The Management of Vascular Malformations of the Airway

Natural History, Investigations, Medical, Surgical and Radiological Management

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KEYWORDS

• Airway • Congenital • Vascular lesion • Lymphatic malformation
• Venous malformation • Arteriovenous malformation

KEY POINTS

- Airway vascular malformations affect all areas of the airway and each have a characteristic appearance and distribution.
- Airway lymphatic malformations involve the supraglottis and above; venous malformations may be transglottic, whereas arteriovenous malformations occur in vascular "choke zones" and are often found in the base of tongue or parapharyngeal spaces.
- Problems with speaking, swallowing, oral competence, glossoptosis, and sleep apnea often coexistent and require simultaneous management.
- Sclerotherapy with bleomycin is a safe and effective agent in both venous and lymphatic airway disease.
- Posttherapy inflammation should be anticipated with proper consideration to airway protection.

UPPER AIRWAY CONGENITAL VASCULAR LESIONS

Congenital vascular malformations of the upper airway present unique problems for the patient and the medical team and often require a multidisciplinary approach.[1,2] The upper airway is defined anatomically from its anterior superior boundary of the

Disclosure Statement: The authors have nothing they wish to disclose.
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Otolaryngol Clin N Am 51 (2018) 213–223
https://doi.org/10.1016/j.otc.2017.09.013
oto.theclinics.com
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lips to the carina of the trachea. This definition covers a large and varied topography including the oral cavity, oropharynx, larynx, glottis, and trachea.

Morbidity includes difficulty with mastication, airway obstruction, speech intelligibility, oral incompetence, sialorrhea, and dysphagia. These concerns necessitate special consideration, with focused evaluation and treatment modalities, which may differ from other locations. Recent studies have shown a high propensity of obstructive sleep apnea in this population with 47% to 85% affected.[1,3,4]

Vascular tumors and malformations may be present in the airway. The most common vascular tumor involving the airway is infantile hemangioma (IH), (see David H. Darrow's article, "Management of Infantile Hemangiomas of the Airway," in this issue). Vascular malformations are defined by their vessel type and may be lymphatic, venous, capillary (port wine stains), or arteriovenous. Each type is discussed with a focus on specific presentation and management.

AIRWAY VASCULAR MALFORMATIONS

Similar to the classification of airway IHs into focal and segmental lesions, airway vascular malformations may also be classified according to their distribution (focal or diffuse).[5] All patients with extensive head and neck cutaneous lesions have a high risk of airway involvement and thus should be evaluated by an otolaryngologist. The treatment is often multidisciplinary. Unlike airway IHs, which proliferate and involute, vascular malformations of the airway persist and enlarge over time.

LYMPHATIC MALFORMATIONS

Patients with lymphatic malformations (LMs) of the head and neck have up to a 73% rate of upper airway involvement.[6] The oral cavity, tongue, oropharynx, base of tongue, and supraglottis are all commonly involved (**Fig. 1**). Lymphatic malformations of the airway may extend from the lips to the supraglottis. To date, we have never seen involvement of the true vocal folds, subglottis, or trachea.[6] Patients with massive bilateral cervicofacial disease will require a tracheostomy in up to 30% of cases. Lesions may be microcystic (<1–2 cm cysts), macrocystic, or mixed. More commonly, in diffuse disease, they are mixed.

Presentation

Upper airway LMs most commonly involve the tongue.[6] These lesions are characterized by mucosal colored or hemorrhagic vesicles on the tongue surface. They have been described as frog's eggs–type vesicles. Patients may also present with tongue swelling, bleeding, pain, halitosis, difficulty eating spicy or acidic foods, speech intelligibility, sleep-disordered breathing, and, in some cases, airway compromise. Tongue base enlargement is an important finding and is often associated with an Ω-shaped edematous epiglottis. Involvement of the floor of mouth, tongue, or tongue base can also result in glossoptosis, which may be characterized by floor-of-mouth involvement, macroglossia, or both. Differentiation of the root cause of glossoptosis is important in the choice of treatment.

A hallmark of LMs is exacerbations and remissions. An exacerbation is characterized by acute swelling and pain with or without the presence of hemorrhagic vesicles. The swelling may resolve fully or may not fully recede after each event. Trauma and hormonal changes also lead to growth and enlargement.[6]

Severe disease with macroglossia is usually detected during prenatal ultrasound fetal screening. In extreme cases, an EXIT procedure (Ex-utero Intrapartum Tracheostomy) may be planned to control the airway at delivery.[6–10]

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