

**TITLE: Vascular Tumors of the Head and Neck**  
**SOURCE: Grand Rounds Presentation, UTMB, Dept. of Otolaryngology**  
**DATE: April 11, 2001**  
**RESIDENT PHYSICIAN: Russell D. Briggs, MD**  
**FACULTY PHYSICIAN: Anna Pou, MD**  
**SERIES EDITOR: Francis B. Quinn, Jr., MD**

[ [Grand Rounds Index](#) | [UTMB Otolaryngology Home Page](#) ]

---

*"This material was prepared by resident physicians in partial fulfillment of educational requirements established for the Postgraduate Training Program of the UTMB Department of Otolaryngology/Head and Neck Surgery and was not intended for clinical use in its present form. It was prepared for the purpose of stimulating group discussion in a conference setting. No warranties, either express or implied, are made with respect to its accuracy, completeness, or timeliness. The material does not necessarily reflect the current or past opinions of members of the UTMB faculty and should not be used for purposes of diagnosis or treatment without consulting appropriate literature sources and informed professional opinion."*

---

## **INTRODUCTION**

Vascular tumors of the head and neck comprise a group of neoplasms that share a common origin with components of the vascular system. While these tumors do have an anatomic origin with the vascular system, they have vastly different presentations, biologic characteristics, and management options between them. A number of authors have classified vascular lesions based on descriptive, pathologic, embryological, or biological characteristics, however no classification scheme has been universally accepted. Vascular tumors of the head and neck can be divided into benign and malignant varieties which allows for comparisons between tumor types. This division of tumor types will be used as the classification scheme for the purposes of this text.

## **BENIGN TUMORS**

### Vascular Birthmarks

Nomenclature is the major obstacle in the understanding and management of vascular birthmarks. Much of the confusion stems from early textbook classifications that offered an array of histologic and descriptive words to document these lesions. Terms describing hemangiomas have included "capillary," "juvenile," "strawberry," and "cavernous," among others. These terms have permeated the medical literature and have lead to a great deal of confusion. Mulliken and Glowacki have revised the classification system of vascular birthmarks into hemangiomas and vascular malformations. According to this system, vascular birthmarks are differentiated into these two categories according to their biologic, cellular, and clinical characteristics.

### *Hemangiomas*

Hemangiomas are the most common tumor of infancy occurring in 8-12% of infants. A female predominance has been reported to range from 3:1 to 5:1. Most of these lesions are small, solitary, and uncomplicated, however 60% do arise on the face presenting a cosmetic concern to the parents.

Clinical presentation is the most important factor in making the proper diagnosis of a hemangioma. A hemangioma is usually not seen at birth, but subsequently arises within the first 8 weeks of life. Many hemangiomas are appreciated at birth as a cutaneous precursor lesion, which may vary from a faint red macule to a pale nevus that may mimic a bruise. Only rarely does a fully developed hemangioma present at birth, and in these rare instances, usually do not proliferate further and begin to regress quite rapidly. For the majority of hemangiomas, however, the natural history consists of a proliferative phase that usually lasts 3 to 9 months, followed by a slow regression phase. Most hemangiomas remain as well-circumscribed lesions 0.5-5 cm in diameter and involve the superficial layer of skin, thus being appropriately called superficial hemangiomas. Some, however, may proliferate into the lower dermis and subcutaneous tissue with little involvement of the superficial dermis. These deeper lesions, once labeled "cavernous" hemangiomas are more appropriately designated as deep hemangiomas. The involution phase of hemangiomas typically progresses at a rate of 10% per year, so that 50% have regressed by five years. After complete regression, normal skin is restored in 50% of patients while 10-38% have permanent changes including scarring caused by ulceration, skin atrophy, puckering, and fibrofatty residuum. However, of the remaining 50% that do not involute by 5 years, 80% will leave a substantial residual cosmetic deformity.

Complications from hemangiomas may occur in 20% of all hemangiomas, but few are life threatening. Ulceration is the most common complication occurring in less than 10% of patients, typically during the rapid proliferation phase. Compromise of function may occur with compression of vital structures such as those hemangiomas that affect the upper eyelid which may impair vision, leading to amblyopia or strabismus. Hemangiomas in the oropharynx, larynx, or nasal passages can lead to respiratory or feeding disturbances.

