Radiological Diagnosis of Head and Neck Vascular Anomalies

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Objectives

- Review ideal sequencing of imaging in a head and neck vascular lesion.
- Variations in presentations of the lesion in head and neck.
- Lesion and anatomical appropriate imaging of a vascular lesion guidelines.
- Outlines of imaging characteristics of each vascular anomaly.

4.1 Introduction

Radiographic evaluation is useful to elucidate the locations, extent, and flow dynamics of lesions. With the advancement of diagnostic radiology, imaging is frequently and widely utilized for the diagnosis and management of vascular anomalies. Among different imaging modalities, ultrasound (US), magnetic resonance imaging (MRI), and magnetic resonance angiography (MRA) are most frequently used, especially in the pediatric population.

Computed tomography (CT) and conventional radiographs are also, though less commonly, used in certain situations compared to MRI and MRA.

Due to its invasive nature, angiogram is now often reserved for interventional planned procedures only.

4.2 Variations in the Radiological Presentations of Vascular Lesions of the Head and Neck Region

Most sporadic lesions may be different in clinical presentation compared to lesions associated with syndromes.

The senior author and editor (Chandra and Ghodke) undertook a retrospective study for reviewing medical and neuroradiology interventional records of all patients with head and neck vascular anomalies, diagnosed and/or surgically treated at the University of Washington medical system from 2006 to 2016. The exclusion criteria were patients with segmental lesions, those associated with syndromes such as Sturge-Weber, and

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S. C. Nair, S. R. Chandra (eds.), *Management of Head and Neck Vascular Lesions*,
https://doi.org/10.1007/978-981-15-2321-2_4

patients with intracranial tissues or viscera involved. A total of 102 patients (Male = 52 and Female = 50) were included in the study. There were 62 patients in the vascular tumor group, and 40 patients in the vascular malformation group. The radiologic appearances varied from unilocular to multilocular, regular to irregular, and well defined to poorly defined margins. Lesion location varied with involvement of the scalp, orbits, nose, maxilla, mandible, and neck regions. Seventy-five patients were symptomatic. Forty-seven patients were surgically treated. Vascular anomalies have varied symptomatic and radiological presentations. So, it is prudent to understand based on the findings from this large study, we should be minimizing unnecessary interventions, biopsies, and therapies. We describe the most salient features of vascular anomalies in Tables 4.1 and 4.2 with ultrasound and magnetic resonance imaging. Ultrasound and MRI are the most frequently used initial review investigational procedures. Some vascular anomalies have a characteristic appearance as described in Table 4.2.

4.3 Imaging Modalities

4.3.1 Ultrasound

- Ultrasound is the first choice for an initial evaluation in most cases. It has a good spatial resolution for superficial lesions. Other advantages include lack of ionizing radiation, wide availability, and relatively low cost. Doppler study can measure blood flow direction and volume in real time, and essential for differentiating low flow from high-flow vascular malformations. High-frequency linear array transducer is preferred in most cases, however,

Table 4.1 US and MRI features of major vascular anomalies [1–8]

Vascular anomalies	US features	MRI features
Vascular tumor		
Infantile hemangioma	Heterogeneous soft tissue mass with mixed echogenicity. Proliferative phase: high vascularity (more than 5 vessels/cm^2) and low RI. Involuting phase: decreased vascularity and high RI	Proliferative phase: well-defined mass with low SI on T1WI, high SI on T2WI, intense contrast enhancement, and flow-voids on gradient echo (GRE) sequence. Involuting phase: high SI on T1WI
Congenital hemangioma	Similar to infantile hemangioma; often contains internal foci of calcifications or hemorrhage	
Low-flow vascular anomalies		
Venous	Hypoechoic cystic lesion with sinusoidal chambers and hyperechoic foci (phleboliths)	Lobulated mass with iso-to-low SI on T1WI, high SI on T2WI, delayed contrast enhancement, lack of flow voids on GRE sequence, and signal voids (phleboliths)
Lymphatic	Macrocystic: anechoic, multiloculated cystic mass, echogenic hemorrhage. Microlytic: Hyperechoic soft tissue thickening. No intracity vascular signals on Doppler	High SI on T2WI with fluid–fluid level and hypodense septations. Macrocystic: rim and septal enhancement; microcystic: no enhancement
Capillary	Clinical diagnosis only; no imaging necessary	
High-flow vascular anomalies		
AVM	Enlarged feeding arteries and dilated draining veins without soft tissue mass	Large flow voids on SE sequences. Early arterial washout and early venous enhancement

