Chapter 76 – Management of Tumors of the Parapharyngeal Space

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The management of patients with tumors of the parapharyngeal space (PPS) is a challenging task. The complex nature of the anatomy of the PPS and the diversity of its contents gives rise to a wide variety of tumors. This presents an opportunity for technically challenging surgery. Surgery of the PPS is a relatively recent activity in our field, because only since the introduction of the computed tomography (CT) scan has the PPS been characterized.[1] Once the imaging techniques became readily available, surgical techniques were devised to remove the tumors identified within the PPS.

ANATOMY

The PPS is described as being an inverted triangle with its base at the base of the skull and the apex at the hyoid bone (Fig. 76-1). The lateral boundary is the mandible and the medial boundary is the buccopharyngeal fascia and the constrictor muscles of the pharynx. Because two sides of the PPS are bone, tumors arising in the PPS expand along the path of least resistance toward the soft tissue of the pharyngeal wall, displacing the wall and the tonsil to present as a submucosal mass in the pharynx.

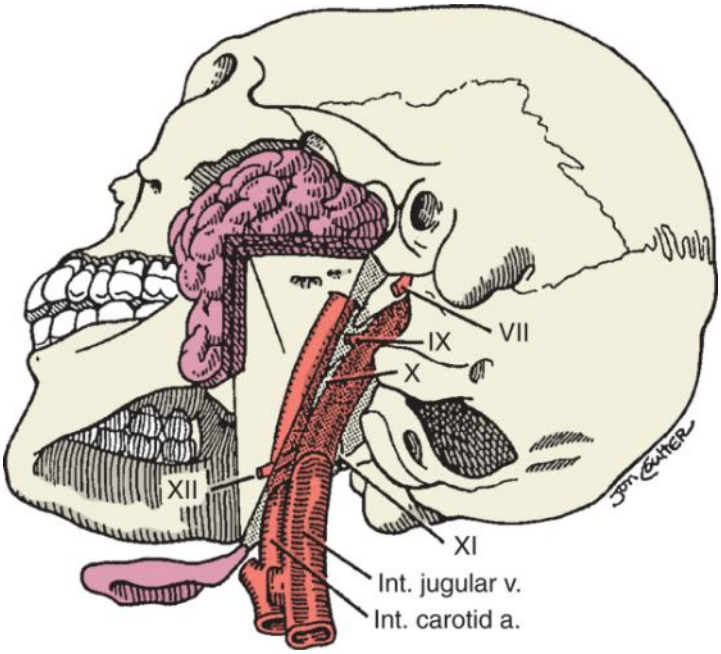

Figure 76-1 Diagram of parapharyngeal space.
(Reprinted with permission from Carrau RL, Myers EN, Johnson JT: Management of tumors arising in the parapharyngeal space. Laryngoscope 100:583-589, 1990.)

Advances in imaging have demonstrated the division of the PPS into prestyloid and poststyloid compartments (Fig. 76-2). The fascia of the tensor veli palatini muscle runs through the PPS from the styloid process to its insertion on the lateral pterygoid plate. This defines the separation between the prestyloid and poststyloid compartments.[1] This information is invaluable in developing a differential diagnosis and plan of management of these tumors.

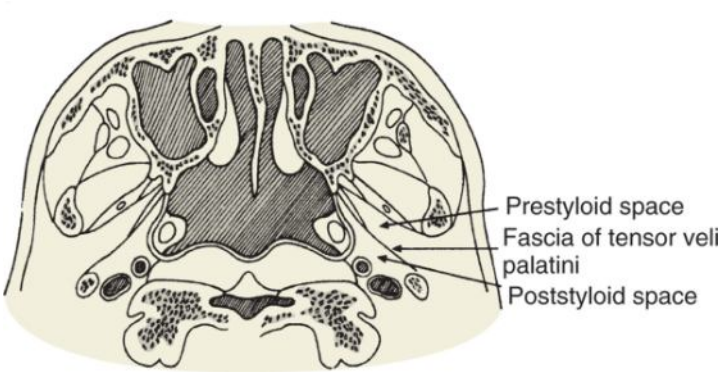

Figure 76-2 The fascia of the tensor veli palatini muscle divides the parapharyngeal space into a prestyloid and a poststyloid compartment.

The prestyloid PPS contains the deep lobe of the parotid gland, adipose tissues, small blood vessels, lymphatics, and minor nerves. The poststyloid PPS contains the carotid sheath and is also traversed by cranial nerves IX, X, and XII. The cervical sympathetic chain lies posterior to the carotid artery.

The clinical correlation of the imaging studies and surgical findings has provided information that allows the clinician to predict the tumor type from the imaging studies with a high degree of accuracy. The tumors of the prestyloid PPS are usually salivary gland in origin. Most of these tumors are pleomorphic adenomas that arise in the deep lobe of the parotid gland and extend through the stylomandibular tunnel into the prestyloid PPS (Fig. 76-3). An accurate CT scan should clearly demonstrate the tumor arising in the deep lobe of the parotid gland going through the stylomandibular tunnel, extending medially, and displacing the parapharyngeal structures intraorally. Less frequently salivary gland tumors arise de novo in the prestyloid PPS (Fig. 76-4).

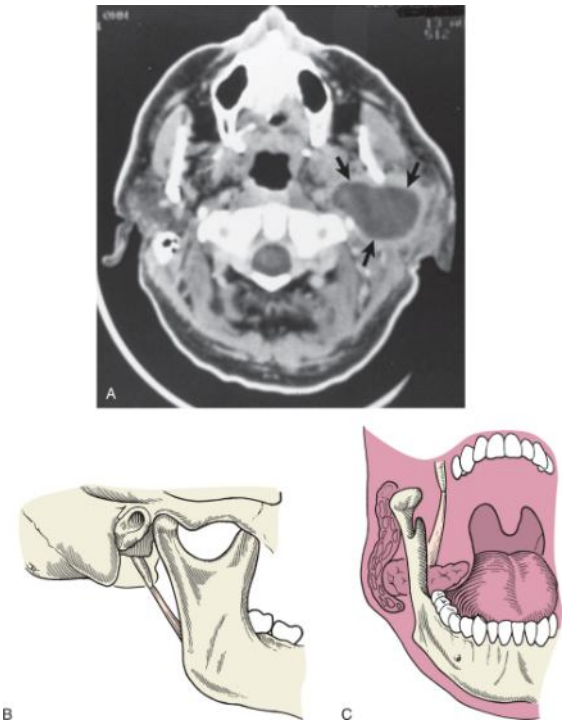

Figure 76-3 **A,** Axial computed tomography scan of a pleomorphic adenoma originating in the deep lobe of the parotid gland with extension through the stylomandibular tunnel into the prestyloid parapharyngeal space *(arrows)*. **B,** A diagram of the relationship of the styloid process, the stylomandibular ligament, and the mandible. **C,** A diagram of a tumor originating in the deep lobe of the parotid gland passing through the stylomandibular tunnel into the prestyloid parapharyngeal space.