Hemangiomas occurring in the larynx usually occur in the subglottis and are found mainly in the pediatric population. These lesions present classically with a previously healthy infant who suddenly develops biphasic stridor during the first few months of life, many times misdiagnosed as croup. These lesions are usually located in the posterolateral subglottis and are submucosal, soft, and compressible masses. These lesions behave like other hemangiomas with most regressing by age 5 years. For this reason, a conservative approach is often employed as long as the airway is acceptable. Approximately 50% of these hemangiomas are associated with cutaneous counterparts, typically in the beard area of the face.

In certain cases such as deep hemangiomas or hemangiomas detected early in neonatal life that have not reached the progressive phase, radiographic investigation of

such lesions may be indicated. Ultrasound with Doppler flow imaging is the most cost effective, however MRI imaging is the most informative noninvasive study, safely providing the information of both CT and angiographic data. MRI can accurately determine the extent of the lesion and the finding of serpentine high-volume flow voids surrounded by nonvascular soft tissue is characteristic of hemangiomas. One particular indication for further radiographic study is for individuals with large facial hemangiomas, usually unilateral, as these may be associated with Dandy-Walker syndrome or posterior fossa malformations.

Indications for treatment have historically been based on tissue destruction or disfigurement, severe bleeding, obstruction of vital functions, high-output cardiac failure, or platelet trapping with a coagulopathy. Recently, many authors have advocated early treatment of facial hemangiomas due to a better understanding of the diagnosis, histopathology, and natural history of these vascular lesions. In addition, these authors contend that advances in laser technology, appropriate use of systemic steroids, safer pediatric anesthesia, and improved surgical techniques have allowed for better cosmetic results and, in turn, improved psychological well-being in these children.

A number of treatment options are available for management of hemangiomas during the proliferative phase including observation, systemic steroid therapy, intralesional steroid therapy, pulse-dye laser therapy, surgical debulking or excision, arterial embolization, alpha 2b-interferon, radiation therapy or combinations of the above.

Most parents desire some method of treatment for their child's hemangioma, and they have difficulty accepting that many lesions do best if they are left alone to involute naturally, leaving either normal or slightly blemished skin. As most will eventually regress, initial management generally consists of observation. In these cases where observation is chosen, serial photographs demonstrating the slow involution are helpful. Regular visits with parental reassurance at 3 to 6 month intervals are sufficient to document the involution of the hemangioma.

Systemic steroid therapy was once considered first-line therapy for complicated hemangiomas. Steroid use, if used cautiously, is effective with minimal long term side effects. This therapy arrests the growth of the lesion and therefore an immature proliferating hemangioma is far more responsive to steroids than a stable or involuting lesion. Systemic steroids may be used in selected infants that 1) have a rapidly growing lesion that seriously distorts facial features, 2) have a lesion causing recurrent bleeding, ulceration, or infection, 3) have a lesion that interferes with normal physiological function (breathing, hearing, vision, or eating). In general, 2 to 4 mg/kg/day of prednisone are given orally in a single daily dose for as many as 6 weeks, followed by a slow taper over 2-3 months. Results are variable with a dramatic response in 30%, an equivocal response in 40%, and no response in 30%. Side effects include cushinoid effects, adrenal suppression, infections, and growth suppression. The child must be closely monitored during the treatment course for any evidence of steroid toxicity.

Intralesional steroid therapy has been used predominately in cases of vision-threatening periorbital hemangiomas, but is also used for cases involving globular tumors of the lips, nasal tip, cheek, or ears. Some authors also utilize intralesional injections for those lesions that rebound from prior systemic steroid therapy and for deep proliferating lesions in patients not candidates for systemic steroid therapy. Typical doses have included 1-2 mL of a combination of beta-methasone (6mg/mL) and triamcinolone (40 mg/mL) given in repeat injections as necessary at 4-6 week intervals. This therapy has the benefit of a rapid action typically within one week, however its success is similar to that of systemic steroids. Complications can be severe and include full thickness eyelid necrosis and blindness.

