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Tumors and Surgery of the Parapharyngeal Space

Kerry D. Olsen, MD

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Tumors and Surgery of the Parapharyngeal Space

Kerry D. Olsen, MD

The parapharyngeal space, an area of complex anatomic relationships, is involved in a wide variety of benign and malignant neoplasms. Because primary parapharyngeal space tumors are rare, it is difficult to obtain surgical experience in this region. This paper reviews the anatomy, presentation, evaluation, surgical approaches, and pathologic features and complications reported in managing patients with parapharyngeal space neoplasms.

Two surgical procedures have been used by the author to treat 44 tumors in the parapharyngeal space. The cervical-parotid approach was used in 35 patients, and the cervical-parotid approach with midline mandibulotomy was used in 9 patients. Of the 44 tumors, 32 were benign lesions and 12 were malignant neoplasms. Forty tumors were primary parapharyngeal space tumors, and 4 cases represented isolated metastases to parapharyngeal nodes. Recurrent tumors accounted for 12 of the 44 cases. Discussion of the indications, surgical technique, and select points pertinent to using these two operative procedures is based on the operative experience gained from these 44 patients. The use of these two operations resulted in low morbidity and provided a safe, efficacious approach to the management of all parapharyngeal space neoplasms encountered.

INTRODUCTION

Tumors of the parapharyngeal space are rare, accounting for only 0.5% of head and neck neoplasms.[1] The wide spectrum of benign and malignant neoplasms encountered in this complex, deep anatomic region contributes to the challenge of surgical treatment of parapharyngeal space tumors. A systematic logical approach to the diagnostic evaluation of patients with parapharyngeal space tumors is necessary to ensure management decisions that minimize morbidity.

The parapharyngeal space is actually a potential space located lateral to the upper pharynx. This bilateral fat-containing region extends from the skull base to the hyoid bone. Various synonyms for parapharyngeal space have been reported in the literature: lateral parapharyngeal space, parapharyngeal space, pharyngomasticatory space, pharyngomaxillary space, pterygopharyngeal space, and pterygomandibular space. The term "parapharyngeal space" is most commonly used in the recent literature.[2]

This paper presents an overview of the anatomy, presentation, evaluation, tumors, and treatment options for parapharyngeal space neoplasms. The author's experience with many tumors in this area led to the use of two surgical approaches for treating tumors in this region: the cervical-parotid approach and the cervical-parotid approach with midline mandibulotomy. The indications, surgical technique, and complications of these procedures will be discussed in detail.

ANATOMY

Surgeons must be knowledgeable about the complex anatomy of the parapharyngeal region to remove tumors from this area safely. Failure to appreciate the regional anatomic relationships can result in the selection of an incorrect surgical approach. The result may be inadequate access with difficulty in tumor removal, damage to vital structures, tumor spillage, and recurrent neoplasms.

The parapharyngeal space is often described as an inverted pyramid with its base at the skull and apex at the greater cornu of the hyoid bone (Fig. 1). The parapharyngeal region is further compartmentalized by thick fascial layers that direct tumor growth. Prior descriptions of these fascial layers varied.[3]

The superior border of the parapharyngeal space is a small portion of the temporal bone. The superior medial wall is enclosed by a fascial connection from the medial pterygoid plate to the spine of the sphenoid. This fascia passes medial to the foramen ovale and the foramen spinosum. These foramina are not included in the superior bony limits of the parapharyngeal space but, rather, are in the infratemporal fossa or masticator space[3] (Fig. 2).

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From the Department of Otorhinolaryngology, Mayo Clinic and Mayo Foundation, Rochester, Minn.

Send Reprint Requests to Mayo Clinic, 200 First St. SW, Rochester, MN 55905.

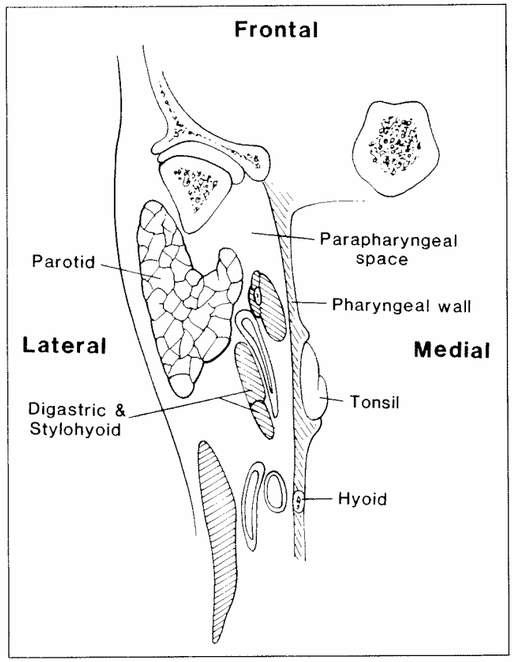

Fig. 1. Inverted pyramidal shape of parapharyngeal space.

The inferior boundary of the parapharyngeal space ends at the junction of the posterior belly of the digastric muscle and the greater cornu of the hyoid bone. The firm fascial attachments in this area limit parapharyngeal space extension inferior to the hyoid bone. This fascia, however, can be weak and may be an ineffective barrier to the spread of infections.

The posterior border of the parapharyngeal space is formed by the fascia over the vertebral column and paravertebral muscles. The anterior limit is composed of the pterygomandibular raphe and medial pterygoid fascia.

The lateral wall of the parapharyngeal space is made up of the fascia overlying the medial pterygoid muscle and the ramus of the mandible. The fascia of the medial pterygoid muscle superiorly incorporates the sphenomandibular ligament that extends from the spine of the sphenoid to the lingula of the mandible. This dense fascia then continues as a firm layer to the skull base. This fascial layer separates the parapharyngeal space from the inferior alveolar nerve, the lateral pterygoid muscle, and the condyle of the mandible.[3] The retromandibular portion of the deep lobe of the parotid gland also forms a small portion of the lateral border, as does a portion of the posterior belly of the digastric muscle.

The medial border superiorly is formed by the approximation of the fascia from the tensor veli palatini muscle to the medial pterygoid muscle. The pharyngobasilar fascia forms the posterior medial border for the retrostyloid space near the levator veli palatini muscle. Inferiorly, the medial border is contiguous with the fascia over the superior constrictor muscle, and contains attachments of the stylopharyngeal aponeurosis. The inferomedial wall continues with fascia that joins the styloglossus and stylopharyngeus muscles.

The parapharyngeal space has been further divided by most authors into a prestyloid and a retrostyloid compartment (Fig. 3). Fascia that extends from the styloid process to the tensor veli palatini muscle crosses posteriorly in the parapharyngeal fat and separates the parapharyngeal space into these two areas. More posteriorly, this fascial plane blends with the styloid muscles.[3] The prestyloid space extends superiorly into a blind pouch formed by the joining of the medial pterygoid fascia to the tensor veli palatini fascia[3] (Fig. 4). This space contains a variable portion of the retromandibular deep lobe of the parotid gland. In addition, a small branch of the fifth cranial nerve crosses this area to reach the tensor veli palatini muscle. Most of the prestyloid parapharyngeal space is composed of fat. Therefore, tumors in this area are generally limited to salivary lesions, lipomas, and rare neurogenic tumors.[3]

The retrostyloid compartment or poststyloid compartment contains the carotid artery and jugular vein located posterolateral to the artery at the skull base. Cranial nerves IX through XII accompany these vessels, with the 10th nerve occupying a position between the artery and the vein. The 11th nerve crosses the vein anteriorly or posteriorly, and the 9th nerve crosses the carotid artery laterally. The 12th nerve ends its vertical course outside the parapharyngeal space.[4] This compartment also contains the sympathetic chain, lymph nodes, and glomus tissue. These structures all serve as a potential source for a retrostyloid tumor. The retropharyngeal space is separated from the retrostyloid space by a thin fascial layer that is a minimal barrier to the spread of tumor or infection.

The fascia uniting the styloid process to the mandibular ramus is called the stylomandibular ligament. This ligament forms one of the boundaries of the stylomandibular tunnel, as described by Patey and Thackray.[5] The remaining borders include the skull base and ascending ramus of the mandible. Extension of tumors through the rigid opening of the stylomandibular tunnel will often be noted by constricted tumor growth in this narrow area. Surgical entry to the parapharyngeal space is improved by dividing the stylomandibular ligament, removing the styloid process, or dislocating the jaw forward.

The parapharyngeal space has numerous lymphatics that drain the paranasal sinuses, the oro-

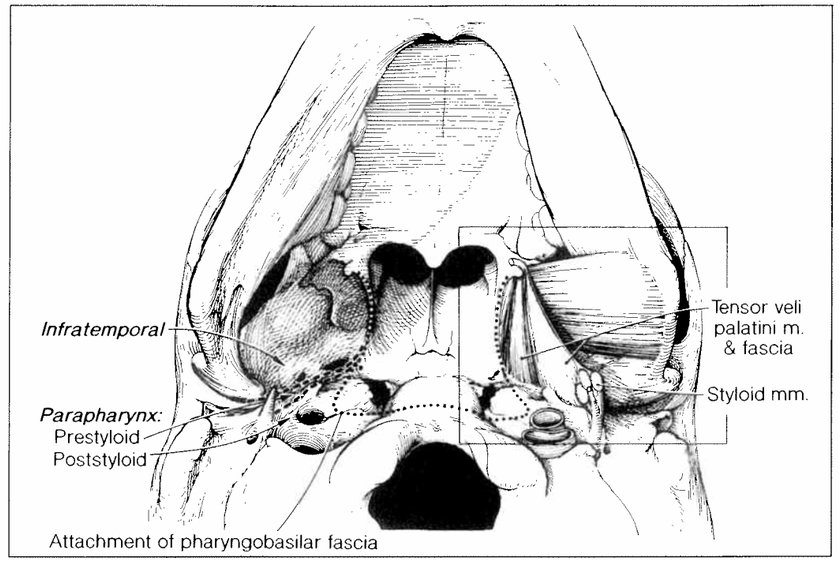

Fig. 2. Area of skull base forming superior border of parapharyngeal space and infratemporal fossa. (By permission of Mayo Foundation.)

pharynx, the oral cavity, and a portion of the thyroid gland. These nodes are connected superiorly to the node of Rouviere, situated in the retropharyngeal space, which drains the nasopharynx, upper oropharynx, and sinuses.[6] Inferiorly, the lymphatic drainage continues to the jugular digastric nodes, which are outside and inferior to the parapharyngeal space.[6] A direct connection from the lymphatics of the thyroid gland to the parapharyngeal nodes was reported.[7] Lymph channels extend along the posterior wall of the pharynx and terminate in the lateral retropharyngeal node. This pathway exists as a result of the embryologic development of the thyroid gland from the tongue base. Similarly, hypopharyngeal and laryngeal tumors that involve the posterior pharyngeal wall may also metastasize to the lateral retropharyngeal nodes.[7]

In summary, the only nonrigid borders of the parapharyngeal space are the medial and inferior areas. In addition, laterally, a small opening occurs in the stylomandibular tunnel. It is in this last area that the more medial fascia of the tensor veli palatini muscle separates from the more lateral fascia of the medial pterygoid muscle, causing a space for parotid tumors to enter the prestyloid area. Additional descriptions of the anatomy of the parapharyngeal space can be found in several excellent reviews.[3,8-12]

Confusion regarding the prestyloid parapharyngeal space relates to the erroneous incorporation of the infratemporal fossa into the prestyloid compartment. Therefore, tumors reported to originate in the parapharyngeal space, in actuality, may not. The infratemporal fossa or masticatory space contains the lateral pterygoid muscle, the third division of the trigeminal nerve, and the condyle of the mandible. Tumors that involve the skull base at the foramen ovale should not be considered parapharyngeal neoplasms. Lesions confined to the jugular foramen also are often included as parapharyngeal space neoplasms. More commonly, all deep lobe parotid tumors are reported as parapharyngeal space lesions. That portion of the parotid gland deep to the facial nerve yet lateral to the ramus of the mandible should not be considered as parapharyngeal. The only parapharyngeal extension of the parotid deep lobe is the retromandibular component. Numerous studies also contain reports of large numbers of carotid body tumors because some authors classify all carotid body tumors as parapharyngeal space neoplasms. Only carotid body tumors that extend above the level of the posterior belly of the digastric muscle should be considered to involve the parapharyngeal space. Carotid body tumors that extend into the parapharyngeal space are rare.[13]

Reports of parapharyngeal space neoplasms that contain a large number of deep lobe parotid tumors, jugular foramen neoplasms, infratemporal fossa tumors, or carotid body tumors should be reviewed critically because they probably refer to tumors that are not neoplasms of the parapharyngeal space.[14] Discrepancies in tumor incidence from various reports can often be explained by these differences in defining the limits of parapharyngeal space tumors.

