A Step-by-step Sonographic Approach to Vascular Anomalies in the Pediatric Population: A Pictorial Essay

Anushka Mittal[1]   Rama Anand[1]   Richa Gauba[1]   Subhasis Roy Choudhury[2]   Pooja Abbey[1]

[1]Department of Radio-Diagnosis, Lady Hardinge Medical College and Associated Hospitals, New Delhi, India
[2]Department of Pediatric Surgery, Lady Hardinge Medical College and Associated Hospitals, New Delhi, India

Address for correspondence Pooja Abbey, MD, Department of Radio-Diagnosis, Lady Hardinge Medical College and Associated Hospitals, New Delhi - 110 001, India (e-mail: pooja_abbey@yahoo.co.in).

Abstract

Keywords
► arteriovenous malformation
► doppler
► hemangioma
► lymphatic malformation
► vascular anomalies
► vascular malformations
► venous malformation

Vascular anomalies are a common cause of soft-tissue masses in children and often referred for ultrasonographic (USG) evaluation. They are broadly classified as vascular tumors (hemangiomas, hemangioendotheliomas, and angiosarcomas) or vascular malformations (venous malformations, lymphatic malformations, and arteriovenous malformations). Findings on USG and Doppler imaging can be used to categorize vascular anomalies into high- or low-flow lesions, which forms the basis for further workup, diagnosis, and management. On careful evaluation of various sonographic features, in conjunction with clinical findings, an accurate clinicoradiological diagnosis can be made in most cases. Further imaging with magnetic resonance (MR) imaging or computed tomography (CT) helps in delineation of lesion extent, whereas MR or CT angiography is useful to map the vascular supply of high-flow lesions. We have illustrated and discussed a step-by-step approach to diagnose vascular anomalies using ultrasound and Doppler imaging.

Introduction

Vascular anomalies are commonly encountered in the pediatric population. They encompass a variety of disorders, from a simple small birthmark to life-threatening entities, which may cause complications like heart failure, Kasabach–Merritt syndrome, or significant airway narrowing. Traditionally, these anomalies have been diagnosed based on descriptive observations, including clinical appearance, location, and fluid contents. This has led to misdiagnosis and improper management of these lesions.[1,2] The term vascular anomaly represents a broad spectrum of vascular pathology, including vasoproliferative tumors and vascular malformations. Vascular tumors demonstrate rapid growth, increased cellular turnover, and endothelial hypercellularity. Examples include infantile hemangioma, congenital hemangioma, Kaposiform hemangioendothelioma (KH), and angiosarcoma. Vascular malformations are not neoplasms; they result from errors in vascular morphogenesis. They usually grow at a rate proportionate to the growth of the child and comprise vascular spaces with normal endothelium. These include slow-flow vascular malformations (venous, lymphatic malformations [LMs]), high-flow malformations (arteriovenous malformations [AVMs]), and various combined malformations.

In 1996, the International Society for the Study of Vascular Anomalies (ISSVA) released a classification system for vascular anomalies, which was updated in 2014 and revised in 2018.[3] The ISSVA classification (►**Table 1**) is widely accepted and used among clinicians managing vascular anomalies. It continues to evolve with our evolving knowledge of the biology and genetic

Table 1 International Society for the Study of Vascular anomalies (ISSVA) classification of vascular anomalies[3]

Tumors	Benign	Infantile hemangioma, congenital hemangioma, tufted angioma, spindle cell hemangioma, epithelioid hemangioma, pyogenic granuloma, etc.
	Locally aggressive	Kaposiform hemangioendothelioma, retiform hemangioendothelioma, papillary intralymphatic angioendothelioma, Dabska tumor, composite hemangioendothelioma, Kaposi sarcoma, etc.
	Malignant	Angiosarcoma, epithelioid hemangioendothelioma, others
Malformations	Simple	Capillary (low flow) port wine stain, telangiectasia, cutis marmorata telangiectatica congenita, nevus simplex, etc.
		Lymphatic (low flow) common (cystic) LM, GLA, channel type LM, primary lymphedema (different types), etc. Venous (low flow) common VM, familial VMCM, blue rubber bleb (bean) syndrome VM, GVM, CCM, etc. Arteriovenous malformation (high flow): sporadic, in HHT, others Arteriovenous fistula (congenital) (high flow): sporadic, in HHT, others
	Combined	CVM, CLM, CAVM, LVM, CLVM, CLAVM, CVAVM, CLVAVM (C: capillary; V: venous; L: lymphatic; AV: arteriovenous)
	Of major named vessels	Affects lymphatics, veins, arteries Anomalies of origin, course, number, length, diameter, persistence (of embryonal vessel)
	Associated with other anomalies	KTS, PWS, Sturge–Weber syndrome, Maffucci syndrome, Proteus syndrome, etc.
Provisionally unclassified vascular anomalies		Intramuscular hemangioma, angiokeratoma, sinusoidal hemangioma, acral arteriovenous "tumor," multifocal lymphangioendotheliomatosis with MLT/CT, PTEN (type) hamartoma of soft tissue/"angiomatosis" of soft tissue (PHOST), FAVA

Abbreviations: CCM, cerebral cavernous malformation; FAVA, fibroadipose vascular anomaly; GLA, generalized lymphatic anomaly; GVM, glomuvenous malformation; HHT, hereditary hemorrhagic telangiectasia; KTS, Klippel–Trenaunay syndrome; LMs, lymphatic malformations; MLT/CT, multifocal lymphangioendotheliomatosis with thrombocytopenia/cutaneovisceral angiomatosis with thrombocytopenia; PTEN, phosphate and tensin homolog; PWS, Parkes–Weber syndrome; VMCM, venous malformation cutaneomucosal.

basis of vascular anomalies. The 1996 classification had fundamentally divided vascular lesions into vascular tumors and vascular malformations (malformations could be simple or combined; combined malformations are defined as more than two malformations in one lesion). The 2014 updated classification expanded the sections on vascular tumors and vascular malformations. Tumors were divided into three categories: benign, locally aggressive/borderline, and malignant. Vascular malformations, apart from simple or combined, were categorized as malformations of major named vessels and those associated with other anomalies like Klippel–Trenaunay syndrome, Sturge–Weber syndrome, etc.

With greater knowledge of the genetic basis of vasoproliferative lesions, the 2018 revised ISSVA classification included a list of causative genes (as an appendix).[3] Most notable among these is the *PIK3CA* gene on chromosome 3, which is associated with several syndromes collectively called the *PIK3CA*-related overgrowth spectrum (PROS). A few vascular lesions—like fibroadipose vascular anomaly (FAVA)—still remain unclassified, because it is unclear whether they are tumors or malformations, or because their clinicopathological characteristics are as yet incompletely understood.

Vascular anomalies are further divided into "high-flow" or "low-flow" groups, based on their flow dynamics. High-flow lesions contain an arterial component. Examples include hemangiomas (infantile and congenital), other vascular tumors, AVMs, and arteriovenous fistulas (AVFs). Low-flow lesions are all other lesions that do not contain an arterial component, including capillary malformations, LMs, venous malformations (VMs), and also involuting hemangiomas.[4]

Sonography (along with Doppler imaging) is the first-line imaging modality employed for diagnosis of vascular anomalies. Usually, patients with these lesions are encountered in a busy routine ultrasound practice, sometimes even in emergencies, where a quick and accurate diagnosis is needed. A simple algorithmic approach on the basis of sonographic findings, in conjunction with clinical features, helps in making a confident diagnosis of these lesions, even by less experienced radiologists. Further imaging using magnetic resonance imaging (MRI) or computed tomography (CT) is helpful in assessment of lesion extent and vascular supply.

Clinical Features

The importance of detailed history and examination in a case of vascular anomaly cannot be emphasized enough, since they form the basis for differentiation. Lesions like superficial hemangiomas often do not need any imaging, being diagnosed solely on the basis of their age of onset, clinical appearance, and growth pattern.

These anomalies may involve any part of the body, most commonly the head and neck, followed by the extremities. Symptoms may range from skin discoloration, swelling, pain, and bleeding to systemic effects like high-output cardiac failure (which may be seen in high-flow lesions). The age of onset and pattern of growth (and involution) help diagnose infantile and congenital hemangiomas and differentiate them from vascular malformations.[5-15]

On examination, deep-seated lesions may not produce any discoloration in the overlying skin. Superficial vascular anomalies are seen in the form of bluish/violet/pink/red-colored lesions with varying morphologies. The important clinical features of common vascular anomalies have been summarized in ►**Table 2**.

Table 2 Clinical features of common vascular anomalies

	Age of onset	Growth	Involution (spontaneous)	Morphology and clinical examination
Infantile hemangioma	2–6 wk after birth	Rapid till 18 mo	Usually by 9 y	Superficial lesion-bright red
Congenital hemangioma	At birth	Variable	RICH by 1 y NICH—no involution PICH—partial involution	NICH—pink/violet with central telangiectasias
Vascular malformations				
High-flow (AVM)	Variable, usually discovered in childhood	In proportion to the growth of child	Absent	Bruit and thrill over the lesion or a pulsatile lesion
Low-flow VM				Superficial lesion: blue/purple, fluctuant, compressible
Low-flow LM				Soft lesions showing transillumination

Abbreviations: LM, lymphatic malformation; NICH, noninvoluting congenital hemangioma; PICH, partially involuting congenital hemangioma; RICH, rapidly involuting congenital hemangioma; VM, venous malformation.

Role of Various Imaging Modalities

Imaging is indicated in vascular anomalies to (1) confirm the clinical diagnosis; (2) characterize the lesion as high or low flow, and further diagnose the type of lesion; (3) know the complete extent of infiltrative, deep-seated, or widespread lesions; and (4) know the vascular supply in high-flow lesions where endovascular management is planned.

Ultrasonography

Ultrasonography (USG) is the first-line imaging modality, with various advantages including lack of ionizing radiation, no need of sedation in small children, cost-effectiveness, portability, and wide availability. Most importantly, grayscale sonography along with color and spectral Doppler can accurately diagnose most of the cases, when interpreted in conjunction with clinical features. The categorization of vascular anomalies as high- or low-flow lesions is best achieved using Doppler-USG. Disadvantages include operator-dependence and inability to adequately assess deep lesions.[16,17]

Contrast-Enhanced Ultrasound

Contrast-enhanced ultrasound combines all the advantages of USG with the ability to evaluate the microvasculature of the lesion and quantify perfusion parameters using a time-intensity curve analysis. Various parameters like time to peak and area under the curve can be studied in different regions of interest in the center and at the periphery of the lesion, and comparison of pretreatment and posttreatment values has a role in monitoring response to therapy.[18]

Magnetic Resonance Imaging

MRI is very frequently the next step after USG for assessment of the extent of a deep-seated or large vascular anomaly, its relation with surrounding vital structures, and the tissue planes involved. Magnetic resonance (MR) angiography provides information about the vascular supply in high-flow lesions. The need for sedation, long scan times, high cost, and limited availability are some disadvantages of MRI.