Pulse-dye lasers have become increasingly popular for the treatment of facial hemangiomas. These lasers, however are only useful for superficial hemangiomas as the laser penetrates only 0.75-1.5 mm deep. It has been quite useful in cases with ulceration as these typically are rapidly proliferating superficial lesions. The laser is relatively vessel specific and thus has a low risk of scarring and is safe to use in young children. It is also quite useful for patients with particular cosmetic defects after the involution phase of the hemangioma is complete (residual telangiectasias).

Surgical therapy for hemangiomas has historically been reserved as a last resort for treatment of complicated lesions. Examples of such indications include eyelid lesions, bulky lesions that will likely leave a sagging bag of skin at involution, lesions involving the vermilion border, nasal tip, and eyebrow. Surgery can also be utilized for correction of redundant skin after spontaneous resolution has occurred. CO<sub>2</sub> laser excision of large subglottic hemangiomas is useful in cases of severe obstruction and those cases that have not responded to systemic steroids.

Arterial embolization is typically reserved only for those alarming hemangiomas that are inoperable, hemorrhaging, or have not responded to other methods. Risks include thrombosis and backflow of material into undesired vessels, unintentional embolization, and even blindness. Even with apparently effective treatment, results are usually temporary.

Alpha 2b-interferon, radiation, and cryotherapy have been used in the past but these are no longer recommended due to serious complications.

Kasabach-Merritt syndrome is a particular entity associated with large hemangiomas or diffuse hemangiomatosis throughout the body. This disorder is associated with severe thrombocytopenia and hemorrhage caused by localized platelet trapping. The coagulopathy associated with this syndrome may require administration of platelets, fresh frozen plasma, and cryoprecipitate. Lesions implicated in this syndrome are typically treated with one of the above options, however, many lesions do not respond to medical therapy and require surgical intervention to correct the coagulopathy.

*Vascular Malformations*

Compared with hemangiomas, vascular malformations are developmental anomalies. Thus, all cutaneous vascular malformations, by definition, are present at birth. These lesions have no gender predilection and are characterized by commensurate growth. These lesions do not have a proliferative and involuting phase, rather, they persist throughout the life of the affected person without treatment. These malformations are categorized into their component vessels: capillary, venous, arterial, lymphatic, or mixed.

Capillary malformations, commonly referred to as “port-wine” stains, occur in approximately 1% of all newborns. Diagnosis is usually straightforward with most occurring on the face, especially in the distribution of the trigeminal nerve. They are classically unilateral with respect to the midline. Initially, capillary malformations are pink, flat and sharply demarcated lesions. In older individuals, they often become more nodular in appearance and a darker, purple color develops. Most of these lesions are isolated anomalies, however some may signal underlying disorders that require additional investigation. Sturge-Weber syndrome is a sporadic disorder associated with a capillary malformation involving at least the V1 distribution with associated ipsilateral leptomeningeal venous malformations, atrophy and calcifications in the adjacent cerebral cortex, seizures, visual field defects, and mental retardation. These patients may also develop retinal detachment and either congenital or acquired glaucoma. Patients diagnosed with this condition should have an early ophthalmologic and neurologic examination with MRI imaging to detect the associated brain anomalies.

Treatment of capillary malformations are varied. Cosmetic concealing makeup are formulated to match the individual skin tones of patients. However, these are only helpful for flat lesions and may take up to 20 minutes to apply. Similarly, tattooing of lesions has fallen out of favor due to a poor cosmetic result secondary to scarring, a mask-like appearance, and poor retention of the pigment. Surgical excision is also losing favor due to the high risk of hypertrophic scarring and permanent depigmentation in grafted areas. Tissue expanders may provide role in moderate sized lesions allowing for closures along facial unit borders. The pulse-dye laser has become the most effective treatment for capillary malformations although up to 50% of successfully treated lesions recur.