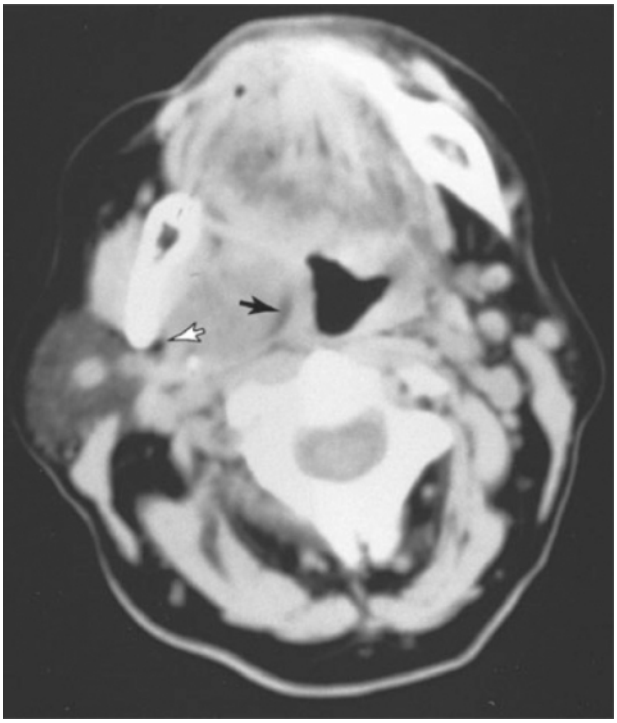

Figure 76-4 Computed tomography scan demonstrating a pleomorphic adenoma arising de novo in the prestyloid parapharyngeal space. The tumor displaces fat medially *(closed arrow)*. A fat plane separates the tumor from the parotid gland *(open arrow)*.

Tumors arising in the poststyloid PPS are almost always neurogenic in origin. The most common of these tumors are paragangliomas (PGL), including the carotid body tumor and glomus vagale. Neurilemmoma arising from the vagus nerve or the superior cervical ganglion may also occur in this area.[2] Ganglioneuroma of the PPS has been reported recently.[3]

A wide variety of other primary tumors may occur in the PPS (Table 76-1). Even though the majority of tumors arising in the PPS are benign (Fig. 76-5), malignant tumors of the PPS are also encountered (Table 76-2). Cancers arising in other sites with metastasis to the PPS have been reported. Aygenc and colleagues[4] reported two cases of papillary adenocarcinoma of the thyroid gland metastatic to the PPS. The authors describe the pathway of spread of the tumor to involve the PPS via the lateral retropharyngeal nodes. This pattern of spread is in keeping with Rouvier's description of lymphatic pathway from the thyroid gland to retropharyngeal nodes in the PPS.

Table 76-1 -- HISTOLOGIC DIAGNOSIS OF TUMORS REMOVED FROM THE PARAPHARYNGEAL SPACE

Histology	No. of Tumors
Paraganglioma	69
Pleomorphic adenoma	32
Squamous cell carcinoma	10
Neurilemmoma	7
Salivary duct carcinoma	3
Sarcoma	3
Schwannoma	3
Adenocarcinoma	2
Adenoid cystic carcinoma	2

Histology	No. of Tumors
Carcinoma ex pleomorphic adenoma	2
Lipoma	2
Lymphangioma	2
Lymphoma	2
Meningioma	2
Hemangioma	1
Hemangiopericytoma	1
Leiomyosarcoma	1
Liposarcoma	1
Lymphoepithelial carcinoma	1
Mucoepidermoid carcinoma	1
Myoepithelioma	1
Neurofibroma	1
Oncocytic papillary cystadenoma	1
Thyroid carcinoma	1
Warthin's tumor	1

From the Department of Otolaryngology, University of Pittsburgh.

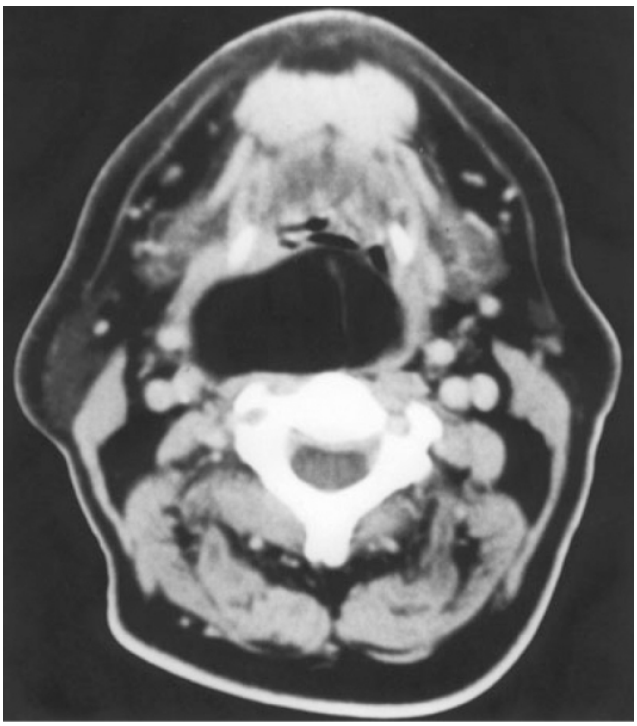

Figure 76-5 Axial computed tomography scan reveals a lipoma involving the parapharyngeal and retropharyngeal space.

Table 76-2 -- HISTOLOGIC DIAGNOSIS OF MALIGNANT TUMORS REMOVED FROM THE PARAPHARYNGEAL SPACE

Tumor	No. of Patients
Carotid body tumor	2
Neurogenic sarcoma	2
Malignant mixed tumor	1
Adenoid cystic carcinoma	1
Neurofibrosarcoma	1
Leiomyosarcoma	1
Liposarcoma	1
Fibrosarcoma	1
Metastatic cancer of the thyroid	1
Malignant meningioma	21
Lymphoma	1

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Chiesa and De Paoli[5] reported metastasis to the PPS from nasopharyngeal cancer. Umeda and associates[6] reported metastasis of squamous cell carcinoma of the maxilla to the PPS. Raut and colleagues[7] reported a case of metastatic breast carcinoma, which presented as a mass in the PPS 15 years after the primary presentation.