RI resistant index, *SI* signal intensity, *T1WI* T1 weighted image, *T2WI* T2 weighted image, *SE* spin echo

Table 4.2 Key features of most common vascular lesions of head and neck. Adapted from Nair et al. JMOS 2016

	US with CDUS	MRI
IH	Hyperechoic or hypoechoic Hypervascular on CDUS	Iso to intermediate signal on T1W, bright signal on T2W, high-intensity flow enhancement on gradient echo, internal flow voids, vigorous enhancement after contrast administration
RICH	Central, non-enhancing, hypodense, hypoechoic, more robust feeding vessels with large diameter than IH	T2W hyperintense component is quite prominent
NICH	Almost similar to IH	Almost similar to IH
VMs	Solid echogenic mass with phleboliths, often multispatial and compressible. Low flow or monophasic or no flow on CDUS	T1W heterogenous intermediate signal, no flow voids, T2 fast spin-echo fat-saturated or short T1 inversion recovery high signal intensity, T1W spin-echo postgadolinium enhancement
LM	Variable multicystic, multispatial masses, with or without fluid or debris levels. No flow pattern on CDUS	T1W low to intermediate signal intensity, T2W high signal intensity, T1W postgadolinium, no enhancement except within septa
AVM	Clusters of vessels with no intervening well-defined mass. High flow (arterial flow) on CDUS. Arterial and venous signals from vessels in the lesions with arterializations of venous structure	T1W and T2W sequences show serpiginous signal voids without a focal mass

US ultrasound, *CDUS* color Doppler ultrasound, *MRI* magnetic resonance imaging, *T1W* T1 weighted, *T2W* T2 weighted, *IH* infantile hemangioma, *RICH* rapidly involuting congenital hemangioma, *NICH* non-involuting congenital hemangioma, *VM* venous malformation, *LM* lymphatic malformation, *AVMs* arteriovenous malformations

depending on the size and depth of the lesion, a combination of different transducers might be required. Limited penetration of ultrasound makes it difficult to evaluate deep structures and the extension of lesions. Also, ultrasound findings are highly dependent on operator experience and thus can vary between different examiners.

4.3.2 Magnetic Resonance Imaging

- When ultrasound findings are equivocal, MRI is the next step. MRI is a favorable exam for most clinicians when evaluating very large lesions or lesions within specific locations (infraorbital or intracranial lesion, etc.) [1]. MRI not only provides valuable information about extension of disease, the depth of lesions, and internal organ involvement, but also guide treatment planning and posttreatment evaluation. MRI does not use ionizing radiation and is preferred in pediatric population. However, sedation is often required for pediatric patients during lengthy MRI exam. Spin Echo sequences and contrast enhancement patterns often provide useful information for diagnosis. MRA can provide hemodynamic information of vascular malformation and is helpful in differentiating high flow from low-flow lesions [2].

4.4 Major Imaging Finding of Vascular Tumors

4.4.1 Infantile Hemangioma

- Most infantile hemangiomas are diagnosed clinically and only a few cases require image studies. The findings vary depending on the clinical phases. In the proliferative phase, it presents as a well-defined lobular mass with high vascular activity. In the involuting phase, fatty replacement occurs with decreased vascular activity.
- Ultrasound demonstrates a well-defined soft tissue mass with variable internal echogenicity. Calcifications are rare. Color Doppler is very helpful in showing the characteristic

