Canalplasty with long-term ventilation tube versus tympanoplasty for chronic tympanic membrane atelectasis

Alaa Abdel Azim Swilam

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.HemangiopericytomaStout 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 6th and 7th 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 SarcomaKaposi'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 TUMORSParagangliomasParagangliomas of the head and neck are typically benign, slow growing tumors arising from widely distributed paraganglionic tissue thought to originate from the neural crest. Paraganglia 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. Paraganglia have been shown to have chemoreceptor roles with modulation of respiratory and cardiovascular function.Carotid bodies are the largest collection of paraganglia 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. Paraganglia 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 paraganglia 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 paragangliomas. Nuclear pleomorphism and cellular hyperchromatism are common in benign paragangliomas 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 paragangliomas 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 nonchrommafin 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 paragangliomasCarotid 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. Mot 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 denerved 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 paragangliomasVagal 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 paragangliomasParagangliomas 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 lesion usually require wide local excision or partial laryngectomy. Radiation has not been effective in controlling these rare entities.Bacillary AngiomatosisAlthough 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.