



# WORLD SKULL BASE E-LEARNING MATERIAL

## Parapharyngeal Space Tumors



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#### Background

The parapharyngeal space (PPS) is a potential space lateral to the upper pharynx.

#### Etiology

Tumors of the parapharyngeal space (PPS) are uncommon, comprising less than 1% of all head and neck neoplasms. Both benign and malignant tumors may arise from any of the structures contained within the parapharyngeal space (PPS). Of parapharyngeal space (PPS) tumors, 70-80% are benign, and 20-30% are malignant. Most parapharyngeal space (PPS) tumors are of salivary or neurogenic origin, although metastatic lesions; lymphoreticular lesions; and a variety of uncommon, miscellaneous lesions may arise in this location.<sup>[1]</sup>



Parapharyngeal space tumor manifesting as an oropharyngeal mass.

#### Salivary gland neoplasms

Neoplasms of salivary gland origin are located in the prestyloid parapharyngeal space (PPS) and account for 40-50% of parapharyngeal space (PPS) lesions. Salivary neoplasms may arise from the deep lobe of the parotid gland, ectopic salivary rests, or minor salivary glands of the lateral pharyngeal wall. The prevalence of neoplasms that arise within the deep lobe of the parotid gland is identical to that of those that arise in the superficial lobe. The most common prestyloid parapharyngeal space (PPS) lesion is pleomorphic adenoma, which represents 80-90% of salivary neoplasms in the parapharyngeal space (PPS).

Other benign salivary lesions, including Warthin tumors and oncocytomas, develop in the prestyloid

parapharyngeal space (PPS), as do malignant salivary lesions. Carcinoma ex pleomorphic adenoma and adenoid cystic carcinoma are the most frequently reported salivary malignancies of the parapharyngeal space (PPS). Approximately 20% of all salivary lesions in the parapharyngeal space (PPS) are malignant.

Common benign neoplasms include pleomorphic adenomas, monomorphic adenomas, and oncocytomas. Malignant neoplasms include adenoid cystic carcinomas, mucoepidermoid carcinomas, adenocarcinomas, and acinic cell carcinomas.

## Neurogenic lesions

Neurogenic lesions are the most common tumors of the poststyloid parapharyngeal space (PPS) and account for 25-30% of parapharyngeal space (PPS) lesions. Neurilemmomas are the most commonly encountered lesions, followed in frequency by paragangliomas and neurofibromas.

Benign neurogenic lesions include neurilemoma (schwannoma), paraganglioma, neurofibroma, and ganglioneuroma. Malignant neurogenic lesions include malignant paraganglioma, neurofibrosarcoma, schwannosarcoma, and sympatheticoblastoma.

Neurilemmomas, or schwannomas, are the most common neurogenic tumors and arise from any nerve surrounded by Schwann cells. In the parapharyngeal space (PPS), the most common sites of origin are the vagus nerve and the sympathetic chain. Neurilemmomas are slow growing and rarely cause palsy of the nerve of origin. They are encapsulated and histologically distinct from the nerve itself. Treatment is by enucleation, and preservation of the nerve of origin is usually possible; however, every patient should be cautioned about the possibility of postoperative paralysis.

Neurofibromas, in contrast, are unencapsulated and intimately involved with the nerve of origin. Neurofibromas are often multiple. They may occur as a manifestation of the neurofibromatosis-1 (NF-1) syndrome, and, in these patients, the incidence of malignant transformation is increased. The nerve of origin usually has to be sacrificed during excision to ensure complete removal of the neoplasm.

Paragangliomas are benign vascular neoplasms that arise from the paraganglia or extra-adrenal neural crest tissue. Paraganglia function as chemoreceptors and are associated with the carotid body, the jugular bulb, and the vagus nerve in the poststyloid parapharyngeal space (PPS). Carotid body tumors, [glomus jugulare](#), and glomus vagale are slow-growing paragangliomas that may not produce symptoms but do cause cranial nerve (CN) deficits, bone erosion, or intracranial extension as they increase in size.

