Head and Neck Vascular Lesions

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KEYWORDS

• Infantile hemangioma • Venous malformation • Lymphatic malformation
• Arteriovenous malformation

KEY POINTS

- Vascular malformations represent a complex and highly variable group of head and neck masses, for which correct diagnosis is of the utmost importance.
- A thorough history and examination can usually lead to diagnosis, which is confirmed with imaging.
- The decision-making process of whether and when to treat these lesions requires expertise across multiple disciplines, and will be dually rewarding and frustrating.
- Using these guidelines, the clinician can set a realistic expectation and plan for each individual patient.

INTRODUCTION

Vascular anomalies represent a broad range of vascular malformations and tumors, which predominantly occur in the head and neck and are seen in approximately 4.5% of children.[1–6] Due to the wide variety of different presentations, growth behavior, and available treatments, which depend on the lesion, accurate diagnosis is of utmost importance. Proper diagnosis will aid in patient and family counseling and expectations, and it is helpful if they understand from the beginning that treatment will tend to be long-term. Using hemangioma as a blanket diagnosis to include vascular malformations (venous, lymphatic, and arteriovenous), with the incorrect

Funding Sources: None.
Conflict of Interest: None.
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Otolaryngol Clin N Am 48 (2015) 29–45
http://dx.doi.org/10.1016/j.otc.2014.09.004 **oto.theclinics.com**
0030-6665/15/$ – see front matter

implication that the lesion will regress over time, will lead to unrealistic expectations and increased the chance of complications from the lesion.

Vascular anomalies are divided into vascular tumors and vascular malformations, according to the International Society for the Study of Vascular Anomalies classification system, which is based on the work of Mulliken and Glowacki[1] in 1982 (**Table 1**). This is an important distinction because malformations and tumors show different growth characteristics. Infantile hemangioma (IH) is the most common and is considered a benign vascular tumor. IH will rapidly enlarge, then slowly regress over time, but many will still need treatment.

Vascular malformations are relatively uncommon, rarely regress, and will continue to enlarge and cause increased complications over time.[2,7–9] This includes venous malformation (VM), lymphatic malformation (LM), and arteriovenous malformation (AVM). The paradigm is shifting toward early treatment of vascular malformations when the lesion is smaller and there has been less distortion or invasion of local tissues.

Vascular anomalies represent an opportunity for collaboration in conjunction with a multidisciplinary vascular anomalies team. The authors encourage an active and frequent dialogue between otolaryngology, dermatology, interventional radiology, plastic surgery, hematology-oncology, pediatric surgery, orthopedic surgery, physical therapy, psychology, and social work. This article discusses the spectrum of common vascular anomalies, with a focus on IH as well as VMs, LMs, and AVMs.

INFANTILE HEMANGIOMAS

Diagnosis

IHs are true benign tumors of infancy and are the most common vascular anomaly. IHs are seen in approximately 4% to 10% of infants, usually before the age of 1 year, and 60% of these will present in the head and neck.[10–12] IHs are more common in whites, low-birth weight infants, and multiple gestations (2–3:1 girl/boy ratio).[2]

Histologically, IHs are composed of proliferating immature endothelial cells and disorganized vessels. The exact pathway is not fully understood, but IHs derive from endothelial stem cells with abnormal angiogenesis and vasculogenesis. Current areas of study have implicated the vascular endothelial growth factor (VEGF) and adrenergic receptor pathway, which may be the mechanism by which propranolol can shrink these tumors.[12] GLUT-1 has been identified as a specific marker for IH,

Table 1
Vascular anomaly classification based on International Society for the Study of Vascular Anomalies guidelines

Vascular Anomalies	
Vascular Tumors	**Vascular Malformations**
Hemangioma	High-flow
Infantile	Arteriovenous
Congenital (RICH or NICH)	Mixed Arterial
KHE	Low-flow
Tufted angioma	Venous
Pyogenic granuloma	Lymphatic
Other rare vascular tumors	Capillary or venular

Abbreviations: KHE, kaposiform hemangioendothelioma; NICH, noninvoluting congenital hemangioma; RICH, rapidly involuting congenital hemangioma.