TUMOR PRESENTATION

Parapharyngeal space tumors generally pre-

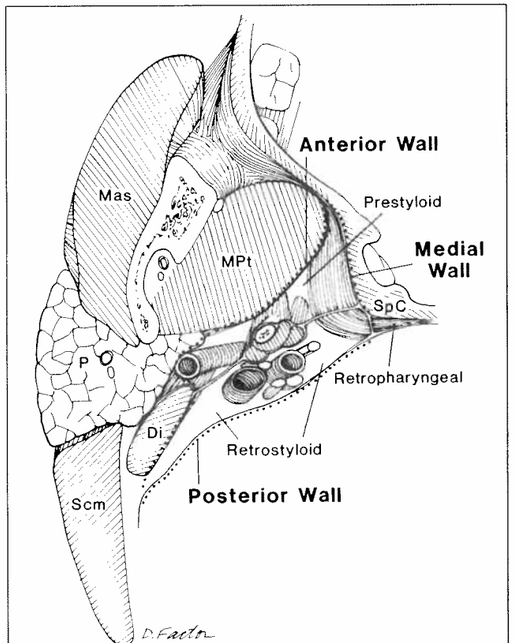

Fig. 3. Prestyloid and retrostyloid compartments of parapharyngeal space. (By permission of Mayo Foundation.)

sent as asymptomatic masses. They often are discovered during a routine physical examination. Their presence should be suspected when a subtle fullness is noted in the soft palate or tonsillar region or when there is mild fullness near the angle of the jaw (Fig. 5). Clinical detection of early parapharyngeal space lesions is difficult. Small tumors cause few symptoms. The tumors must grow to at least 2.5 to 3.0 cm before a mass can be detected clinically.[15]

As tumors enlarge and extend superiorly, they may cause symptoms related to the eustachian tube and, rarely, middle ear effusions. As tumors expand medially, voice change, nasal obstruction, aspiration, and dyspnea can occur. Rarely, tumors have been found that require immediate tracheotomy for relief of upper airway obstruction. As tumors enlarge, they may compress the 9th, 10th, 11th, or 12th cranial nerve, causing hoarseness, dysphagia, or dysarthria. Horner's syndrome may also be produced by tumor pressure on the superior cervical ganglion.[16] For a benign tumor to cause nerve deficits, it must generally enlarge to a considerable degree. Pain, trismus, or cranial nerve paralysis often suggests malignancy.

Several reports[2,17,18] described obstructive sleep apnea symptoms secondary to parapharyngeal space lesions. The symptoms include loud snoring, restless sleep, and daytime hypersomnolence from the airway encroachment secondary to palatal and tonsillar displacement from pleomorphic adenomas and cystic hygromas.

Malignant tumors of the parapharyngeal space can cause carotid sinus hypersensitivity and glossopharyngeal neuralgia. Asystole, bradycardia, and hypertension were reported[19,20] from inflammatory or malignant conditions in the parapharyngeal space. These symptoms may be due to neural irritation of the glossopharyngeal afferent fibers.

Tumors of the parapharyngeal space are often misdiagnosed as infections or tonsil tumors. Three of 23 patients reported by Allison, *et al.*[2] underwent tonsillectomy because of unilateral tonsillar swelling. The actual problem was a parapharyngeal space lesion. Another case of a neurofibroma in the parapharyngeal space was initially misdiagnosed as quinsy.[21] Patients often complain of a mild sore throat, a lumplike feeling in the throat, or dysphagia. The swelling in the tonsillar and soft palatal region may be misdiagnosed as a peritonsillar abscess. Delays in diagnosis have also occurred because patients were being treated for presumed nasal obstruction, eustachian tube dysfunction, or serous otitis media. Patients have also been misdiagnosed with temporomandibular joint pathologic condition when in actuality they had a parapharyngeal space lesion.

The performance of a complete head and neck examination is one of the most important aspects of the evaluation of parapharyngeal space tumors. However, the anatomic location of the parapharyngeal space makes it difficult to assess accurately tumor presence and size. For a mass to be noted on clinical examination, it generally must be at least 3 cm.[22] Only a mass of considerable size will cause a visible bulge or palpable abnormality of the lateral pharyngeal wall or external neck.[23] Swelling of the medial wall of the oropharynx is generally the first sign of a parapharyngeal space lesion. Parotid tumors displace the tonsil and neurogenic tumors often more characteristically displace the posterior portion of the pharynx and posterior tonsillar pillar. As tumors enlarge superiorly, they fill the space between the heads of the tensor veli palatini muscle, causing soft palate and nasopharyngeal swelling.[24] As a tumor extends inferiorly, it presents as a palpable mass near the angle of the jaw. Cranial nerve function should be noted.

Bimanual palpation with one of the physician's fingers in the patient's mouth and the physician's other hand on the patient's neck assesses mobility, pulsation, and tumor extent and may help determine the tumor's origin.[11,25] The finding of a pulsatile mass is generally not a helpful differentiating characteristic because of the frequency of transmitted pulsations. Clinical examination, however, remains difficult because the parapharyngeal space lies deep to the muscles of mastication, the mandible, and the parotid glands. Usually, the overlying pharyngeal mucosa is normal. If a preauricular mass is noted at physical examination in addition to a lateral pharyngeal mass, this indicates a parotid tumor extending through the stylomandibular tunnel.

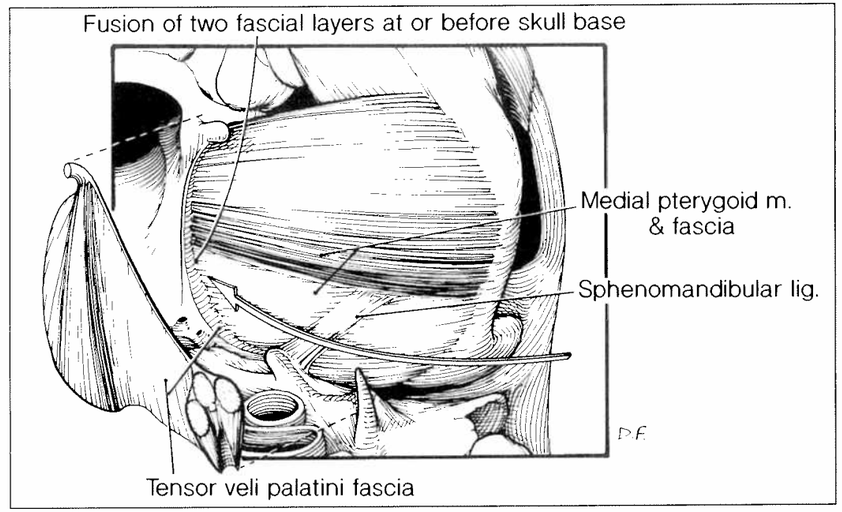

Fig. 4. Superior extent of prestyloid compartment closed by fusion of fascia from medial pterygoid and tensor veli palatini muscles. (By permission of Mayo Foundation.)

PARAPHARYNGEAL SPACE TUMORS

Tumors of the parapharyngeal space include primary neoplasms, direct extension from adjacent regions, and metastatic disease (Table I). Malignant tumors can invade the parapharyngeal space from the nasopharynx, oropharynx, mandible, maxilla, oral cavity, or parotid gland. The node of Rouviere is often the first site of metastasis of nasopharyngeal or antral carcinoma.[1] This review will focus on primary neoplasms of the parapharyngeal space and not on tumors that involve the parapharynx by direct extension from adjacent areas. Primary parapharyngeal tumors, however, can extend intracranially through the jugular foramen or into the retropharyngeal space.[26]

Reports in the literature[27] indicate that 80% of parapharyngeal space tumors are benign and 20% are malignant. The most common primary tumors are salivary gland neoplasms that originate from the deep lobe of the parotid gland or from minor salivary gland tissue. Two percent of parotid lesions are said to present as parapharyngeal space tumors.[22] Neurogenic tumors, primarily neurilemomas and paragangliomas, are the second most frequent group of neoplasms.

Overall, however, parapharyngeal space tumors are rare. Work and Hybels[28] noted only 40 parapharyngeal space neoplasms in a 10-year period. McIlrath and co-workers[29] reported 112 patients with parapharyngeal space tumors in a 30-year period at the Mayo Clinic. In a 10-year period, 5 cases of parapharyngeal space neoplasms were identified at two Southern Illinois University teaching hospitals.[30]

Considering the complexity of the contents of the parapharyngeal space, it is not surprising that there is a wide variety of neoplastic tumors. A compilation[31] of 318 cases of parapharyngeal space neoplasms reported in the literature showed 45.6% salivary gland origin, 23.3% neurogenic, 15.1% involvement of lymph nodes, and 16.1% miscellaneous. Another report of eight studies in the literature reviewed by John, *et al.*[32] found the following tumor distribution: 44.6% salivary gland tumors, 29.4% neurogenic tumors, 8.3% lymphomas, 4.6% infective cause, 2.3% metastatic, and 10.8% miscellaneous. A report (1990)[22] from the University of Pittsburgh showed 30% mixed tumors, 28% carotid body tumors, 13% glomus vagale, and 29% miscellaneous tumors. Two other reports[27,33] found similar distributions, with major and minor salivary gland tumors accounting for 40% to 50%; neurogenic tumors, 27% to 40%; and miscellaneous, 10% to 33%. Overall, approximately 50% of tumors of the parapharyngeal space are salivary gland in origin and 20% are neurogenic.[25]

Salivary Gland Neoplasms

In the majority of reports, pleomorphic adenoma is the most common parapharyngeal space tumor. This tumor generally originates from the deep

Fig. 5. Clinical appearance of soft palate swelling from prestyloid pleomorphic adenoma.

lobe of the parotid gland but also can occur from extraparotid salivary tissue. Bond (1916)[34] reported the first documented case of a mixed tumor involving the parapharyngeal space. New (1920)[35] added four additional parapharyngeal space tumors to the literature in a review of 68 pleomorphic adenomas. Stein and Geschickter (1934)[36] found parapharyngeal extension in 7 of 113 parotid tumors.

Approximately 10% of all parotid neoplasms are thought to originate from the deep parotid lobe. Of these tumors, only a small percentage, less than 1%, extend into the parapharyngeal space.[25,37] The major symptom in these patients is awareness of a mass in the throat.

The most common deep lobe parotid tumors arise from the gland deep to the facial nerve, yet remain lateral to the mandible. These tumors, in the majority of cases, do not involve the parapharyngeal space. They present as an external parotid mass indistinguishable clinically from most superficial parotid lesions.

However, a deep-lobe parotid tumor may have an external component palpable anterior to the tragus and have a pharyngeal component that extends through the stylomandibular tunnel into the parapharyngeal space. This "dumbbell" tumor was first described by Patey and Thackray.[5] The pharyngeal component of these tumors is usually the smallest and may not be clinically visible. It may only be appreciated by bimanual palpation. One half of the pleomorphic adenomas in the series by Work and Hybels[28] showed a dumbbell-type configuration. Another report[37] of 481 cases of parotid tumors showed that 21 were parapharyngeal and 15 of the 21 were the dumbbell variety. This is very different from the author's experience and other reports[25] in which the dumbbell, mixed tumor type was not the most common.

Pleomorphic adenomas can also arise from the retromandibular portion of the parotid gland. Medial extension occurs where the external carotid pierces the parotid fascia inferior to the stylomandibular ligament.[25] The tumor expands into the parapharyngeal space and displaces the tonsil and palate. The typical tumor configuration is more round. As these tumors grow and enlarge, they may also become palpable below the angle of the jaw. They do not expand laterally to present as a pretragal mass, as do dumbbell tumors.[25] These tumors can enlarge to such degree that they displace the soft palate and obstruct the nasopharynx. They cause surprisingly few symptoms and generally have no palpable component in the neck.

TABLE I.
Tumors of the Parapharyngeal Space.

Primary tumors
Direct extension from adjacent structures
Mandible
Maxilla
Nasopharynx
Neck
Oral cavity
Oropharynx
Temporal bone
Metastatic tumors
Follicular thyroid carcinoma
Medullary thyroid carcinoma
Osteogenic sarcoma
Papillary thyroid carcinoma
Squamous cell carcinoma

A final origin of parotid parapharyngeal space tumors is the tail of the superficial lobe. These tumors can form round lesions that grow medially and cranially to present as a parapharyngeal space mass. The pharyngeal component is generally the largest, but the tumors also have an external palpable component situated posterior and inferior to the angle of the jaw.[25]

Extraparotid salivary tissue also is a source for parapharyngeal space salivary neoplasms; the reported[15] benign-to-malignant ratio is 9:1. These salivary gland tumors usually are benign pleomorphic adenomas. These tumors may arise in ectopic salivary rests in lymph nodes or from ectopic salivary gland lateral to the superior pharyngeal constrictor muscle. This salivary tissue lying in the parapharyngeal fat may be the source for parapharyngeal space tumors with no obvious connection to the deep lobe of the parotid gland and no extension through the constrictor muscle.[15] Parapharyngeal salivary tumors also may arise from serous glands located in the pharyngeal mucosa medial to the superior constrictor muscle. Warrington, *et al.*[38] reported nine cases of pleomorphic adenomas of the parapharyngeal space. Five of these were situated medial to the superior constrictor muscle and were thought to arise from minor salivary glands in the pharyngeal mucosa.

Removal of parapharyngeal pleomorphic adenomas, in most cases, involves, by necessity, a capsular excision and enucleation. Tumor recurrence is possible, especially if the capsule is broken and tumor spillage occurs. Previous biopsy or attempts at transoral removal also increase the risk of recurrence. However, the loose areolar tissue in the parapharyngeal space may permit capsular dissection with little risk of tumor spillage. This may account for a lower recurrence of parapharyngeal pleomorphic adenomas than would occur in a comparable series of superficial lobe parotid tumors removed via enucleation.[24]

Other benign salivary tumors have occurred in the parapharyngeal space. These include 2 cases of Warthin's tumors and 2 oncocytomas of 17 benign salivary lesions in one study.[24] A benign lymphoepithelial lesion has also been reported in the parapharyngeal space.[6]

The frequency of malignant parapharyngeal salivary tumors compared to benign salivary tumors varied from a low of 24% to a high of 75%.[15,24] The reported malignant tumors include mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, malignant mixed carcinoma, squamous cell carcinoma, adenocarcinoma, and a case of a malignant Warthin's tumor.[15,24,37,39]

Neurogenic Tumors

The most common neurogenic neoplasm found in the parapharyngeal space is the neurilemoma or schwannoma. The site of origin is generally the vagus nerve or sympathetic chain.[40] Overall, approximately 30% of all neurilemomas occur in the head and neck area, with the majority originating from the vagus nerve.[41] The vagus nerve has been reported to be the nerve of origin in 50% of parapharyngeal neurilemomas.[42] In a review of vagus nerve tumors, however, the most frequent neoplasm of the vagus nerve was a paraganglioma in 50%.[43] A neurilemoma was noted in 31%, neurofibroma in 14%, and neurofibrosarcoma in 6%.[43] Cranial nerves IX through XII and the cervical sympathetic chain are all encased by Schwann cells and can also give rise to neurilemomas.

