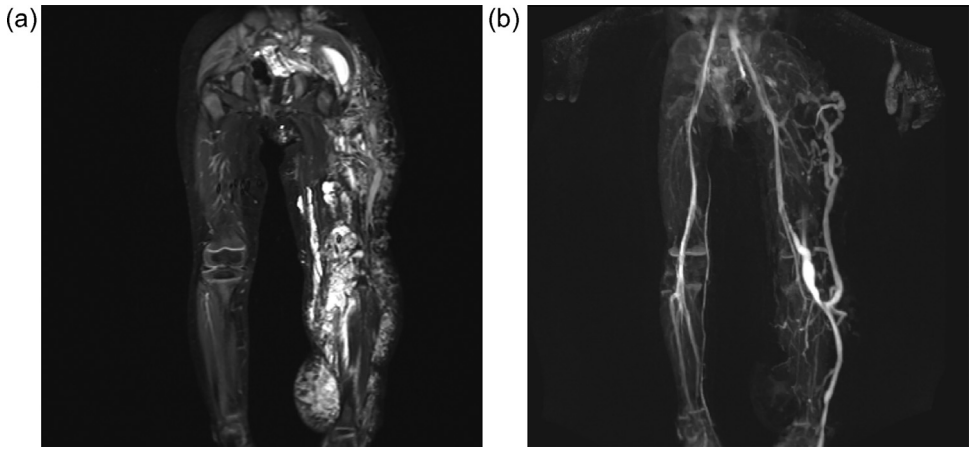

Figure 12 Klippel-Trénaunay syndrome. A 2-year-old girl presents with lifelong left leg enlargement, pain and discoloration. Coronal short tau inversion recover (STIR) MR image (**a**) demonstrates left leg hypertrophy and extensive venolymphatic malformation. Coronal venous phase image from time resolved MR angiography (**b**) demonstrates aneurysmal dilation of the popliteal vein as well as a large lateral marginal vein.

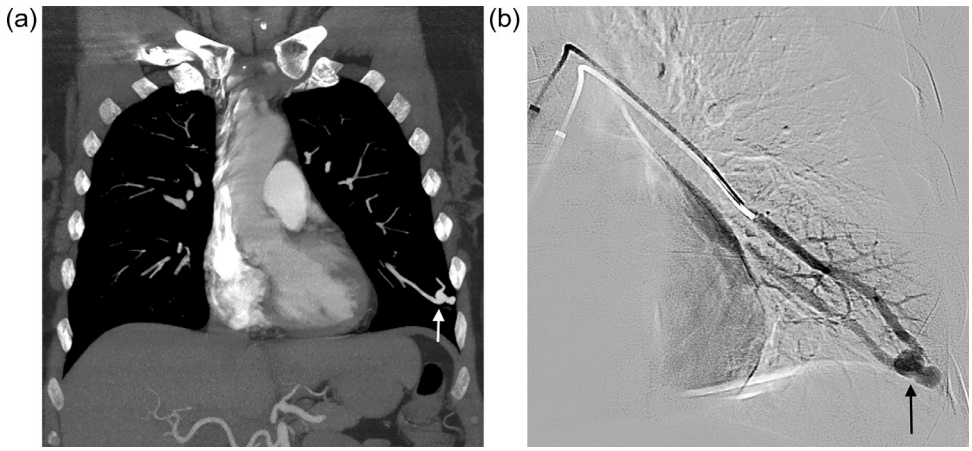

Figure 13 Pulmonary arteriovenous malformation in hereditary hemorrhagic telangectasia. A 50-year-old woman presents with epistaxis and intermittent chest pain. Coronal maximum intensity projection CT image (**a**) demonstrates a pulmonary arteriovenous malformation in the periphery of the left lung base (arrow). Angiography prior to embolization (**b**) confirms brisk shunting from a single segmental pulmonary artery to pulmonary vein through the malformation (arrow).