PATIENT SELECTION

The signs and symptoms associated with tumors that arise in the PPS are numerous and depend on both the location within the PPS as well as the histologic type.

The most commonly encountered presentation of the neoplasm involving the prestyloid PPS is an asymptomatic mass in the lateral wall of the oropharynx with or without a mass in the parotid or the neck (Fig. 76-6). Expansion of these tumors may interfere with the proper fit of a denture and on occasion may become symptomatic based on the large size, producing interference with breathing and swallowing. Submucosal masses are usually asymptomatic and they are often found on routine physical examination. The presence of a mass in the parotid is often associated with the mass in the oral cavity. The presence of pain or neuropathy should direct the clinician to suspect a primary or metastatic cancer.

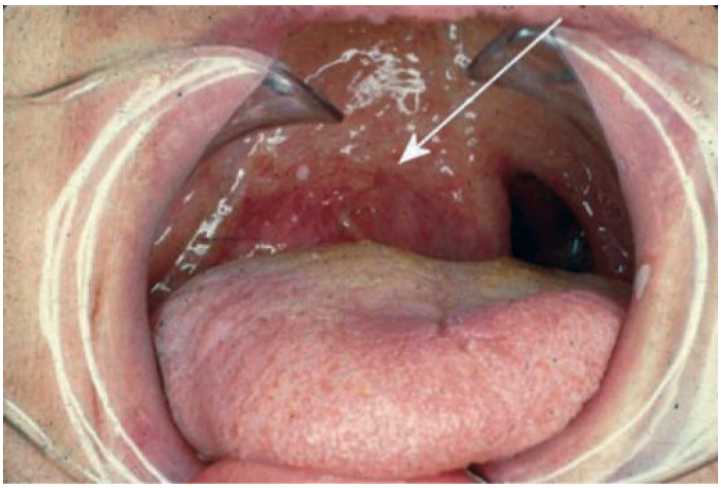

Figure 76-6 Typical appearance of a tumor of the parapharyngeal space displacing tonsil and uvula medially *(arrow)*.

Patients with a tumor involving the poststyloid PPS may present with a mass in the neck. Lesions of the posterior PPS may also present with a neuropathy. For example, a tumor arising from the vagus nerve may result in paralysis of the vocal cord with resultant hoarseness and aspiration. A tumor originating in the cervical sympathetic chain may produce Horner's syndrome.

PREOPERATIVE EVALUATION

All patients being evaluated for a tumor of the PPS should undergo a thorough history and physical examination (Table 76-3). The history should be inclusive, such as that done with other patients with tumors in the head and neck and should emphasize the possibility of cranial nerve neuropathies. In patients suspected of having PGL, a family history should be sought, particularly in patients with multiple PGLs. The history of palpitation, flushing, or difficult to control hypertension also should be sought, and these patients should be evaluated preoperatively for the presence of a pheochromocytoma.[8] Physical examination should include inspection of the head and neck, and direct or indirect laryngoscopy to evaluate the motion of the vocal cords and direct palpation of the tumor both intraorally and extraorally. We do not advocate fine needle aspiration biopsy or incisional biopsy before surgery.

Table 76-3 -- PRESENTING SIGNS AND SYMPTOMS FROM THE PARAPHARYNGEAL SPACE

Sign/Symptoms	No.	(%)
Neck mass	76	47.5
Hoarseness or vocal cord paralysis	16	10.0
Hearing loss	13	8.1
Oropharyngeal mass	11	6.9
Dysphagia	10	6.3
Pain	9	5.6
Tinnitus	9	5.6
Otalgia/aural fullness	7	4.4
CN XII dysfunction	6	3.8
Dyspnea/postnasal drip	5	3.1
Headache	4	2.5
Facial weakness/paralysis	3	1.9
Dizziness	2	1.3
Trismus	2	1.3
Bleeding	1	0.6
Horner's syndrome	1	0.6
Sleep apnea	1	0.6
Trigeminal dysesthesia	1	0.6
Weight loss	1	0.6

From the Department of Otolaryngology, University of Pittsburgh.

IMAGING

All patients suspected of a tumor in the PPS should undergo imaging studies. The distinction between tumors of the prestyloid and poststyloid space may be made radiographically based on a clear understanding of the anatomy in this region. In our department, CT scan with contrast has been used almost exclusively with the exception of certain patients when there is suspicion of intracranial extension who benefit from magnetic resonance imaging (MRI) studies.

The distinction between tumors of the prestyloid and poststyloid space is the most important aspect of the evaluation in that clinical correlation has very clearly defined what type of tumor the patient has and provides us with a very clear plan of management based on the location and histology. In most cases, we use the information from imaging studies as a substitute for a biopsy. Table 76-4 and 76-5 summarize our observations on the correlation of imaging studies with surgical findings.[8]

Table 76-4 -- CORRELATION OF IMAGING STUDIES AND OPERATIVE FINDINGS IN TUMORS OF THE PARAPHARYNGEAL SPACE

Computed Tomography Scan
• Accurately defined position of the tumor relative to the styloid in 96% (43/45) of the cases.
• Correct identification of the origin of salivary gland tumors (parotid vs. nonparotid) in 88% (14/16) of the cases.
• Assessment of the tumor vascularity correlated with the angiographic findings (28/28) as well as with the surgical findings (45/45).

Table 76-5 -- CORRELATION OF SURGICAL SPECIMENS WITH ANGIOGRAPHY

Angiography
• Performed for all vascular tumors
• More specific than computed tomography scans for vascular lesions yielding a correct diagnosis in 93% (26/28) of the cases

Recently other types of imaging studies have emerged. Brink and colleagues[9] describe the high degree of accuracy of FDG-PET scanning in the detection of adrenal gland malignancies, which are often associated with carotid body tumors. The authors found that the sensitivity of FDG-PET in paragangliomas seems to be slightly inferior to DOPA-PET and MIBG-scintigraphy. The authors feel that co-registration or image fusion of ^{18}F-DOPA-PET or ^{123}I-MIBG with MRI/CT scan may improve diagnostic accuracy.

Tumors that arise de novo in the PPS independent of the parotid gland have a fat plane separating it from the deep lobe of the parotid gland. This is a very important point in the differential diagnosis, because the tumors independent of the parotid gland may be removed through a cervical approach, whereas tumors arising in the deep lobe of the parotid gland with extension into the PPS must be approached through a parotid-submandibular approach in order to avoid injury to the facial nerve.

