Vascular Anomalies and Airway Concerns

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Abstract

Keywords

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Vascular anomalies, both tumors and malformations, can occur anywhere in the body, including the airway, often without any external manifestations. However, vascular anomalies involving the airway deserve special consideration as proper recognition and management can be lifesaving. In this article, the authors discuss vascular anomalies as they pertain to the airway, focusing on proper diagnosis, diagnostic modalities, and therapeutic options.

Vascular anomalies, composed of both tumors and malformations, remain a diagnostic and therapeutic challenge despite significant advancements made over the years. However, these anomalies present the greatest challenge when they compromise function due to visceral involvement, especially those that affect the airway, which can be life threatening. It is, therefore, important to discuss the presentation, associated conditions, diagnostic approach and treatment of such lesions.

The classification system established by the International Society for the Study of Vascular Anomalies (ISSVA) is now a widely accepted system used to categorize vascular anomalies into two types: (1) vasoproliferative or vascular neoplasms such as hemangioma, and (2) vascular malformations.¹ The distinction between the two is based on histopathological assessment of increased cell turnover where vascular tumors represent true neoplasms with pathologic cell proliferation. An in-depth review of vascular anomalies is beyond the scope of this article and will be provided in separate articles. Here we will focus solely on vascular anomalies pertaining to the airway specifically tumors and malformations.

Vascular Tumors

Subglottic Hemangioma

The most common vascular tumor of the head and neck is the infantile hemangioma (▶ Fig. 1). When located in a subglottic

position, its progression during the proliferative phase can result in airway compromise (▶ Fig. 2). These hemangiomas follow the predicted course as for all hemangiomas. As such, neonates are asymptomatic at birth and remain so for months until the lesion grows to the point of airway constriction. In the case of a 2- to 6-month-old child with progressive inspiratory and expiratory stridor or persistent croup, consideration should be given to the possibility of a subglottic hemangioma. As in all cases of infantile hemangioma, it is more common in girls and Caucasians, so extra clinical suspicion is warranted in these groups. Fifty percent of these children will also have cutaneous hemangiomas in a facial or beard distribution. Diagnosis should be made with direct laryngoscopy and bronchoscopy.^{2,3}

Special consideration should generally be given to a child with three or more cutaneous hemangiomas.⁴ In these children, an abdominal ultrasound should be obtained to evaluate for visceral hemangiomas, such as hepatic hemangioma. If the screening ultrasound is abnormal, MRI of the entire body is indicated to detect other internal hemangiomas. In addition, PHACE syndrome should be considered in patients with large facial hemangioma (> 5 cm) (▶ Fig. 3).^{5,6} The acronym refers to a constellation of findings to include posterior fossa anomalies, hemangioma, arterial lesions, cardiac abnormalities/aortic coarctation, and abnormalities of the eye. Although PHACE syndrome does not necessarily involve the airway, the recognition of such associations can lead to improved outcomes through proper diagnosis and treatment of all systems involved.

Fig. 1 Infantine hemangioma involving the lip and the oral mucosa.

Subglottic and other airway hemangioma can be temporized with medical therapy, just as with hemangioma in other locations, until natural involution occurs. For large or symptomatic lesions with pending airway compromise, β – blockers and/or steroids can be given.⁷ They may not necessarily decrease the size of these lesions, but may be able to prevent further growth. If airway patency cannot be accomplished with this approach, subglottic hemangiomas can be treated with endoscopic debulking. Although techniques vary, the CO₂ laser is frequently used for this purpose. A tracheostomy is rarely required. Typically, debulking is attempted at the time of diagnostic bronchoscopy and medical therapy is initiated.^{8–11} Open surgical excision is usually reserved for difficult or refractory cases.

Fig. 2 Subglottic hemangioma.

Fig. 3 Possible PHACE (posterior fossa anomalies, hemangioma, arterial lesions, cardiac abnormalities/aortic coarctation, and abnormalities of the eye) syndrome should be considered in a patient with a large hemangioma of the lower face and neck.

Pyogenic Granuloma

Another vascular tumor, which may be confused with the hemangioma of infancy, is the pyogenic granuloma. The pyogenic granuloma is histologically described as lobular capillary hemangiomas and may present as 1 to 10 mm sessile or pedunculated lesions. They are frequently associated with prior trauma and can bleed impressively with the slightest provocation. In children, pyogenic granulomas are most commonly located in the head and neck region with ~12% involving the oral mucosa or conjunctiva. A nocturnal hemorrhage of an intraoral lesion could compromise a child's airway and should be managed with surgical excision. Unlike other vascular malformations, imaging is unnecessary as the diagnosis is typically made clinically and later confirmed by histologic evaluation.¹²

Vascular Malformations

Vascular malformations are characterized as having an organized histology, which differs from the pathognomonic proliferation of vascular tumors. This subset of vascular anomalies is further divided into two categories based on flow characteristics of the lesion as either slow flow or fast flow.¹³ Slow-flow lesions include capillary malformations (CMs), venous malformations (VMs), and lymphatic malformations (LMs), whereas arteriovenous malformations (AVMs) are fast-flow lesions. Moreover, mixed lesions, such as lymphaticovenous malformations (LVMs) are common, further complicating both diagnosis and treatment of these lesions. Each of these lesions presents a unique treatment challenge in the setting of airway compromise.

