

**TITLE: Primary Parapharyngeal Tumors**

**SOURCE: Grand Rounds Presentation, UTMB, Dept. of Otolaryngology**

**DATE: March 22, 2006**

**RESIDENT PHYSICIAN: Jing Shen, MD**

**FACULTY PHYSICIAN: Shawn Newlands, MD, PhD**

**SERIES EDITORS: Francis B. Quinn, Jr., MD and Matthew W. Ryan, MD**

---

*"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."*

---

The parapharyngeal space is a complex anatomical area. Primary parapharyngeal tumors are rare tumors and only account for 0.5% of all the head and neck tumors. About 80% of these tumors are benign and the other 20% are malignant. The differential diagnosis of the primary parapharyngeal tumor includes salivary gland neoplasm, neurogenic tumor and many miscellaneous tumors.

## **Review of Anatomy:**

The parapharyngeal space is often described to be a deep potential neck space shaped as an inverted pyramid. The base of the pyramid is at the skull base and the apex is at the greater cornu of the hyoid bone. The boundaries of the parapharyngeal space are as following:

- 1) Superior boarder: small portion of the temporal bone. There is a fascia connecting the medial pterygoid plate to the spine of the sphenoid. This fascia lies medial to the foramen ovale and foramen spinosum. These foramina are not considered to be in the parapharyngeal space.
- 2) Inferior boarder: junction of the posterior belly of the digastric muscle and the greater cornu of the hyoid bone
- 3) Medial boarder: pharyngobasilar fascia and pharyngeal wall. Superiorly it is formed by the approximation of the fascia from the tensor veli palatine muscle to the medial pterygoid muscle. Inferiorly the medial boarder is contiguous with the fascia over the superior constrictor muscle.
- 4) Lateral boarder: medial pterygoid muscle fascia, the ramus of the mandible, retromandibular portion of the deep lobe of the parotid gland and posterior belly of the digastric muscle. Sphenomandibular ligament extends from the spine of the sphenoid to the lingual of the mandible. The medial pterygoid muscle fascia incorporates this ligament superiorly to the skull base as a dense fascia. This fascia separates the inferior alveola nerve, lateral pterygoid muscle and mandibular condyle from the parapharyngeal space. Stylomandibular ligament extend from the styloid process to the angle of the mandible. This ligament is a part of the stylomandibular tunnel. The other two boundaries of the stylomandibular tunnel are ascending mandibular ramus and skull base.

Primary tumors of the deep lobe of the parotid gland can grow into the parapharyngeal space through this tunnel. The constriction of this tunnel on tumor growth gives a characteristic “dumbbell” shape on the CT scan.

- 5) Posterior boarder: vertebral fascia and paravertebral muscles.
- 6) Anterior boarder: pterygomandibular raphe and medial pterygoid fascia.

Clinically, the parapharyngeal space should be considered in two spaces: pre-styloid space and post-styloid space. Fascia from the styloid process to the tensor veli palatine muscle divides the parapharyngeal space into these two compartments. The pre-styloid space is anterolateral and contains retromandibular portion of the deep lobe of the parotid gland, minor or ectopic salivary gland, a small branch of the CN V to the tensor veli palatine muscle, ascending pharyngeal artery and pharyngeal venous plexus. The majority of the pre-styloid space is actually fat. The post-styloid compartment is posteromedial and contains internal carotid artery, internal jugular vein, CN IX to XII, cervical sympathetic chain, lymph nodes and glomus bodies.

### **Differential diagnosis of primary parapharyngeal tumors:**

Most of the tumors of the parapharyngeal space are metastatic disease or direct extension from adjacent spaces. Primary parapharyngeal tumor is uncommon. Per literature, 80% tumors of the parapharyngeal space are benign and 20% are malignant. The location of the tumor can be helpful for differential diagnosis. Tumors in the pre-styloid space are most likely to be salivary gland tumor, lipoma, or rare neurogenic tumors. And all the structures in the post-styloid compartment are potential sources for post-styloid tumors.