Computed Tomography

The presence of ionizing radiation, lack of soft-tissue contrast, and contrast-related side effects are some important drawbacks of CT. It is performed in select cases where an urgent evaluation needs to be done, for example, in evaluation of airway compromise caused by a lesion, and when MRI is not available or the child cannot be sedated for MRI.[19]

Step-by-Step Doppler-USG Approach to the Diagnosis of Vascular Anomalies

A step-wise approach to sonographic diagnosis of vascular anomalies has been summarized in ►**Fig. 1**.

First step: Assess the presence/absence of arterial flow
The presence of arterial flow suggests a high-flow lesion, whereas low-flow lesions do not show arterial flow.

- When arterial flow is detected within the lesion on color Doppler imaging, the following features should be evaluated:
- *Vessel density:* A semiquantitative assessment of vessel density is possible using color Doppler imaging.[17,20] The area of greatest vascularity should be identified within the lesion, and the number of vessels counted in an area of 1 cm^2. Less than 2 vessels/cm^2 is described as low vessel density, 2 to 4 vessels/cm^2 as moderate, and ≥5 vessels/cm^2 as high vessel density. High vessel density is typically seen in "high-flow" vascular anomalies, like hemangiomas, other vascular tumors, and high-flow vascular malformations (AVMs, AVFs).
- *Peak arterial systolic velocity:* Peak systolic velocity (PSV) of the arterial vessels within the lesion should be measured, after appropriate angle correction. High values of arterial PSV are seen in high-flow lesions. A study by Paltiel et al found no significant difference in mean arterial PSV values between hemangiomas and AVMs.[20]
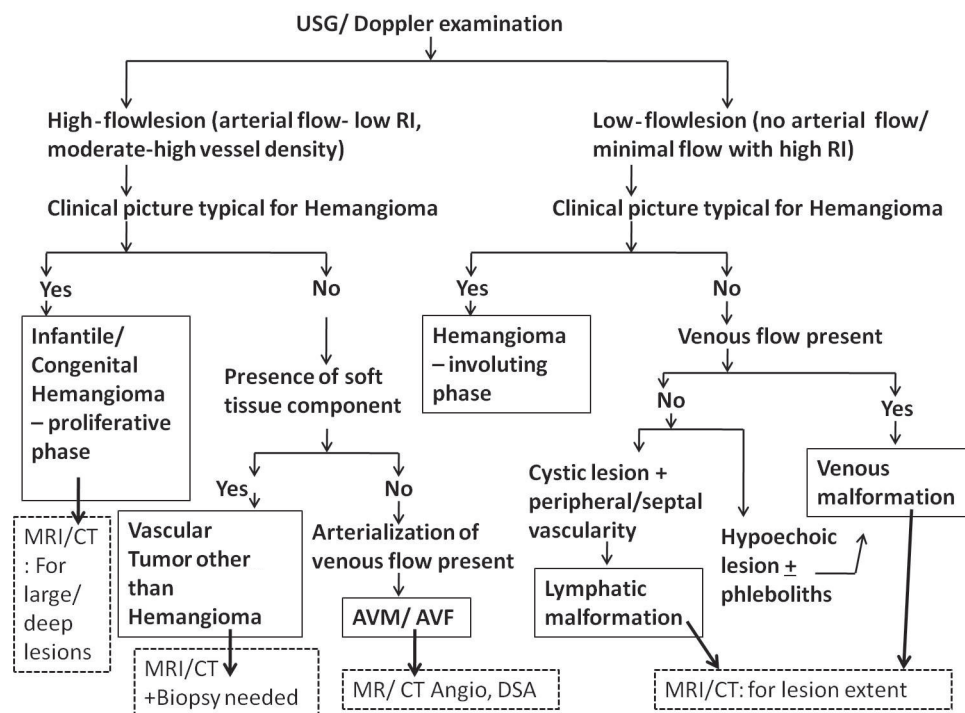
Fig. 1 A step-by-step ultrasonographic (USG) Doppler approach to the diagnosis of vascular anomalies. This is a simplified approach. Combined malformations and those associated with other anomalies/syndromes would show a combination of/additional findings (AVF, arteriovenous fistula; AVM, arteriovenous malformation; CT, computed tomography; DSA, digital subtraction angiography; MRI, magnetic resonance imaging; RI, resistive index).

- *Resistive index:* The arteries within high-flow vascular anomalies show low-resistance flow, with low values of resistive index (RI). In some instances, a single or a few areas of arterial flow may also be seen within low-flow vascular malformations—for instance, venous and LM often infiltrate around normal structures, and a few arterial vessels may be seen passing through these lesions. Also, LMs may show areas of arterial flow within the septa/walls of their constituent cysts. These foci of arterial flow in low-flow lesions show high RI values, in contrast to arteries within high-flow lesions.

Imaging Approach if Arterial Flow Is Present (High-Flow Lesion)

Second Step: Is the Lesion Typical for Hemangioma or Not

As the name suggests, congenital hemangiomas are present at birth, whereas infantile hemangiomas develop soon after birth. An initial phase of rapid growth followed by involution is typically seen in infantile hemangiomas or rapidly involuting congenital hemangiomas (RICH).

On clinical examination, superficial hemangiomas are characteristic in appearance—red/blue/violet in color without any bruit/thrill/transilluminance.

Hemangiomas, which are generally high flow in nature (►Fig. 2), may not show arterial flow in their involuting phase/posttreatment (►Fig. 3). Such hemangiomas are, thus, low-flow lesions.

Third Step: Assess the Presence of Soft-tissue Component within the High-flow Lesion

Intralesional soft tissue is seen in vascular tumors, whereas lesions without a soft-tissue component are high-flow malformations (AVM/AVF). Presence of soft-tissue mass is the most reliable predictor for differentiating hemangiomas (or other vascular tumors) from AVMs.[20]

The most common vascular tumors are infantile hemangiomas. Lesions with atypical imaging/clinical findings may be vascular tumors other than hemangiomas. These usually require biopsy for diagnosis.

Fourth Step: Does the Lesion Show Arterialization of Venous Flow and Low-Resistance Arterial Waveform?

Such a lesion, which lacks a soft-tissue component, is likely to be an AVM/AVF. The term arterialization of venous flow refers to high venous peak velocities seen within these lesions. Mean venous peak velocities in AVMs are significantly higher than those seen in hemangiomas and VMs.[20]

Imaging Findings of Various High-Flow Lesions

Vascular Tumors
Infantile Hemangioma

Infantile hemangiomas are the most common vascular tumors, may be single or multiple, and are seen in ~4 to 10% of infants.[21] They express the glucose transporter isoform 1 (GLUT1) protein,[6] which is not expressed by any other normal
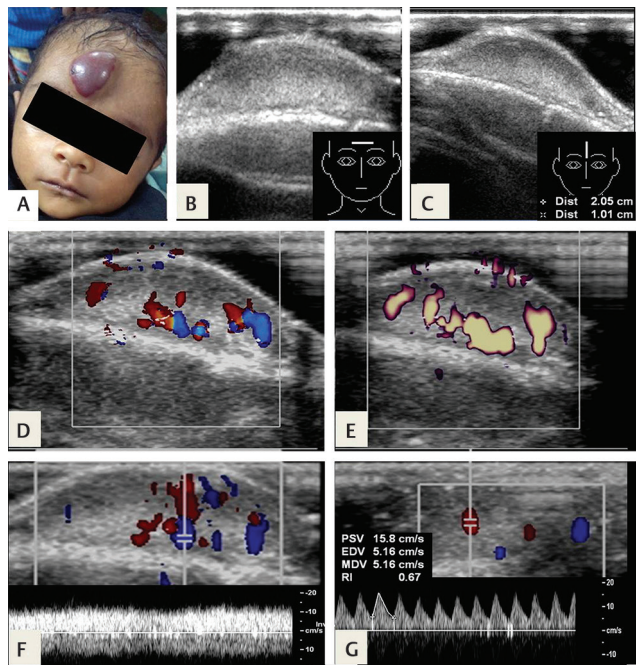
Fig. 2 (**A**) Infantile hemangioma in a 2-month-old infant with forehead swelling since age of 15 days. Ultrasonographic (USG) gray scale (**B** and **C**—using standoff pad) shows a hyperechoic lesion in the subcutaneous plane, (**D,E**) with vascularity on color and power Doppler. The lesion shows both (**F**) venous and (**G**) arterial waveforms.
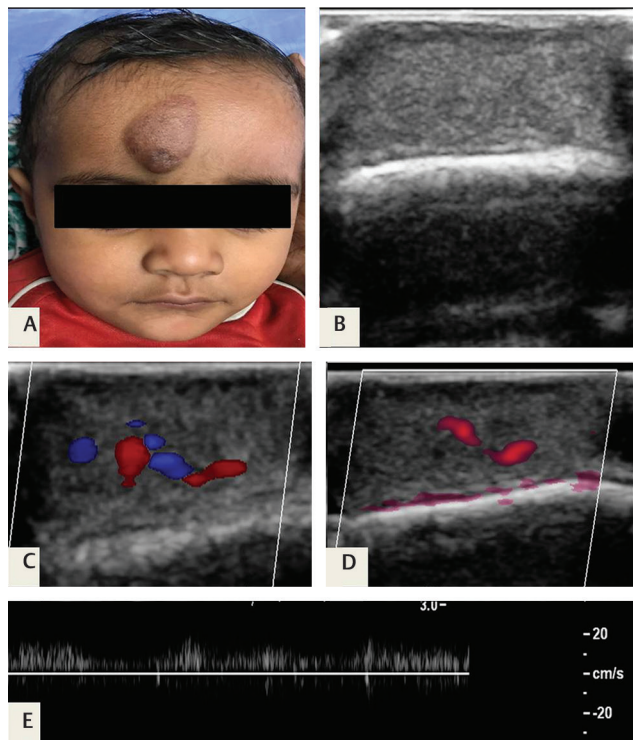

Fig. 3 Same case as Fig. 2. (**A**) There is reduction in size and discoloration of the hemangioma after propranolol therapy. (**B–D**) Follow-up Ultrasonography (USG) after 6 months shows significant reduction in the vascularity, (**E**) with only venous flow.

tissue or vascular tumor. Skin is the most common location, with multiple cutaneous hemangiomas in 10 to 25% cases. These lesions are most commonly located in the subcutaneous

plane and may infiltrate into the deeper tissue planes when large. Liver is the most common extracutaneous site (►**Fig. 4**).[22]

The typical Doppler-USG appearance is of a well-circumscribed, homogenous, hypo-/hyperechoic lesion with internal arterial as well as venous flow on color Doppler (►**Fig. 5**). In the proliferative phase, hemangiomas typically show a high vessel density (>5 vessels/cm^2) and high Doppler shift (>2 kHz).[17] Involuting lesions may show mixed echogenicity due to the intermixing of fat and may not show arterial flow within. Small, superficial lesions with typical clinical as well as USG features do not usually require further imaging, unless there is suspicion of associated visceral involvement. Uncommonly, large hemangiomas may be infiltrative in appearance (►**Fig. 6**). Such lesions require further imaging evaluation using MRI/CT. On MRI, hemangiomas typically show multiple flow voids and intense enhancement on postcontrast images. In large facial hemangiomas, MRI of the brain is indicated to rule out PHACES syndrome (P—posterior fossa malformations, H—hemangiomas, A—arterial anomalies, C—coarctation of aorta and cardiac anomalies, E—eye/ocular anomalies, S—sternal defects). Although the majority of hemangiomas are self-limiting, treatment is required in the presence of complications like heart failure or Kasabach–Merritt syndrome.