Venous malformations are low-flow lesions that are deeply invasive and may worsen slowly through the course of the patient’s life. As with other vascular malformations, these are present at birth. Diagnosis is usually clinical as these usually compress easily with pressure and swell when dependent or with increased venous pressure. Management of these lesions varies greatly depending on the size, location, and clinical presentation. Asymptomatic lesions can be observed, however when loss of function or cosmesis becomes unacceptable, treatment often entails multiple procedures over several years, using a combination of sclerotherapy and surgical excision.

Lymphatic malformations , also termed cystic hygromas or lymphangiomas, are vascular tumors arising from maldevelopment of the lymphatic system. As with other vascular malformations, the lesions are usually slow growing but sometimes expand

rapidly with respiratory tract infections which can threaten the airway in patients with head and neck lesions. Complete excision is the recognized primary treatment modality, but this goal can be achieved in only one-third of cases if vital and functionally important structures are not sacrificed. Definitive management is indicated when vital structures are endangered or when episodic hemorrhage ensues. Another indication for surgical management is associated macroglossia which may lead to articulation, feeding, dental occlusion, or airway problems. Nonsurgical treatment with diathermy, radiation therapy, and most sclerosing agents has met with only limited clinical success. A newer sclerosing agent, Picibanil (OK-432) has shown promising preliminary results. MRI can be used to establish the diagnosis and estimate the extent of the lesion. It is also helpful for following patients after treatment to detect recurrence.

Arteriovenous malformations are high-flow malformations resulting from persistent abnormal direct arterial and venous connections. While present at birth (congenital type), many of these lesions are not recognized until late childhood, even into adulthood. Violaceous color of overlying skin, palpable thrills, and bruits should suggest such lesions. Also hyperhidrosis, hypertrichosis, and hyperthermia may be present in the area. Although usually clinically apparent, these lesions can be diagnosed with Doppler ultrasound exams or with MR imaging with angio-sequences which can better define the anatomic extent. Angiography can establish a nidus for such lesions but is usually reserved for therapeutic efforts. Embolization can be used as a therapeutic modality, however many lesions recur. Embolization with operative excision within 48 hours allows for decreased bleeding complications and allows for improved elimination of the lesion.

### Angiofibromas

Nasopharyngeal angiofibromas are the most common benign tumors of the nasopharynx yet represent only 0.5% of all head and neck tumors. In the past, the term juvenile nasopharyngeal angiofibroma was used, however, this term is inappropriate because the neoplasms occur in older patients as well. It is a benign, yet highly vascular tumor that occurs almost exclusively in adolescent boys. These lesions may occur at any time, but are most commonly diagnosed between the ages of 14 and 25.

The usual presenting complaints are painless, progressive nasal obstruction and recurrent epistaxis for no apparent reason. Clinically, the lesion appears as a firm, friable mass in the nasopharynx or nose. As the tumor enlarges, the tumor interferes with eustachian tube function producing a conductive hearing loss. Persistent, unilateral middle ear effusions in this patient population should alert the physician to this possibility and the need for appropriate nasopharyngeal examination. As the tumor enlarges, more obvious signs may develop related to the direction of tumor spread. These include orbital findings such as proptosis or limitation of ocular movements, cranial nerve deficits, sinusitis, noticeable cheek swelling, or meningitis. These tumors have a characteristic for aggressive local growth and a tendency for base of skull and intracranial invasion. It is believed that nasopharyngeal angiofibromas arise in the

fibrovascular stroma normally present in the posterolateral wall of the roof of the nasal cavity where the sphenoidal process of the palatine bone meets the horizontal ala of the vomer. This is located at a point just above the sphenopalatine foramen, thus allowing for extension into the nose, nasopharynx, sinuses, temporal or infratemporal fossa, or cranium.