In general, the most common primary parapharyngeal tumor is salivary gland neoplasm from the deep lobe of the parotid gland or from minor salivary gland tissues. Hughes reviewed 172 patients with parapharyngeal space neoplasm and found that pleomorphic adenoma was the most common neoplasm (40%), followed by paraganglioma (20%), neurogenic tumor (14%), malignant salivary gland tumor (13%), miscellaneous malignant tumors (7%), and miscellaneous benign tumors (6%). This result is consistent with most reports. However, there are few literatures reported neurogenic tumors as the most frequent entities. In one of the most recently published study on primary parapharyngeal tumors, Luna-Ortiz reported that neurogenic tumors represented 57% of all tumors in his study population.

Pleomorphic adenoma is the most common salivary gland tumor in the parapharyngeal space. It can originate in the deep lobe of the parotid gland as well as from the minor salivary gland tissue. A deep lobe parotid gland tumor can extends through the stylomandibular tunnel into the parapharyngeal space. This kind of tumor will give a characteristic “dumbbell” appearance on the CT scan. Also tumor can arise from the retromandibular portion of the parotid gland. It then can expand into the parapharyngeal space and displace tonsil and soft palate and cause obstruction of the nasopharynx. This kind of tumor is more rounded and will not give a “dumbbell” appearance. There is ectopic salivary tissue in the parapharyngeal fat lateral to the superior pharyngeal constrictor muscle. Salivary gland tumors arise from these tissues will not connect to the deep lobe of the parotid gland. The frequency of malignant parapharyngeal salivary neoplasm varied greatly in the literature. The reported malignant tumors include mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, malignant mixed carcinoma, squamous cell carcinoma, adenocarcinoma, and a case of malignant Warthin’s tumor.

In most of the reports neurogenic tumor is the second most common tumor in the parapharyngeal space. Schwannoma is the most common type of neurogenic tumor. All of the cranial nerves and sympathetic chain in the parapharyngeal space are capable to give rise to schwannoma, but the most common ones are the vagus nerve and the cervical sympathetic chain. The vagus nerve has been reported to be the nerve of origin in 50% of parapharyngeal schwannoma. Schwannomas in general are slow growing and usually do not affect the nerve of origin. The symptoms are mainly compressive.

Paraganglioma is the second most common neurogenic tumor in the parapharyngeal space. They are either vagal paragangliomas or carotid body tumors. Vagal paragangliomas are parapharyngeal in location in 2/3 of the cases. In contrast, carotid body tumors rarely extend above the posterior belly of the digastric muscle. According to Olsen, the occurrence varies from 8% to 30% in different studies. Only the extension above the level of the posterior belly of the digastric muscle justifies its inclusion as parapharyngeal tumor.

Neurofibroma is the third most common neurogenic tumor in the parapharyngeal space. Malignant neurogenic tumors have been reported in the parapharyngeal space include malignant schwannoma, malignant paraganglioma, malignant neuroblastoma, or sympatheticoblastoma.

There are numerous benign and malignant tumors reported as primary parapharyngeal tumors. Again majority of these tumors are rare and mentioned only in case reports.

## **Clinical presentation and Evaluation:**

Clinical detection of parapharyngeal tumor is difficult. The tumor has to grow to 2.5 to 3.0 cm to be detected clinically. The presentation can be very subtle. Generally it presents as an asymptomatic mass causing mild bulging in the soft palate or tonsillar region, or fullness near the angle of the mandible. Parotid tumors can displace the tonsil and neurogenic tumors often displace the posterior portion of the pharynx and posterior tonsillar pillar. With tumors enlarging superiorly, they can cause soft palate and nasopharyngeal swelling. And inferior growth of tumors result palpable masses at the angle of the mandible. The ensuing symptoms depend on the affected site. The pre-styloid lesion can present as serous otitis media, voice change, nasal obstruction, aspiration or dyspnea. The post-styloid lesion can compress the 9<sup>th</sup>, 10<sup>th</sup>, 11<sup>th</sup> and 12<sup>th</sup> nerve and cause hoarseness, dysphagia, dysarthria, or Horner's syndrome by tumor pressure on the superior cervical sympathetic ganglia. Cranial nerve palsy, pain, and trismus often suggest malignancy. Parapharyngeal tumors can be mistaken as peritonsillar abscess or tonsil tumors and cause delaying in diagnosis.