Congenital Hemangioma

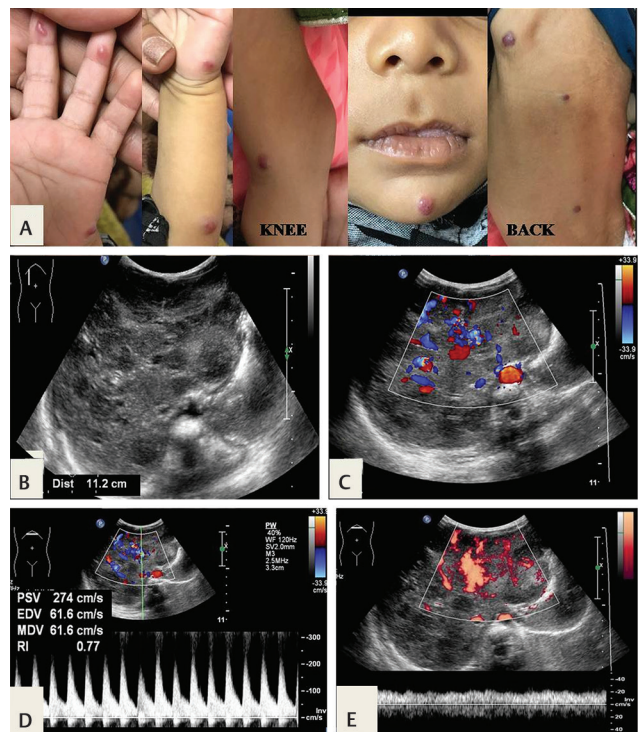Congenital hemangiomas are much less common than infantile hemangiomas, usually solitary, and test negative for

Fig. 4 (**A**) A 3-month-old infant with multiple cutaneous hemangiomas over the palm, elbow, back, left leg, chin. The patient was evaluated for abdominal distension. (**B**) Ultrasonography (USG) revealed hepatomegaly with (**C–E**) multiple hypoechoic liver lesions showing marked internal vascularity, consistent with infantile hepatic hemangioma.
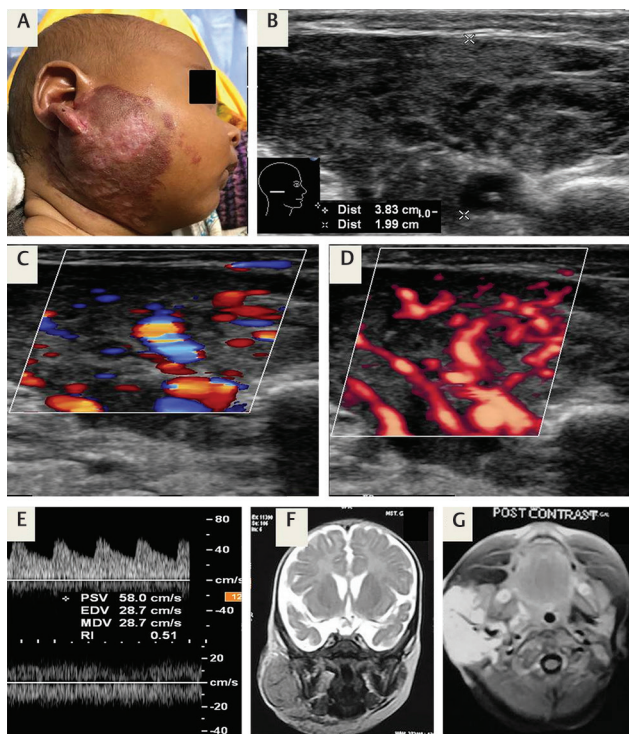
Fig. 5 Parotid gland hemangioma in (**A**) 2-month-old infant. (**B**) Grayscale ultrasonography (USG) showed an enlarged, heteroechoic right parotid gland, (**C,D**) with increased internal vascularity showing (**E**) both arterial and venous flow. (**F,G**) Magnetic resonance imaging (MRI) showed an enhancing lesion limited to the parotid gland, with no intracranial abnormality.

Fig. 6 (**A**) A large, infiltrative facial hemangioma in a 6-month-old infant, which developed soon after birth and grew to involve the entire left face. (**B,C**) Ultrasonography (USG) shows an (**D,F**) infiltrative, hyperechoic high-flow lesion. (**G,H**) Axial and (**I**) coronal T2-weighted (T2W) magnetic resonance (MR) images confirm an infiltrative lesion with multiple flow voids (*arrow*), involving the left parotid gland, buccal, and masticator spaces, (**J**) with intense enhancement.

the immunohistochemical marker GLUT1. They are of two major types—RICH and noninvoluting congenital hemangiomas (NICH). Imaging findings are broadly similar to those of infantile hemangiomas.[8,9] Presence of calcification, which is not a feature of infantile hemangiomas, may be seen in congenital hemangiomas (17% NICH, 37.5% RICH vs. none in infantile hemangiomas).[23] A partially involuting congenital hemangioma has also been recently described, which shows initial rapid involution like RICH, but does not completely involute and persists instead, like a NICH.[24]

Pyogenic Granuloma/Lobular Capillary Hemangioma

These are common, benign vascular tumors diagnosed clinically in most cases. They are seen as small growths, usually blood red in color, with a "minced meat"–like surface. However, they have an increased tendency to bleed, which may sometimes make it difficult to differentiate them from AVMs. In such cases, imaging may help by demonstrating a well-defined, homogenous, vascular mass (►**Fig. 7**), without the flow voids or the nidus typical of an AVM.[25] Final diagnosis is established on histopathology.

Solitary Fibrous Tumor/Hemangiopericytoma

A solitary fibrous tumor (previously referred to as a hemangiopericytoma) is a mesenchymal tumor, extremely rare in children. Most tumors are benign, but some may show variable malignant potential. On USG, these are solid or rarely cystic masses, well demarcated, with homogeneous or heterogeneous echogenicity. On Doppler, the presence
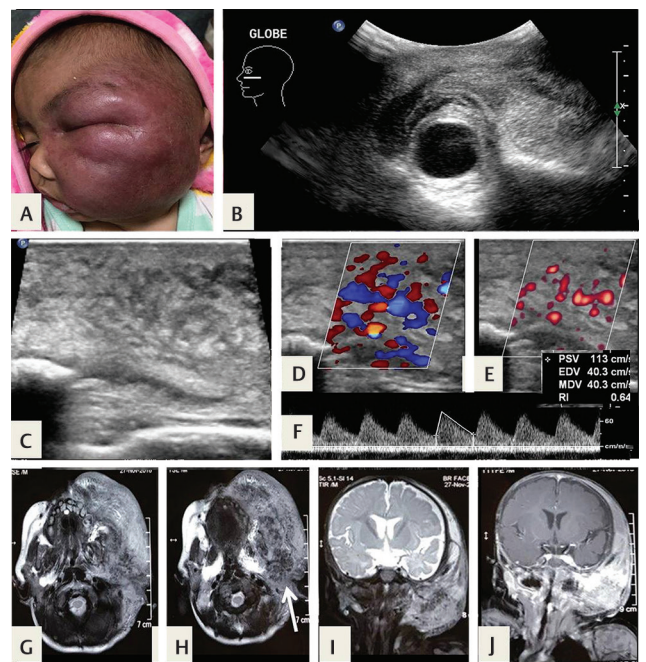
of intratumoral arteriovenous shunting can be diagnosed based on the presence of low-resistance flow in the feeding arteries, arterialization of the venous flow, and large internal vessels with low vascular impedance (►**Fig. 8**). Presence of soft tissue helps differentiate from an AVM.[26-28] Further imaging with MRI/CT may demonstrate a locally aggressive soft-tissue lesion with marked contrast enhancement (►**Fig. 8**). Biopsy is needed for diagnosis.

Kaposiform Hemangioendothelioma

KH is a rare, locally aggressive, vasoproliferative tumor, which has a low malignant potential and usually presents shortly after birth. It may be seen in the head and neck, trunk, extremities, retroperitoneum, and rarely in other locations. KH has both vascular and lymphatic components and may be associated with Kasabach–Merritt phenomenon, a term that describes profound thrombocytopenia caused by platelet sequestration resulting from a consumptive coagulopathy.[29]

The imaging appearance of various vasoproliferative tumors overlaps with each other. The diagnosis of hemangiomas can be usually made in the presence of typical clinical features. If the history, clinical examination, and/or Doppler-USG findings are atypical, then further imaging is indicated. If a vascular tumor other than hemangioma is suspected, biopsy is required for diagnosis.

Vascular Malformations: AVM/AVF

An AVM typically shows an abnormal cluster of arterial channels communicating with venous channels via a nidus. An AVF is similar to an AVM except for the lack of a nidus.
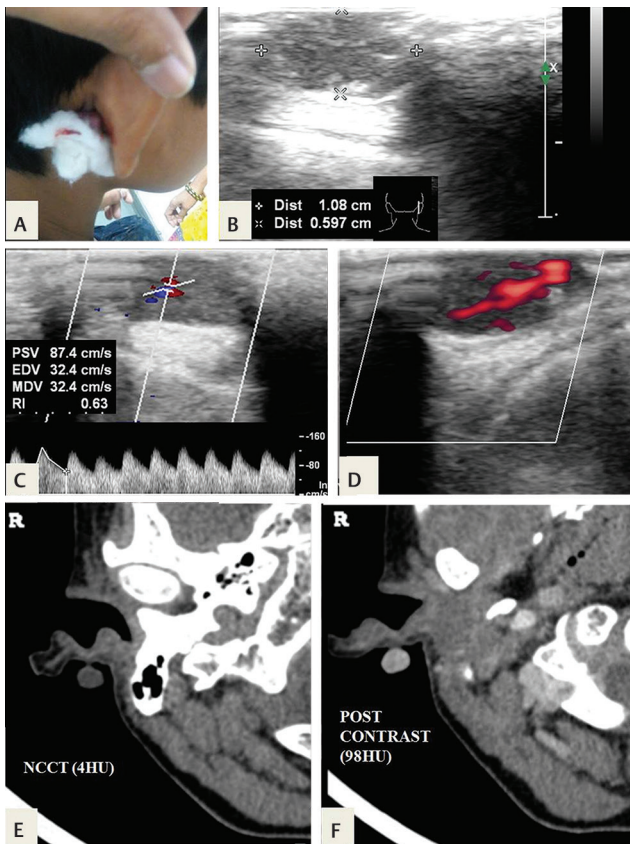
Fig. 7 (**A**) A pyogenic granuloma in a 13-year-old adolescent boy with bleeding from a swelling behind the ear. (**B**) Ultrasonography (USG) shows a circumscribed hypoechoic soft-tissue lesion with internal arterial vascularity. (**E,F**) Computed tomography (CT) shows intense enhancement, and absence of nidus. Possibility of a vascular tumor was given; final diagnosis made on excision biopsy.