In general, parapharyngeal neurilemomas are uncommon. The first case of a neurilemoma in the parapharyngeal space was reported by Figi[44] in 1933. Guggenheim[45] reviewed the world literature in 1953 and reported 34 cases of neurilemoma in the parapharyngeal space. In 1981, 114 cases of parapharyngeal schwannomas had been reported.[46] By 1984, Chang and Schi[47] could find only 61 neurilemomas specifically involving the vagus nerve in the parapharyngeal space.

Neurilemomas are generally benign slow-growing tumors and present as masses. They usually do not affect their nerve of origin. With time they can cause obstructive symptoms and loss of nerve function. Pain is uncommon. There have been many reports of large neurilemomas without associated nerve dysfunction. In a report[29] of 16 patients with neurilemomas of the parapharyngeal space whose ages ranged from 24 to 72 years, half the patients were asymptomatic. Only 1 patient had Horner's syndrome and 1 had a fixed cord. The carotid artery is usually displaced anteromedially by most parapharyngeal neurilemomas. The most common enhancing extraparotid lesion on computed tomographic (CT) scan is the schwannoma.[48]

The cervical sympathetic chain is the second most common nerve of origin for neurilemomas of the parapharyngeal space.[49] Preoperative Horner's syndrome is rare but can occur. The glossopharyngeal nerve has also been reported[41,42] as the source of parapharyngeal space neoplasms. When large parapharyngeal neurilemomas involve adjacent nerves, it may be difficult to determine the true nerve of origin. Hypoglossal neurilemomas have also been reported[50] with a frequency greater than ninth nerve tumors.

Tumors can extend through the jugular foramen intracranially. The true distinction between a jugular foramen tumor and a parapharyngeal space neurilemoma as to the site of origin is often unclear.[42] Extension through the jugular foramen intracranially, however, is not common.

Treatment of schwannomas is enucleation or tumor removal with preservation of the involved nerve if possible. Often, however, the large size of the tumor precludes nerve preservation. Nerve grafting is difficult when tumors extend up to the jugular foramen area. However, if possible, nerve grafting should be performed. A successful case of a nerve graft placed after removal of a parapharyngeal neurilemoma of the 12th nerve and return of function noted 15 months later was reported by McCurdy, *et al.*[50] Less than 1% of all schwannomas undergo malignant change.

Chemodectomas or paragangliomas that involve the parapharyngeal space originate from either the vagal or carotid bodies. Stout[51] reported the first paraganglioma arising from the vagus nerve. Since that time, nearly 150 cases of paraganglioma of the vagus nerve have been reported in the literature.[52] Glomic tissue of neural-crest origin has been found in or around the surface of the nodose ganglion. This finding was first described by White[53] and has been termed the "glomus intravagale" or "paraganglion intravagale." Vagal paragangliomas are parapharyngeal in location in two thirds of cases.[29]

In contrast, carotid body tumors that extend into the parapharyngeal region are much rarer, occurring in various series from 8% to 30% of the time.[25,29] Carotid body tumors can present with an

associated pharyngeal mass that displaces the tonsil medially. Extension into the parapharynx above the level of the posterior belly of the digastric muscle justifies inclusion as parapharyngeal space tumors. The usual presentation is a painless mass located below the angle of the jaw, and these tumors are not considered parapharyngeal in origin. Overall, paragangliomas are multicentric in 10% to 20% of cases and have been reported to be more common in females.[43,52] In addition, there can be a strong familial tendency with an autosomal dominant inheritance.[52]

Vagal paragangliomas often cause bulging of the lateral pharyngeal wall. Symptoms due to vagal nerve involvement, *i.e.,* hoarseness or dysphonia, may be present.[24] Up to 30% of vagal paragangliomas present with cranial nerve deficits manifesting as vagal paralysis or jugular foramen syndrome.[52] The malignant rate was reported to be 10%, with cervical nodes positive in two thirds of those cases.[52]

Three documented cases of vagal paragangliomas producing catecholamines were reported.[43] Therefore, all potential paragangliomas must be screened for possible secretion of bioactive amines.

Glomus vagale tumors can extend intracranially and produce a fatal outcome.[26] Special consideration regarding surgical removal has to be directed to their vascularity, which can encompass and surround the internal carotid artery. In up to 10% of cases, major vessels have been reported to be resected with the tumor, and associated nerve injury is possible.[6]

Carotid body tumors that interfere with swallowing or speaking, compress the palate or pharynx, or experience an aggressive growth pattern should be considered for removal. It is unusual for a patient to die of an untreated carotid body tumor, and metastasis is rare.[11] Surgical removal is generally done in individuals who are less than age 70 years and in good health.

Additional neurogenic tumors include neurofibromas and ganglion cell tumors and their malignant counterparts: malignant paragangliomas and schwannosarcomas.[54] Neurofibromas are the third most common neurogenic tumor occurring in the parapharyngeal space. Neurofibromas generally occur as multiple lesions. They intimately involve the nerve and removal of the tumor with preservation of the nerve is generally not possible. Cranial nerve deficits occur more often with neurofibromas than with schwannomas.[25] Neurofibromas in the parapharyngeal space have been reported[21,55] to involve the vagus nerve, the glossopharyngeal nerve, and the spinal accessory nerve. A ganglioneuroma of the parapharyngeal space was described in two series.[55,56]

Several malignant nerve tumors have also been described in the parapharyngeal space. Malignant neuroblastoma or sympathicoblastoma was reported by Brandenburg.[55] Malignant schwannomas have also been described in several papers.[16,40,57]

Miscellaneous Tumors

Nine cases of temporal bone meningiomas extending into the parapharyngeal space were reported by Nager, *et al.*[58] These tumors may represent extracranial extension of a primary intracranial tumor, a neoplasm arising in the jugular foramen, or a tumor originating from arachnoid cell clusters within the trunk of a cranial nerve or from its perineural sheath. Metastasis also can occur from a primary intracranial meningioma.[58] Additional cases of meningiomas of the parapharyngeal space were reported by Rose, *et al.*[59] and Lau, *et al.*[60] An additional case report[61] of an extracranial meningioma that filled the parapharyngeal space was thought to be an ectopic meningioma.

Numerous vascular lesions were reported as primary neoplasms of the parapharyngeal space. These include hemangiomas and lymphangiomas.[6,25,40,62,63] Arteriovenous malformation of the parapharyngeal space was described in one report.[64] Internal carotid aneurysms were also described, with the saccular type presenting as a mass lesion.[25,52] Hemangiopericytoma and hemangioendothelioma were also found.[29] Isolated myeloma was described in the parapharyngeal space.[62]

Several additional lesions were described in multiple reports: lipomas,[15,24,48] hibernoma,[13,40] extracardiac rhabdomyomas,[40,65] branchial cleft cysts,[15,30,65] teratomas,[48,66] and dermoid tumors.[6,48] Isolated lesions reported in single reports include amyloid tumor,[6] Kimura's disease,[60] fibromatosis,[60] leiomyoma,[13] giant lymphoid hyperplasia,[67] inflammatory pseudotumor,[68] granular cell myoblastoma,[69] amyloblastoma,[70] extracranial chondroma,[71] and carcinoid.[72]

Miscellaneous malignant tumors also are found in the parapharyngeal space. The most commonly reported lesion is isolated lymphoma. The largest series reported in the literature was from the Mayo Clinic in 1963.[29] This series excluded lymphomas involving Waldeyer's ring and excluded any lymphomas in the parapharyngeal space that also had involvement of the neck or systemic involvement of the reticuloendothelial system. The Mayo Clinic series reported on 25 patients ranging in age from 3 to 82 years. The patients often complained of a sore throat at the time the mass was first discovered. Physical examination typically revealed an irregular firm mass that had rapidly increased in size.[29] Eight additional cases of lymphoma of the parapharyngeal space were reported in another study,[15] and isolated case reports of lymphoma confined to the parapharyngeal space were also described by Ogasawara,[73] Shoss,[72] and Bass.[30]

TABLE II.
Primary Parapharyngeal Space Tumors.

	Benign	Malignant
Salivary gland	Benign lymphoepithelial disease Monomorphic adenoma Oncocytoma Pleomorphic adenoma Warthin's	Acinic cell carcinoma Adenocarcinoma Adenoid cystic carcinoma Carcinoma expleomorphic adenoma Malignant Warthin's Mucoepidermoid carcinoma Squamous cell carcinoma Undifferentiated carcinoma
Neurogenic	Ganglioneuroma Neurilemoma Neurofibroma Paraganglioma	Malignant paraganglioma Neurofibrosarcoma Schwannosarcoma Sympathicoblastoma
Miscellaneous	Ameloblastoma Amyloid tumor Arteriovenous malformation Branchial cleft cyst Carotid aneurysm Chondroma Choroid plexus tumor Dermoid Desmoid Giant lymphoid hyperplasia Granular cell myoblastoma Hemangioendothelioma Hemangioma Hibernoma Inflammatory pseudotumor Kimura's disease Leiomyoma Lipoma Lymphangioma Meningioma Rhabdomyoma Teratoma Venous angioma	Chondrosarcoma Chordoma Ectomesenchymoma Fibrosarcoma Fibrous histiocytoma Hemangiopericytoma Liposarcoma Lymphoma Malignant meningioma Malignant teratoma Plasmacytoma Rhabdomyosarcoma Synovial cell sarcoma

Ectomesenchymoma is a rare malignant parapharyngeal tumor that consists of mixed malignant fibrous histiocytoma and a primitive neuroectodermal tumor. The neural crest may be the site of origin, and recurrence and metastasis are common. Two series[74,75] reported a total of three cases of this unusual neoplasm. Chordoma remains an unusual neoplasm when isolated to the parapharyngeal space. It can occur even without evidence of cervical bone involvement.[76,77] Malignant fibrous histiocytoma confined to the parapharyngeal space was also described in two reports.[64,78]

Additional descriptions of unusual malignant tumors originating in the parapharyngeal space include malignant teratoma,[79] chondrosarcoma,[70] rhabdomyosarcoma,[80] malignant hemangiopericytoma,[81] meningiosarcoma,[40] fibrosarcoma,[6,22] liposarcoma,[22] malignant meningioma,[22] and an unusual case of undifferentiated carcinoma involving the parapharyngeal space with a plasmacytoma.[27] A rare malignant carotid body tumor was also shown to involve the parapharyngeal space.[22] Reported parapharyngeal space tumors are listed in Table II.

Squamous cell carcinoma can metastasize to lymph nodes in the parapharyngeal space. This is not uncommon with nasopharyngeal tumors, oral cavity tumors, oropharyngeal tumors, and hypopharyngeal tumors. However, it is rare to have parapharyngeal metastasis present as the initial manifestation of a tumor. In contrast, several reports[7,60,82] in the literature described a parapharyngeal mass that was found to be metastatic thyroid carcinoma as the first manifestation of a thyroid tumor. This included papillary thyroid cancer, medullary carcinoma of the thyroid, and metastatic follicular carcinoma of the thyroid.

EVALUATION OF PARAPHARYNGEAL SPACE TUMORS

Radiographic study of all parapharyngeal space tumors is essential. A computed tomography (CT) scan with and without contrast medium or a magnetic resonance imaging (MRI) study with gadolinium should be performed in all cases. Angiographic procedures may be necessary and fine-needle aspiration can also be done in select cases. The results of these studies provide the necessary information for treatment planning.[83]

Computed tomography imaging is capable of displaying the soft tissues of the parapharyngeal space extremely well.[8] CT scans may be helpful in separating prestyloid deep-lobe parotid tumors from extraparotid salivary neoplasms. The best way to distinguish between these two lesions is the finding of a fat plane between the deep lobe of the parotid gland and the posterolateral aspect of a mass. The fat represents compressed fibrofatty supporting tissue in the parapharyngeal space and, when seen, the tumor is extraparotid.[15] Unfortunately, for lesions larger than 4 cm, the fat plane is obliterated and it is difficult to determine whether a tumor originates in a minor salivary gland or in the parotid gland.

Benign prestyloid salivary tumors can cause erosion of the pterygoid plate, and this finding is not pathognomonic for malignant lesions. Radiographically, low-grade malignancies are difficult to distinguish from benign parapharyngeal space tumors.

In general, prestyloid tumors are usually salivary gland neoplasms that displace the carotid sheath contents posteriorly. It is also important to look for evidence of skull base or cervical vertebral erosion and extension through the jugular foramen into the cranial cavity.

Retrostyloid tumors generally displace the internal carotid artery in an anteromedial direction. This displacement occurred in two thirds of the parapharyngeal neurilemomas evaluated by CT.[15] However, schwannomas can displace the carotid artery in many different directions because of the unpredictable tumor position between the great vessels and the site of origin of the tumor. Vessel displacement depends on the nerve of origin and whether the tumor arises near the base of the skull or in the inferior portion of the parapharyngeal space.[28]

Large pleomorphic adenomas have a less homogeneous appearance on CT and contain irregular areas of minimal enhancement which can give an appearance similar to that of many neuromas. Neurilemomas also often have areas of hemorrhage, cystic necrosis, and fatty deposition.[15] Lesions that show enhancement on CT with contrast medium include paragangliomas, hemangiomas, hemangiopericytomas, aneurysms, and neurilemomas (Fig. 6).