Capillary Malformations

Capillary malformations can involve the airway, but usually are asymptomatic. Bleeding from CMs, however, can lead to airway compromise if not properly managed. Often, these symptomatic CMs are associated with hereditary

Fig. 4 Venous malformation of the lip and cheek causing disfigurement.

hemorrhagic telangiectasia (HTT) or Osler-Weber-Rendu disease.⁴ The diagnosis of HTT is often missed, but early recognition can be life saving. The malformation first manifests in the capillary beds and progresses to the capillary venous shunting. Lesions can be found in the skin, mucous membranes, lungs, liver, and brain. Nosebleeds and gastrointestinal bleeding are common sequelae. Appreciating the telangiectasias on the tongue is a simple observation that can lead to early diagnosis. Multiple modes of medical therapy are under evaluation for mucosal CMs. However, there is no adequate systemic therapy in wide acceptance at this time. For the treatment of epistaxis, septal dermoplasty can reduce severity by 75%.⁴ This procedure involves replacing the nasal mucosa with autologous skin grafts, but

Fig. 6 A child with diffuse lymphatic malformations after initial tracheostomy and gastrostomy tube placement.

carries a risk of CM recurrence within the grafted areas. Photocoagulation laser treatment may be used, but several sessions may be needed for symptomatic improvement. Septal perforation is a common significant complication of this therapy. For treatment of severe acute epistaxis, some patients undergo endovascular embolization. Unfortunately, this modality often requires multiple treatments and frequently need eventual surgical intervention. Septectomy

Fig. 5 A child with venous malformation involving right cheek and lips (A,B) undergoing laser treatment as a staged procedure (C) for possible surgical debulking.

Fig. 7 A child with diffuse lymphatic malformations involving the tongue and the mouth managed with a combination of sclerotherapy and surgery in a top-down fashion. Patient tolerated the procedure well without complications and the tracheotomy and gastrostomy tube were successfully removed. This is the same child in ▶Fig. 6 after serial management using a “top-down” strategy.

combined with septal dermoplasty is aggressive, but has been employed in patients with severe transfusion-dependent epistaxis. Nasal closure is a simple, very effective, and reversible procedure, but can be difficult to gain patient acceptance.

Patients with HHT should be screened for cerebral and pulmonary AVMs. All first-degree relatives should also have genetic testing for HHT. Many unexplained cerebral AVMs are thought to be manifestation of undiagnosed HHT.

Venous Malformations

Venous malformations (VMs) are generally most problematic when located in dependent extremities. However, VMs of the head and neck may thrombose or engorge, becoming disfiguring, painful, and obstructive to the airway (▶Fig. 4). In the event that airway obstruction results from a VM, a temporary tracheotomy is often necessary. Interventional radiology can be considered for both primary treatment, such as staged sclerotherapy or embolization, or used in conjunction with surgery via preoperative or intraoperative sclerotherapy or embolization. When sclerotherapy is used alone, recurrence

is common. When sclerotherapy is used in combination with surgery, cure is the goal. Venous malformations can also respond very well to laser treatment, including KTP and Nd:YAG lasers.¹⁴ This may be used as a primary treatment to shrink the VM, or used in combination with sclerotherapy and surgery (▶Fig. 5).

Lymphatic / Lymphaticovenous Malformations

Lymphatic and lymphaticovenous malformations (LM/LVMs) present a unique clinical picture as they are frequently diagnosed in utero.^{15,16} The increased use of ultrasound has led to the diagnosis of patients with LM prenatally and subsequent treatment dilemmas in early life. Anterior and lateral neck swellings identified on fetal ultrasound, which remain persistent on repeat studies, likely represent congenital LMs, and are sometimes massive. Posterior neck swellings are unlikely LMs and may be associated with chromosomal abnormalities.⁴ Children with massive congenital LMs are usually born by an exit procedure, in which the airway is stabilized by intubation, bronchoscopy, or tracheostomy.

Fig. 8 (A) A child with a localized lymphatic malformation involving left neck region. (B) MRI shows a discrete lymphatic malformation causing mass effect and encroaching on the airway.