Histologically, AVMs and AVFs consist of dysplastic arteries that drain into arterialized veins, bypassing capillary beds. Unlike other high-flow lesions, no soft-tissue component or lesion matrix is seen in these lesions. Typically, most AVMs are congenital, whereas most AVFs are acquired. A congenital AVF may be sporadic or occur as a part of hereditary hemorrhagic telangiectasia.[3]

Doppler-USG demonstrates low-resistance flow in the involved arteries, arterialization of flow in the venous system, and high peak velocities in both the arterial and venous channels (►**Fig. 9**). It may show the site of arteriovenous communication in an AVF (►**Fig. 10**). CT or MR angiography helps by demonstration of the vascular anatomy of the lesion (►**Fig. 11**), which helps in planning its management.[30]

Imaging Approach if Arterial Flow Is Absent (Low-Flow Lesion)

Second Step: Is the Lesion Typical for Hemangioma or Not?
It is important to remember that hemangiomas may not show arterial flow in the involuting phase, or on treatment. These lesions will have a typical history and should not be misdiagnosed as VMs. In typical cases, no further imaging is necessary and the patient can be managed conservatively.
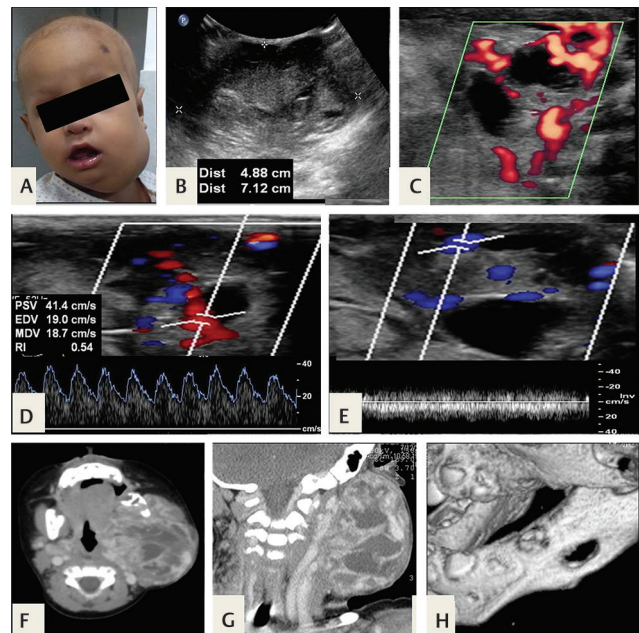

Fig. 8 Biopsy-proven solitary fibrous tumor/ hemangiopericytoma (**A**) A 4-month-old infant presented with a rapidly growing swelling of recent onset, not typical of hemangioma. (**B,C**) Ultrasonography (USG) shows a large soft-tissue lesion with cystic areas, internal vascularity, (**D,E**) both arterial and venous flow. (**F,G**) Contrast-enhanced computed tomography (CECT) confirms a vascular soft-tissue lesion with (**H**) associated lytic lesion in left hemimandible, suggesting a locally aggressive tumor.

Third Step: Look for the Presence or Absence of Venous Flow
If venous flow is seen, the lesion is a VM. However, not all VMs will show flow on Doppler-USG. In case no flow is seen, the lesion may be either a lymphatic or a VM (with very slow flow or thrombosis). Approximately 16% of all VMs show no detectable flow on Doppler-USG.[31]

Fourth Step: Look for Phleboliths
Calcified phleboliths, if seen, are highly specific for the diagnosis of VMs, even in the absence of flow (►**Fig. 12**). Phleboliths are seen in ~16% of all VMs.[29]

Fifth Step: Is the Lesion Cystic with Peripheral Septal Vascularity?
LMs are cystic, usually multilocular cystic lesions, lack phleboliths, and show no flow or only peripheral/septal flow. Internal echoes may be seen. VMs, on the other hand, are often hypoechoic and not multicystic; they may comprise venous spaces or ectatic/dysplastic veins.

Imaging Findings of Various Low-Flow Lesions

Common low-flow lesions are vascular malformations including capillary, venous, and LMs, and their combinations. Capillary malformations typically involve superficial layers of skin—like the facial malformations seen in Sturge–Weber syndrome and are clinically diagnosed. Venous, lymphatic, and venolymphatic malformations are the most common
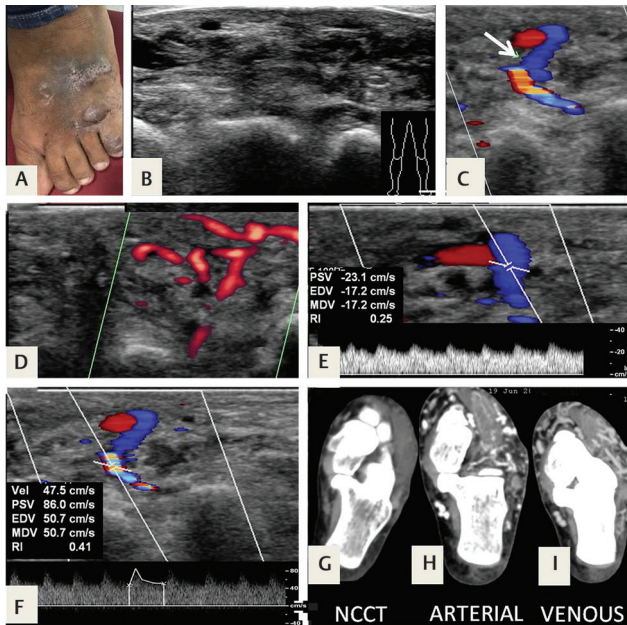
Fig. 9 (**A**) An arteriovenous malformation (AVM) in an 18-year-old man, who presented with a vascular swelling over his foot. (**B–D**) Ultrasonography (USG) showed multiple vascular channels with both arterial and venous flow. (**E**) There was arterialization of venous flow, whereas the arteries showed low-resistance flow with (**F**) a resistive index (RI) of 0.41. (**G–I**) Computed tomography (CT) angiography showed multiple enlarged vessels, and early filling of venous channels in the arterial phase.
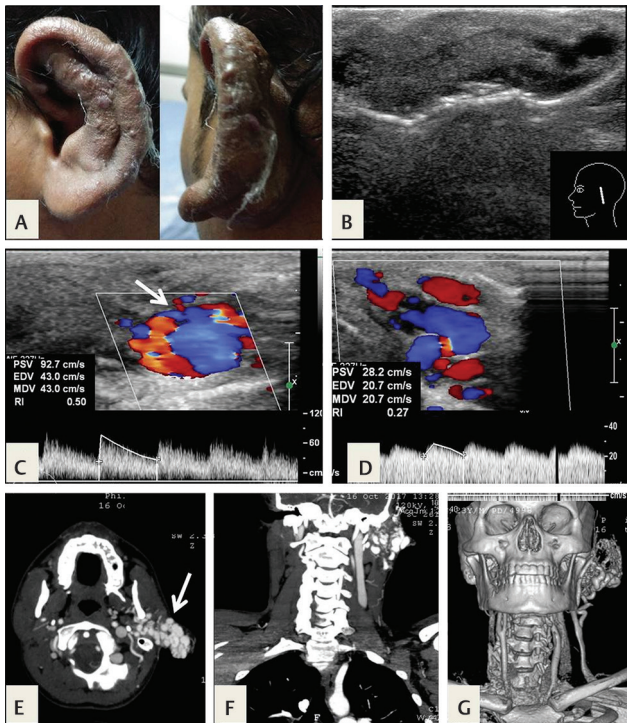

Fig. 10 Intrahepatic arterioportal fistula in a 6-month-old infant with Down's syndrome and abdominal distension. (**A,B**) Ultrasonography (USG) shows an aneurysmally dilated left portal vein, with an abnormal intrahepatic arterioportal communication (*arrow* in **B**). (**C,D**) Color Doppler shows turbulent intralesional flow. Spectral Doppler confirms (**E**) a high-velocity, low-resistance flow in the feeding artery and (**F**) chaotic, arterialized flow in the dilated vein.

Fig. 11 (**A**) An arteriovenous malformation (AVM) of the pinna in a 16-year-old adolescent boy who had a swollen pinna showing multiple dilated vascular channels over it. (**B–D**) Ultrasonography (USG) showed multiple dilated vessels showing arterialization of venous flow, diagnostic of AVM. (**E–G**) Computed tomography (CT) angiography well depicted the tangle of vessels, with arterial supply from branches of the left external carotid artery. The patient was referred for embolization.
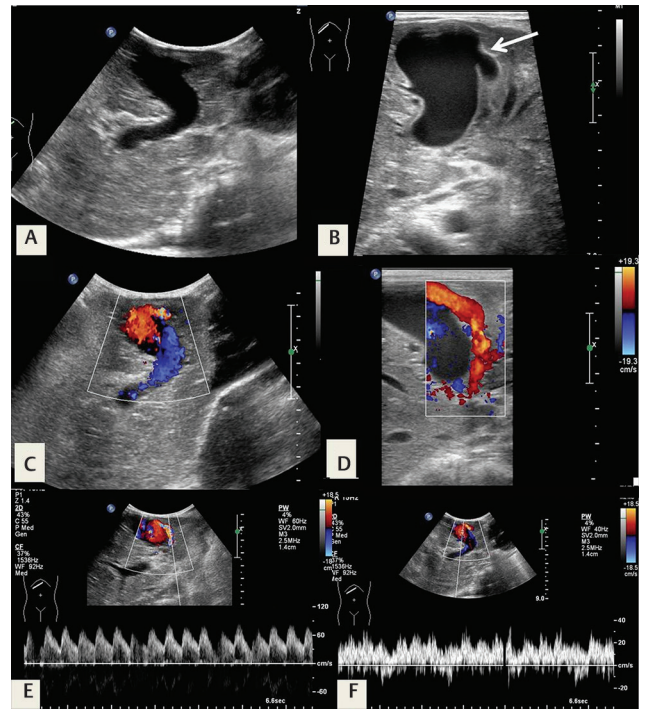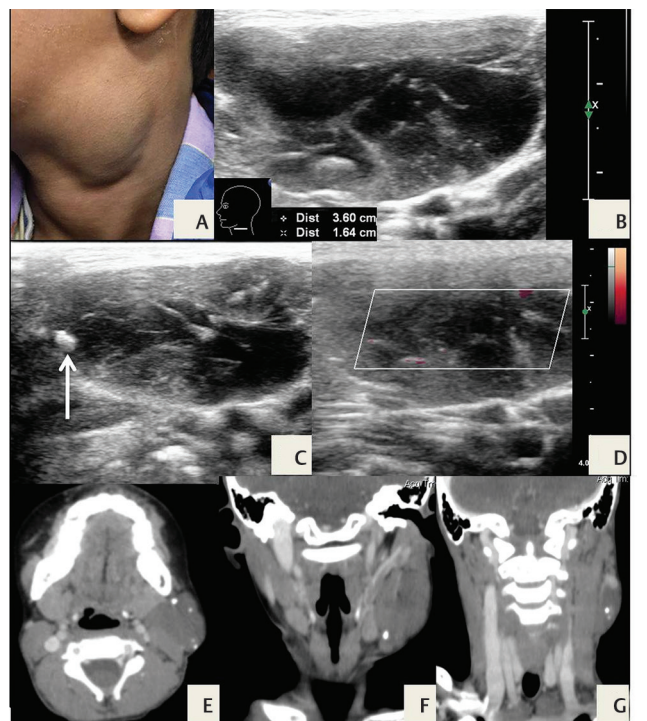