Indications of malignancy in the parapharyngeal space include irregular tumor margins, spread into surrounding tissues and fat planes on CT, and evidence of enlarged necrotic nodes in the cervical area or in the retropharyngeal space.[23] CT scan has been shown to be helpful in assessing the parapharyngeal space as an area of extension of nasopharyngeal carcinomas. The parapharynx is often involved directly by nasopharyngeal tumors, and CT aids in the assessment of this region in patients who have nasopharyngeal carcinoma.[84] Excellent examples of parapharyngeal pathologic features in CT axial studies were presented.[10]

Currently, MRI provides the most useful preoperative information about the extent of the tumor and its relationship to surrounding structures. The capability of imaging the coronal and sagittal planes directly with MR scan is a significant advantage over CT.[85] MRI also has the advantage of outlining the position of the internal carotid artery relative to the tumor. In the future, MR angiography may replace formal angiography for many lesions. As with CT studies, MR scans also help to distinguish between intraparotid and extraparotid neoplasms by noting the retention or absence of a fat plane separating the lesion from the parotid gland. In general, the distinction between intraparotid and extraparotid masses is said to be more clearly defined on MR than CT scans because of better definition of the soft tissue interface available with MR imaging.[86]

The most common parapharyngeal neoplasms can be differentiated from each other by their appearance on MR scans or by the direction in which the parapharyngeal fat or the internal carotid artery (or both) are displaced.[86] Although it is difficult to distinguish between neurilemomas and minor

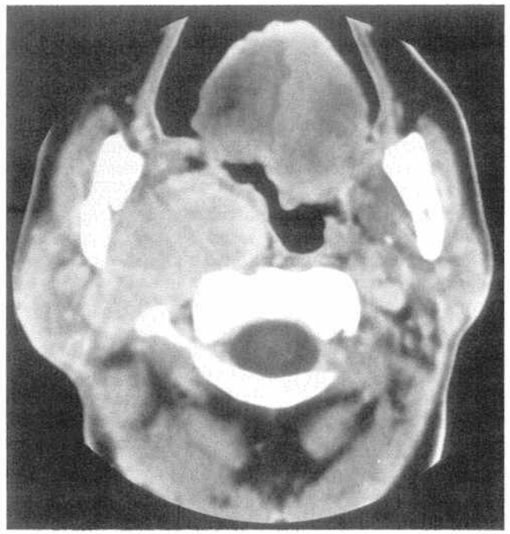

Fig. 6. Computed tomographic scan shows enhancing large retrostyloid vagal neurilemoma.

salivary gland tumors on the basis of their MR scans, neurilemomas generally displace vessels anteriorly. MRI can identify paragangliomas greater than 2 cm by the presence of signal flow void characteristics.[86] The presence of vascular flow voids distinguishes paragangliomas from the less vascular schwannomas. Metastatic renal cell cancers and thyroid carcinomas must be ruled out by history.[86] It still is extremely difficult to distinguish malignant parotid tumors from benign tumors unless there are marked irregular margins seen on MR scan (Fig. 7). There was little specific information provided by adding CT to MR study.[85]

If on the basis of history, examination, and imaging studies, a paraganglioma is suspected, patients should undergo testing for possible catecholamine secretion. Glomus jugulare tumors, carotid body tumors, and vagal paragangliomas all can secrete catecholamines. Historical points that may indicate the presence of catecholamine production incude labile hypertension, tremulousness, headache, pallor, palpitations, and sweating. Urine should be tested for the presence of vanillylmandelic acid, metanephrines, dopamine, epinephrine, and norepinephrine. Serum catecholamine levels can also be analyzed.[14,22] Failure to discover a secreting tumor preoperatively can have dire consequences during surgical removal of the lesion. If the secreting tumor is found, preoperative blockade with propranolol and phenoxybenzamine may help control intraoperative arrhythmias and hypertension.[26]

From CT or MR scans, it is difficult to determine if the carotid artery is invaded by tumor. En-

Fig. 7. Magnetic resonance scan shows irregular prestyloid extension from deep lobe of parotid gland. Tumor was adenoid cystic carcinoma.

casement of the carotid artery by a known malignancy generally indicates carotid-wall involvement. Ultrasonography was reported[87] to be more accurate than CT in determining invasion of a vessel wall.

Angiography should be done when carotid artery involvement is suspected. Bilateral carotid angiograms are also recommended whenever paragangliomas are found. Angiography helps to further delineate the tumor and its blood supply and to rule out any secondary lesions. Angiography is also performed for other highly vascular tumors and for tumors that invade the cervical vertebral bodies, or for vascular tumors that extend to the skull base or intracranially.

Prior to surgery, it may be necessary to perform a carotid occlusion study to determine if the patient can tolerate loss of the carotid artery. This must always be done whenever a malignant tumor involves the retrostyloid portion of the parapharyngeal space or for extensive vascular tumors that surround the carotid artery at the level of the skull base. A cerebral perfusion agent, technetium 99m-hexamethylpropylene amine oxime (HMPAO), is injected intravenously with a balloon inflated. The balloon is then deflated and a single photon-emission CT image of the brain is obtained. If there is evidence of hypoperfusion of the ipsilateral cerebral hemisphere, a repeat scan without test occlusion is performed within 24 to 48 hours. This helps determine if hypoperfusion was due to temporary balloon occlusion or to a preexisting abnormality. During the study, the patient also undergoes electroencephalographic monitoring and is observed for clinical evidence of neurologic dysfunction. A carotid occlusion study may best identify patients who are at greater risk of cerebral infarction developing after permanent occlusion.[88,89]

Tumor embolization is used for paragangliomas with intracranial extension. Embolization is performed 1 day prior to surgery, and then the tumor is removed in a single operation. Embolization is not used by the author for carotid body tumors or for vagal paragangliomas that do not extend through the jugular foramen.

Transoral biopsy prior to surgery is contraindicated for most parapharyngeal space tumors. This technique causes the pharyngeal mucosa to adhere to the tumor capsule, and subsequent removal is more difficult. In addition, there is a risk of hemorrhage and tumor rupture, with a greater risk of subsequent recurrent lesions. Fine-needle aspiration prior to surgery is an easy procedure that can be performed transorally without difficulty. It generally is accurate in confirming the presence of pleomorphic adenoma. A report by Allison, *et al.*[2] of 12 patients who underwent fine-needle aspiration of nonvascular tumors found no complications, but their results were diagnostic for pleomorphic adenomas only. In addition, when metastasis is suspected, fine-needle aspiration biopsies guided by ultrasound or CT scan can be helpful in cases of nonpalpable lesions.

SURGICAL APPROACHES

Many surgical procedures have been described for the treatment of parapharyngeal space lesions. This may be indicative of the problems in managing various tumors in an anatomic area that is difficult to access. The goal of parapharyngeal surgery should be to provide adequate tumor visualization that ensures complete tumor removal with preservation of surrounding nerves and vessels and control of any hemorrhage. The operation must be planned so it can extend to adjacent regions if necessary. Morbidity and mortality should be nearly zero.

Most authors recognize that the best surgical approach used for tumors of the parapharyngeal space is an external one.[30] An external operation can be augmented with mandibulotomy. Division of the mandible was first reported by Ariel, *et al.* (1954).[90] They discussed resection of the posterior aspect of the ascending ramus or actually dividing the ramus with a saw. Retraction of the mandibular ends allowed improved access to the parapharyngeal space. However, the authors reported that they did not have to utilize mandibulotomy in any of the parapharyngeal tumors, even with lesions up to 11 × 9 × 9 cm. They did not believe that the morbidity from mandibulotomy (*i.e.,* inferior alveolar nerve

anesthesia, scarring, or possible need for tracheotomy) justified its use for benign tumors. Mandibulotomy or mandibulectomy, however, may have a role in managing malignant tumors or for extensive benign tumors with invasion of adjacent structures. It is thought to be necessary in fewer than 10% of patients.[1,2]

Reported surgical approaches to the parapharyngeal space include the following: the cervical approach, the cervical-parotid approach, the transparotid approach, the transoral approach, the combined transoral-external approach, a combined cervical-transpharyngeal approach, and any of the above in combination with a mandibulotomy. Many surgical descriptions indicated ways to improve access to the parapharyngeal space: dislocation of the mandible,[25,88] division of the styloid process or stylohyoid ligament,[5,91] use of osteotomy,[30,92] or resection of part of the mandibular ramus or body.[90,93,94] Some of the surgical approaches described in the literature will be reviewed.

Cervical Approach: With or Without Mandibular Osteotomy

In 1955, Morfit[95] first described the submandibular incision and removal of the submandibular gland with blind tumor enucleation as a way to remove 12 parapharyngeal space neoplasms. Variations of this approach have been reported since then.

The cervical approach has been recommended by many authors[11,30,37,38,40,60,66,96,97] as the best access route for removal of parapharyngeal space tumors. The cervical approach uses a transverse incision at the level of the hyoid bone. The submandibular gland is often removed to gain access to the parapharynx. This approach does not include visualization of the facial nerve. Bass[30] described the removal of the submandibular gland with identification of the marginal mandibular branch of the facial nerve. An incision through the fascia deep to the submandibular space allowed for blunt dissection of the tumor. An extension of this approach to include division of the digastric, stylohyoid, and styloglossus muscles from the hyoid bone further improved access to the parapharyngeal space. Dissection using the fingers is then necessary.[48] Warrington, *et al.*[38] described the cervical approach by retracting the sternocleidomastoid muscle and carotid sheath backward while elevating the mandible forward. The styloid process and stylomandibular ligament were divided. Lau, *et al.*[60] also stated that, in the transcervical approach, the digastric and stylohyoid muscles were retracted or divided to allow access to the parapharyngeal space tumor. The submandibular gland was generally retracted, and the inferior pole of the parotid gland was retracted upward.

The cervical approach combined with various mandibular osteotomies and occasional partial segmental mandibulectomies has been used to improve exposure of the parapharyngeal space. Various osteotomies have been described: straight, step, or angled osteotomies in the mandibular body,[95,98,99] angle,[37,92,97] ramus,[90,100,101] or parasymphyseal/symphyseal[48] region. Morfit[95] divided the mandible in the premolar region through an extraction socket. Posterior mandibular osteotomies at the angle of the jaw were described as oblique,[37] step-wise,[98] or angular.[97] A case report by Attia, *et al.*[102] described a cervical approach with double osteotomies of the mandible; an intact inferior dental nerve was preserved and vessels were elevated with the ascending ramus and body of the mandible. This was done as a means of approaching the pterygomaxillary and parapharyngeal space. The mandible was elevated with the attached masseter muscle and overlying skin. This case report[102] described removal of a large angiofibroma. A step osteotomy of the mandibular angle was described[94] with the complete removal of the ramus and section of the condyles for access to remove a desmoid of the pterygopalatine fossa. The multitude of osteotomies described were designed to limit inferior alveolar nerve dysfunction, avoid entrance to the mouth, decrease morbidity with closure, and improve access to tumor. Repair of the mandible has generally included wiring or plating techniques. A report by Pinsolle, *et al.*[103] described five cases that involved a transcervical approach with a stair-step osteotomy at the angle for parapharyngeal parotid tumors. This defect was repaired with miniplates.

Heeneman and Maran[11] described a cervical approach that included identification of the facial nerve and exposure of the vessels of the carotid sheath early. They placed tapes around them to ensure hemostasis in an emergency. They also described removing the submandibular gland and tail of the parotid.

Cervical-Parotid Approach

A cervical approach was also described[1,83] that included an extension of the cervical incision in front of the ear in a standard parotidectomy fashion. The main trunk of the facial nerve is identified, with exposure of the lower division of the facial nerve. The posterior belly of the digastric muscle is then divided, exposing the internal and external carotid arteries, jugular vein, and adjacent nerves. The stylomandibular ligament, styloglossus muscle, and stylohyoid muscle are followed to the styloid process where they are divided close to the styloid process. The styloid is then removed with the bone-cutting forceps. According to Stell,[1] this approach was used successfully in 47 surgical procedures. Four individuals required either mandibulotomy or mandibulectomy. Cook and Ranger[99] described a mandibular osteotomy via a standard parotid incision extending into the submandibular region. They divided the mandible at the second premolar area and repaired it with plates. This did cause inferior dental nerve anesthesia. de Campora and colleagues[104] used an osteotomy in the parasymphyseal region through an incision from the mastoid process to the hyoid bone. A transcervical approach with a subsigmoid osteotomy (osteotomy at the notch dividing the condyle from the mandible) was described for removal of a calcified pleomorphic adenoma by Pedlar and Ravindranathan.[105]

Transparotid Approach

The transparotid approach is commonly used for deep-lobe parotid tumors. This is especially applicable for dumbbell-shaped lesions. Bass[30] described the transparotid approach. First, a superficial parotidectomy was done and then the facial nerve was isolated and dissected free from the deep lobe of the gland. Dissection then continued posteriorly and inferiorly around the mandible and, if mandibulotomy was necessary, he recommended performing this posterior to the entrance of the inferior alveolar nerve in the body of the mandible. A similar transparotid approach with initial superficial lobectomy and retraction of the facial nerve was followed, if necessary, by sectioning the styloid process and dislocation of the mandible anteriorly to allow for blunt tumor resection.[48] The transparotid incision alone was used by Allison, *et al.*[2] for the removal of five tumors. The transparotid approach was also used by Davidge-Pitts, *et al.*[70] and has been described in other publications.[106] Finally, the transparotid approach was further extended by elevating the soft tissues off the mandible and performing an L-shaped osteotomy, as described by Flood and Hislop.[100] These osteotomy cuts were designed to leave the inferior dental nerve undisturbed on the distal fragment. The proximal fragment of the mandible is rotated upward and the distal fragment is retracted anteriorly to gain access to tumors of the parapharyngeal space.[100] Use of the transparotid approach alone has generally been reserved for deep lobe parotid lesions.