Fig. 9 A discrete lymphatic malformation involving the tongue can be managed with laser resurfacing or coblation.

These neonates should not undergo massive dissection unless symptoms dictate necessity due to high rate of complications, which can include recurrence, poor wound healing, damage to neurovascular structures, and death. If symptoms do not dictate a need for immediate therapy, delayed sclerotherapy may be the optimal initial treatment.

Lymphatic malformations may cause airway compromise through compression from mass effect. Lymphatic malformations are categorized by the size of the lymphatic chamber: macrocystic (> 2 cm), microcystic (< 2 cm) or mixed.¹⁷ However, from a treatment standpoint, LMs can be divided into either a diffuse or discrete lesion. Diffuse microcystic cervicofacial LMs deserve special consideration as the management is complex and may be a lifelong endeavor (►Fig. 6). These LMs not only cause mass effect, but can cause bony changes in the form of mandibulomaxillary hypertrophy. This is due to involvement of the bone and growth of the LM within the bone. From a treatment perspective, a secure airway is

essential, and tracheostomy is often necessary to avoid acute respiratory problems. It is important not to create iatrogenic injuries, such as cranial nerve damage, that can increase the morbidity of the disease. The problem should be addressed by anatomical zones in an incremental fashion until each zone is free of disease, as recurrence is more likely to occur if gross malformation remains. A typical boundary used to divide these massive problems into several more manageable pieces is the mylohyoid.⁴ It is advisable to approach the components from the top-down to minimize swelling of the untreated zones. For example, attempt to deal with the tongue before the floor of mouth, and then approach the neck (►Fig. 7).

Both surgery and sclerotherapy are treatment options depending on the type, size, and location of the disease. Several sclerosants are available for use, including doxycycline, bleomycin, 3% sodium tetradecyl sulfate (STS), absolute alcohol, and OK-432.^{17–20} Alcohol, though an effective sclerosant, has frequent complications, including soft tissue necrosis, and some have been hesitant to inject it near the carotid sheath.²¹ OK-432 is most commonly used worldwide. Because of limited availability of OK-432 in the United States, doxycycline is the most commonly used agent in United States.

Bleomycin is also an important tool because it causes limited swelling after injection, which is unique to this agent. Sclerotherapy is especially useful for lesions that are not amenable to complete surgical excision, such as those that extend into the retropharyngeal space. In addition, many of the diffuse cervicofacial LM patients will require maxillomandibular reconstruction for bony overgrowth.

A discrete LM, in contrast, presents as a localized malformation (►Fig. 8). The clinical sequelae are related to its size and the anatomy involved. Lesions within the tongue or

Fig. 10 (A) Airway obstruction secondary to lymphatic malformation. (B) Immediate postop after debulking the obstruction with a CO₂ laser. (C) Resolved airway obstruction after complete healing.

Fig. 11 A massively enlarged tongue generally requires surgical reduction.

glottis can result in significant airway compromise. Lesions on the tongue can manifest clinically as small blebs that bleed and become infected. These cases can usually be managed with laser resurfacing or coblation (►Fig. 9). Glottic involvement is best managed by debulking the airway obstruction with a CO₂ laser (►Fig. 10). A tracheostomy tube should always be in place for this type of airway procedure. If the tongue is massively enlarged, laser therapy is inadequate and surgical reduction is generally required (►Fig. 11).

Both types of LMs can be complicated by swelling caused by systemic viral or bacterial infections. This typically subsides with the resolution of infection, but occasionally the LM itself will become infected and require antibiotic and steroid therapy.^{15,16,18,22}

Arteriovenous Malformations

Arteriovenous malformations (AVMs) are uncommon, but are most often found in the head and neck region as pulsatile masses.²³ They can sometimes present as massive bleeding after dental extraction. Arteriovenous malformations follow four distinct stages (quiescent, growing, symptomatic, and decompensating) and the stage of the disease dictate treatment plan.⁴ As with other types of vascular malformations already discussed, treatment involves a multidisciplinary approach and can involve both interventional radiology and surgery. Moreover, treatment can be used palliatively to relieve pain or other symptoms, or as part of a combined treatment plan with intent to completely eliminate the lesion.^{24,25} Ligation of feeding vessels should never be done as this leads to rapid recruitment of collaterals and heightens vascularity.

Conclusion

It is important to recognize the signs and symptoms of vascular anomalies of the airway as the presentation can be elusive. It is equally imperative to recognize that the initial treatment for each of these lesions is the creation of a secure,

patent airway. The specific treatment for these lesions depends on a number of factors, including the type, size, and location of the lesion. Treatment can be palliative in nature or performed with intent for cure. Recurrence and incomplete treatment is common, but satisfactory results can be achieved with proper management through a multidisciplinary approach.

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