Title: Parapharyngeal Space Tumors
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Introduction

The parapharyngeal space is located deep within the neck lateral to the pharynx and medial to the ramus of the mandible. This is an area of complex anatomy, which is difficult to examine clinically. Tumors of the parapharyngeal space are rare, approximately 0.5% of all head and neck tumors. The majority of tumors in this region are benign, and surgical excision is the primary treatment. In order to reduce patient morbidity, and to provide adequate preoperative counseling, the surgeon must have a comprehensive understanding of treatment of tumors in this space. This paper will review the anatomy, pathology, clinical evaluation, radiologic evaluation, and surgical treatment of parapharyngeal space tumors.

Anatomy

Understanding the anatomy of the parapharyngeal space is important in making a correct diagnosis and surgical plan for excision of tumors in this region. The parapharyngeal space is a potential space, which is often described as being in the shape of an inverted pyramid with the floor at the skull base and it’s tip at the greater cornu of the hyoid bone.

Boundaries

The superior limit of the parapharyngeal space is a small portion of the temporal and bone and the sphenoid bone. It includes the carotid canal, jugular foramen, and hypoglossal foramen. There is a fascial connection from the medial pterygoid plate to the spine of the sphenoid at the superior medial wall, which crosses medial to the foramen ovale and foramen spinosum, which are not included in this space but rather in the infratemporal fossa. The inferior border is the junction of the posterior belly of the digastric muscle and the greater cornu of the hyoid bone. Medially, the boundary is made up of the buccopharyngeal or visceral fascia overlying the superior pharyngeal constrictors. The lateral boundary is made up of the fascia over the medial pterygoid muscle, the ramus of the mandible, the posterior belly of the digastric
muscle, and the fascia over the retromandibular deep portion of the parotid gland. Anteriorly the limit is the pterygomandibular raphe. The posterior limit is the dorsal layer of fascia making up the carotid sheath. The internal carotid artery, jugular vein, cranial nerves IX-XII, and sympathetic chain all course through this space.

**Prestyloid versus Retrostyloid Tumors**

A key anatomical division of the parapharyngeal space is into prestyloid and retrostyloid compartments. Fascia which extends from the styloid process to the tensor veli palatini muscle, called the tensor-vascular-styloid fascia, because it also contains the ascending palatine artery and vein, divides the parapharyngeal space into an anteromedial or prestyloid, and a posterolateral, or retrostyloid compartments. The prestyloid compartment contains fat, a portion of the retromandibular parotid gland, and some lymph nodes. The retrostyloid compartment contains the internal carotid artery, internal jugular vein, cranial nerves IX-XII, sympathetic chain, and lymph nodes. The parapharyngeal lymph nodes have numerous lymphatics, which along with the retropharyngeal nodes drain the soft palate, paranasal sinuses, posterior oral cavity, base of tongue, and a portion of the thyroid gland. The parapharyngeal nodes superiorly are connected to the node of Rouviere in the lateral most retropharyngeal space, this can be a site of metastasis from the nasopharynx, upper oropharynx, and sinuses.

**Stylomandibular tunnel**

The stylomandibular tunnel is bounded by the posterior ramus of the mandible, the skull base and the stylomandibular ligament. Deep parotid tumors can enter the parapharyngeal space posterior to the stylomandibular ligament resulting in a round shaped lesion or can enter through the stylomandibular tunnel resulting in a “dumbbell” shaped mass.

**Pathology**

Neoplasms of the parapharyngeal space fall into three categories: primary, metastatic, and extension from adjacent structures. Approximately 80% are benign and 20% malignant. Direct extension can occur from the mandible, maxilla, nasopharynx, neck, oral cavity, oropharynx, and temporal bone. Metastatic tumors include: follicular thyroid carcinoma, medullary thyroid carcinoma, papillary thyroid carcinoma, osteogenic sarcoma, and squamous cell carcinoma. Primary parapharyngeal space neoplasms fall into three categories: salivary gland neoplasms, neurogenic tumors, and miscellaneous tumors.

**Salivary Gland Neoplasms**

Salivary gland neoplasms are the most common parapharyngeal tumors accounting for 40-50% neoplasms. They are located in the prestyloid space and can arise from either the deep lobe of the parotid gland or from minor salivary glands in the parapharyngeal space. Pleomorphic adenoma is the most common tumor type and mucoepidermoid is the most common malignant neoplasm. Less than 5% of parotid tumors involve the parapharyngeal space. A fat plane on CT or MRI between the mass and the parotid can distinguish a tumor of minor salivary origin from a deep lobe of the parotid tumor. Also, on CT or MRI these lesions displace the
carotid artery posteriorly. Masses arising from the deep lobe of the parotid can pass behind the stylomandibular ligament, forming a round tumor, or through the stylomandibular tunnel, forming a “dumbbell” shaped tumor as mentioned previously.