Transoral Approach and Transoral-Transcervical (Transparotid) Approach

A series from the Mayo Clinic[29] of parapharyngeal space tumors removed prior to 1958 reported that one half the neurilemomas and one half the pleomorphic adenomas were excised by the transoral route. This was done in conjunction with ligation of the external carotid artery prior to excision. Since 1958, at the Mayo Clinic, all parapharyngeal space tumors have been removed via an external approach.[29] Removal of parapharyngeal space tumors through the intraoral approach also has been described by Ehrlich[107] and Thompson.[108] Allison, *et al.*[2] reported three tumors that were removed transorally. However, tumor spillage occurred in two of these cases.

The transoral approach may be useful for small pleomorphic adenomas arising in the palate or lateral pharyngeal wall. Problems with the transoral approach have included blind surgical removal, with increased risk of subtotal removal, tumor rupture, seeding of tumor cells, damage to vascular structures, hemorrhage, increased risk of infection, and increased risk to the facial nerve.[72]

The transoral approach has recently been reported as a route to remove benign, highly selected minor salivary gland tumors that originate in the parapharyngeal space. CT scan and fine-needle aspiration should be done prior to this surgery. Goodwin and Chandler[109] described their transoral method for avascular benign tumors that were not palpable in the neck or parotid region. They thought this approach was safe and effective. Tumor recurrence, however, was noted in one of their four patients who was followed for more than 5 years. They reserved this approach for pleomorphic or monomorphic adenomas that were palpable only in the oropharynx. They also stated that it may be necessary to transform the operation to an external incision with mandibulotomy and tracheotomy.

Allison, *et al.*[2] also described a combination transoral-cervical approach or transoral-transparotid approach that was used in 9 patients. Four of the 9 had tumor rupture. A combined transoral-transcervical approach was used in 3 patients by Lau, *et al.*[60] They abandoned the transoral approach because of tumor rupture and inadequate exposure. A combined transoral-external approach was used by Berdal and Hall.[37] In 5 cases, they reported that the tumor was removed by the transoral approach alone; however, this has not been done since 1957.

Cervical-Transpharyngeal Approach

The cervical-transpharyngeal approach has been used by several surgeons for the removal of large malignant or highly vascular tumors in the parapharyngeal space.[83] The cervical incision is combined with a lower lip incision to gain access to the pharynx via a mandibulotomy. A mandibulotomy is generally performed in the midline and is often referred to as the mandibular swing approach. This procedure was first described by Roux.[110] He divided the lower jaw and lip in the midline for access to floor-of-mouth and tongue tumors. Spiro, *et al.* (1981)[110] further defined this procedure and used it primarily for the removal of tongue and tonsillar cancers. Allison further described this approach for the removal of malignant lesions in the parapharyngeal space. Som, *et al.*[33] described it for the removal of vascular tumors at the cranial base level. de Campora, *et al.*[104] further described their experience with the cervical-transpharyngeal approach. They, however, used a parasymphyseal mandibular osteotomy in a zigzag manner. Intraorally, their ex-

TABLE III.
Surgical Approaches to the Parapharyngeal Space.

Transoral
Transoral-cervical
Transparotid
Cervical
Cervical-parotid
Cervical-pharyngeal, with or without mandibulotomy
Infratemporal fossa approach
Subtemporal preauricular infratemporal fossa

cision extended along the floor of the mouth and alveolar lingual sulcus onto the anterior tonsillar pillar. The tonsil and upper constrictor muscles were moved medially, giving access to parapharyngeal space lesions that extend to the skull base and to highly vascular tumors.

Similar procedures for removing parapharyngeal space tumors were described by other authors.[6,48] Krespi[111] also described the midline transmandibular approach for skull base lesions. He reported the removal of 6 parapharyngeal space tumors via this approach: 3 glomus vagales, 1 vascular schwannoma, 1 internal carotid aneurysm, and 1 choroid plexus tumor. This approach was chosen primarily for vascular parapharyngeal space tumors. The cervical-transpharyngeal approach via a midline mandibulotomy has also been used for lateral skull base surgery for the removal of cancers that involve the parapharyngeal space.[111]

The importance of the removal of retropharyngeal nodes for tonsil, oropharyngeal, and hypopharyngeal lesions was described.[111] Lymph nodes located between the superior constrictor and prevertebral muscles consisted of two main groups: the superior lateral nodal group (Rouviere) at the skull base close to the internal carotid foramen and jugular foramen and the inferior medial group located at the level of the oropharynx deep to the superior constrictor muscle. Several authors[111,112] thought that removal of these lymph nodes from the skull base should be done for all tumors of the tonsillar fossa with superior extension and for lateral and posterior pharyngeal wall tumors and for nasopharyngeal tumors. Using the mandibular swing approach, Krespi[111] and Omura, *et al.*[112] followed the internal carotid artery to the skull base and resected the structures medial to this vessel. Refinements in preventing complications with the mandibular swing osteotomy were reported by Cohen, *et al.*[113]

Infratemporal Fossa Approach

The lateral infratemporal fossa approach gives exposure of the skull base as well as the parapharyngeal space. This procedure was described by Ugo Fisch[114] for parapharyngeal space lesions that involve the temporal bone and was used in several series.[89,114]

Smith and Sharkey[115] reported their experience with removal of parapharyngeal space cancer via an infratemporal fossa approach. This was a modification of the technique used by Ugo Fisch. It combined a posterior auricular parotid flap with a parasymphyseal osteotomy for removal of the involved mandible or maxilla (or both). Another variation of the infratemporal fossa approach was described by Sekhar, *et al.*[116] who used a subtemporal preauricular incision for the treatment of seven patients with lesions of the parapharyngeal space. Unfortunately, no specific indication or description of the tumor types for these seven cases was given. The operation used a preauricular incision with neck and coronal extension and a temporal craniotomy to gain control of the petrous internal carotid artery. They stated that this approach was generally used for tumors that involve the lateral and posterior cranial base; sphenoidal clival area; and medial part of the petrous temporal bone, infratemporal fossa, nasopharynx, and retropharyngeal and parapharyngeal area. This did allow for direct access to the ipsilateral petrous and upper cervical internal carotid artery. There was minimal brain retraction and facial nerve function was maintained.[116] Surgical approaches described in the literature are shown in Table III.

COMPLICATIONS

Complications of surgery in the parapharyngeal space result from lack of familiarity with the anatomy, improper preoperative assessment, poor selection of patients, or the use of an inappropriate surgical approach. Few reports of parapharyngeal surgical cases in the literature describe complications. One series[22] of 54 parapharyngeal space tumors found a 48% complication rate, but permanent deficits occurred in only 11% of patients. Paraganglioma removal was associated with the greatest frequency of complications. Another report[1] of 52 cases described 1 temporary facial palsy, a permanent Horner's syndrome, loss of vagus nerve function, major blood vessel injury requiring repair, and 1 case of severe postoperative hemorrhage. In a report by Lau, *et al.*,[60] 5 of 15 patients had complications after parapharyngeal space surgery, including Frey's syndrome in 2, Horner's syndrome in 1, and loss of the vagus nerve with resulting hoarseness, dysphagia, and aspiration necessitating a gastrostomy tube in 2 patients. In general, complications after parapharyngeal space surgery are greatest with the removal of paragangliomas, malignancies, and neurogenic tumors.

Surgery for malignant lesions that extend into the parapharyngeal space also has caused significant morbidity. A report by Smith and Sharkey[115] described results after surgical removal of malignant lesions that involved the parapharyngeal space. Facial nerve dysfunction occurred in all survivors, and 4 of the 6 surviving patients required permanent gastrostomy due to dysphagia and inability to maintain dental obturators. The long-term efficacy of surgical tumor ablation and relief of symptoms from cancer that involves the parapharyngeal space is still unknown. Fortunately, most parapharyngeal space tumors are benign lesions and expected morbidity and mortality should be nearly zero. Unfortunately, other complications can occur. A list of complications associated with tumors of the parapharyngeal space is shown in Table IV.

Recurrence

Tumor recurrence is most likely initiated during the removal of parapharyngeal pleomorphic adenomas. Rupture of the tumor capsule with tumor spillage can result in recurrent neoplasms that may not be noted until 10 to 20 years after the initial surgery. The surgery for recurrent parapharyngeal pleomorphic adenoma has an increased risk to the facial nerve and should always be done by an experienced surgeon aided by facial nerve monitoring. In addition, highly vascular tumors or tumors that extend into the jugular foramen area may recur due to the surgeon's inability to totally remove the lesion secondary to problems with bleeding or exposure.

Nerve Injury

During the cervical-parotid or transparotid approach, the facial nerve is isolated and a mild paresis may occur. Permanent facial nerve injury should be extremely rare and steps should be taken to isolate and preserve the nerve and its branch at greatest risk for injury. When the facial nerve must be transposed because of intracranial tumor extension, meticulous technique and preservation of a large cuff of normal tissue around the nerve in the region of the stylomastoid foramen help minimize postoperative facial weakness. Permanent injury to the seventh nerve during parapharyngeal space surgery is extremely rare, with the exception of malignant neoplasms that involve the parotid gland.

Additional cranial nerves at risk for injury during parapharyngeal space surgery are nerves X, XI, and XII, and the cervical sympathetic plexus. Often during the course of removal of a neurogenic tumor, disruption in nerve continuity is unavoidable. If possible, nerve grafting can be performed. If the vagus nerve is injured during tumor resection, microscopic reanastomosis or nerve grafting should be done if possible. If the 12th cranial nerve alone is divided, postoperative problems generally are minimal.

However, if cranial nerves X and XII are both disrupted and had normal preoperative function, significant problems with swallowing and speech can occur. Transection of the vagus nerve above the inferior (nodose ganglion) is especially debilitating because of the additional loss of the pharyngeal branches and the superior laryngeal nerve. When normal functioning vagus and hypoglossal nerves are divided during tumor removal, most patients, especially elderly ones, have significant problems with swallowing. They should be treated by placing a nasogastric tube and a cuffed tracheotomy tube and by performing a cricopharyngeal myotomy and vocal cord augmentation with Teflon® at the time of operation. The tracheotomy tube should be changed slowly to a metal tube and then corked and removed only when the patient can handle secretions without aspiration. Oral feedings should not begin until the patient is able to swallow saliva without difficulty. Swallowing techniques similar to those used for supraglottic laryngectomy patients may be helpful.

TABLE IV.
Complications of Surgery of the Parapharyngeal Space.

Hematoma
Seroma
Airway obstruction
Infection
Tumor recurrence
First bite pain
Frey's syndrome
Leakage of cerebrospinal fluid
Meningitis
Nerve injury
Greater auricular
Facial
Glossopharyngeal
Vagus
Hypoglossal
Spinal accessory
Cervical sympathetic
Vessel injury
Stroke
Hemorrhage
Death
Osteotomy site complications

Fortunately, compensation for unilateral loss of vagal and hypoglossal nerve function occurs with time. When the vagus nerve alone is divided, it is best to assess the patient's degree of morbidity before proceeding with any additional treatment. Young individuals may have voice change but no difficulty swallowing. It is best to wait several months to determine the final cord position. This knowledge helps optimize the results of subsequent vocal cord augmentation with Teflon or thyroplasty. If aspiration or cough occurs, immediate vocal cord augmentation with Teflon or cord medialization is indicated.

If the spinal accessory nerve is injured, reanastomosis can be done, and an immediate physical therapy program should be initiated in the postoperative period. The sympathetic plexus can be injured during removal of a neurogenic tumor of the

nerve or ganglion. Likewise, the great auricular nerve is generally divided during the cervical-parotid or transparotid approach, and patients should be told preoperatively about the numbness that will occur.

Vessel Injury

Carotid artery injury is the greatest potential risk of operation in the parapharyngeal space. Uncontrolled bleeding can cause death from hemorrhage or stroke. Parapharyngeal space tumors can directly invade the jugular vein, internal or external carotid arteries, and vertebral artery. Vessel injury is most likely to occur during the removal of paragangliomas, recurrent tumors, and malignant lesions or after removal of tumor when the patient had received radiation therapy. Preoperative assessment of the tumor, the use of angiography when appropriate, embolization for vascular lesions that extend intracranially, and placement of permanent balloons to occlude carotid blood flow have all been found to decrease the incidence of complications from vessel injury. Surgical procedures that put the patient at high risk for vessel injury should also be done by a team of physicians capable of handling all possible sequelae.

Osteotomy Complications

Osteotomy complications include loss of lower teeth, malunion, and nonunion. If there is any concern about adjacent periapical dental disease, a preoperative Panorex radiograph should be taken.[113] Direct wiring of the osteotomy site can be done in combination with the insertion of a preformed lingual splint for the edentulous patient. Reconstruction plates can also be used; however, before mandibular fixation is performed, a drill should be used to remove uneven bone from the osteotomy site so that reapproximation can be tight and secure.[113] Intermaxillary fixation is not necessary. Patients who undergo a midline mandibulotomy are generally fed by a nasogastric tube for 2 weeks.