Approximately 2% of head and neck paragangliomas secrete catecholamines and may cause paroxysmal symptoms of catecholamine excess. Ten percent of paragangliomas are multiple and associated with paraganglioma at other locations. Ten percent of paragangliomas are hereditary, associated with a familial paraganglioma syndrome. In patients with hereditary paraganglioma, the prevalence of multicentricity is 35%.

Hypertension and flushing are suggestive of either a secreting paraganglioma or an associated pheochromocytoma. If these symptoms are present, obtain urinary catecholamine levels. If the level of catecholamines is elevated, rule out a concomitant pheochromocytoma. Malignant transformation occurs in fewer than 10% of patients and is associated with rapid growth and development of metastatic disease.

## Lymphoreticular lesions

Lymphoreticular lesions comprise 10-15% of parapharyngeal space (PPS) lesions. The lymphatics of the parapharyngeal space (PPS) may be involved primarily or secondarily by carcinoma, or they may be involved by infectious or inflammatory processes. Lymphoma is the most common malignant lymphoid process, but metastases from thyroid cancer, osteogenic sarcoma, squamous cell carcinoma, renal cell carcinoma, hypernephroma, and meningioma may also appear as parapharyngeal space (PPS) masses. The most common lymphoreticular lesions are lymphomas and metastases.

## Miscellaneous lesions

- Aneurysm
- Ameloblastoma

- Amyloid tumor
- Arteriovenous malformation
- Branchial cleft cyst
- Chondroma
- Chondrosarcoma
- Chordoma
- Choroid plexus tumor
- Dermoid
- Desmoid
- Ectomesenchymoma
- Fibrosarcoma
- Fibrous histiocytoma
- Granular cell myoblastoma
- Hemangioendothelioma
- Hibernoma
- Inflammatory pseudotumor
- Leiomyoma
- Liposarcoma
- Malignant meningioma
- Malignant teratoma
- Meningioma
- Rhabdomyoma
- Rhabdomyosarcoma
- Sarcoma
- Teratoma
- Venous angioma

A variety of more unusual lesions may occur in the parapharyngeal space (PPS), and these lesions comprise 10-15% of parapharyngeal space (PPS) masses. While the pathologist usually makes the final determination and diagnosis in such cases, recognizing that vascular lesions, such as hemangiomas, arteriovenous malformations, and internal carotid artery aneurysms, may occur in the parapharyngeal space (PPS) is important. Imaging studies of this region must be performed before attempting to obtain a biopsy or to excise the lesion.

## Presentation

Clinical presentations may include the following:

- Neck mass
- Oropharyngeal mass
- Unilateral eustachian tube dysfunction
- Dysphagia
- Dyspnea
- Obstructive sleep apnea
- CN deficits
- Homer syndrome
- Pain
- Trismus
- Symptoms of catecholamine excess

Patients with tumors of the parapharyngeal space (PPS) most commonly present with a neck or oropharyngeal mass that does not cause symptoms detectable on physical examination because only the inferior and medial boundaries of the parapharyngeal space (PPS) are distensible as seen in the images below.



Parapharyngeal space tumor manifesting as an oropharyngeal mass.



Parapharyngeal space tumor manifesting as a neck mass.

Tumors must be at least 2 cm in size before the bulge or abnormality is palpable. Often, a parapharyngeal space (PPS) lesion is discovered incidentally on routine physical examination. Unilateral eustachian tube dysfunction may result from significant medial extension, causing soft palate and nasopharyngeal swelling. Oropharyngeal bulging from an underlying parapharyngeal space (PPS) mass may cause significant displacement of the ipsilateral tonsil and may create the appearance of a primary tonsillar lesion. An ill-fitting denture may be the first symptom of a benign prestyloid lesion.

Symptoms of dysphagia, dyspnea, and obstructive sleep apnea may result from distortion of the lateral pharyngeal wall by a massive parapharyngeal space (PPS) lesion. In such cases, tracheostomy has been recommended for relief of airway obstruction.