The diagnosis is typically made by clinical exam and radiographic findings. On imaging, opacification of one or paranasal sinuses is usually seen. The presence of anterior bowing of the posterior wall of the maxillary sinus is pathognomic. Computed tomography or MRI can document the extent of the lesion and provide the diagnosis in most cases. Biopsy is rarely recommended, however, if necessary should be done in a controlled setting under general anesthesia with preparations made for tumor removal. Angiography can delineate the vascular supply to the tumor, however is typically used for embolization of the tumor prior to surgery. When possible, diagnostic angiography and embolization should be carried out simultaneously 48 hours prior to surgery. While most common feeding vessels are from the external carotid system (internal maxillary and ascending pharyngeal arteries), internal carotid contribution may occur with large tumors and therefore angiography should include bilateral internal and external carotid systems.

Surgery is the recommended treatment for this tumor, however, because the major vascular supply arises from the internal and external carotid arteries, surgery can result in extensive blood loss. Most surgeons utilize preoperative arterial embolization to decrease intraoperative blood loss and some prescribe stilbestrol for 5 weeks preoperatively to reduce the vascularity. Additional measures of managing blood loss include autologous blood banking preoperatively and the use of a Cell Saver device during the procedure. Depending on the location, a number of surgical approaches have been utilized. Transnasal endoscopic removal of tumor is generally limited to those tumors confined to the nasopharynx, nose, or paranasal sinuses. A lateral rhinotomy or midfacial degloving procedure combined with a medial maxillectomy or LeFort I osteotomy provides exposure of the nasal cavity, ethmoid sinuses, maxillary sinuses, and pterygomaxillary fossa. Transpalatal and transpharyngeal routes achieve access inferiorly and preauricular subtemporal access to the infratemporal fossa laterally. A facial translocation or maxillary swing can be extended to provide further access to the infratemporal fossa and skull base. Intracranial extension may be removed with a formal craniotomy, if necessary. The recurrence rate after surgical treatment varies from 20% up to 40% in some studies, typically due to incomplete resection. Recurrent lesions may be managed with additional surgery, however radiotherapy and chemotherapy can also be used. Because of the effects of radiotherapy on the growth of the craniofacial skeleton and the potential carcinogenic effects of radiotherapy, many clinicians reserve radiotherapy for unresectable, life threatening tumors. Chemotherapy is usually offered for unresectable disease in which radiation is ineffective.

## **MALIGNANT TUMORS**

Angiosarcoma

Angiosarcoma of the head and neck is extremely rare. These tumors represent less than 1% of all sarcomas in humans with 50% of the cases involving the head and neck region. The most common region in the head and neck is the scalp and facial skin, with the neck, oropharynx, and sinonasal tract following in decreasing order. Most of these tumors are rapidly growing requiring prompt diagnosis and treatment if long term survival is to be expected. The prognosis is very much dependent on tumor size and degree of cellular differentiation. Treatment consists of wide surgical resection with tumor margins being another important prognostic factor. Unfortunately, accurate identification of margins are difficult due to the presence of anastomosing vascular channels dissecting the underlying stroma. Histologically, they are classified as either high or low grade depending on the number of mitoses observed and the overall appearance of the lesion. Radiation therapy is minimally effective and usually given as an adjunct to surgical therapy or as palliative treatment. Angiosarcomas arising in the sinonasal tract have been found to behave less aggressively than those found in other areas, thereby allowing for improved survival in these areas. Overall survival is poor, less than 50% at five years.

### Hemangiopericytoma

Stout and Murray first described hemangiopericytomas in 1942. These tumors arise from the pericytes of Zimmerman, which are cells that normally give mechanical support to capillaries and regulate luminal size. Overall, 25% of hemangiopericytomas arise in the head and neck, however they represent only a small portion of all head and neck tumors. Many series on this lesion in the head and neck literature involve the sinonasal tract as the primary site. Similar to the angiosarcomas, hemangiopericytomas arising in the sinonasal tract behave less aggressively and have been termed “hemangiopericytoma-like” when arising in this tissue.

The tumor typically presents as a painless mass in all age groups predominately in the 6<sup>th</sup> and 7<sup>th</sup> decades of life, with no sex predilection. The etiology is unknown, although these lesions have been linked to trauma, prolonged steroid use, and hormonal imbalances. The treatment of choice is wide surgical excision which are known to insidiously recur even years later (57%). Well-differentiated tumors rarely metastasize, however given their propensity to recur locally, these lesions are considered malignant. Similar to angiosarcomas, these lesions are graded as high or low grade tumors with survival rates improved in the latter group. Radiation therapy is usually reserved for recurrent lesions not amenable to surgical excision or those with a more active histology. These tumors are poorly chemosensitive, however a recent report documented a good response to alpha-interferon in two patients.