Other Complications

Large parapharyngeal space tumors can impinge on the airway by displacing the pharyngeal wall medially or by obstructing the choana. Tracheotomy may be necessary and should always be done with a midline mandibulotomy. Postoperative hematoma can also compromise the airway because of bleeding into the neck. Meticulous hemostasis and suction drainage are required in all cases. Portable suction equipment should be used to maintain constant negative pressure, even when the patient is ambulatory.

Leakage of cerebrospinal fluid and meningitis are possible complications after removal of parapharyngeal space tumors that extend into the jugular foramen, extend intracranially, or cause destruction of cervical vertebrae. Leakage of cerebrospinal fluid is most likely after removal of tumors that extend intracranially. In a report[117] of neuromas removed from the jugular foramen area, there was a 45% incidence of postoperative leakage of cerebrospinal fluid. After tumor excision, the dura should be closed so that it is watertight. If a dural defect remains, a dural graft that uses the temporalis fascia is sutured into place. If the dural closure is tenuous, muscle or fat is also packed into the area. A local muscle flap or free flap is used to reconstruct the defect and augment the skull base closure.[26] Spinal drainage is used for approximately 1 week if there is concern about the repair. It is not used routinely in the author's practice.

Because of the large dead space that occurs after removal of a parapharyngeal space tumor, a seroma can form, and infection is possible. The patient is maintained on a broad-spectrum cephalosporin for several days postoperatively. When the oral cavity is entered, metronidazole is also used for prophylaxis.[26]

If the superficial portion of the parotid gland has to be removed, complications reported for parotidectomy would also apply for parapharyngeal surgery.

When Not to Operate

It is unlikely that patients will die of an untreated benign parapharyngeal space tumor. One exception may be paragangliomas. The treatment goals should be to remove the tumor and to cause minimal morbidity. The aim is to avoid subsequent neurogenic damage, dysphagia, airway obstruction, hearing loss, or cosmetic problems that occur from tumor enlargement. In all cases, the risks of the operation must be weighed against the risks of inaction. This is especially true for malignant tumors, neurogenic tumors, and vascular tumors in the elderly.

Neurogenic tumors can be present for years without causing nerve deficits. Functionally, gradual paralysis is less debilitating than sudden loss caused by a nerve transection. In young patients, neurogenic tumors are removed to prevent future loss of function of adjacent nerves and to try to preserve the nerve of origin. However, isolated asymptomatic parapharyngeal neuromas in elderly patients with no nerve deficits should probably be watched. Parapharyngeal vagal paragangliomas can cause progressive nerve paralysis and extend intracranially with significant morbidity. Again, elderly patients should not undergo surgical removal of these lesions if they have been present for many years and have not caused cranial nerve deficits, because the risks of operation outweigh inaction. Most prestyloid salivary tumors can be removed with little morbidity and, therefore, there is little restriction on the age of patients who undergo operation, because of the risks of tumor enlargement and possible malignant degeneration of pleomorphic adenomas.

PERSONAL SERIES

During the past 10 years, the author surgically removed 44 tumors that involved the parapharyngeal space. Tumors that extended into the parapharyngeal space from adjacent structures were excluded. Parapharyngeal space tumors treated with biopsy alone, observation, or radiation therapy are not included. Parapharyngeal space tumors removed by colleagues at the author's institution also are excluded. Of these 44 patients, 40 underwent surgery in the past 5 years. The lesions removed include 40 primary parapharyngeal space tumors and 4 cases of isolated metastasis to parapharyngeal nodes.

The mean age of these 44 patients was 46 years, with a range from 17 to 80 years. The male-to-female ratio was 17:27. Twenty-four of the patients presented with an asymptomatic mass discovered on physical examination or noted by the patient.

Six patients described pain present in the deep parotid area or diffusely in the mandibular region. All six patients proved to have parapharyngeal cancers. Three had adenoid cystic cancers, 1 squamous cell carcinoma, 1 neurofibrosarcoma, and 1 chordoma.

Mild aching in the throat and discomfort in the ear were noted in four patients. These individuals all had benign lesions: 1 neurilemoma, 1 vagal paraganglioma, and 2 pleomorphic adenomas. One individual described paresthesias of the third division of the fifth cranial nerve. This individual had an adenoid cystic carcinoma. She also complained of pain. Trismus, generally of a mild nature, was described by 4 people. Two had benign lesions (hemangioma and cystic fibroma) and 2 had carcinomas (squamous cell cancer and adenoid cystic carcinoma).

Dysphonia and chronic cough were complaints of the 6 individuals who preoperatively had 10th nerve paralysis. Decreased hearing was noted in 2 individuals secondary to eustachian tube dysfunction from a pleomorphic adenoma in 1 and a vagal paraganglioma in the other.

One patient with a carotid body tumor had a positive family history, and one of the individuals with a vagal paraganglioma proved to have a catecholamine-secreting tumor. This patient described preoperative lightheadedness, headaches, sweats, nausea, and flushing episodes. Another individual with symptoms that suggested catecholamine-secreting tumor noted intermittent dizziness characterized by spells of near loss of consciousness. She had a pleomorphic adenoma that may have been impinging on the ninth cranial nerve as the cause for her symptoms.

The duration of symptoms in these patients varied but, generally, symptoms were present for several months up to 5 to 6 years. Prior to the diagnosis of a parapharyngeal space lesion, four individuals were thought to have peritonsillar abscess, cellulitis, or tonsillar inflammation; two individuals underwent tonsillectomy elsewhere prior to diagnosis of their tumor. Two patients were being treated for temporomandibular joint disturbance prior to the diagnosis of their parapharyngeal-space lesion.

Radiographic studies were performed in all cases and included 32 CT scans, 16 MRI scans, 18 angiograms, and 4 balloon occlusion studies. Preoperative embolization was done in one individual with a vagal paraganglioma that extended intracranially. A permanent balloon occlusion of the carotid artery was placed in one individual with an osteogenic sarcoma of the retrostyloid space. Fine-needle aspiration was performed in 9 individuals. Seven patients had a diagnosis of pleomorphic adenoma made accurately prior to surgery, 1 individual with a pleomorphic adenoma had a negative aspirate, and another individual with a chordoma had a hypocellular, nondiagnostic result.

Pathologic Findings

Of the 44 patients who underwent surgery, 12 were treated for recurrent tumors of the parapharyngeal space. All these individuals had surgery at another institution.

Overall, there were 32 benign lesions and 12 malignant tumors. Of the malignant tumors, 4 represented isolated metastasis to the parapharyngeal space without direct extension. If these cases are excluded from the primary tumors, the benign-to-malignant ratio for the remaining 40 individuals would be 4:1. The overall tumor distribution for the entire group was 16 of salivary gland origin, 18 neurogenic, and 10 miscellaneous tumors.

Pleomorphic adenoma was the only benign salivary gland tumor encountered in this series. Seven tumors were thought to originate from the retromandibular portion of the deep lobe of the parotid gland and 4 from minor salivary glands. Four malignant tumors originated from the retromandibular portion of the deep lobe of the parotid gland; 3 were adenoid cystic cancers and 1 was a squamous cell carcinoma. A single case of an adenoid cystic cancer arising from a minor salivary gland was also treated.

Eighteen neurogenic tumors were removed, and these included 3 neurilemomas of the vagus nerve. Two of these patients had preoperative paralysis of the vagus. One additional individual had a tumor that was either a neurilemoma or a neurofibroma that originated from a probable branch of the ninth

cranial nerve. There were 7 vagal paragangliomas removed; 6 were benign and 1 was malignant. The patient with the malignant vagal paraganglioma had undergone a prior attempt at surgical removal of the tumor and radiation therapy elsewhere. Six carotid body tumors were included that clearly extended into the parapharyngeal space. Although these tumors may not be considered to be primary parapharyngeal space in origin, most series do include carotid body tumors. The author included only those carotid body lesions that definitely extended into the parapharynx and were found clinically to displace the tonsil and soft palate, were retromandibular on imaging studies or, at the time of surgery, clearly extended above the level of the posterior belly of the digastric muscle. One of these six individuals had two prior attempts at surgical removal of the lesion at another institution that were abandoned because of excessive bleeding. Two of the patients with carotid body tumors had multiple paragangliomas noted on angiographic studies. The final neurogenic tumor was a neurofibrosarcoma of the vagus nerve.

Five benign miscellaneous lesions were removed from the parapharyngeal space. These included a recurrent venous angioma, a hemangioma, a lymphangioma, a carotid artery aneurysm, and an unusual case of reactive lymphoid tissue in the retrostyloid area causing 10th and 12th nerve paralysis. Histopathologic findings from specimens of tumors removed showed no evidence of lymphoma on pathologic review, and symptoms gradually resolved over time.

The malignant miscellaneous lesions encountered in the parapharyngeal space consisted of 1 primary chordoma and 4 cases of isolated metastases to parapharyngeal nodes. Two were from squamous cell carcinomas of the tonsil that had noncontiguous tumor, and 1 individual had nasopharyngeal carcinoma involving the superior parapharynx after radiation therapy. There was also no evidence of direct tumor extension in this patient. The last patient had metastatic osteogenic sarcoma to the parapharyngeal space.

Treatment

The diagnosis and surgical procedures used for each patient are shown in Table V. Thirty-five patients underwent a cervical-parotid approach and nine underwent the cervical-parotid approach with midline mandibulotomy. One patient with a vagal paraganglioma extending intracranially had preoperative tumor embolization. This individual and one other patient with a neurilemoma extending intracranially also had a suboccipital craniotomy performed at the time of the cervical-parotid approach to remove the intracranial tumor extension at the same surgery. One patient with recurrent osteogenic sarcoma confined to the retrostyloid portion of the parapharyngeal space preoperatively underwent balloon occlusion of the carotid artery.

The cervical-parotid approach with midline mandibulotomy was used for malignant lesions or for extensive benign tumors. The pathologic findings for the malignant lesions included the three cases of isolated metastases. These all occurred in the superior parapharyngeal area. Three individuals undergoing this operation had extensive vagal paragangliomas that circumferentially involved the carotid artery at the level of the skull base. One was a malignant tumor that had previously undergone two attempts at removal and radiation therapy. A chordoma was excised by this approach also in an individual whose tumor invaded the cervical vertebral bodies and involved the vertebral artery. The only pleomorphic adenoma removed via mandibular osteotomy was a recurrent tumor that had undergone transoral removal elsewhere several years earlier. The tumor recurrence was quite extensive at the level of the nasopharynx and skull base, again surrounding the internal carotid artery on scan.

Follow-up

Of the patients with primary tumors, two had recurrences. One patient with an adenoid cystic carcinoma of the superior parapharyngeal space has local and distant metastasis. She currently is alive with persistent disease and is asymptomatic. A second individual with extensive adenoid cystic carcinoma of the parapharyngeal space underwent a full course of radiation therapy after his surgery. He had no evidence of local or regional disease but has extensive pulmonary metastases. He currently is alive and doing well with stable pulmonary lesions. The remaining individuals with malignant tumors have, at this time, no evidence of recurrent disease; however, the follow-up is less than 2 years in the majority of these cases. There was no known recurrence of any of the benign tumors, and no patient was lost to follow-up.

Complications

The most frequent complication in these patients was transient facial nerve paresis, which occurred in 10 individuals. All 10 recovered completely and the paresis primarily involved the marginal mandibular branch of the facial nerve. Seven patients had permanent loss of the vagus nerve, which occurred in cases of 10th nerve neurilemoma or paraganglioma tumor surgery. Of the 7 individuals who lost their vagus nerve, 2 were treated with vocal cord augmentation with Teflon, and 2 patients underwent thyroplasty. Three individuals required no further treatment. Three patients, in addition to losing the 10th cranial nerve, also lost the 12th cranial nerve at the time of surgery, which was for extensive paragangliomas. These patients all had prolonged difficulty with

TABLE V.
Author's Parapharyngeal Surgical Procedures.