Other than a complete head and neck exam, imaging studies such as CT, MRI and angiogram are important modalities to support the diagnosis of parapharyngeal tumors and distinguish pre-styloid tumors from those in the post-styloid space.

On CT scan, salivary gland tumor is in pre-styloid space and displaces the carotid artery posteriorly. Also if there is a fat plane between the lesion and the deep lobe of the parotid gland, the mass is extraparotid. The most common enhancing extraparotid lesion on CT scan is the schwannoma. The schwannoma on the CT scan is in the post-styloid space and usually displaces the carotid artery anteromedially. Lesions that show enhancement on CT scan include

paragangliomas, hemangiomas, hemangiopericytomas, aneurysms, and schwannomas.

MRI provides the most useful preoperative information about the extent of the tumor and its relationship to surrounding structures. The relationship between the tumor and the carotid artery can be more clearly seen with MRI. If carotid involvement is suspected, an angiogram should be done. It can be used to accurately define the vascular anatomy and collateral circulation, to perform the carotid occlusion study and possibly to embolize the tumor preoperatively. Carotid occlusion test must always be done whenever a malignant tumor involves the post-styloid space or for extensive vascular tumors that surround the carotid artery at the level of the skull base.

In recent years increasing number of patients underwent fine needle aspiration of nonvascular lesions. Some investigators have found it to be very helpful for accurate diagnosis preoperatively.

### **Surgical approaches:**

The goal of parapharyngeal surgery is to provide adequate tumor visualization to achieve complete tumor removal, while preserve the surrounding nerves and vessels and control of any hemorrhage. Many surgical approaches have been reported in the literature. Overall, transcervical and transparotid approach are the two main approaches. They have been reported to be used alone or in combination of each other. They have also been used with mandibulotomy to increase exposure. Transcervical approach is usually used for post-styloid tumors and transparotid approach for pre-styloid tumors.

Transparotid approach is commonly used for deep lobe parotid tumors. It starts with a superficial parotidectomy with facial nerve preservation. The facial nerve is then separated from the deep lobe of the parotid gland and retracted. The dissection continued posteriorly and inferiorly around the mandible. Mandibulotomy can be performed if necessary to improve exposure. Bass recommended placing the mandibulotomy site posterior to the entrance of the inferior alveolar nerve in the body of the mandible. Or the styloid process can be removed with dislocation of the mandible anteriorly to allow blunt dissection. Again, transparotid approach generally is used for deep lobe parotid lesions.

Transcervical approach starts with a transverse incision at the level of the hyoid bone. The submandibular gland is often removed or retracted anteriorly. An incision through the fascia deep to the submandibular space allowed for entry into the parapharyngeal space and blunt dissection of the tumor. Many modifications have reported. Some surgeons divide the digastric, stylohyoid, and styloglossus muscles from the hyoid bone to improve exposure. The styloid process and the stylomandibular ligament can also be divided to elevate the mandible anteriorly to improve access. This approach frequently involves blind finger dissection in the parapharyngeal space and does not provide enough exposure for larger benign lesions extending cranially or those with a more aggressive growth pattern.