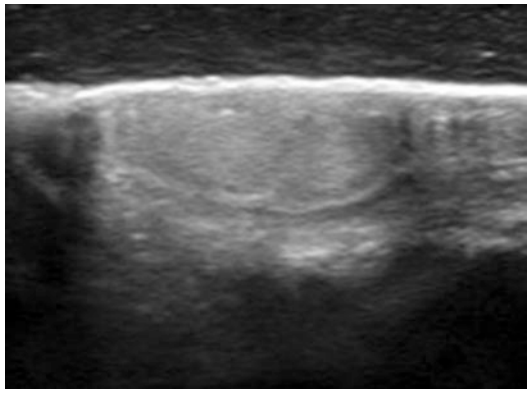

Fig. 4.1 Grayscale ultrasound shows hyperechoic subcutaneous soft tissue mass overlying the right lambdoid suture

high vascularity with both arterial and venous flow. Hypervascularity in proliferative phase is defined as more than **5 vessels per square centimeter with low resistive index** (Figs. 4.1 and 4.2). The involuting phase, on the contrary, has decreased vascularity and increased resistive index.

- MRI is mainly used for lesions involving critical organs (i.e., infraorbital or intracranial extension) or treatment planning. In the proliferative phase, lesions are well-defined with low signal intensity on T1-weighted image (T1WI), high signal intensity on T2-weighted image (T2WI), and intense contrast enhancement. In the involuting phase, there is high signal intensity on T1WI due to fatty replacement.

4.4.2 Congenital Hemangioma

- Both ultrasound and MRI findings of congenital hemangioma are similar to infantile hemangioma, characterized by soft tissue mass with hypervascularity. It often contains internal foci of calcifications or hemorrhage [1–3].

4.4.3 Kaposiform Hemangioendothelioma

- Ill-defined, infiltrating soft tissue mass with diffuse enhancement involving multiple tissue planes. Calcifications are common. Perilesional edema makes it difficult to define tumor margins. On MRI, diffuse high signal intensity on T2WI mix with foci of intermediate to low signal intensity. Feeding/draining vessels are usually smaller and along the tumor margin other than within the lesion.

4.5 Low-flow Vascular Malformations

4.5.1 Venous Malformation

- Venous malformation usually presents as an asymptomatic soft tissue mass. In imaging study, it appears as a well-defined lobulated lesion.
- Ultrasound is very useful for the evaluation of superficial lesions. It shows a characteristic hypoechoic cystic lesion with sinusoidal chambers and hyperechoic foci (phleboliths). It is compressible with low-velocity venous flow or less commonly no flow on color Doppler which confirms the venous component.
- One of the few lesions that CT could be helpful for diagnosis. The presence of phleboliths within a soft tissue mass on CT is characteristic of venous malformation (Fig. 4.3). However, CT utilizes ionizing radiation which limits the use in pediatric patients.
- MRI demonstrates a lobulated mass with iso-to-low signal intensity on T1WI, high signal intensity on T2WI due to its cystic nature, delayed contrast enhancement, and signal voids suggestive of phleboliths.

4.5.2 Lymphatic Malformation

- There are two main subtypes of lymphatic malformation, macrocystic and microcystic. Each subtype has distinguished radiologic appearances. Macrocystic lymphatic malformation is a well-defined lobulated soft tissue mass with multiple septations and fluid–fluid level. Microcystic lesion is typically an