Patient	Age (yr)	Sex	Diagnosis	Surgical Approach
1	32	M	Vagal neurilemoma	Cervical-parotid
2	29	F	Vagal paraganglioma	Cervical-parotid with suboccipital craniotomy
3	70	F	Vagal paraganglioma	Cervical-parotid
4	25	M	Recurrent venous angioma	Cervical-parotid
5	36	F	Vagal neurilemoma	Cervical-parotid
6	66	F	Carotid body tumor	Cervical-parotid
7	50	M	Pleomorphic adenoma (minor salivary gland)	Cervical-parotid
8	55	M	Recurrent pleomorphic adenoma (parotid gland)	Cervical-parotid
9	41	F	Carotid body tumor	Cervical-parotid
10	25	F	Carotid body tumor	Cervical-parotid
11	24	F	Hemangioma	Cervical-parotid
12	57	F	Pleomorphic adenoma (minor salivary gland)	Cervical-parotid
13	45	F	Pleomorphic adenoma (parotid gland)	Cervical-parotid
14	60	F	Carotid body tumor	Cervical-parotid
15	80	F	Recurrent pleomorphic adenoma (parotid gland)	Cervical-parotid
16	45	M	Pleomorphic adenoma (parotid gland)	Cervical-parotid
17	18	M	Recurrent lymphangioma	Cervical-parotid
18	49	F	Reactive lymphoid tissue	Cervical-parotid
19	49	M	Recurrent carotid body tumor	Cervical-parotid
20	47	M	Squamous cell carcinoma (parotid gland)	Cervical-parotid
21	62	M	Adenoid cystic carcinoma (parotid gland)	Cervical-parotid
22	39	F	Recurrent adenoid cystic carcinoma (parotid gland)	Cervical-parotid
23	17	M	Carotid body tumor	Cervical-parotid
24	31	M	Recurrent metastatic osteogenic sarcoma	Cervical-parotid
25	42	M	Pleomorphic adenoma (parotid gland)	Cervical-parotid
26	63	F	Carotid aneurysm	Cervical-parotid
27	53	F	Vagal paraganglioma	Cervical-parotid
28	50	F	Vagal paraganglioma	Cervical-parotid
29	48	F	Vagal neurofibrosarcoma	Cervical-parotid
30	37	M	Recurrent vagal neurilemoma	Cervical-parotid
31	20	F	Neurilemoma/neurofibroma (unknown nerve of origin)	Cervical-parotid with suboccipital craniotomy
32	68	F	Pleomorphic adenoma (minor salivary gland)	Cervical-parotid
33	18	M	Adenoid cystic carcinoma (parotid gland)	Cervical-parotid
34	75	F	Pleomorphic adenoma (parotid gland)	Cervical-parotid
35	54	F	Pleomorphic adenoma (parotid gland)	Cervical-parotid
36	65	F	Chordoma	Cervical-parotid with midline mandibulotomy
37	65	F	Recurrent malignant vagal paraganglioma	Cervical-parotid with midline mandibulotomy
38	69	F	Recurrent adenoid cystic carcinoma (minor salivary gland)	Cervical-parotid with midline mandibulotomy
39	55	F	Metastatic squamous cell carcinoma from tonsil	Cervical-parotid with midline mandibulotomy
40	57	F	Recurrent pleomorphic adenoma (minor salivary gland)	Cervical-parotid with midline mandibulotomy
41	73	M	Recurrent metastatic nasopharyngeal carcinoma	Cervical-parotid with midline mandibulotomy
42	42	F	Vagal paraganglioma	Cervical-parotid with midline mandibulotomy
43	40	M	Metastatic squamous cell carcinoma/tonsil primary	Cervical-parotid with midline mandibulotomy
44	40	M	Vagal paraganglioma	Cervical-parotid with midline mandibulotomy

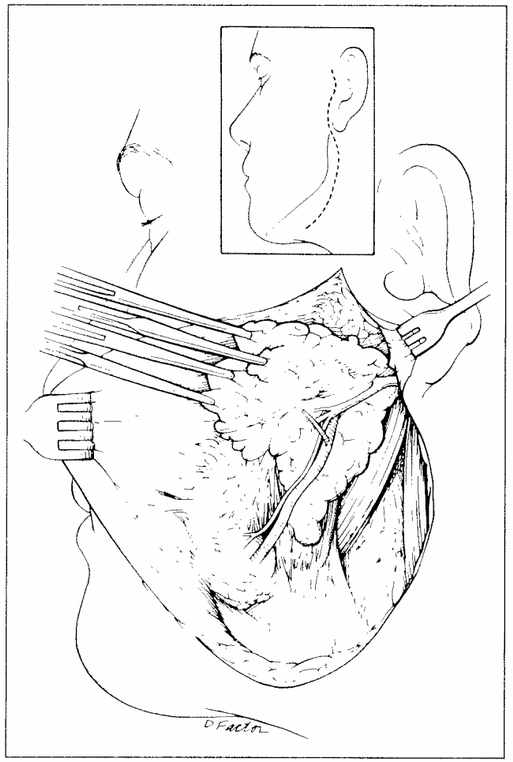

Fig. 8. Exposure of facial nerve and its inferior division: cervical-parotid approach. (By permission of Mayo Foundation.)

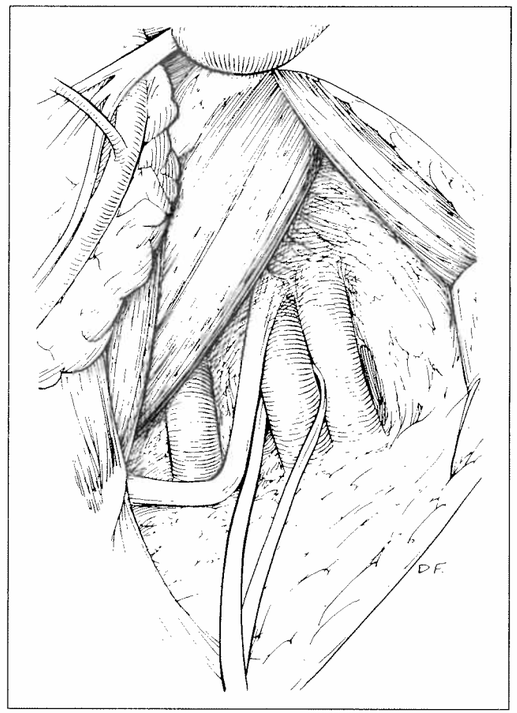

Fig. 9. Exposure of vessels and nerves in upper neck: cervical-parotid approach. (By permission of Mayo Foundation.)

swallowing and eventually were able to eat without supplementation, but one patient was not able to eat normally until 5 months after operation. Sympathetic nerve injury with Horner's syndrome was noted in two patients. One individual also had a temporary accessory nerve weakness, and one had a permanent loss of the third division of the fifth cranial nerve.

Other complications noted in this series included 3 cases of postoperative infection, 4 hematomas (2 requiring simple drainage procedures), and first-bite pain related to mobilization of the tail of the parotid gland.

AUTHOR'S RECOMMENDED SURGICAL APPROACHES

The author has used two surgical approaches during the past 10 years to manage successfully and safely a wide variety of benign and malignant parapharyngeal space tumors. These procedures have the additional flexibility to manage neoplasms that extend into adjacent structures or into the intracranial cavity. The indications and operative technique of the cervical-parotid approach and the cervical-parotid approach with midline mandibulotomy are reviewed in detail.

Cervical-Parotid Approach: Surgical Technique

The cervical-parotid approach used by the author is similar to the surgical technique described by Stell, *et al.*[1] An incision is made in front of the ear and extended beneath the mandible. A parotid skin flap and inferior neck flap are raised. The inferior and posterior areas of the parotid gland are separated from the sternocleidomastoid muscle, from the cartilaginous ear canal, and from the posterior belly of the digastric muscle. The main trunk of the facial nerve is identified (Fig. 8). If it is obvious that the parapharyngeal space tumor is a dumbbell variety of deep-lobe parotid tumor, a superficial parotidectomy is performed. In other cases, the inferior division of the facial nerve is isolated and followed out to the level of the submandibular gland.

The sternocleidomastoid muscle is retracted laterally and the upper jugular nodes are removed to allow identification of deeper structures. The spinal accessory nerve, internal jugular vein, internal and external carotid artery, and cranial nerves X, XI, and XII are isolated. Vessel loops are placed around the internal and external carotid arteries for security (Fig. 9).

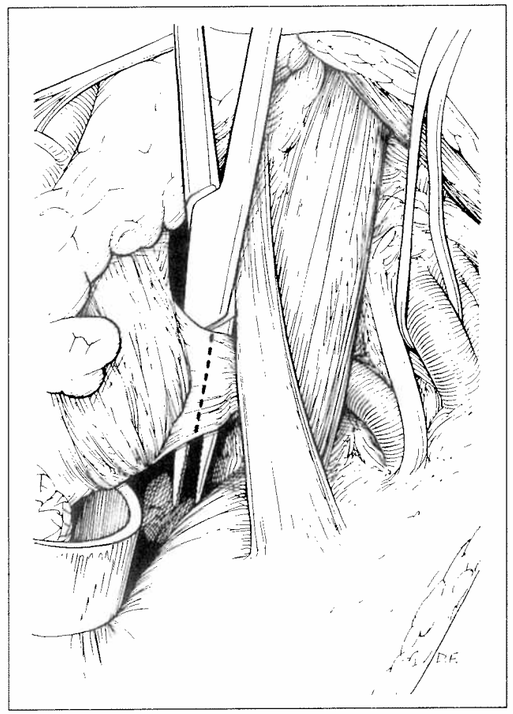

Fig. 10. Division of stylomandibular ligament: cervical-parotid approach. (By permission of Mayo Foundation.)

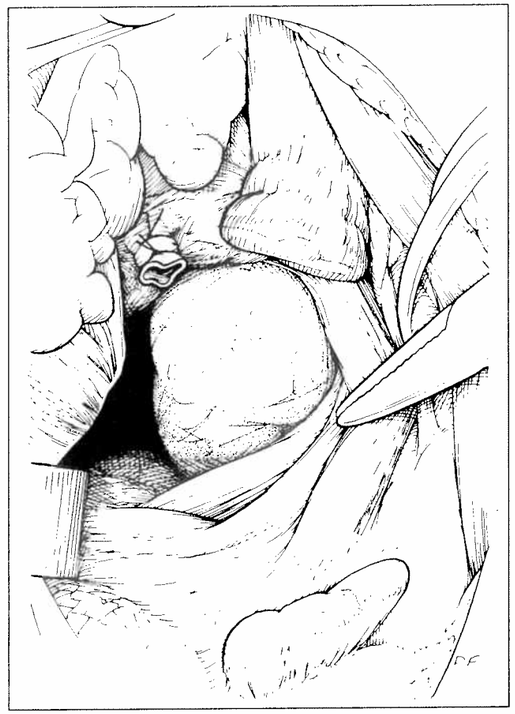

Fig. 11. Tumor exposure: cervical-parotid approach. (By permission of Mayo Foundation.)

The posterior belly of the digastric muscle is isolated completely down to its insertion on the hyoid bone. The dense stylomandibular fascia between the inferior parotid gland and the submandibular gland is divided so the submandibular gland can be easily retracted medially if necessary. The posterior belly of the digastric muscle and the stylohyoid muscle are separated from their attachments at the mastoid tip and from the styloid process, and are reflected medially. This gives further superior exposure of the internal carotid artery, jugular vein, and adjacent nerves. The external carotid artery is now easily seen passing into the parotid tissue in front of the styloglossus muscle. This artery and its corresponding vein are divided. The angle of the mandible is retracted anteriorly. This stretches the stylomandibular ligament so that it can easily be palpated and visualized (Fig. 10). This ligament is divided, creating a wide opening into the parapharyngeal space. The parapharyngeal space tumor is now easily visualized and can generally be removed under direct vision (Fig. 11). After tumor removal, a Hemovac drain is inserted and the incision is closed in layers (Fig. 12).

Indications

The cervical-parotid approach has been used to remove the majority of lesions encountered in the parapharyngeal space. All deep-lobe parotid neoplasms and extraparotid salivary tumors are excised via this approach. In addition, many retrostyloid tumors, including most neurogenic tumors and small paragangliomas, can be removed in the same manner. Low-grade malignant tumors of the deep lobe of the parotid gland that extend into the parapharyngeal area can also be excised by this operation. For tumors that are known prior to surgery to extend intracranially, the cervical-parotid approach is combined with a suboccipital craniotomy by a postauricular incision. The cervical-parotid approach was used in 80% of the author's cases.

Important Points

1. The cervical-parotid approach can be combined with a mandibulotomy or transoral incision to

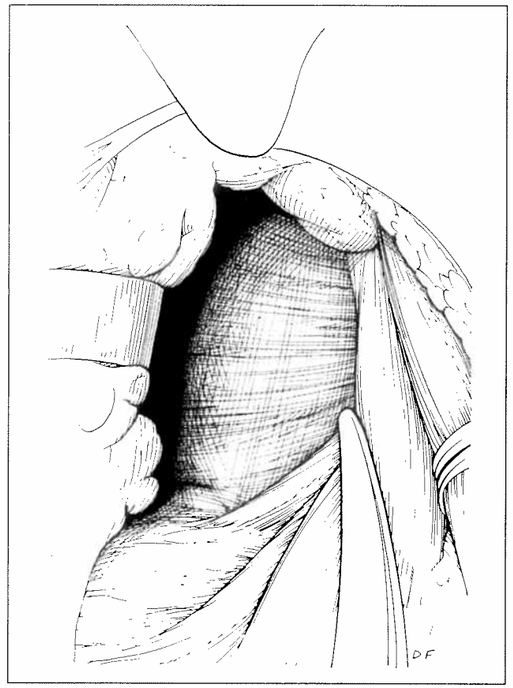

Fig. 12. Tumor removal and remaining defect: cervical-parotid approach. (By permission of Mayo Foundation.)

improve exposure of the parapharyngeal space. However, this was not necessary in any of the cases in the author's series.

2. For certain tumors of the retrostyloid space and for prestyloid tumors that extend around the styloid process, division of the styloid muscles and removal of the styloid process are often helpful.

3. The cervical-parotid approach can be extended easily via a postauricular incision to enable the surgeon to perform a suboccipital craniotomy. This should be planned whenever there is radiographic evidence of tumor that extends intracranially or if tumor cannot be removed completely from the jugular foramen region.

4. It is not necessary to perform a superficial or total parotidectomy for most cases of deep-lobe parotid parapharyngeal tumors.

5. Several authors stated that the preoperative differentiation of an extraparotid neoplasm from a deep-lobe parotid neoplasm is important, because extraparotid tumors can be approached through a cervical operation without the need to identify the facial nerve. The author does not agree with this. Extraparotid parapharyngeal space neoplasms often approach the main trunk of the facial nerve at the level of the stylomastoid foramen. By identifying the facial nerve in all cases, there is less chance of its injury.

6. For deep-lobe tumors that extend through the stylomandibular tunnel into the parapharyngeal space, an initial superficial parotidectomy can be performed.