SURGICAL APPROACHES

Management of Tumors of the Prestyloid Parapharyngeal Space

There are a variety of surgical approaches that may be used for the management of tumors of the PPS (Table 76-6).

Table 76-6 -- SURGICAL APPROACHES USED IN SURGERY OF THE PARAPHARYNGEAL SPACE

Surgical Approaches
• Transoral • Transcervical submaxillary • Transcervical • Transmandibular • Transparotid • Infratemporal fossa

Transoral excision has been advocated for neoplasms arising in the prestyloid PPS.[10] This technique does not afford the surgeon enough exposure, thereby depriving the surgeon of the opportunity to identify the closely related neurovascular structures or Stensen's duct. It may also compromise the wound through contamination by oral secretions or seed the tumor into the mucosa of the palate. Because of these adverse features we do not recommend that tumors of the PPS be removed through the transoral approach.

The prestyloid PPS is ideally approached using the transcervical submandibular technique (Fig. 76-7). The skin incision is made in the major transverse skin crease in the neck and carried down through subcutaneous tissue and platysma muscle. The mandibular branch of the facial nerve must be identified and preserved. Identification of the anterior border of the sternocleidomastoid muscle facilitates identification of the posterior belly of the digastric muscle. The submandibular gland is then dissected free of the surrounding tissues. The facial artery is clamped, transected, and doubly ligated with silk suture. The submandibular gland is then mobilized anteriorly and pedicled on the submaxillary ganglion and Wharton's duct. It is not necessary to remove the submandibular gland as has been reported by Malone and colleagues,[11] but it is important to retract the gland anteriorly because this exposure provides direct access to the apex of the prestyloid PPS. Once the gland has been mobilized and retracted, the tumor is identified. Using sharp and blunt dissection, the surgeon separates the tumor from the surrounding tissues. Cohen and colleagues[12] described specific maneuvers that were used to increase the exposure provided by the transcervical approach. The authors suggest that dividing the stylomandibular ligament and anterior dislocation of the mandible can increase exposure by 50%. The stylomandibular tenotomy was also emphasized by Orabi and associates.[13] The styloid musculature and the posterior belly of the digastric muscle may be removed if necessary, thereby further enhancing exposure.

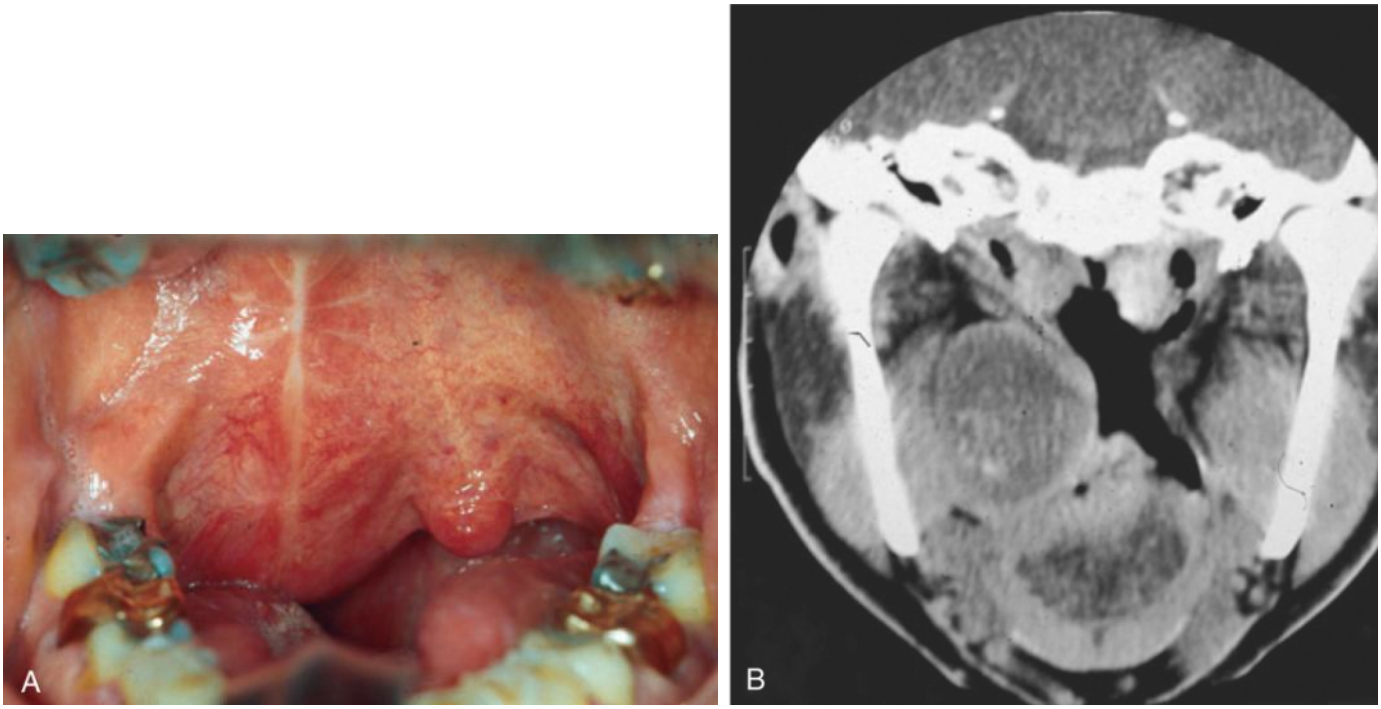

Figure 76-7 **A,** Tumor of parapharyngeal space—previously biopsied. **B,** Computed tomography scan demonstrating tumor of parapharyngeal space. **C,** Diagram of transcervical approach to prestyloid parapharyngeal space. Submandibular gland ret
(C, Reprinted with permission from Rabuzzi DD, Johnson JT: Diagnosis and Management of Deep Neck Infections. Washington, DC, American Academy of Otolaryngology-Head and Neck Surge

Tumors in the prestyloid PPS are usually pleomorphic adenomas that do not invade the surrounding soft tissue. However, to prevent recurrence, every effort should be made to preserve the capsule of the tumor. A suction drain is placed in the depths of the wound and brought out just posterior to the skin incision in order to collapse the dead space. The wound is then closed and a pressure dressing applied.