**Neurogenic Tumors**

The neurilemoma or schwannoma is the most common neurogenic neoplasm of the parapharyngeal space and are the most common retrostyloid masses. The most common site of origin is the vagus nerve, while the sympathetic chain is second. These can also originate from cranial nerves IX, XI, and XII. These lesions are benign and slow growing, often presenting as asymptomatic masses. They generally do not affect their nerve of origin but can extend up through the jugular foramen intracranially. Less than 1% undergo malignant change. By CT or MRI these tumors displace the carotid artery anteriorly.

Paragangliomas (also chemodectomas or glomus tumors) are the second most common neoplasm of the parapharyngeal space. They arise from the nodose ganglion of the vagus nerve, extend into the parapharyngeal space from the carotid body, or extend inferiorly from the jugular bulb. These are also retrostyloid masses. The glomus vagale from the vagus nerve is the most common type. Paragangliomas can be bilateral 10% of the time, and in a patient with a family history of paragangliomas 26-35% of the time. Malignancy has been reported as 10%. These tumors are of neural crest origin and can secrete catecholamines 2% of the time, causing hypertension and flushing. Glomus vagale tumors displace the carotid anteriorly while carotid body tumors will splay the internal and external carotids resulting in the “lyre” sign.

Neurofibromas are the third most common neurogenic tumor of the parapharyngeal space. They originate from the Schwann cells and perineural fibroblasts. These tumors are unencapsulated and intimate involvement of the nerve of origin is common. Sites of origin include the vagus nerve, glossopharyngeal nerve, sympathetic chain, and spinal accessory nerve. This lesions are often multiple and can be associated with Neurofibromatosis type I, in which case there is a higher instance of malignancy.

**Miscellaneous Tumors**

A wide variety of benign and malignant neoplasms can occur in the parapharyngeal space and account for 20% of lesions. A comprehensive list is beyond this review. Some possible masses include: lymphoma, hemangioma, teratoma, lipoma, branchial cleft cyst, arteriovenous malformation, and internal carotid artery aneurysm.

**Clinical Evaluation**

The signs and symptoms of parapharyngeal space neoplasms can be subtle and clinical evaluation of this space is difficult. Lesions most often present as an oropharyngeal bulge or as a neck mass. Tumors must reach a size of 2.5-3 cm before becoming palpable as a mass because of the deep location in the neck, so they can reach a large size prior to patient presentation. Other signs and symptoms include: dysphagia, dyspnea, unilateral middle ear effusion, pulsatile tinnitus, bruit, thrill, otalgia, airway obstruction, hoarseness, foreign body sensation, true vocal
cord palsy, Horner’s syndrome, dysarthria, and symptoms of catecholamine excess like hypertension and flushing.

All patients should undergo a comprehensive history and complete head and neck examination. If the lesion is large enough to be palpated, bimanual palpation is important. Paragangliomas are classically described as compressible and mobile in an anterior-posterior direction but not in a vertical direction. Patients in whom a paraganglioma is suspected should undergo a 24-hour urine collection for vanillylmandelic acid and metanephrines. If urinary catecholamines are elevated a metaiodinated benzylguanidine (MIBG) scan should be obtained to look for multiple lesions. In some cases fine needle aspiration of the lesion can be useful if a malignant tumor is suspected, however this should only be performed after imaging to rule out a vascular lesion.

**Imaging**

CT, MRI, and angiography are all used to evaluate parapharyngeal neoplasms. A CT or MRI is most commonly the first study obtained. Angiography is performed for all enhancing lesions.

*CT Scan*

CT scan with and without contrast is useful in the evaluation of parapharyngeal masses. Information obtained from this study is the location of a tumor in either the prestyloid or retrostyloid compartments, the presence or absence of a fat plane between the deep lobe of the parotid and a prestyloid mass, and if the mass enhances with contrast. Schwannomas enhance because of the extravascular accumulation of contrast while a paraganglioma is a hypervascular mass. Displacement of the internal carotid artery posteriorly is characteristic of prestyloid tumors while displacement anteriorly is indicative of a poststyloid mass. Bone erosion by malignant tumors can be seen. CT does have limitations in soft tissue detail in comparison to MRI and exposes the patient to radiation. If a retrostyloid mass is found or if malignancy is suspected an MRI should be obtained.