### Kaposi's Sarcoma

Kaposi's sarcoma is a multicentric proliferation of vascular and spindle cell components, first described in 1872. It is now considered to be a viral-induced tumor and it is unclear as to whether the lesion is a true tumor or hyperplasia. It is strongly

affiliated with AIDS and its course is greatly influenced by the immune status of the individual. Kaposi's sarcoma has four distinct clinical entities: classic, endemic, transplant-associated, and AIDS-related. Classic Kaposi's is not associated with HIV and typically affects elderly men of Italian heritage. Endemic Kaposi's is seen in endemic portions of Africa among young black children. This entity is rapidly progressive and affects lymph nodes and internal organs diffusely. Transplant-associated Kaposi's affects transplant recipients that are immunosuppressed and is correlated with loss of cellular immunity. AIDS related Kaposi's is found primarily in male homosexuals with 40% of affected AIDS patients developing this entity. Lesions occur in many cutaneous locations, especially along lines of cleavage and on the tip of the nose. Various treatments have been used with Kaposi's with varied success. Small lesions can be surgically excised but more recent therapies have concentrated on low-dose radiation and intralesional chemotherapy and sclerosing solutions. For larger lesion, chemotherapy is effective but can be quite morbid to the HIV infected individual.

## **OTHER VASCULAR TUMORS**

### **Parangliomas**

Parangliomas of the head and neck are typically benign, slow growing tumors arising from widely distributed paraganglionic tissue thought to originate from the neural crest. Paranglia in the head and neck region are closely aligned with the distribution of the parasympathetic nervous system and often have a close spatial relationship with neural or vascular structures. Paranglia have been shown to have chemoreceptor roles with modulation of respiratory and cardiovascular function.

Carotid bodies are the largest collection of paranglia in the head and neck region and appear as small ovoid structures on the medial aspect of the carotid bifurcation on each side of the neck. Paranglia are also located in other locations in the head and neck including the middle ear, jugular bulb, ganglion nodosum of the vagus nerve, larynx, and base of the heart. Histologically, their appearance is similar to the normal histology of the paranglia and includes two cell types. Type I cells (chief) cells are APUD type cells with copious cytoplasm and large round or oval nuclei. Their cytoplasm contains dense core granules that store and release catecholamines. Type II (sustentacular) cells are elongated cells that closely resemble Schwann cells although their function is not entirely clear. The two cell types are arranged into clusters with a core of chief cells surrounded by sustentacular cells embedded in a fibrous stroma. These clusters make up the fundamental histologic structure (termed "Zellballen") and may be somewhat enlarged in parangliomas. Nuclear pleomorphism and cellular hyperchromatism are common in benign parangliomas and should not be considered evidence of malignancy. In fact, there are no clear histologic characteristics of malignancy in these lesions. Malignancy is based on the clinical finding of metastasis, not on histologic examination.

Terms used in the past to describe parangliomas have included glomus tumors (general term used to describe a cluster of specialized cells and more appropriately

applied to tumors of the skin and superficial tissues of the extremities), chemodectomas (describing a tumor from chemoreceptor origin), carotid body tumors, and nonchromaffin tumors (related to staining characteristics). Currently, the correct terminology is paraganglioma based on the anatomical location. The predominant paragangliomas of the head and neck include carotid paragangliomas, jugulotympanic paragangliomas, vagal paragangliomas, and laryngeal paragangliomas. For the purposes of this text, jugulotympanic paragangliomas will be excluded as these have been discussed in other grand rounds presentations.