7. In the author's experience, most deep-lobe parotid tumors are not the dumbbell type, but are more often round and originate from the retromandibular portion of the deep lobe of the parotid gland. There often is a narrow attachment of parotid tissue to the tumor that can be separated easily under direct vision without necessitating removal of the superficial portion of the gland. Even with greater extension into the deep lobe, by identification and retraction of the main trunk of the facial nerve, a wide cuff of deep-lobe parotid tissue can be removed with the tumor. This should always be done because the site of origin is most at risk for rupture of the tumor capsule and tumor spillage.

8. When the superficial and deep lobes of the parotid gland are removed with a large parapharyngeal space tumor, consideration should be given to using a gracilis free flap to repair the significant depression that occurs from loss of the entire parotid gland and surrounding structures.

9. The carotid artery can be followed easily to its foramen by reflecting the muscles off the styloid process and removing the styloid process.

10. It is not necessary to remove the submandibular gland to excise parapharyngeal space tumors.

11. Division of all of the stylomandibular ligament is essential to opening the parapharyngeal space.

12. For recurrent tumors of the parapharyngeal space, especially if a superficial parotidectomy was previously performed, these operations should be done with cranial nerve monitoring, especially facial nerve monitoring.

13. After the operation, it is essential to obtain complete hemostasis. A large Hemovac drain should be left in the defect for at least 2 to 3 days postoperatively to maintain the patient on constant suction drainage. This is done to decrease dead space and subsequent seroma formation and possible infection.

14. When paragangliomas extend intracranially, preoperative embolization should be performed the day prior to surgery.

15. The cervical-parotid approach provides excellent exposure of the superior extent of most carotid body tumors or internal carotid artery lesions.

16. The stylohyoid ligament is often a thick fibrous band or calcified band that should be divided.

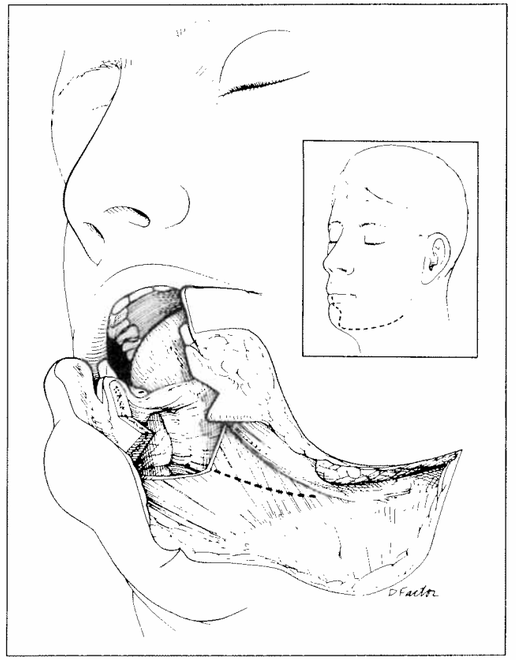

Fig. 13. Midline mandibular stair-step osteotomy. (By permission of Mayo Foundation.)

17. For individuals who have had previous transoral excisions, biopsies, or tonsillectomies, it is best to let the tonsillar fossa heal for several weeks prior to surgical therapy. A cuff of pharyngeal mucosa should be included if a transoral biopsy has been done. The transoral wound can be closed easily at surgery.

18. If there is not a loose areolar plane surrounding a prestyloid tumor, the lesion is probably malignant.

19. Rupture of a prestyloid pleomorphic adenoma is most likely to occur with traction of the tumor against the styloid process, pterygoid plates, or mastoid tip, or if a cuff of parotid tumor is not included with the connnection to the deep lobe of the parotid gland.

20. Removal of the mastoid tip may be done for pleomorphic adenomas that are tightly compressed into the stylomastoid foramen area.

21. During the removal of prestyloid tumor, care must be taken because a tortuous internal carotid artery often has contact with the tumor at a level superior to the styloid process.

22. For suspected malignant lesions, the initial node removed as part of the exposure should always be sent for pathologic study. If a malignant tumor is encountered, an appropriate select or modified neck dissection can be performed, if indicated.

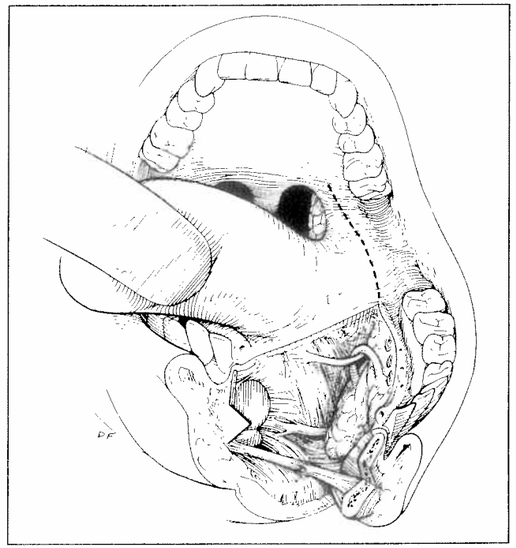

Fig. 14. Intraoral incision and identification and preservation of lingual and hypoglossal nerves. (By permission of Mayo Foundation.)

23. During closure, the styloid muscles and posterior belly of the digastric muscle can be reattached.

Cervical-Parotid Approach With Midline Mandibulotomy: Surgical Technique

All individuals undergoing a cervical-parotid approach with midline mandibulotomy have an initial tracheotomy. Then the steps of the cervical-parotid approach are performed. After this is completed, the incision is extended around the chin and used to split the lower lip in the midline. The anterior mandible is exposed. The digastric tendon is separated from the hyoid bone, and the mandible is divided in the midline in a stair-step manner, preserving the incisor teeth if good dentition is present (Fig. 13).

An incision is made intraorally in the floor of the mouth by using cautery and including the submandibular duct orifice with the mandible. The incision extends back onto the anterior tonsillar pillar and up to the level of the hard palate. Further extension onto the hard palate is occasionally necessary for nasopharyngeal lesions.

The hypoglossal nerve is identified and followed into the tongue base. The mylohyoid muscle is divided over the hypoglossal nerve. The tongue musculature is reflected medially and the lingual nerve is identified and preserved as it stretches across the

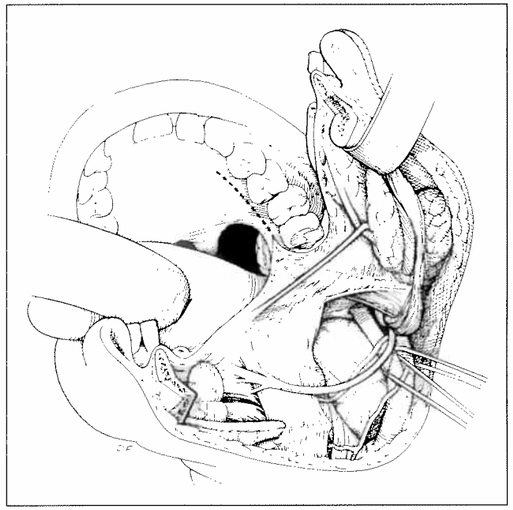

Fig. 15. Tumor exposure and isolation of carotid contents to the skull base. (By permission of Mayo Foundation.)

field (Fig. 14). The tongue is retracted medially and the parapharyngeal space is exposed back to the retropharyngeal space lateral to the constrictor muscles. The styloglossus and stylopharyngeus muscles are identified, tagged, and divided.

The parapharyngeal and retropharyngeal space is then opened widely (Fig. 15). Depending on the extent of the tumor and the histologic findings, the superior portion of the operation can be expanded as necessary. The mandible is retracted laterally and superiorly, widely opening the parapharyngeal space. Tumor removal can now be performed safely (Fig. 16). After excision of the tumor, the intraoral incision is closed in layers. A feeding tube is inserted. The mandibulotomy is wired or plated. Drains are placed in the neck, and the neck incision is closed in layers.

Indications

The cervical-parotid approach with midline mandibulotomy is used for all vascular tumors that extend into the superior portion of the parapharyngeal space or surround the carotid artery at its foramen. Also, this approach gives the best access for tumors that are confined solely to the superior aspects of the parapharyngeal space, *i.e.*, in the eustachian tube area and skull base. When malignant tumors have invaded the skull base or vertebral bodies, this approach is used. Finally, for exposure and removal of metastatic nodes or for nasopharyngeal, tonsillar, oropharyngeal, or hypopharyngeal tumor excision, this approach can be extremely helpful.

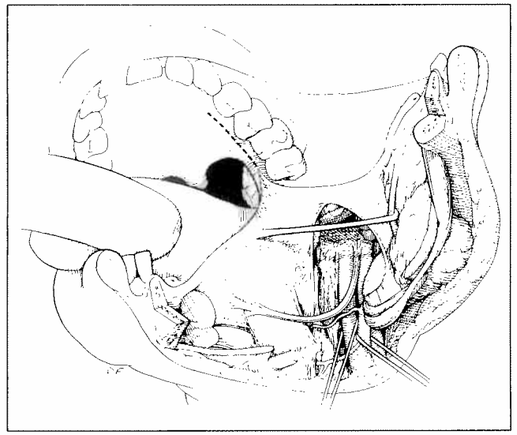

Fig. 16. Defect and operative exposure after tumor removal by cervical-parotid approach with midline mandibulotomy. (By permission of Mayo Foundation.)

Important Points

1. A tracheotomy and a feeding tube are always necessary with a midline mandibulotomy.

2. It is not necessary, in all cases, to split the lower lip and, if cosmesis is an issue, the incision can be extended across to the opposite side of the neck beneath the mandible, and the tissues can be raised off the mandible as a visor flap. A lower lip split incision, closed carefully and extended around the natural border of the chin, results in a cosmetically acceptable scar in most cases.

3. For individuals who have lower teeth, the mandibular osteotomy can be performed by using an osteotome between the incisor teeth, dividing this portion of bone as the last cut. The remaining bone cuts are made with a Stryker saw.

4. If the teeth are crowded, it is simpler to remove one tooth and make the osteotomy through that site.

5. For individuals with teeth, preoperative impressions are made and a pre-made lingual splint is fabricated, which is wired into position at surgery and removed, generally, in 6 weeks. This further helps to ensure adequate healing of the osteotomy site and preservation of the lower incisor teeth.

6. The osteotomy is done in a stair-step fashion and can be reapproximated at the end of the operation by using wires or miniplates.

7. The divided mylohyoid, styloglossus, and stylopharyngeus muscles are reattached as part of the closure.

8. In most cases, the lingual nerve can be preserved.

9. It is helpful to enter the parapharyngeal and retropharyngeal space inferiorly just above the 12th cranial nerve and to extend the incision and dissection in a superior direction.

10. If, as a result of tumor removal, the 10th and 12th cranial nerves lose preoperative functioning, consideration should be given to performing a cricopharyngeal myotomy and Teflon augmentation prior to closure.

11. Neck dissection can be performed in conjunction with the cervical-parotid approach for tumors that are known to be malignant or when approaching malignancies that extend into the parapharyngeal space.

12. For tumors that involve the nasopharynx, the superior incision can be extended onto the hard palate to allow removal of the eustachian tube area and adjacent muscles.

13. The decision to attempt removal of a malignant tumor that has invaded the retrostyloid space by direct extension or metastatic involvement is always difficult. A complete evaluation must be done first to rule out any evidence of distant metastasis. The patient's overall health must be assessed and the surgeon should have a frank discussion with the patient about expected goals and risks of surgery. If the decision is made to proceed with surgery, a balloon occlusion study of the carotid artery must be performed first. If this study shows the carotid artery can be occluded safely, then, 2 days prior to surgery, permanent detachable balloons are placed in the internal carotid artery. The patient is placed on anticoagulant therapy for 48 hours prior to surgery, and the operation is performed.

14. Hospitalization generally averages 10 to 14 days, in contrast to a cervical-parotid approach, in which hospitalization is 2 to 3 days. Edema can be significant after the midline mandibulotomy, and a short course of steroid therapy initially is helpful in decreasing oral swelling.

15. Swallowing is often difficult after a midline mandibulotomy because of interruption of the branches of the pharyngeal plexus. Care should be taken in initiating an oral diet.

INTRAOPERATIVE MONITORING

Intraoperative monitoring of regional cranial nerves is now done routinely for tumors that require mandibulotomy or craniotomy. Intraoperative monitoring of cranial nerves VII, X, XI, and XII is simple and may decrease the incidence of nerve injury. Monitoring has been most helpful for surgery in which tumors extend intracranially through the jugular foramen or for tumors requiring reoperation for a recurrent tumor. Monitoring has been used most often when operating on recurrent parapharyngeal parotid tumors when a prior parotidectomy has been done. In this case, intraoperative monitoring of the seventh nerve is done alone.

For tumors with intracranial extension, obvious erosion of the skull base or cervical spine, or extensive vascular features, the author performs these operations in conjunction with a neurosurgical colleague. Use of the combined talents of two surgical disciplines ensures maximum ease of tumor removal with the least possible morbidity for the patient. If intracranial exposure is necessary or grafting of the carotid artery is necessary, appropriate help is readily available.

SUMMARY

The wide variety of tumors encountered in the parapharyngeal space contributes to unique management problems. The surgeon must be well versed in the complex anatomy of this area to plan for safe removal of tumor. Imaging studies and the capability to predict preoperatively the results of carotid balloon-occlusion studies render many of the tumors operable. Surgery in the parapharyngeal space should not be considered as an operation performed in an inaccessible area with high morbidity. The use of two surgical procedures, the cervical-parotid approach and the cervical-parotid approach with midline mandibulotomy, has met the goals of surgery of the parapharyngeal space. These goals are excellent tumor visualization, preservation of surrounding nerves and vessels, control of bleeding, and complete tumor removal with low morbidity.

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