Fig. 12 (**A**) A venous malformation in an 11-year-old boy with a long-standing neck swelling. Ultrasonography (USG) reveals (**B**) a circumscribed lesion with cystic and hypoechoic areas and (**C**) a phlebolith, but (**D**) no flow even on power Doppler. (**E–G**) Computed tomography (CT) confirms intralesional phleboliths with only minimal enhancement.

types of vascular malformations; their prevalence is ~1% in the general population.[4]

Simple Low-flow Vascular Malformations

Venous Malformations

VMs are usually present at birth but may not be apparent. They grow in proportion to the growth of the child. They are composed of vascular channels, sometimes containing intraluminal thrombi, lined by thin endothelium. Based on the pattern of venous drainage, these may be of different types—isolated malformations without venous drainage; those draining into normal veins, or into dysplastic veins; or malformations comprising primarily of venous ectasia.[19]

On Doppler-USG, VMs are often hypoechoic, may be infiltrative lesions involving multiple tissue planes (►**Fig. 13**), or may be well marginated (►**Fig. 14**). They may comprise venous spaces or ectatic/dysplastic veins and show venous flow or no flow. Phleboliths (well defined, round, calcific foci), if seen, are characteristic (►**Fig. 15**). These represent areas of spontaneous thrombosis.

MRI is the preferred examination to evaluate the extent of large or infiltrative lesions. Fat-suppressed T2-weighted or short tau inversion recovery images can depict the lesions, which are hyperintense, well. On postcontrast CT or MRI, VMs have a wide range of contrast enhancement patterns: from homogeneous to heterogeneous, faint to vivid, and rapid to delayed.[13-34] Phleboliths within the lesion may be missed on MRI and are best depicted on CT images.

Lymphatic Malformations

LMs comprise dilated lymphatic channels, forming cystlike structures, isolated from the normal lymphatic system. They are congenital, and the majority (80–90%) of these lesions are discovered by the age of 2 years. They are subclassified on the basis of the size of the lymphatic spaces into macrocystic (cysts >1 cm in diameter), microcystic (cysts <1 cm in diameter), and combined (►**Fig. 16**).[20,35,36] They commonly involve the subcutaneous tissues, and most often occur in the head and neck region. They are often trans-spatial and appear infiltrative, involving multiple tissue planes (►**Fig. 17**).
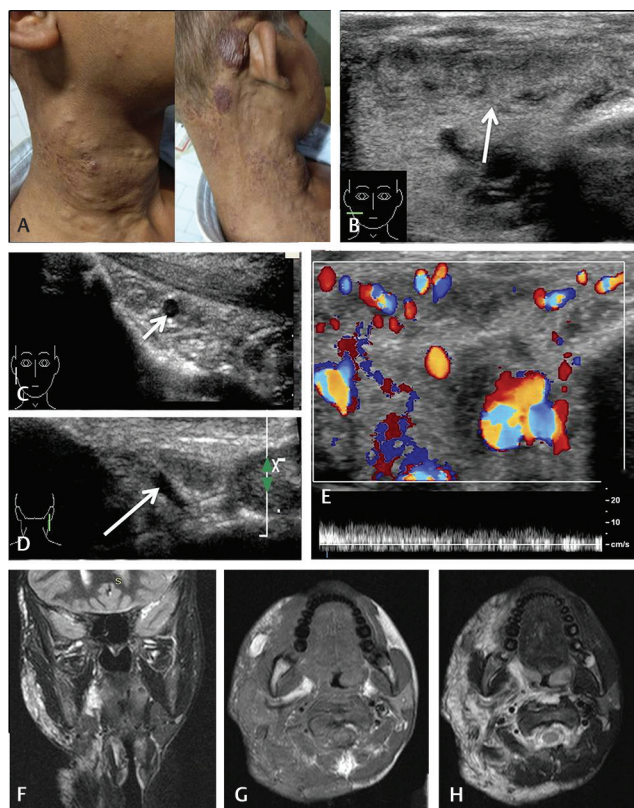

Fig. 13 (**A**) An infiltrative venous malformation of the neck in a 12-year-old boy. (**B–D**) Ultrasonography (USG) showed an infiltrative lesion with multiple abnormal vessels showing (**E**) venous flow. (**F**) Magnetic resonance imaging (MRI) coronal short tau inversion recovery (STIR) and (**G,H**) axial T1-weighted (T1W) and fat-saturated T2W images clearly demonstrated an infiltrative lesion involving the right buccal, parotid, masticator, carotid, prevertebral, and perivertebral spaces.
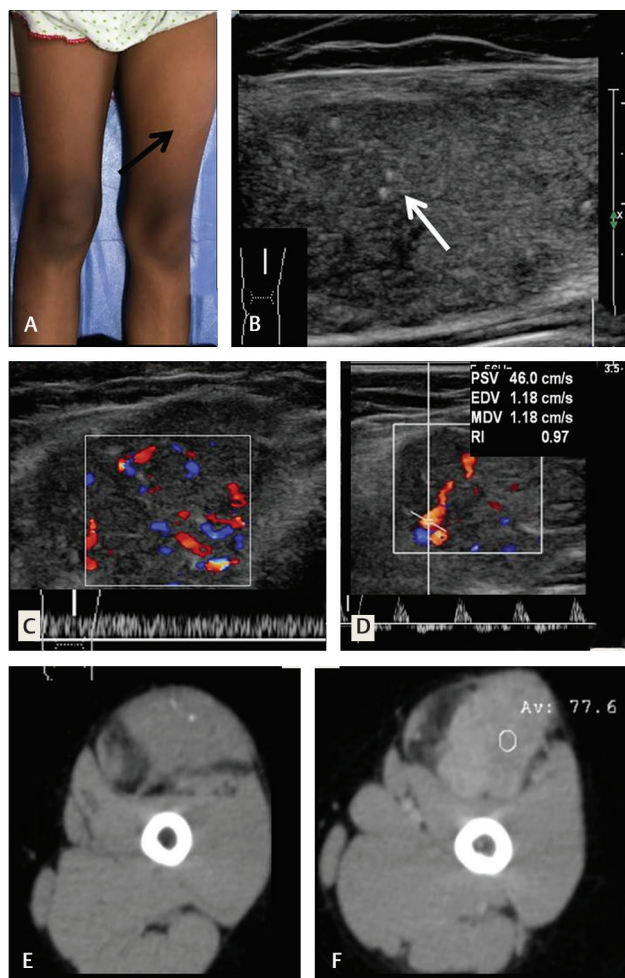

Fig. 14 (**A**) Intramuscular venous malformation of the left thigh in a 6-year-old girl. Ultrasonography (USG) shows a well-circumscribed lesion with (**B**) phleboliths and (**C**) venous flow. (**D**) An arterial branch is seen within the lesion, with high resistive index (RI) and biphasic flow, suggestive of a muscular arterial branch. Computed tomography (CT) confirms (**E**) phleboliths (non–contrast-enhanced CT [NCCT]) and (**F**) enhancement within the lesion.
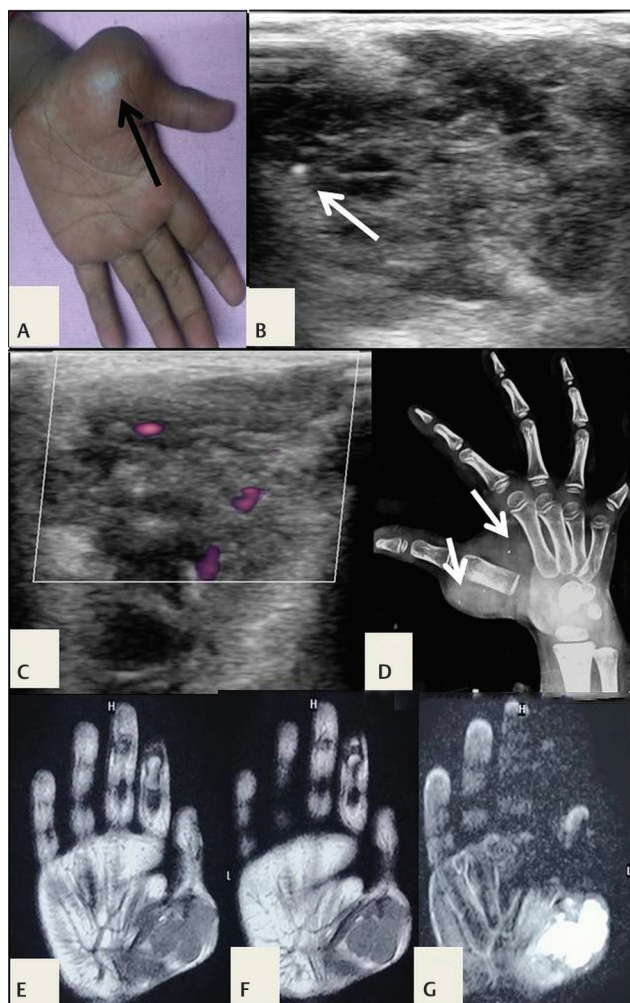
Fig. 15 (**A**) A venous malformation along the thenar eminence of palm in a 4-year-old boy. Ultrasonography (USG) showed (**B**) a phlebolith and (**C**) low flow within. (**D**) Radiograph also confirmed intralesional phleboliths. (**E,F**) Magnetic resonance imaging (MRI) coronal T1-weighted (T1W) and (**G**) short tau inversion recovery (STIR) images well depicted the lesion extent.

LMs can show periods of sudden growth, in response to immunological stimuli like the common cold, or in the presence of hemorrhage or infection. On USG, LMs appear as multilocular cystic masses with increased through-transmission. They may show low-level internal floating echoes (►**Fig. 18**) or a fluid–fluid level representing chyle, blood, or pus. On Doppler study, no internal vascularity is noted (►**Fig. 19**); however, peripheral vascularity may be visible within the fibrous septations (►**Fig. 18**) and capsule.[20,35,36] If arterial flow is seen within the septa or cyst wall, it shows high-resistance flow and high RI values.