musculature of the lower extremity or forearm. While some MR features may cause difficulty distinguishing common VMs and FAVAs, FAVAs are less T2 hyperintense and contain conspicuous fatty elements (Fig. 14). Furthermore, US will demonstrate a hyperechoic, noncompressible mass rather than the anechoic or hypechoic and compressible channels of a VM.[50] Occasionally, mistaken for VMs, FAVAs respond poorly to sclerotherapy. While surgical resection may be necessary, cryotherapy has shown promising early results for symptom control.[51] Other entities in this provisionally unclassified category include intramuscular hemangioma, sinusoidal hemangioma, angiokeratoma, and multifocal lymphangioendotheliomatosis with thrombocytopenia/cutaneous angiomatosis with thrombocytopenia.

Conclusion

Standardized nomenclature within the widely accepted ISSVA classification system serves as a foundation for the study of vascular anomalies. Accurate characterization affords expedited diagnosis and appropriate treatment. The 2018 update keeps pace with the rapidly expanding knowledge regarding histopathology and genetic origins of these conditions, serving as a platform for the next era of discovery.

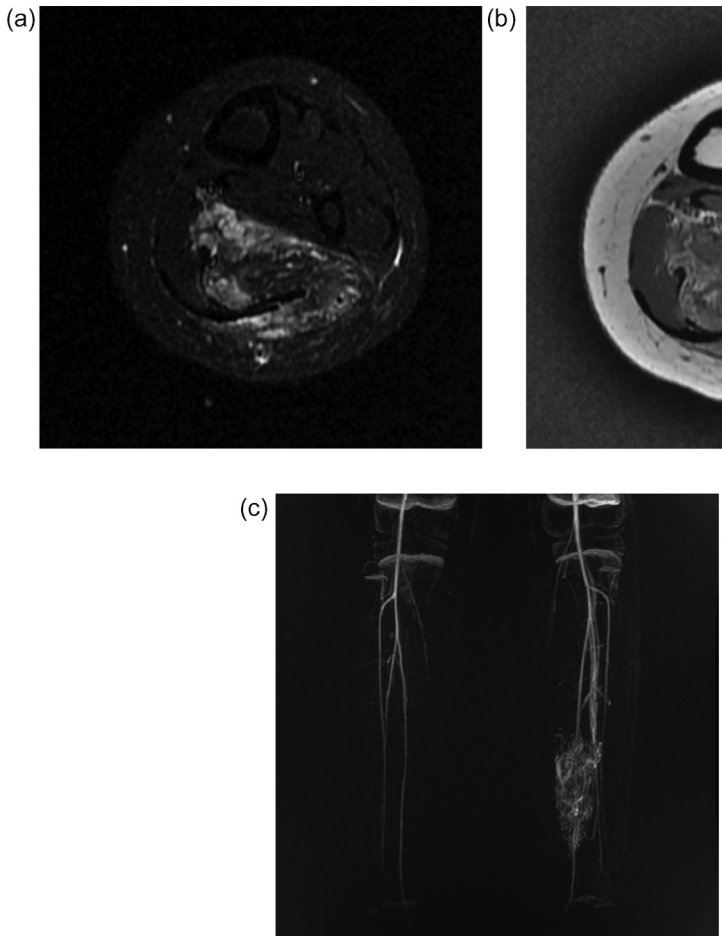

Figure 14 Fibroadipose vascular anomaly. An 11-year-old boy presents with progressive pain and stiffness of the right lower extremity. Axial STIR MR image (**a**) demonstrates a heterogeneously hyperintense lesion centered within the soleus, extending into the lateral gastrocnemius and posteriorly through the fascia into the subcutaneous fat. Axial T1-weighted image (**b**) demonstrates intermixed fatty elements. Coronal time resolved MR angiogram image (**c**) demonstrates moderate patchy enhancement of the lesion with faint early venous drainage.

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