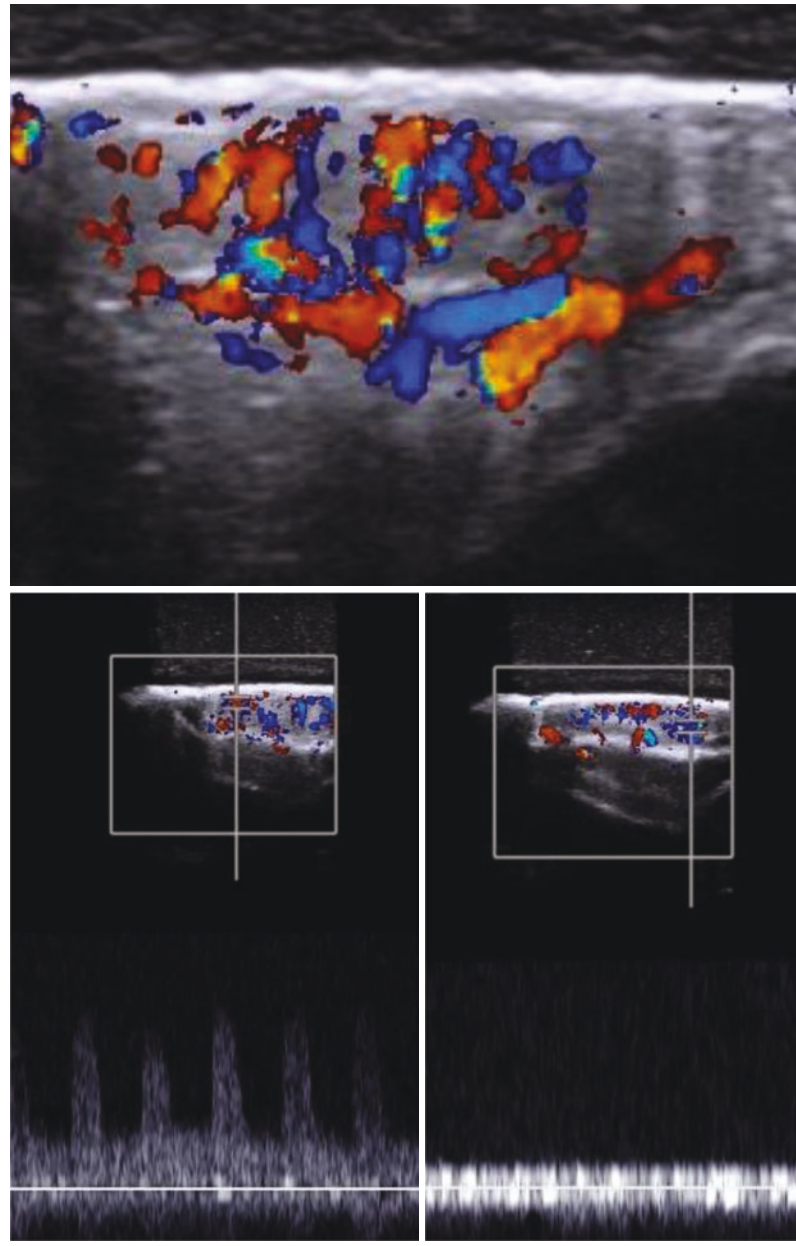

Fig. 4.2 Doppler study shows intralesional hypervascularity with both arterial and venous flow, compatible with infantile hemangioma

ill-defined, infiltrative, solid mass that involves multiple tissue planes [4, 5].

- On US, macrocystic lymphatic malformation is presented as an anechoic, multiloculated cystic mass, often with echogenic intralesional hemorrhage (Fig. 4.4). However, US often cannot show the extent of large lesions. Microcystic lymphatic malformation is often presented as an ill-defined hyperechoic soft tissue thickening. No intra-lesions vascular signals were seen on Doppler.
- On MRI, lymphatic malformation has high SI on T2WI with fluid–fluid level and hypodense thin septations (Fig. 4.5). Macrocystic and microcystic lesions have different contrast enhancement patterns. Macrocystic lesions have rim and septal enhancement while microcystic lesions have no enhancement.