MRI is the preferred imaging modality for further evaluation of the extent of the lesion. These lesions are typically hyperintense on T2-weighted images and may show variable signal intensity on T1-weighted images, depending on cyst content. CT may be useful if MRI cannot be done. No enhancement is seen within LMs (►**Fig. 20**), and this is very useful to differentiate them from VMs.
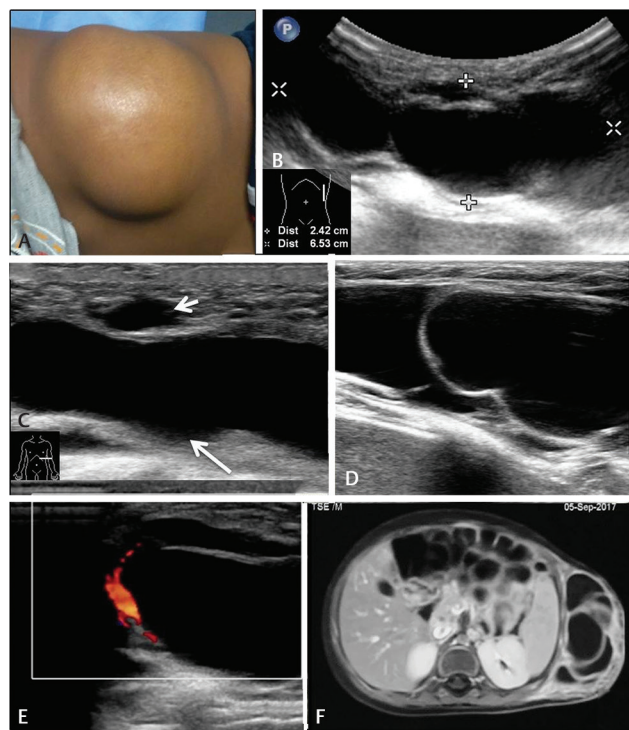

Fig. 16 (**A**) Lymphatic malformation of the abdominal wall in a 9-month-old infant. (**B–D**) Ultrasonography (USG) revealed a well-defined lesion with multiple macrocysts (*large arrow*) and few microcysts (*small arrow*) and (**E**) septal flow. (**F**) Contrast-enhanced magnetic resonance imaging (MRI) also showed peripheral and septal enhancement.
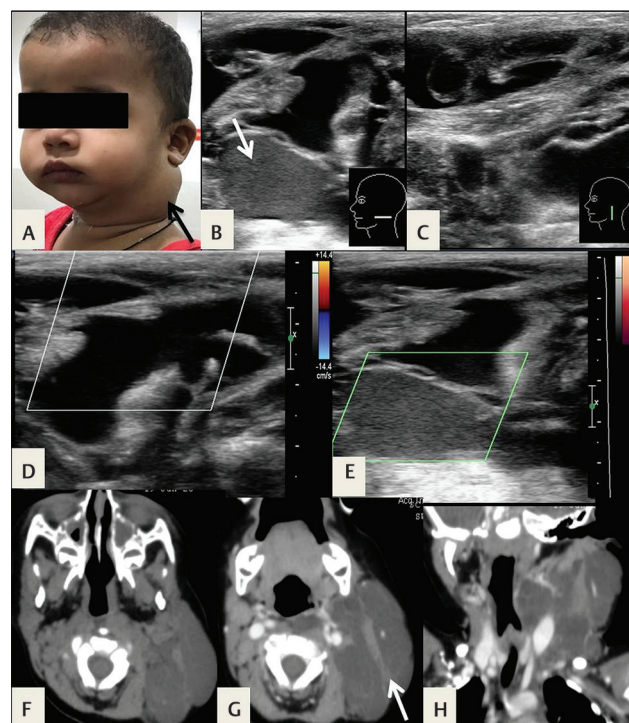

Fig. 17 (**A**) Multispatial lymphatic malformation of the neck in an 11-month-old infant. (**B,C**) Ultrasonography (USG) showed a multicystic lesion with internal echoes and (**D,E**) no color flow. (**F-H**) Computed tomography (CT) depicted the extent of the lesion both anterior and posterior to the left sternocleidomastoid muscle (arrow), with a component in the prevertebral space.
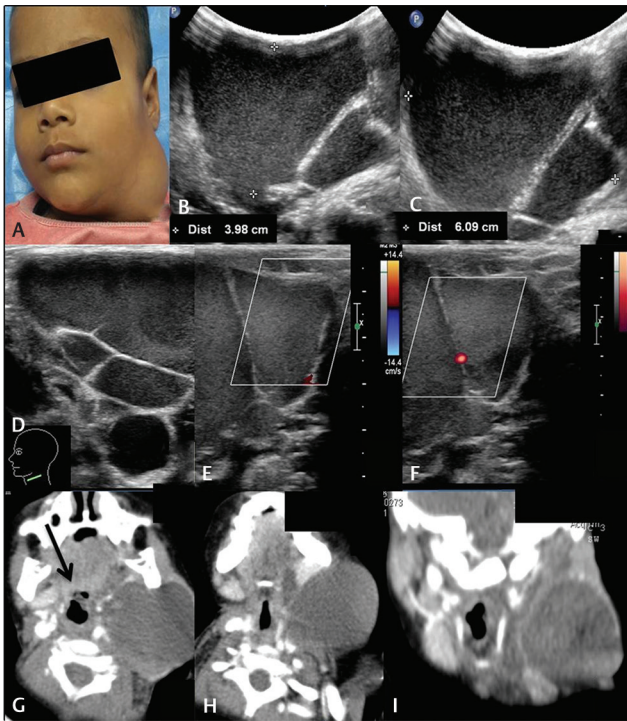
Fig. 18 (**A**) Lymphatic malformation in a 3-year-old girl with neck swelling. (**B–D**) Ultrasonography (USG) showed multiple cystic areas with (**E,F**) internal echoes and septal flow. (**G–I**) Computed tomography (CT) better showed the extent, with a small component in the preepiglottic space (**G**) and the left submandibular space.
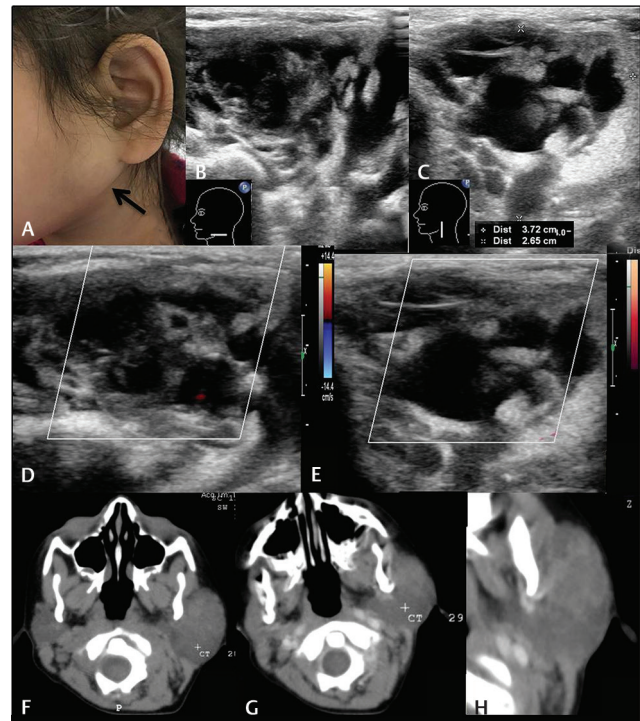

Fig. 20 (**A**) Lymphatic malformation of the left parotid in a 3-year-old girl. (**B,C**) Ultrasonography (USG) revealed multiple cystic areas within the parotid, (**D,E**) with no internal vascularity. (**F–H**) Computed tomography (CT) confirmed a nonenhancing lesion confined to the parotid gland.

Complex Lymphatic Anomalies

Rare systemic lymphatic anomalies that can involve multiple organs and have significant morbidity and mortality include generalized lymphatic anomaly, Kaposiform lymphangiomatosis, Gorham–Stout disease, and central conducting lymphatic anomaly (channel type LM, lymphangiectasia).[37] USG has a limited role in evaluation of these lesions, and MRI or CT evaluation is indicated, depending on the site(s) of involvement.

Combined Vascular Malformations

Combined vascular malformations comprise two or more malformations in a single lesion. They may occur in sporadic or syndrome forms and are named by hyphenation of the names of the types of component tissues—for example, CLM for capillary lymphatic malformations. Combined venolymphatic malformations may occur, which show imaging features of both VMs and LMs (►**Fig. 21**).

Malformations of Major Named Vessels

The malformations of major named vessels are also known as the "channel-type" or "truncal" vascular malformations and include a wide range of anomalies of origin, course, number, and diameter of vessels as well as residual embryonal vessels.[3]
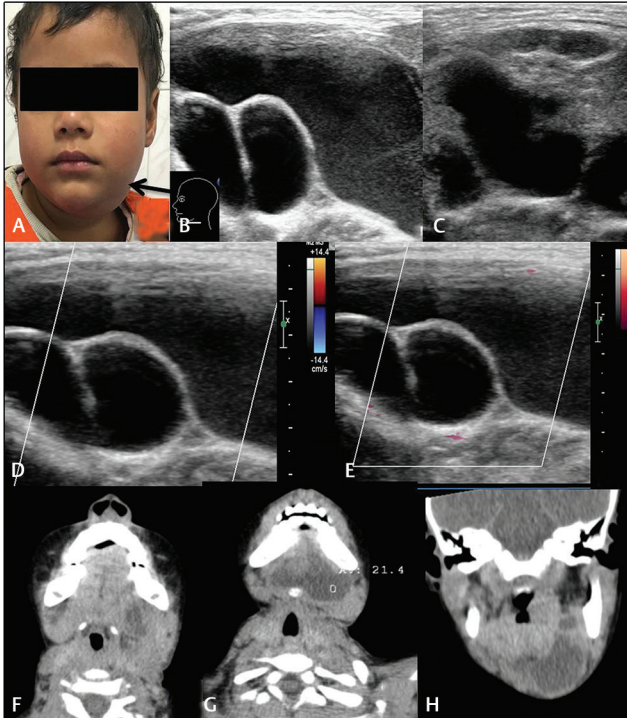

Fig. 19 Same patient in Fig. 18. (**A**) on postsclerotherapy follow-up, the patient showed reduction in the size of the lesion clinically. (**B–E**) The same was observed on ultrasonography (USG) as well. (**F–H**) Computed tomography (CT) revealed interval enlargement of the deeper aspect of the lesion, in the submental and bilateral submandibular spaces (left > right).
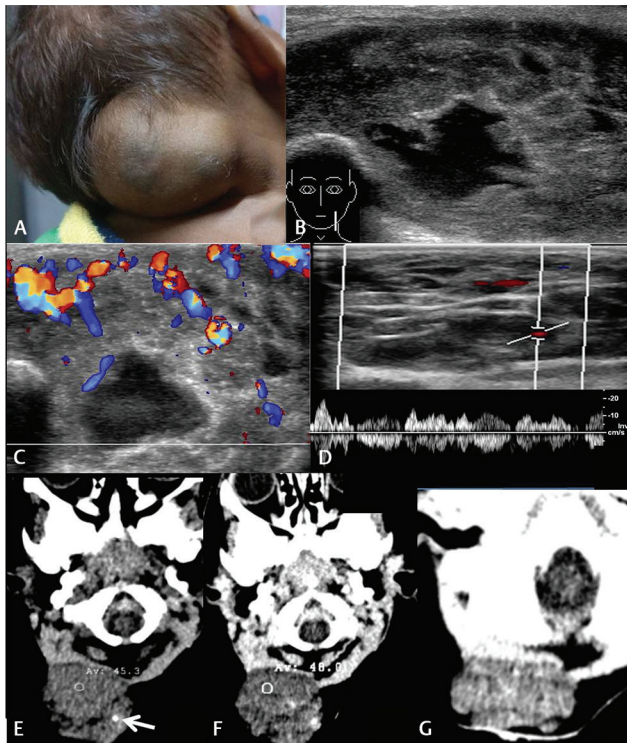
Fig. 21 (**A**) Venolymphatic malformation over the nape of the neck in a 6-month-old infant. (**B**) Ultrasonography (USG) revealed a part-hypoechoic and part-cystic lesion, (**C,D**) showing venous flow in its superficial portion and no flow in the deeper, cystic component. (**E–G**) Computed tomography (CT) confirmed the nonenhancing deeper component, whereas the superficial portion showed enhancement and phleboliths, suggestive of a venolymphatic malformation.

