

Parapharyngeal Space Tumors

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.

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.

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 most common prestyloid parapharyngeal space (PPS) lesion is pleomorphic adenoma, which represents 80-90% of salivary neoplasms in the parapharyngeal space (PPS).

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.

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.

Neurofibromas, in contrast, are unencapsulated and intimately involved with the nerve of origin. Neurofibromas are often multiple. 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.

Lymphoreticular lesions

Lymphoreticular lesions comprise 10-15% of parapharyngeal space (PPS) lesions.

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.

A variety of more unusual lesions may occur in the parapharyngeal space (PPS), and these lesions comprise 10-15% of parapharyngeal space (PPS) masses.

Presentation

Clinical presentations may include the following:

- Neck mass (must be at least 2 cm in size before the bulge or abnormality is palpable)
- Oropharyngeal mass (may cause significant displacement of the ipsilateral tonsil and may create the appearance of a primary tonsillar lesion)
- Unilateral eustachian tube dysfunction
- Dysphagia
- Dyspnea
- Obstructive sleep apnea
- CN deficits V, X, XI, or XII, resulting in symptoms of hoarseness, dysarthria, and dysphagia
- Horner syndrome
- Pain (more often indicative of malignancy with infiltration of the skull base)
- 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

Diagnosis

Laboratory Studies

- Twenty-four-hour urine collection for catecholamines:
- Vanillylmandelic acid (VMA), or 4-hydroxy-3-methoxymandelic acid
- Metanephrine

Imaging Studies

- Radiologic studies are essential in the evaluation of a patient with a suspected parapharyngeal space (PPS) mass. Performing these studies before considering biopsy is important because, given the differential diagnosis of a parapharyngeal space (PPS) lesion, one can often make a diagnosis on the basis of imaging studies without the need for fine-needle aspiration biopsy (FNAB) or open biopsy.
 - Imaging studies should answer the following questions:
 - Is the mass prestyloid or poststyloid?

- What is the relationship to the parotid gland?
 - What is the relationship to the great vessels?
- Computed tomography (CT) scanning and magnetic resonance imaging (MRI) have equal efficacy in localizing the lesion to the prestyloid or poststyloid space (see [Images 3-6](#)).
- Angiography
 - Angiography is recommended in the workup of most vascular lesions. Angiography is also used if malignancy is suspected and if carotid sacrifice is possible during resection. If carotid resection is considered, angiography is combined with balloon occlusion testing to measure cerebral blood flow (see Balloon occlusion test in [Other Tests](#)).
- Biopsy
 - Under most circumstances, a presumptive diagnosis can be made on the basis of the findings of the imaging studies described above. Under no circumstances should biopsy of a parapharyngeal space (PPS) mass be performed prior to obtaining results from the radiologic studies. Complete surgical excision is the mainstay of treatment and is recommended for both diagnostic and therapeutic purposes.

Treatment

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. However, when surgery is contraindicated, alternatives to surgical therapy consist of observation or radiation therapy.

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.

Surgical Therapy

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.

Intraoperative Details

- Transoral approach
- Transcervical approach
- Transcervical-transparotid approach
- Transcervical-transmandibular approach
- Infratemporal fossa approach

Outcome and Prognosis

The recurrence rate of benign parapharyngeal space (PPS) neoplasms following surgical extirpation ranges from 0-9%. Malignant tumors of the parapharyngeal space (PPS) have a much higher rate of recurrence (25-77%), depending on histology, extent of resection, and duration of follow-up.