CASE REPORT

The parotid-submandibular incision is used in patients whose tumor arises from the deep lobe of the parotid gland (Fig. 76-8). This approach was used in a 45-year-old man who had been treated for sleep apnea and “heroic snoring” for 2 years before being diagnosed with a deep lobe parotid tumor with PPS extension. Physical examination revealed a large mass seen through the oral cavity that completely occupied the posterior aspect of the pharynx, shifting the tonsil and the uvula all the way over to the contralateral tonsil with almost complete airway obstruction. The patient also had a large mass in the parotid gland. The MR scan revealed a large mass almost completely occluding the oropharynx seen in the sagittal image, whereas in the axial image, the tumor was noted to arise from the deep lobe of the parotid gland and the impression made by the stylomandibular ligament as the tumor extended into the PPS could be seen. An extended parotid-submandibular incision was made, the skin flaps were elevated, and the main trunk of the facial nerve was identified by using the landmarks of the anterior border of the sternocleidomastoid muscle, the mastoid tip, and the cartilaginous external auditory meatus. The dissection was then carried anteriorly along the main truck of the facial nerve. All of the branches were dissected free of the capsule of the tumor and mobilized. The superficial lobe was pedicled anteriorly and preserved. The submandibular gland was then mobilized by ligating the facial artery and displacing the gland anteriorly. After the deep lobe of the parotid gland was dissected free of the mobilized branches of the facial nerve, blunt digital dissection was used to enter the PPS through the submandibular space. Using this three-dimensional approach, the mass was removed completely, and under direct vision one could see crossing this encapsulated mass was the impression of the stylomandibular ligament. At the end of the procedure, after hemostasis was obtained and the wound irrigated, the superficial lobe of the parotid gland was then replaced in its anatomic position in order to ensure that the contour of the face was preserved. The wound was then closed in layers over a suction drain and a dressing applied.

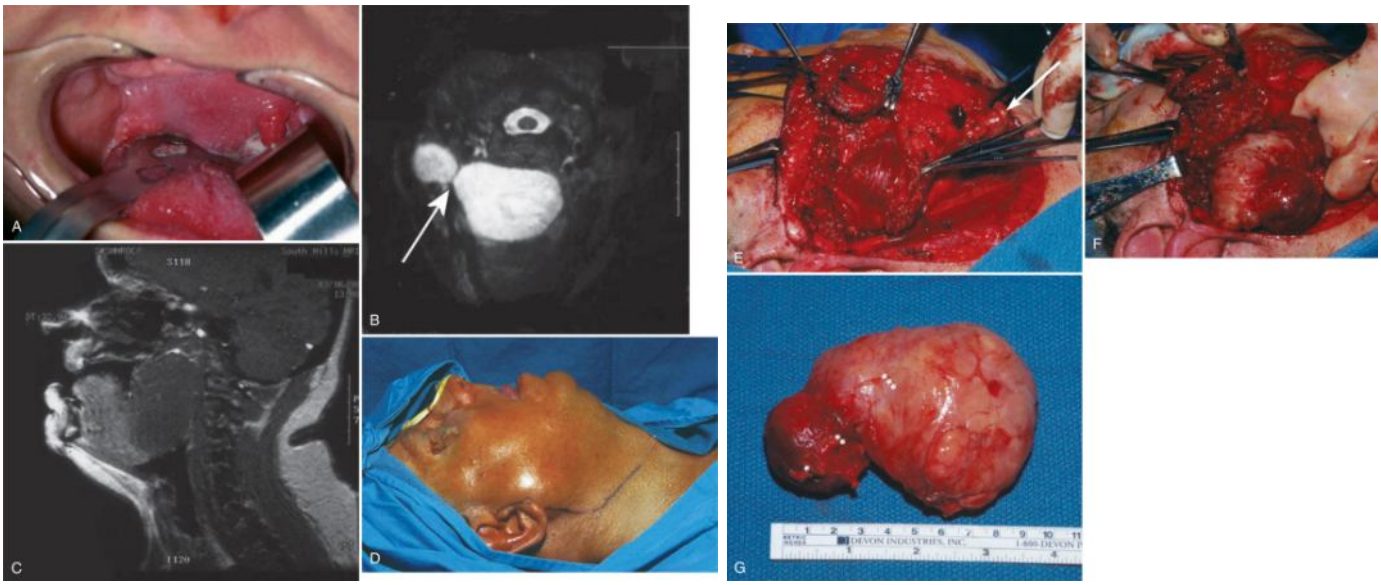

Figure 76-8 **A,** Large mass in parapharyngeal space displacing uvula to contralateral tonsil. **B,** Axial magnetic resonance (MR) scan revealing a tumor originating in the deep lobe of the parotid gland with massive extension into the parapharyngeal spa line *(arrow)* reflects the impression of the stylomandibular ligament. **C,** Sagittal MR scan demonstrating almost complete obstruction of airway. **D,** Large parotid mass noted. The parotid submandibular incision is outlined. **E,** Facial nerve dissected free fro lobe of parotid gland. Submandibular gland retracted anteriorly *(arrow)*. **F,** Blunt dissection through the submandibular space into the parapharyngeal space releases the tumor. **G,** Specimen revealing tumor originating in deep lobe of parotid gland (left), ir stylomandibular ligament, and parapharyngeal component of the tumor (right).

Although the literature is replete with descriptions of techniques, including mandibulotomy, by which exposure of the PPS may be improved, we do not feel that this approach is necessary except perhaps in very unique circumstances. Mandibulotomy can be carried out either directly through the body of the mandible or anteriorly with retraction of the mandible laterally. Teng and associates[14] described a subcutaneous mandibulotomy that the authors feel affords excellent access to the PPS without the lip-split, chin-split, and floor of the mouth incisions, which are part of the usual midline mandibulotomy. They propose the approach as an intermediate step for extensive PPS tumors that are inaccessible through the transcervical approach yet do not require a full labiomandibuloglossotomy for safe and complete removal.

MANAGEMENT OF TUMORS OF THE POSTSTYLOID PARAPHARYNGEAL SPACE

Most of the tumors that originate in the poststyloid PPS are tumors of neural origin. Most of these are PGLs (see Table 76-1), the most common of which is the carotid body tumor (CBT). It is usually possible to tell preoperatively, by imaging studies, whether these tumors are in fact CBT, glomus vagale tumors, or schwannomas. The CT scan with contrast is very helpful in the differential diagnosis. The CBT is located at the bifurcation of the carotid body, whereas the glomus vagale tumor, because of its site of origin from the vagus nerve, arises posterior to the carotid artery.