*MRI*

Soft tissue resolution is better with MRI than with CT scan. It is probably the most useful study to evaluate parapharyngeal neoplasms. The relationship between the mass and the carotid can be more clearly seen with MRI than with CT scan. There are characteristic appearances of different tumors on MRI scan. Pleomorphic adenomas have low signal intensity on T1 images and high on T2 images and displace the carotid posteriorly. Schwannomas have higher signal intensity on T2 images like pleomorphic adenomas, but displace the carotid anteriorly. Paraganliomas have a characteristic “salt and pepper” appearance on T2 weighted images because of flow voids.

*Angiography*

In the past all enhancing lesions were evaluated with angiography. Advances in MRI imaging have decreased the use of angiography in these tumors. This study can demonstrate the
relationship of the tumor to the great vessels and distinguish between neurogenic and vascular lesions. Carotid body tumors cause splaying at the bifurcation resulting in the “lyre” sign mentioned above. Carotid artery balloon occlusion tests should be performed if malignancy is suspected and there is the possibility of carotid sacrifice. Tumor embolization can be performed for paragangliomas 1 day prior to surgery to reduce blood supply. Some authors report that this increases fibrosis around the tumor making dissection more difficult, and do not use it.

**Surgical Approaches**

**Transoral**

The transoral approach has been used for selected small benign lesions of the prestyloid space presenting as an oropharyngeal mass. Problems with this approach are limited exposure, increased risk of tumor spillage, and possibility of neurovascular injury.

**Cervical with or without mandibulotomy**

Approach recommended by many authors. A transverse incision at the level of the hyoid bone with either removal or displacement of the submandibular gland is performed. Incision of the fascia deep to the posterior submandibular gland is incised. To increase exposure the digastric muscle, stylohyoid, and styloglossus muscles can be released from the hyoid bone and the stylomandibular ligament can be divided. Tumors may often be bluntly dissected with a finger.

Further exposure can be obtained by performing mandibulotomy of the body, ramus or the angle. Tracheostomy is necessary with this approach. After removal of the tumor the mandibulotomy is repaired with plates and screws.

**Cervical-Parotid**

A cervical approach incision may be extended superiorly in front of the ear. This allows identification of the facial nerve trunk and lower division of the facial nerve. The posterior belly of the digastric muscle is divided exposing the internal carotid, jugular vein, and nerves. The stylomandibular ligament, styloglossus muscle, and stylohyoid muscle are divided close to the styloid process. This approach may be combined with mandibulotomy to improve exposure.

**Transparotid**

For deep lobe parotid tumors a transparotid approach may be used. A superficial parotidectomy is performed and the facial nerve retracted. Dissection is carried around the mandible. If necessary, mandibulotomy may be used posterior to the entrance of the inferior alveolar nerve.
Cervical-Transpharyngeal

For large or highly vascular tumors a cervical incision is extended to midline lip incision. Mandibulotomy is performed either in the midline or anterior to the mental foramen and the incision is carried along the floor up to the anterior tonsillar pillar. The mandible is swung laterally and the tonsil and superior constrictors moved medially. This gives wide access to the parapharyngeal space. Tracheostomy is necessary.

Infratemporal Fossa

A preauricular lateral infratemporal fossa approach may be used for tumors involving the skull base or extending into the infratemporal fossa. This may be combined with a frontotemporal craniotomy for tumors with significant intracranial extension.

Transcervical-Transmastoid

The cervical incision is extended postauricularly and a mastoidectomy is performed. The mastoid tip is removed and the jugular fossa exposed. Sometimes the facial nerve may need to be dissected out of the fallopian canal and transposed. Transient facial paresis should be expected.

Nonsurgical Management

In some cases of patients who are poor surgical candidates, who fail balloon occlusion, are elderly, have unresectable lesions, or have benign slow growing tumors that would require sacrifice of multiple cranial nerves surgery may not be the only option. Other options include observation, or for paragangliomas, radiation. Paragangliomas tend to grow 1.0-1.5 mm per year. Mortality is less than 10% per year for untreated paragangliomas. Although radiation is not curable for paragangliomas, local control rates of 96-100% have been reported.

Conclusions

The parapharyngeal space is an area of complex anatomy where clinical exam is difficult. The surgeon must understand the pathology of tumors of this space and the proper use of imaging studies to make a preoperative diagnosis. This allows for planning of a surgical approach and preoperative counseling.
Bibliography


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