Vascular Malformations Associated with Other Anomalies

PIK3CA-Related Overgrowth Spectrum

The *PIK3CA* gene on chromosome 3 is important for growth regulation and mutations in this gene are associated with several cancers. This gene is also implicated in a spectrum of overgrowth syndromes/phenotypes, many of which have vascular anomalies as a part. PROS lesions include congenital lipomatous overgrowth, vascular malformations, epidermal nevi, scoliosis/skeletal and spinal syndrome, and CLAPO syndrome (which stands for—Capillary malformations of the lower lip, cervicofacial LM, Asymmetry and Partial or generalized Overgrowth), fibroadipose hyperplasia or overgrowth, macrodactyly, and Klippel–Trenaunay syndrome.[3,38]

Klippel–Trenaunay Syndrome

Klippel–Trenaunay syndrome classically comprises a triad of capillary malformations, VMs, and soft-tissue/bone overgrowth, most commonly involving (one) lower limb. A wide spectrum of venous anomalies may be present, including ectasia, aplasia/hypoplasia of deep veins, and VMs involving the superficial and deep venous systems. LMs may also be present. USG is useful as an initial investigation, whereas MRI is helpful for delineating the extent of abnormality (►**Fig. 22**).
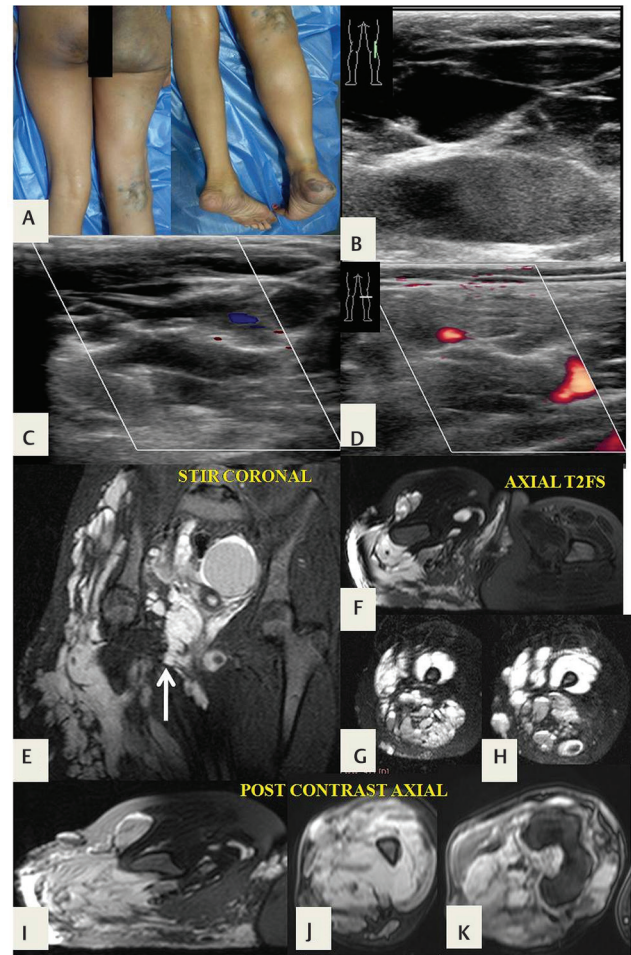

Fig. 22 A 14-year-old girl with Klippel–Trenaunay syndrome. (**A**) The right lower limb is enlarged, with port wine stains and varicose veins. (**B–D**) Ultrasonographic (USG) Doppler shows multiple dilated vascular channels, with slow flow. (**E**) Short tau inversion recovery (STIR) coronal and (**F–H**) fat-suppressed axial T2-weighted magnetic resonance (MR) images demonstrate soft-tissue hypertrophy and extensive venous malformations involving the limb muscles, with right gluteal and intrapelvic component (*arrow* in **E**). These lesions are T2 hyperintense and (**I–K**) show enhancement on delayed postcontrast fat-suppressed T1 axial images.

Provisionally Unclassified Vascular Anomalies: Fibroadipose Vascular Anomaly

The 2018 ISSVA classification has expanded the list of provisionally unclassified entities. FAVA is a recently described, complex vascular malformation that has certain typical clinical and imaging findings.[39]

Adolescent females are most often affected and present with constant pain. FAVA lesions occur within the muscle, most often of the lower limb (gastrocnemius, soleus). On imaging, they may be focal mass–like, focal infiltrative, or diffuse infiltrative. Variable amounts of fat and fibrous tissue are present within the lesion. Phlebectasia is a characteristic imaging finding (►**Fig. 23**) that may be seen on USG, MRI, or venography. On USG, presence of an echogenic soft-tissue component helps differentiate FAVA from VM. Macroscopic
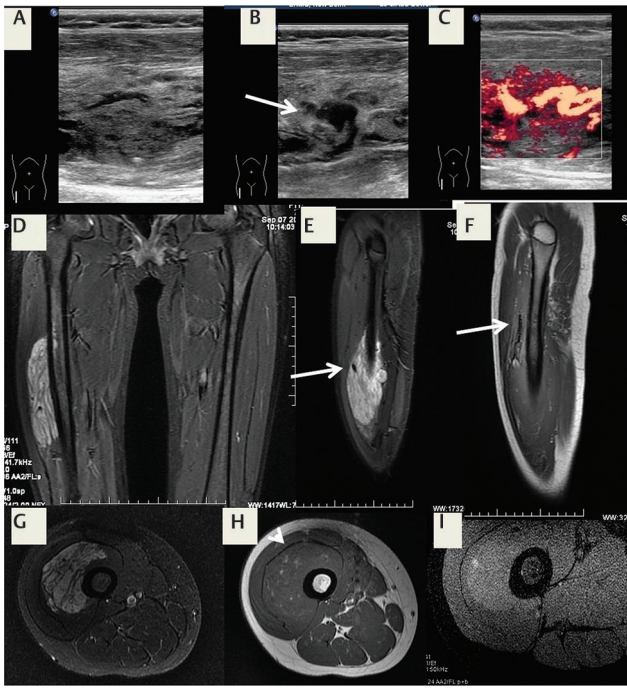
Fig. 23 Suspected fibroadipose vascular anomaly (FAVA) in an 11-year-old girl with pain. (**A**) Ultrasonography (USG) reveals a heterogeneous intramuscular lesion with (**B**) a dilated intralesional vein and (**C**) venous channels. (**D**) Magnetic resonance imaging (MRI) short tau inversion recovery (STIR) coronal image shows the hyperintense lesion; the dilated vein is seen on (**E**) sagittal STIR and (**F**) T1 images. The lesion shows mild hyperintensity on axial (**G**) T2-weighted fat-suppressed image, (**H**) areas of hyperintensity on T1, and (**I**) mild contrast enhancement. The patient is awaiting surgical resection.

Fig. 24 (**A**) Plexiform neurofibroma presenting as a facial swelling in a 5-year-old boy with family history of neurofibromatosis type I. (**B,C**) Ultrasonography (USG) shows a lobulated, serpiginous, trans-spatial hypoechoic lesion partly within and partly surrounding the right parotid gland, (**D**) without significant intralesional vascularity. (**E**) The normal right submandibular and (**F**) left parotid glands are shown for comparison.

venous channels and dilated veins may be seen. On MRI, hyperintense areas are seen on T1-weighted images (due to intralesional fat), and these lesions are less hyperintense on T2-weighted images compared with conventional VMs. Postcontrast enhancement is seen.

FAVA lesions may be mistaken for VMs on imaging, but it is important to diagnose them correctly as the management is different. They respond poorly to sclerotherapy and may require surgical resection or ablative/cryotherapy for pain relief.

Management of Various Vascular Anomalies

The management of most vascular anomalies requires a multidisciplinary approach and close cooperation between clinicians and radiologists. Increasingly, interventional radiology techniques have an important role to play in the management of these lesions.[40] Uncomplicated infantile hemangiomas may be managed conservatively or with oral propranolol, whereas complicated lesions can be managed surgically with laser treatment or with resection.[41,42] RICH presenting with acute cardiac failure may require emergency embolization to reduce the risk of arteriovenous shunting. NICH, usually small, do not require treatment but can be surgically treated. In most cases of congenital hemangiomas, conservative management with a "watch-and-wait" approach is
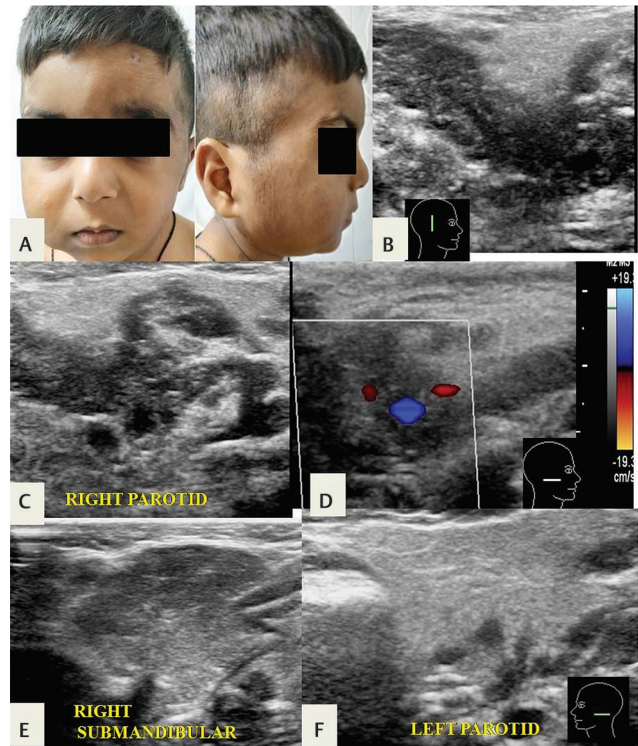
adopted. However, cases complicated by hemorrhage, ulceration, airway obstruction or ophthalmic involvement, heart failure, and those causing severe psychological distress or disfigurement warrant further discussion and consideration for treatment. Most common treatment for solitary fibrous tumor/hemangiopericytoma is wide surgical excision, whereas for AVM, it is embolization focused on obliteration of the nidus, which can be achieved percutaneously, via an endovascular approach, or surgically.[11] The mainstay of treatment for VMs or LMs is percutaneous sclerotherapy, or surgery when feasible.