On CT scanning, both neurilemomas and CBT may look vascular. If there is a degree of uncertainty based on the anatomic site, then angiography can be the deciding factor in making the diagnosis (Fig. 76-9). Conventional angiography using the transfemoral artery approach, in our opinion, remains the most accurate technique for identification of the tumor and assessment of its vascularity. MR angiography (MRA) scanning has been used, but errors in diagnosis can be made. We recently had a patient in whom a diagnosis was made of a CBT by history, physical examination, and a vascular blush on MR scanning. An MRA was carried out, which confirmed this diagnosis. Unfortunately, at the time of surgery, an aneurysm of the internal carotid artery rather than a CBT was identified. This mistake could have been avoided had a conventional angiogram been carried out (Fig. 76-10).[15]

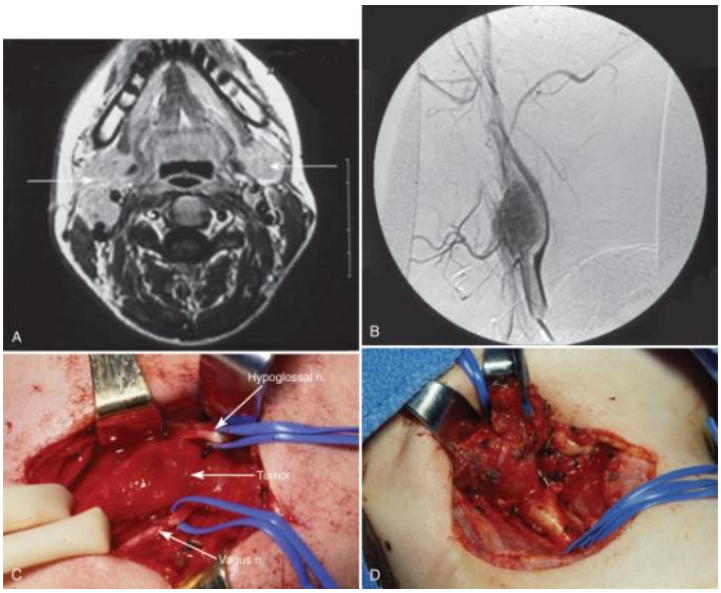

Figure 76-9 **A,** Magnetic resonance scan of a 21-year-old woman with bilateral carotid body tumors *(arrows)*. **B,** Angiogram of same patient demonstrates a carotid body tumor. **C,** The operative field, including tumor. The carotid artery and cranial nerves XI and XII have been identified; vessel loops are placed for safety. **D,** Dissection is carried along the carotid artery in a subadventitial plane up to the bifurcation of the artery. The external carotid artery is then ligated.

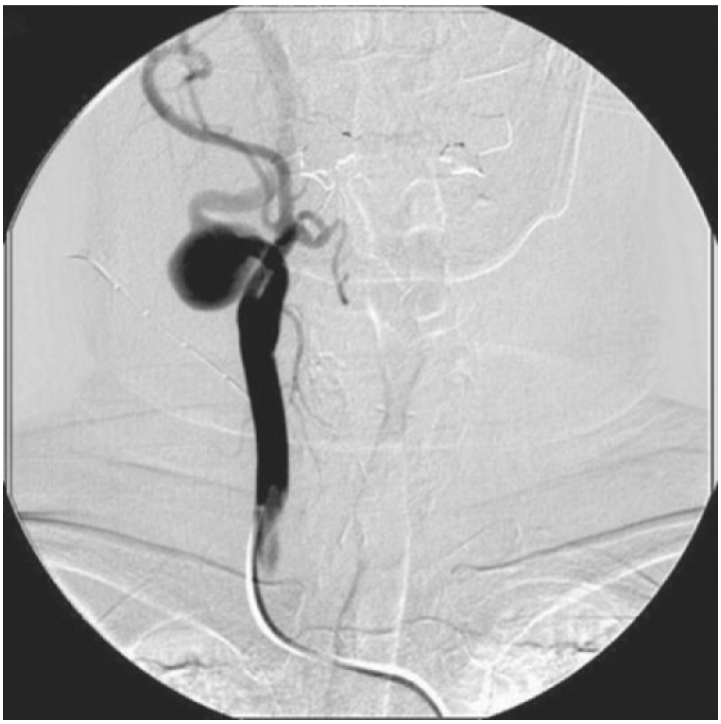

Figure 76-10 Magnetic resonance angiogram of a mass in the neck mistakenly diagnosed as a carotid body tumor. Transfemoral angiography revealed aneurysm of the internal carotid artery.

The Shamblin classification[16] of CBT and its relationship to the carotid artery (Fig. 76-11) has proved to be very useful in the assessment of CBT. The Shamblin I and Shamblin II types of tumors are amenable to surgery by dissecting the tumor off of the carotid artery. However, the Shamblin III tumors in which the carotid artery is encased cannot be removed safely by this technique. Not only is the artery encased in the tumor, but also the tumor actually invades into the muscularis of the artery and cannot be dissected free. Further dissection then creates an opening in the wall of the carotid artery. Patients with Shamblin III tumors should undergo a complete resection of the artery by a vascular surgeon with reconstruction using either a vein graft or a synthetic graft (Fig. 76-12).

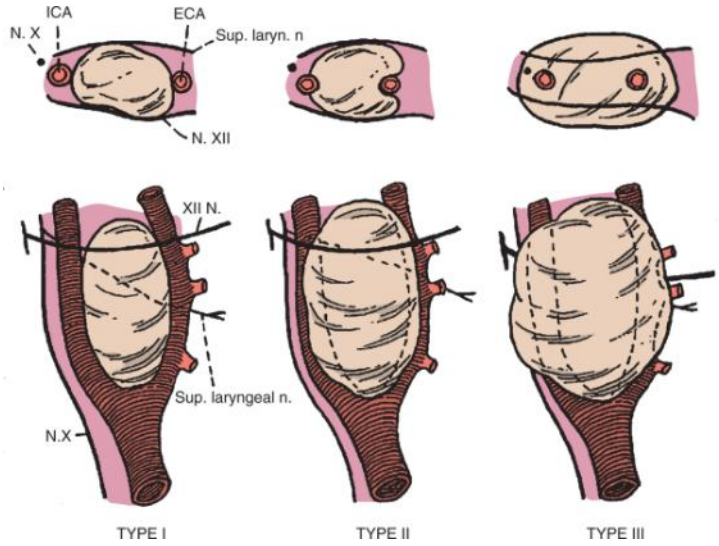

Figure 76-11 Carotid body paragangliomas. Shamblin's classification scheme: type I—small and easy to dissect, type II—medium and more intimately attached to carotid, and type III—large with transmural invasion of the wall of the carotid requiring resection and grafting.
(Reprinted with permission from Shamblin WR, ReMine WH, Sheps SG, et al: Carotid body tumors. Am J Surg 122:733-739, 1971.)