Cranial neuropathies may result from enlargement of parapharyngeal space (PPS) lesions with compression of CN IX, X, XI, or XII, resulting in symptoms of hoarseness, dysarthria, and dysphagia. Horner syndrome (ie, ptosis, miosis, anhidrosis) has been described as resulting from pressure on the cervical sympathetic chain. With the exception of glomus vagale tumors, which have been associated in the literature with a high frequency of vagal paresis, most benign lesions of the parapharyngeal space (PPS) do not result in cranial nerve dysfunction. Pain is unusual with benign lesions and may be due to compression or hemorrhage into the lesion; however, pain and neurologic dysfunction are more often indicative of malignancy with infiltration of the skull base. Under these circumstances, CN VII may be involved. Trismus results from malignant invasion of the pterygoid musculature or involvement of the coronoid process of the mandible.

Physical examination findings may suggest the origin and nature of the tumor. The most common physical finding is a painless oropharyngeal or neck mass. Pay careful attention to the oropharynx, tonsillar area, pharynx, and neck. Lesions arising from the deep lobe of the parotid may often be localized using bimanual palpation. Perform a full cranial nerve evaluation, including laryngoscopy, to test the motor and sensory innervation of the larynx. The vagus nerve is the most commonly involved cranial nerve, and vagal palsy is suggestive of either a paraganglioma or a malignancy.

A neck mass that is pulsatile or has a thrill to auscultation suggests a vascular tumor, although carotid pulsations may be transmitted through an overlying mass and may be misleading. Paragangliomas are typically mobile in an anteroposterior direction but not in a vertical direction. Any patient with aural symptoms should undergo thorough audiologic evaluation as well as careful examination of the nasopharynx.

## Indications

Complete surgical excision is the mainstay of treatment and is recommended for both diagnostic and therapeutic purposes. The choice of surgical approach is dictated by the size of the tumor, its location, its relationship to the great vessels, and the suspicion of malignancy (see Surgical therapy). However, when surgery is contraindicated, alternatives to surgical therapy consist of observation or radiation therapy.

## Relevant Anatomy

The parapharyngeal space (PPS) is a potential space lateral to the upper pharynx. The parapharyngeal space (PPS) is shaped like an inverted pyramid, extending from the skull base superiorly to the greater cornu of the hyoid bone inferiorly.

The superior border of the parapharyngeal space (PPS) comprises a small area of the temporal and sphenoid bones, including the carotid canal, jugular foramen, and hypoglossal foramen. The parapharyngeal space (PPS) is limited anteriorly by the pterygomandibular raphe and pterygoid fascia and posteriorly by the cervical vertebrae and prevertebral muscles. The medial border of the parapharyngeal space (PPS) is the pharynx, and the lateral border is comprised of the ramus of the mandible, the medial pterygoid muscle, and the deep lobe of the parotid gland. Below the level of the mandible, the lateral boundary consists of the fascia overlying the posterior belly of the digastric muscle.

The fascia from the styloid process to the tensor veli palatini divides the parapharyngeal space (PPS) into an anteromedial compartment (ie, prestyloid) and a posterolateral (ie, poststyloid) compartment. The prestyloid compartment contains the retromandibular portion of the deep lobe of the parotid gland, adipose tissue, and lymph nodes associated with the parotid gland. The poststyloid compartment contains the internal carotid artery, the internal jugular vein, CNs IX- XII, the sympathetic chain, and lymph nodes.

These lymphatics receive afferent drainage from the oral cavity, oropharynx, paranasal sinuses, and thyroid. The distinction between the prestyloid and poststyloid space is more than just semantic because imaging studies can delineate between the 2 compartments and can assist in reaching the correct diagnosis preoperatively.

## Contraindications

Surgery may be contraindicated and nonoperative management of parapharyngeal space (PPS) lesions considered for patients who are poor surgical candidates because of comorbid disease; those who are elderly; those in whom balloon occlusion fails; those who have unresectable lesions; and those who have benign, slow-growing tumors that would carry a significant risk of sacrifice of multiple cranial nerves if resected. The risks and benefits of surgery must be weighed in every case.

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