Pitfalls in USG Evaluation/Diagnosis of Vascular Anomalies

- There are several pitfalls that must be kept in mind while using sonography and Doppler imaging for the evaluation of vascular anomalies. Some of these are the following:
- Optimal Doppler settings are essential for the evaluation of any vascular anomaly. For evaluation of low-flow lesions, it is imperative to utilize low-flow settings on color Doppler. Power Doppler imaging is also a valuable tool, as it has greater flow sensitivity. Sometimes, presence of low flow may be seen as moving echoes on real-time grayscale USG.

- All hemangiomas do not show arterial flow. Lesions in the involuting phase may show venous flow or no flow on Doppler study. These patients need to be diagnosed based on typical clinical appearance and response to treatment.
- In LMs/VMs, sometimes a few arterial channels may be seen traversing through the lesion. Such arteries on spectral evaluation show high-resistance and low-velocity flow, in contrast to low-resistance flow within the arterial component of a high-flow vascular anomaly.
- Some LMs may not appear cystic and instead appear hypoechoic or heteroechoic on sonography. This may be due to the presence of multiple internal echoes or due to their microcystic nature. These should not be confused with VMs or solid tumors. The presence of increased through-transmission establishes their cystic nature.
- Certain cystic tumors (like teratomas) and tumorlike lesions (like epidermoid/dermoid cysts) may mimic LMs on ultrasound, especially in the head and neck region. The presence of a solid component, along with demonstration of calcification and/or fat within the lesion, suggests the diagnosis of a teratoma rather than an LM.
- VMs, in some cases, show a very slow flow that is not detected by Doppler-USG. Absence of flow does not preclude the diagnosis of a VM. VMs with thrombosis may show absence of flow on Doppler and may even show absence of enhancement on postcontrast MRI or CT.
- Certain entities can mimic VMs on imaging; a common example is a plexiform neurofibroma, seen in patients with neurofibromatosis type I (►Fig. 24). On USG, these are hypoechoic, trans-spatial lesions, which may mimic infiltrative VMs. On MRI, they are T2-hyperintense lesions, typically showing avid postcontrast enhancement.

Conclusion

Sonography can accurately diagnose most vascular anomalies if examined in a systematic manner and should be the initial imaging modality of choice, especially in the pediatric population. Doppler sonography helps in categorizing vascular anomalies into high- and low-flow lesions, which forms the basis of imaging diagnosis and guides further management. Sonographic findings must be interpreted in conjunction with history and clinical examination findings. Further imaging evaluation using MRI or CT has a role in assessment of lesion extent, and in angiographic evaluation of high-flow lesions.

Conflict of Interest
There are no conflicts of interest.

References

1   Fevurly RD, Fishman SJ. Vascular anomalies in pediatrics. Surg Clin North Am 2012;92(3):769–800, x

2   Marler JJ, Mulliken JB. Vascular anomalies: classification, diagnosis, and natural history. Facial Plast Surg Clin North Am 2001;9(4):495–504

3   International Society for the Study of Vascular anomalies. ISSVA classification of vascular anomalies. 2018. Available at: issva.org/classification. Accessed April 6, 2021

4   Eifert S, Villavicencio JL, Kao TC, Taute BM, Rich NM. Prevalence of deep venous anomalies in congenital vascular malformations of venous predominance. J Vasc Surg 2000;31(3):462–471

5   Mulliken JB, Enjolras O. Congenital hemangiomas and infantile hemangioma: missing links. J Am Acad Dermatol 2004;50(6):875–882

6   Moore CW, Lowe LH, Hepatic tumors and tumor-like conditions. In: Slovis TL, ed. Caffey's Pediatric Diagnostic Imaging. Philadelphia, PA: Mosby Elsevier; 2008 1929–1948

7   Starkey E, Shahidullah H. Propranolol for infantile haemangiomas: a review. Arch Dis Child 2011;96(9):890–893

8   Krol A, MacArthur CJ. Congenital hemangiomas: rapidly involuting and noninvoluting congenital hemangiomas. Arch Facial Plast Surg 2005;7(5):307–311

9   Berenguer B, Mulliken JB, Enjolras O, et al. Rapidly involuting congenital hemangioma: clinical and histopathologic features. Pediatr Dev Pathol 2003;6(6):495–510

10 Burrows PE, Mulliken JB, Fellows KE, Strand RD. Childhood hemangiomas and vascular malformations: angiographic differentiation. AJR Am J Roentgenol 1983;141(3):483–488

11 Mahady K, Thust S, Berkeley R, et al. Vascular anomalies of the head and neck in children. Quant Imaging Med Surg 2015;5(6):886–897

12 Flors L, Leiva-Salinas C, Maged IM, et al. MR imaging of soft-tissue vascular malformations: diagnosis, classification, and therapy follow-up. Radiographics 2011;31(5):1321–1340, discussion 1340–1341

13 Morrow MS, Oliveira AM. Imaging of lumps and bumps in pediatric patients: an algorithm for appropriate imaging and pictorial review. Semin Ultrasound CT MR 2014;35(4):415–429

14 Stavros AT, Rapp CL, Parker SH, Nonmalignant breast disorders that have complex cystic phases. In: Stavros AT, Rapp CL, Parker SH, eds. Breast Ultrasound. Philadelphia, PA: Lippincott Williams and Wilkins; 2004 435–437

15 Liu AS, Mulliken JB, Zurakowski D, Fishman SJ, Greene AK. Extracranial arteriovenous malformations: natural progression and recurrence after treatment. Plast Reconstr Surg 2010;125(4):1185–1194

16 Dubois J, Garel L. Imaging and therapeutic approach of hemangiomas and vascular malformations in the pediatric age group. Pediatr Radiol 1999;29(12):879–893

17 Dubois J, Patriquin HB, Garel L, et al. Soft-tissue hemangiomas in infants and children: diagnosis using Doppler sonography. AJR Am J Roentgenol 1998;171(1):247–252

18 Wiesinger I, Jung W, Zausig N, et al. Evaluation of dynamic effects of therapy-induced changes in microcirculation after percutaneous treatment of vascular malformations using contrast-enhanced ultrasound (CEUS) and time intensity curve (TIC) analyses. Clin Hemorheol Microcirc 2018;69(1-2):45–57

19 Legiehn GM, Heran MK. Classification, diagnosis, and interventional radiologic management of vascular malformations. Orthop Clin North Am 2006;37(3):435–474, vii–viii

20 Paltiel HJ, Burrows PE, Kozakewich HP, Zurakowski D, Mulliken JB. Soft-tissue vascular anomalies: utility of US for diagnosis. Radiology 2000;214(3):747–754

21 Bruckner AL, Frieden IJ. Hemangiomas of infancy. J Am Acad Dermatol 2003;48(4):477–493, quiz 494–496

22 Nord KM, Kandel J, Lefkowitch JH, et al. Multiple cutaneous infantile hemangiomas associated with hepatic angiosarcoma: case report and review of the literature. Pediatrics 2006;118(3):e907–e913

23 Rogers M, Lam A, Fischer G. Sonographic findings in a series of rapidly involuting congenital hemangiomas (RICH) Pediatr Dermatol 2002;19(1):5–11

24 Nasseri E, Piram M, McCuaig CC, Kokta V, Dubois J, Powell J. Partially involuting congenital hemangiomas: a report of 8 cases and review of the literature. J Am Acad Dermatol 2014;70(1):75–79

25 Lee GK, Suh KJ, Lee JH, et al. Lobular capillary hemangioma in the soft tissue of the finger: sonographic findings. Skeletal Radiol 2010;39(11):1097–1102

26 Stout AP, Murray MR. Hemangiopericytoma: a vascular tumour featuring Zimmermann's pericytes. Ann Surg 1942;116(1):26–33

27 Bhutia O, Roychoudhury A. Hemangiopericytoma of mandible. J Oral Maxillofac Pathol 2008;12:26–28

28 Sinha A, Rawson K, Singh G. A rare case of hemangiopericytoma in the maxilla of a 4 year-old child. J Indian Acad Oral Med Radiol 2010;22:64–66

29 Sarkar M, Mulliken JB, Kozakewich HP, Robertson RL, Burrows PE. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with Kaposiform hemangioendothelioma and not with common infantile hemangioma. Plast Reconstr Surg 1997;100(6):1377–1386

30 Lowe LH, Marchant TC, Rivard DC, Scherbel AJ. Vascular malformations: classification and terminology the radiologist needs to know. Semin Roentgenol 2012;47(2):106–117

31 Trop I, Dubois J, Guibaud L, et al. Soft-tissue venous malformations in pediatric and young adult patients: diagnosis with Doppler US. Radiology 1999;212(3):841–845

32 Dubois JM, Sebag GH, De Prost Y, Teillac D, Chretien B, Brunelle FO. Soft-tissue venous malformations in children: percutaneous sclerotherapy with Ethibloc. Radiology 1991;180(1):195–198

33 Puig S, Casati B, Staudenherz A, Paya K. Vascular low-flow malformations in children: current concepts for classification, diagnosis and therapy. Eur J Radiol 2005;53(1):35–45

34 Puig S, Aref H, Chigot V, Bonin B, Brunelle F. Classification of venous malformations in children and implications for sclerotherapy. Pediatr Radiol 2003;33(2):99–103

35 Donnelly LF, Adams DM, Bisset GS II. Vascular malformations and hemangiomas: a practical approach in a multidisciplinary clinic. AJR Am J Roentgenol 2000;174(3):597–608

36 Dubois J, Alison M. Vascular anomalies: what a radiologist needs to know. Pediatr Radiol 2010;40(6):895–905

37 Ozeki M, Fukao T. Generalized lymphatic anomaly and Gorham-Stout disease: overview and recent insights. Adv Wound Care (New Rochelle) 2019;8(6):230–245

38 Monroe EJ. Brief description of ISSVA classification for radiologists. Tech Vasc Interv Radiol 2019;22(4):100628

39 Amarneh M, Shaikh R. Clinical and imaging features in fibro-adipose vascular anomaly (FAVA) Pediatr Radiol 2020;50(3):380–387

40 Mulligan PR, Prajapati HJS, Martin LG, Patel TH. Vascular anomalies: classification, imaging characteristics and implications for interventional radiology treatment approaches. Br J Radiol 2014;87(1035) :20130392

41 Hartzell LD, Buckmiller LM. Current management of infantile hemangiomas and their common associated conditions. Otolaryngol Clin North Am 2012;45(3):545–556, vii

42 Frieden IJ, Haggstrom AN, Drolet BA, et al. Infantile hemangiomas: Current knowledge, future directions. Proceedings of a Research Workshop on Infantile Hemangiomas, April 7-9, 2005, Bethesda, Maryland USA. Pediatr Dermatol 2005;22(5):383–406