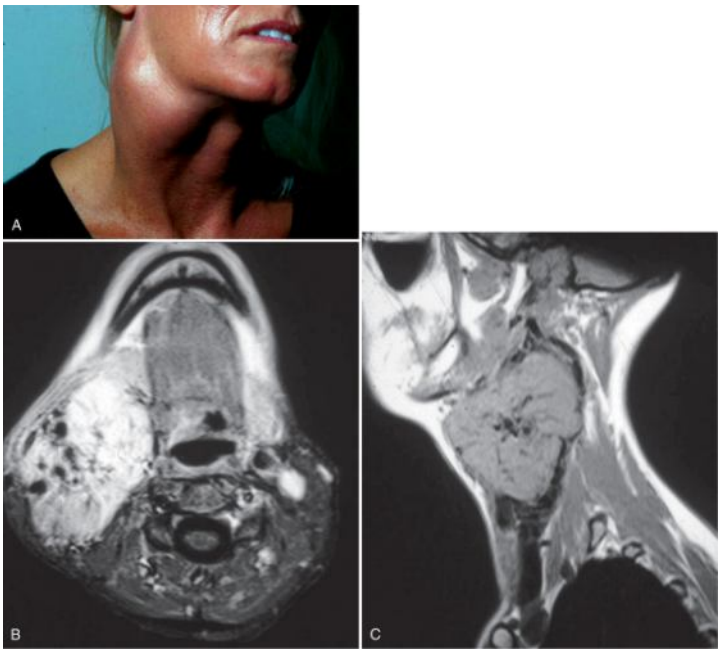

Figure 76-12 **A,** Patient with a mass in the neck, which has increased in size over a 10-year period. **B,** Magnetic resonance imaging demonstrates a large carotid body tumor with numerous flow voids. **C,** Lateral view of mass extending to prevertebral fascia.

There is a subset of PGLs referred to as the familial form (PGL1). These PGLs are multiple and usually involve the carotid body so that there may be unilateral CBT or bilateral CBT with additional PGLs arising from unnamed arteries in the vicinity. The familial PGL group, in addition to being multiple, is also more likely to be malignant and more likely to have an associated pheochromocytoma.

The familial form of PGL has an autosomal dominant inheritance pattern. Baysal and colleagues in *Science* reported the analysis of families carrying the *PGL1* gene, which revealed mutations in the *SDHD*, a mitochondrial complex 11 gene in hereditary paraganglioma, on chromosome 11q23.[17] In this paternal inheritance pattern, the father may pass the gene along to the son who has a 50% chance of having a PGL and passing it along to successive males. However, it may also pass to the female, who may develop a tumor but cannot pass the gene to her offspring because of genetic imprinting.[18] The families of patients who have PGL1 should undergo genetic testing. For patients who do not have the gene, this is reassuring and no further testing is required. Patients who test positive should undergo MRI screening. If the MRI discloses a tumor, this may be removed. If the MRI is negative, it should be repeated annually so that if a tumor develops it may be removed safely. Further information is avail-able at our website: http://www.upmccancercenters.com/paraganglioma/study.html.

INDICATIONS FOR SURGERY

Small tumors that have recently been discovered should be removed, as should tumors demonstrating growth, whether accelerated or steady, with or without involvement of the lower cranial nerve. Patients with a CBT may have this tumor removed safely if it falls into the Shamblin I or II classification. Patients demonstrating neuropathy, such as vocal cord paralysis or paralysis of the tongue, should have the tumor removed. These are usually Shamblin type III and require resection of the carotid artery. Patients who are elderly, have severe comorbidities, refuse surgery, or fail the balloon occlusion test should be considered for observation, including yearly CT scans, or radiation therapy.

We have not used preoperative embolization because the lesions in which the carotid artery is not encased can be dissected off of the muscularis of the carotid artery using dissection in the subadventitial plane. We believe that the preoperative embolization may induce an intense inflammatory response that may lead to obliteration of the subadventitial plane, making dissection of the tumor dangerous.

Patients diagnosed with any paraganglioma should be questioned about symptoms related to secretion of catecholamines, including sudden flushing, palpitations, and labile or difficult to control hypertension. All patients who have the preoperative diagnosis suggestive of paraganglioma should have examination of a 24-hour urine specimen for catecholamines before surgery. If a patient is found to have a pheochromocytoma, it should be removed before removing the paraganglioma in order to avoid a potentially fatal intraoperative hypertensive crisis. If the abnormal catecholamines are present but no pheochromocytoma is identified, it must be assumed that an unusual catecholamine-secreting PGL is present. The anesthesiologist should be consulted and a program for the management of a potential hypertensive crisis planned in advance.

SURGICAL TECHNIQUES

Tumors of the poststyloid PPS may be approached through a cervical incision. The incision is placed in a natural skinfold in the neck and carried through subcutaneous tissue. The skin flaps are elevated and the sternocleidomastoid muscle is identified and retracted. There is no need to dissect the submandibular triangle in this approach.

Carotid Body Tumor

Once the sternocleidomastoid muscle has been retracted, the carotid sheath will be identified. The jugular vein is then identified and dissected free of the carotid artery. The vagus and hypoglossal nerves are also skeletonized (see Fig. 76-9). Arterial loops are placed around these structures for the purpose of identification and retraction. The common carotid artery is then identified, skeletonized, and a Penrose drain is placed around the artery. There is usually a large venous plexus that goes approximately halfway down from the bifurcation to the clavicle. At the point where the carotid artery is clear of this plexus, subadventitial dissection is begun. This is carried superiorly until the bifurcation of the artery is encountered. At this time dissection is carried superiorly along the external carotid artery approximately 1 to 2 cm above the bifurcation (Fig. 76-13). The artery is then clamped, transected, and ligated with silk tie and a suture ligature. The larger CBTs usually extend deep and go down to the prevertebral fascia (see Fig. 76-12) and if the external carotid artery is not ligated it is very difficult to gain entry for dissection in this deep plane (see Fig. 76-13). The tumor is then dissected off the internal carotid artery and removed. Identification and retraction of the neural structures and the jugular vein usually result in a very clean, safe, uncomplicated, and relatively bloodless removal of the tumor.