### *Carotid paragangliomas*

Carotid paragangliomas are the most common paragangliomas of the head and neck comprising approximately 60% of the total. Their incidence is rare, occurring in approximately 0.12% of all surgical specimens. They can occur at any age but mean age at diagnosis is 45-50 years. A slightly higher female predominance persists. These tumors may be multicentric 10% of the time with bilateral carotid body lesions being the most common combination in multicentric lesions. Carotid paragangliomas are familial in 20% of cases in an autosomal dominant fashion with a higher propensity for multicentric lesions in familial types. Malignancy occurs in approximately 10% of cases which ranks carotid paragangliomas as the most frequently malignant of all paragangliomas in the head and neck.

The lesions usually present as a painless mass that is slow growing along the anterior border of the sternocleidomastoid muscle. They tend to splay the carotid bifurcation as they enlarge and can extend along the carotid artery to the skull base. Patients typically have noted the mass for many years, on average 2-8 years. Very large lesions may present with vocal cord paralysis or dysphagia. On examination, the masses are freely mobile laterally however they are immobile in a cephalad-caudad direction. The mass may be pulsatile and a bruit may be auscultated. Carotid paragangliomas may also produce catecholamines, therefore, patients with symptoms of catecholamine excess should be screened for urinary metanephrines and VMA (vanillyl mandelic acid) as well as circulating catecholamines.

Diagnosis can be made with either CT or MRI which shows a mass arising from the carotid bifurcation and displaces the internal and external carotid arteries. The diagnosis is confirmed with arteriography by revealing a characteristic tumor blush at the carotid bifurcation called the lyre sign. This modality can establish the diagnosis, demonstrate multiple lesions, determine the size and vascularity of the tumor, as well as evaluate its blood supply. Additionally, it can be modified to include selective, controlled balloon occlusion of the internal carotid artery to evaluate cerebral cross-flow. This information is important in preoperative counseling of the patient as to the relative risk of surgery. Since embolization is not often used with carotid body paragangliomas, MRA may be an appropriate alternative to angiography in selected cases. Biopsy, including fine needle aspiration is unnecessary and contraindicated in the evaluation of paragangliomas. Some argue for routine screening of urinary metanephrines and VMA

and serum catecholamines for all cases, however others recommend these tests only for familial forms or in the presence of catecholamine excess.

Surgery is the mainstay of treatment for carotid paragangliomas. The recurrence rate is approximately 10% with the mortality rate intraoperatively of up to 8%. Because of their close approximation to important vessels and nerves, there is a real risk of morbidity (usually CN X-XII and vascular injuries). Tumor size is important because those greater than 5 cm in diameter have a markedly higher incidence of complications (67% vs. 15%). An extensive preoperative workup is essential for safe resection of these tumors. In cases where the internal carotid artery may require resection, a vascular surgeon should be available for assistance. Perioperative alpha and beta adrenergic blockers should be available for all catecholamine producing tumors. Most authors do not recommend embolization preoperatively as this interferes with the subadventitial dissection necessary for removal of these tumors. Complications include permanent nerve palsy (20%), baroreceptor failure (bilateral), and “first-bite” syndrome (from denervated parotid myoepithelial cells). Radiation is usually reserved for incompletely excised tumors (with intracranial extension), recurrent tumors, or poor surgical candidates. The mortality rate of untreated carotid paragangliomas is estimated at 8% per year, indicating the indolent nature of this tumor. Malignancy rate in carotid paragangliomas is estimated between 2-10%. There is no histologic criteria for malignancy, only the finding of spread to regional lymph nodes or distant sites.

### *Vagal paragangliomas*

Vagal paragangliomas arise most commonly at the level of the nodose ganglion but may occur anywhere along the course of the vagus nerve in the neck. Mean age at presentation is 50 years and there is a slight female predominance. The presentation is usually one of a painless slow growing mass located behind the angle of the mandible that has been present for many years. The patient may complain of tongue weakness, hoarseness, or have a Horner’s syndrome. Imaging studies such as a CT or MRI may delineate the tumor from surrounding structures with angiography classically demonstrating a tumor blush that displaces the carotid artery anteriorly and medially. Catecholamine producing vagal paragangliomas are virtually nonexistent however are multicentric in approximately 25% of sporadic lesions. Vagal paragangliomas arising in the familial form exhibit multicentricity in 78% of patients. Malignancy in these tumors is estimated at 18%.