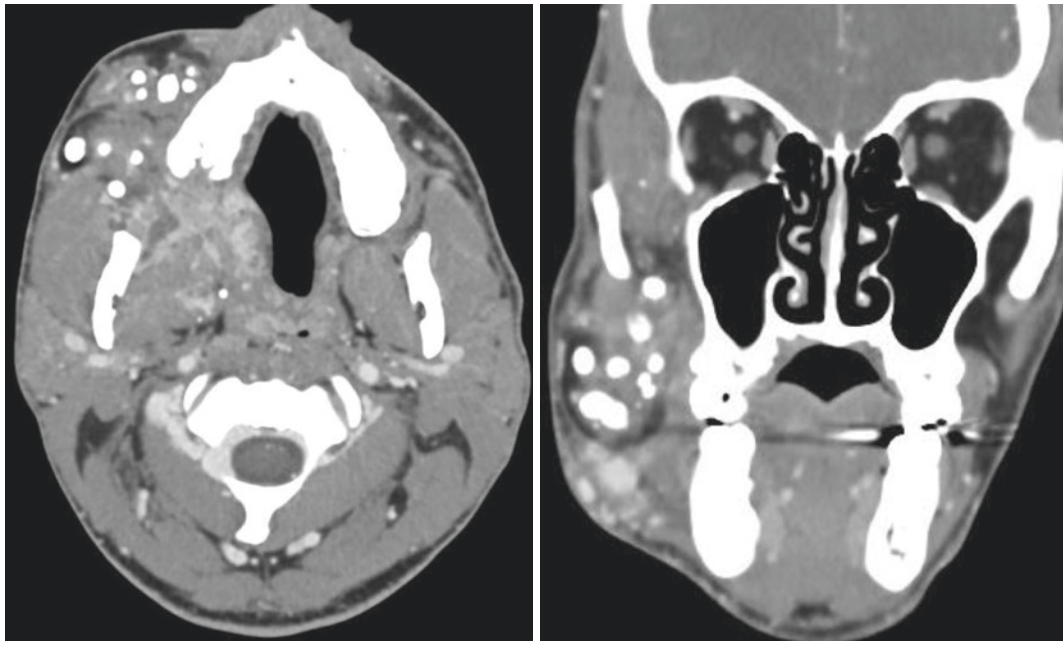

Fig. 4.3 Contrast-enhanced axial and coronal CT images show soft tissue mass with serpiginous enhancement and numerus phleboliths involving multiple compartments of the right face, suggestive of low flow lympho-venous malformation

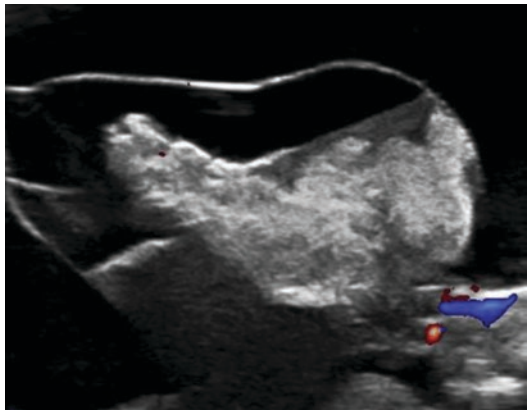

Fig. 4.4 Large multiloculated left neck mass on ultrasound with fluid–fluid level and echogenic debris from recent hemorrhage. No internal flow on color Doppler. Findings are characteristics of a macrocystic lymphatic malformation

4.5.3 Capillary Malformation

- Capillary malformation is a clinical diagnosis. Imaging is not necessary for diagnosis [4].

4.6 High-flow Vascular Malformations

4.6.1 Arteriovenous Malformation (AVM)

- Ultrasound is usually the first choice for diagnosis. Tangles of enlarged feeding arteries and dilated draining veins without discrete soft tissue mass suggest the diagnosis. The feeding arteries have low resistant waveform on color Doppler due to shunting.
- On MRI, the enlarged feeding arteries and draining veins are shown as large flow voids on echo spin sequences. AVM has a characteristic contrast enhancement pattern due to shunting. Early arterial washout and early venous enhancement are typically for AVM. Figure 4.6 depicted a T2WI MRI and CT angiogram of a large arterial venous malformation within the right frontal parasagittal white matter with tangles of enlarged feeding arteries and a large draining vein [6–8].