Transcervical approach can be combined with mandibulotomy. Various locations for osteotomy have been reported, including mandibular body, angle, ramus, and parasymphyseal. The key is to try to limit injury to the inferior alveola nerve while providing access to the

parapharyngeal space. The risk of mandibulotomy includes inferior alveolar nerve anesthesia, loss of dentition, malocclusion, mandibular malunion or nonunion, and possibly requires a tracheotomy. It is thought to be necessary in less than 10% of patients. Olsen recommended mandibulotomy for vascular tumors extending into the superior parapharyngeal space, solid tumors that are confined to the superior aspect of the parapharyngeal space and malignant tumors invading the skull base.

Many surgeons prefer the transcervical-transparotid approach. A standard parotidectomy incision is made and carried into the lateral neck. The main trunk and lower division of the facial nerve are identified. The posterior belly of the digastric and stylohyoid muscle are divided, allowing for visualization of the internal and external carotid arteries, internal jugular vein, CN IX, X, XI, XII, and sympathetic chain. The styloid process and stylomandibular ligament may be transected to give a wide opening into the parapharyngeal space. Olsen reported using this technique in 80% of his case series. He recommended this approach for all deep-lobe parotid tumors, extraparotid salivary tumors and most of the post-styloid neurogenic tumors. Hughes also reported transcervical-transparotid approach is the preferred procedure for removal of most parapharyngeal tumors in his review of 172 patients with primary parapharyngeal tumors.

For vascular tumors that extending into the superior portion of the parapharyngeal space, Olsen recommended cervical-parotid approach with midline mandibulotomy. Midline lip splitting is used to expose anterior mandible. Then the mandible is divided in the midline. An intraoral incision is then made in the floor of the mouth extending back to the anterior tonsillar pillar and up to the level of the hard palate. The hypopharyngeal nerve and lingual nerve are preserved. The styloglossus and stylopharyngeus muscles are divided. The mandible is retracted laterally and superiorly to give wide exposure to the parapharyngeal space. Tracheotomy is necessary for this approach.

Because primary parapharyngeal tumors are exceedingly rare, few large scale studies are available. Shahab recently published his review of 114 parapharyngeal tumors for over 27 years of experience, the second largest study in literature. The 5-year and 10 year survival for benign parapharyngeal tumor is 100%. For malignancies the 5 year survival was 93%, but fall to 57% at 10 years. This study showed that a patient is highly unlikely to die of a benign parapharyngeal space tumor, therefore careful consideration of surgical treatment and discussion with the patient are crucial. While surgery is the mainstay of the treatment for parapharyngeal tumor, radiation therapy should be considered in elderly patients with paragangliomas. Also isolated asymptomatic parapharyngeal schwannomas in elderly patients with no nerve deficit should probably be observed.

## **Conclusion:**

Primary parapharyngeal tumors are rare and locate in a complex anatomical region. The clinical presentation of these tumors can be subtle. Therefore radiographic study provides important information for diagnosis and surgical planning. Majority of the tumors are benign with salivary gland neoplasm being the most common tumor. Surgical resection is the mainstay of treatment. Transcervical-transparotid approach with or without mandibulotomy is preferred by most surgeons.

## **Bibliography:**

Hughes KV., Olsen KD., McCaffrey TV. Parapharyngeal space neoplasms, *Head Neck* 1995; 17; 124-130.

Luna-Ortiz K, Navarrete-Aleman JE., Granados-Garcia M., Herrera-Gomez A. Primary parapharyngeal space tumors in a Mexican cancer center. *Otolaryngology and Head and Neck Surgery* 2005; 132(4); 587-591.

Olsen KD. Tumors and surgery of the parapharyngeal space. *Laryngoscope* 1994; 104: 1-28.

Shahab R., Heliwell T., Jones AS. How we do it: A series of 114 primary pharyngeal space neoplasms. *Clinical Otolaryngology* 2005; 30; 364-367.

Shields G., Bailey, B. Parapharyngeal space tumor. Dr. Quinn's Online Textbook 2002. Available at [www.utmb.edu](http://www.utmb.edu)