

Vascular Anomalies of the Neck




David E. Webb, Maj, USAF, DC ^{a,*}, Joseph McDermott, MD ^b,
David Gover, MD ^c

KEYWORDS

• Vascular anomalies • Head and neck surgery • Embolization • Imaging

KEY POINTS

- The key to good surgery is good access, and the key to good access is good surgery. This is never more true than when dealing with vascular lesions—which demand unrestricted exposure.
- Hypervascular tumors or those known to have a high degree of blood loss during surgery may benefit from preoperative embolization.
- If embolization is necessary, the embolic agent can be determined by using the following 3-question algorithm:
 - Is the vessel to be embolized large or small?
 - How long (temporally) is the vessel to remain occluded?
 - Is the tissue supplied by the vessel to remain viable after the embolization?
- Surgery is usually performed the day after embolization, although some investigators suggest resection can be delayed up to 8 days.

 **Video of the flammable nature of Onyx during electrocautery of the post-embolized facial artery accompanies this article at <http://www.oralmaxsurgeryatlas.theclinics.com/>**

Introduction

A multidisciplinary team is imperative to the diagnosis and successful management of vascular anomalies of the head and neck. Patients routinely require the services of a radiologist (both imager and proceduralist) and an experienced surgeon. Hematologist-oncologists and dermatologists can also be of assistance. A patient's history cannot be over-emphasized, as it aids the radiologist in deciding on imaging modalities, tailoring studies to the needs of the patient, and making the diagnosis.

This article discusses the classification of vascular anomalies, provides a clinicopathologic review of a few vascular lesions, reviews imaging principles of vascular anomalies and image-guided (interventional) procedures, and concludes with treatment pearls.

Classification of vascular anomalies

The whole team should share a common lexicon. The International Society for the Study of Vascular Anomalies (ISSVA)

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^a Department of Oral and Maxillofacial Surgery, David Grant USAF Medical Center, 101 Bodin Circle/SGDD, Travis AFB, CA 94535, USA

^b Department of Pathology, David Grant USAF Medical Center, 101 Bodin Circle/SGQC, Travis AFB, CA 94535, USA

^c Department of Radiology, David Grant USAF Medical Center, 101 Bodin Circle/SGQX, Travis AFB, CA 94535, USA

* Corresponding author.

E-mail address: david.webb.5@us.af.mil

provides such a framework for classifying “Vascular Tumors.” These tumors, along with common examples, are listed in [Table 1](#).

In addition to these vascular lesions, this article also considers “vascular-like tumors”: hypervascular neoplasia such as hemangiopericytomas, paragangliomas (carotid body tumors and glomus tumors), and hypervascular metastasis.

Clinicohistopathologic description

Hemangiomas

Hemangiomas are common, comprising 7% of benign pediatric age tumors and affecting 4.5% of children. One-third of hemangiomas are found in the head and neck, and 14% within the oral cavity. Hemangiomas of childhood are divided into infantile and congenital types, based on whether they are fully developed at birth. Hemangiomas that develop in adulthood tend to be small and are often located within the oral cavity or pharynx. Mucosal hemangiomas develop in areas that are often traumatized, such as the lip, buccal mucosa, and tongue ([Figs. 1–5](#)).

Infantile hemangiomas

Infantile hemangiomas are the most common vascular neoplasms, and include capillary and cavernous hemangiomas of any organs. These hemangiomas develop more often in infants who are female, Caucasian, premature, or of low birth weight. Most infantile hemangiomas present between 2 weeks and 2 months after delivery, and grow with the child; they present as an erythematous macule, patch, or plaque, but may also be present within deep soft tissues. Most do not require medical

Table 1 ISSVA classification for vascular anomalies

| Vascular Tumors | | | Vascular Malformations | |
|---|--|----------------------|------------------------|----------|
| Benign | Locally Aggressive or Borderline | Malignant | Simple | Combined |
| Infantile hemangioma | Kaposiform hemangioendothelioma | Angiosarcoma | CM | CVM |
| Congenital hemangioma | Retiform hemangioendothelioma | Epithelioid | LM | CLM |
| Rapidly involuting (RICH) | Papillary intralymphatic angioendothelioma | hemangioendothelioma | VM | CAMV |
| Noninvoluting (NICH) | (PILA), Dabska tumor | | AVM | LVM |
| Partially involuting (PICH) | Composite hemangioendothelioma | | AVF | CLVM |
| Tufted angioma | Kaposi sarcoma | | | CLAVM |
| Spindle-cell hemangioma | | | | CVAVM |
| Epithelioid hemangioma | | | | CLVAVM |
| Pyogenic granuloma (lobular capillary hemangioma) | | | | |

Abbreviations: AV, arteriovenous; C, capillary; F, fistula; L, lymphatic; M, malformation; V, venous.

or surgical intervention. For those requiring medical intervention, propranolol and glucocorticoids are often the first-line therapy.

Capillary hemangiomas are the most common soft-tissue neoplasm in infants and children, and consist of numerous intertwining capillary-sized blood vessels. These tumors grow rapidly in first year of life, then undergo gradual involution over the next 1 to 7 years. By age 7, 75% to 90% regress.

Cavernous hemangiomas are composed of larger vascular spaces filled with erythrocytes, are generally larger than capillary hemangiomas, and may involve deeper tissue. Although they are usually well circumscribed, they may be locally destructive. Unlike capillary-type infantile hemangiomas, they do not spontaneously regress.

Congenital hemangiomas

Congenital hemangiomas are much rarer than infantile hemangiomas. Unlike infantile hemangiomas they are fully formed

at birth, occur equally in males and females, and stain negative for GLUT1 by immunohistochemistry. Congenital hemangiomas occur as solitary lesions on the head or near limb joints, and are categorized by the tendency to spontaneously involute. Rapidly involuting congenital hemangiomas regress completely within 2 years of birth; noninvoluting congenital hemangiomas grow proportionately with the infant and may display partial involution, but not full regression.

Vascular malformations

Vascular malformations are vascular proliferations arising from disordered angiogenesis, either during development or following trauma, and are categorized as either high-flow lesions, which involve an artery, or low-flow lesions, which do not. Vascular malformations may also be categorized by the number of vessel types involved (see [Table 1](#)). Simple malformations are composed of only a single type of vessel, and



Fig. 1 A 40-year-old man presents with remote history of lip trauma. The lesion is firm and nonpulsatile.



Fig. 2 In-office ultrasonography (US) performed to rule out a high-flow lesion/arterialization before incisional biopsy.

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