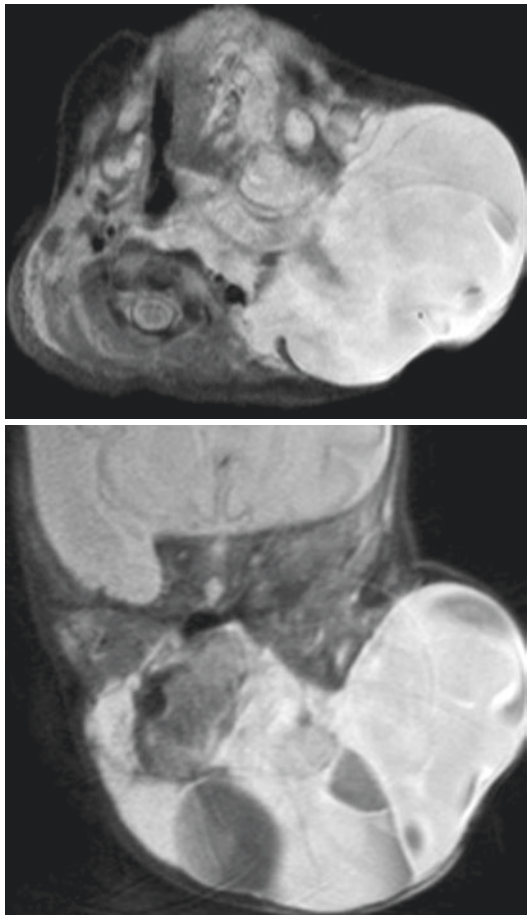

Fig. 4.5 Axial and Coronal T2-weighted MR images show a large multiloculated and multispatial left neck mass with fluid–fluid levels and associated mass effect, compatible with lymphatic malformation with recent hemorrhage

- CT Angiography or MR Angiography shows details of feeding arteries, nidus, and draining veins, and are often used for treatment planning.

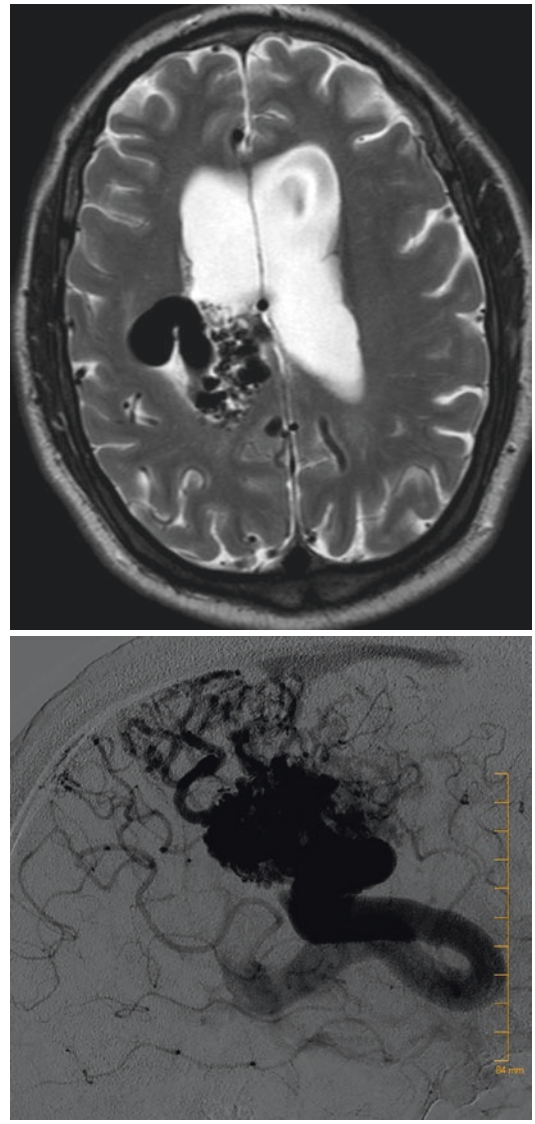

Fig. 4.6 T2WI MRI and CT angiogram show a large arterial venous malformation within the right frontal parasagittal white matter with tangles of enlarged feeding arteries and a large draining vein

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