Management is complicated due to their propensity for multicentricity. Some argue that surgical treatment is the mainstay however cranial nerve deficits can be expected with their removal. Radiation can be used but responses are often suboptimal.

### *Laryngeal paragangliomas*

Paragangliomas of the larynx usually arise from the superior laryngeal paraganglia above the anterior part of the vocal folds near the aryepiglottic fold.

Hoarseness and dysphagia are the most common complaints and these are associated with high rates of malignancy. Laryngeal lesions usually require wide local excision or partial laryngectomy. Radiation has not been effective in controlling these rare entities.

### Bacillary Angiomatosis

Although not clinically a tumor, bacillary angiomatosis can clinically mimic many of the other previously described vascular tumors, particularly Kaposi's sarcoma. This skin lesion is a vasoproliferative response to infection by *Bartonella* species of bacteria. It is this bacteria that causes cat scratch disease in immunocompetent children, however bacillary angiomatosis typically occurs in immunocompromised individuals. Treatment of this condition involves appropriate antibiotic therapy, usually erythromycin.

## References

- Carrau RL, Snyderman CH. Juvenile nasopharyngeal angiofibroma. In: Myers EN, Ed. Operative Otolaryngology-Head and Neck Surgery. Philadelphia: W.B. Saunders, 1997.
- Johnson JT. Carotid paraganglioma. In Myers EN, Ed. Operative Otolaryngology-Head and Neck Surgery. Philadelphia: W.B. Saunders, 1997.
- Maves MD, Stevens, C. Vascular tumors of the head and neck. In Bailey BJ, Ed. Head and Neck Surgery-Otolaryngology. Philadelphia: Lippincott-Raven, 1998.
- Willing SJ. Atlas of Neuroradiology. Philadelphia: W.B. Saunders, 1995.
- Gnepp DR. Diagnostic and Surgical Pathology of the Head and Neck. Philadelphia: W.B. Saunders, 2001.
- Mulliken JB, Young AE. Vascular Birthmarks: Hemangiomas and Malformations. Philadelphia: W.B. Saunders, 1988.
- Fagan JJ, Snyderman CH, Carrau RL, et al. Nasopharyngeal angiofibromas: selecting a surgical approach. Head and Neck, August 1997, 391-398.
- Danesi G, Panizza B, Mazzoni A, et al. Anterior approaches in juvenile nasopharyngeal angiofibromas with intracranial extension. Oto-HNS, February 2000, 277-283.
- Brown TJ, Firedman J, Levy ML. The diagnosis and treatment of common birthmarks. Clinics in Plastic Surgery, October 1998, 509-525.
- Greinwald JH, Burke DK, Sato Y, Poust RI, Kimura K, et al. Treatment of lymphangiomas in children: An update of Picibanil (OK-432) sclerotherapy. Oto-HNS, October 1999, 381-387.
- Williams EF, Stanislow P, Dupree M, Mourtzikos K, Mihm M, Shannon L. Hemangiomas in infants and children. Archives in Facial Plastic Surgery, April-June 2000, 103-111.
- Netterville JL, Jackson CG, Miller FR, et. al. Vagal paraganglioma: a review of 46 patients treated during a 20 year period. Archives of Oto-HNS, October 1998, 1133-1140.
- Ungkanot K, Byers RM, Weber RS, Callender DL, Wolf PF, Goepfert H. Juvenile nasopharyngeal angiofibroma: an update on therapeutic management. Head and Neck, January-February 1996, 60-66.

Netterville JL, Reilly KM, Robertson D, Reiber ME, et al. Carotid body tumors: a review of 30 patients with 46 tumors. *Laryngoscope*, February 1995, 115-125.

Carew JF, Singh B, Kraus DH. Hemangiopericytoma of the head and neck. *Laryngoscope*, September 1999, 1409-1411.

Billings KR, Fu YS, Calcaterra TC, et al. Hemangiocyoma of the head and neck. *American Journal of Otolaryngology*, July-August 2000, 238-243.