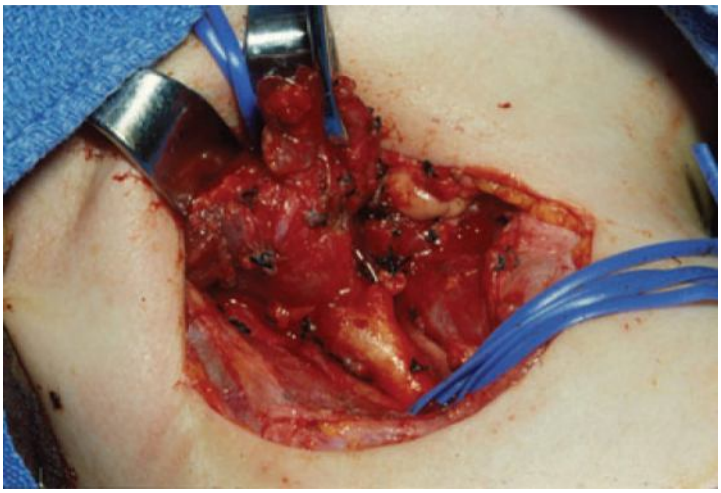

Figure 76-13 Dissection of carotid body tumor in subadventitial plane. Note ligation of external carotid artery.

Glomus Vagale Tumor

The glomus vagale tumor is adjacent to but does not involve the carotid artery. It is important initially to identify the carotid artery, skeletonize it, separate it from the tumor, and place a rubber drain around it for retraction purposes and to gain control of hemorrhage in case of an inadvertent injury to the artery. The hypoglossal nerve is identified and mobilized off of the tumor. A small vascular loop is placed around it for purposes of retraction and identification. The vagus nerve is then identified in the inferior aspect of the neck, and dissection of the nerve is carried superiorly retracting gently on the carotid artery and the nerve in order to develop a plane of dissection between the carotid artery and the tumor. The tumor, which is usually fusiform in shape, is identified as dissection is carried superiorly, gently separating the tumor from the carotid artery and the surrounding tissues. Once the dissection is carried superior to the tumor itself, the nerve can be transected. The tumor takes its origin from the nerve itself and can be abnormal, even distal to the tumor itself (Fig. 76-14). After being fully mobilized, the mass is excised, with care taken to include any abnormal-appearing nerve. Because this creates a high vagal injury with resultant vocal cord paralysis, aphonia, and potential for aspiration, we always carry out a type I thyroplasty intraoperatively to medialize the vocal cord. This avoids postoperative dysfunction and restores the patient's ability to speak and swallow.[19]

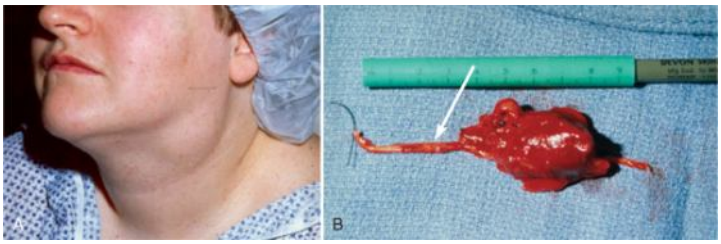

Figure 76-14 **A,** Patient with glomus vagale tumor. **B,** Glomus vagale tumor with obvious intravagal tumor *(arrow)*.

Neurogenic Tumors

The most common neurogenic tumors are the schwannoma of the vagus nerve and the superior cervical ganglion. The same incision is used as for PGL to identify and isolate these tumors. Schwannoma of the vagus nerve is removed in a manner exactly the same as that described above with the glomus vagale tumor. An immediate thyroplasty is also carried out.

In the case of the schwannoma of the superior cervical ganglion, a procedure similar to that described above isolating the neural and vascular structures is carried out and then the carotid artery is mobilized and retracted so that the tumor, which arises deep to the bifurcation of the carotid artery, can be removed intact. This procedure invariably results in Horner's syndrome, which is unavoidable. These tumors can be localized by preoperative scanning and identified as a tumor of the superior cervical ganglion. Patients must be counseled accordingly with respect to Horner's syndrome.

COMPLICATIONS

Prestyloid Parapharyngeal Space

The most common complication of dissection of this space relates to the facial nerve. If the tumor arises from an embryonic rest, then no complications should be encountered. If, however, this is a PPS extension of a deep lobe parotid tumor, then it is possible that injury to the facial nerve, either from inaccurate dissection or traction on the nerve, may occur. Patients who have weakness or paresis from traction on the facial nerve in whom the nerve is anatomically intact should expect return of function within 3 months.[12] Proper eye care is essential for patients with facial nerve injury. In the immediate postoperative period, a moisture chamber is used. Patients who are expected to have a prolonged recovery phase benefit from insertion of a gold weight into the upper eyelid.

First Bite Syndrome

First bite syndrome may occur in patients with dissection of prestyloid PPS tumors.[20] This is characterized by a crampy pain in the parotid region after the first bite of each meal. The pain lasts a few seconds, improves with each bite, and is extinguished over the next several bites. Symptoms are at their worst with the first bite of food in the meal. There is no really good medical treatment. Surgical treatment requires transmeatal excision of Jacobson's plexus on the promontory of the middle ear.

Poststyloid Parapharyngeal Space

The potential for injury to the carotid artery is minimized by preoperative evaluation to rule out the possibility of encasement of the carotid artery and invasion of the artery by the tumor. Even very precise dissection in the subadventitial plane is dangerous and should not be attempted in this setting.

Inadvertent injury to the hypoglossal in itself is not severe and requires no reconstruction. Inadvertent transection of the vagus nerve requires immediate thyroplasty. However, if both the hypoglossal and the vagus nerve are injured and the thyroplasty is not carried out, the patient will have substantial problems with a weak voice and aspiration.

PEARLS

- Tumors in the prestyloid parapharyngeal space are usually of salivary gland origin.
- Tumors in the poststyloid parapharyngeal space are usually neurogenic in origin.
- Modern imaging techniques permit the surgeon to formulate a plan of management without a preoperative biopsy.
- Excision of a tumor in the deep lobe of the parotid gland with extension to the parapharyngeal space requires that the branches of the facial nerve be mobilized off of the tumor.
- Intraoperative thyroplasty is used routinely when the vagus nerve is taken or injured at surgery.

PITFALLS

- Attempting to remove a Shamblin III type of carotid body tumor using a subadventitial approach will eventuate in massive bleeding due to invasion of the wall of the artery by tumor.
- Failure to evaluate for abnormal catecholamines in patients with paraganglioma (PGL) may result in intraoperative hypertensive crisis in the presence of a secreting PGL or pheochromocytoma.
- Failure to preserve the lower cranial nerves in excision of carotid body tumor will result in hoarseness and aspiration in the postoperative period.

- Facial nerve injury may be the result of failure to recognize that the tumor in the prestyloid parapharyngeal space may actually be extension of a tumor in the deep lobe of the parotid gland.
- Transoral biopsy of a tumor of the parapharyngeal space will result in contamination of the mucosa and make eventual transcervical resection more difficult.