Management of Tumors of the Parapharyngeal Space

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Benign and malignant tumors can arise from any of the structures contained within the parapharyngeal space. Such tumors are very rare, however. Also, malignant tumors from adjacent areas (eg, the pharynx) can extend into the parapharyngeal space by direct growth, or distant tumors may metastasize to the lymphatics within the space. Although the history and physical examination can provide clues to the site of origin and nature of a parapharyngeal space tumor, imaging studies are more useful for defining the site of origin and extent of the mass, as well as its vascularity and relationship to the great vessels of the neck and other neurovascular structures. Surgery is the mainstay of treatment. The surgical approach chosen should facilitate complete tumor extirpation with minimal morbidity. Irradiation is administered as primary therapy in patients with unresectable tumors, poor surgical candidates, and selected other patients. Radiation therapy is also used after surgery for high-grade malignancies or when wide surgical margins cannot be achieved.


ABSTRACT: Benign and malignant tumors can arise from any of the structures contained within the parapharyngeal space. Such tumors are very rare, however. Also, malignant tumors from adjacent areas (eg, the pharynx) can extend into the parapharyngeal space by direct growth, or distant tumors may metastasize to the lymphatics within the space. Although the history and physical examination can provide clues to the site of origin and nature of a parapharyngeal space tumor, imaging studies are more useful for defining the site of origin and extent of the mass, as well as its vascularity and relationship to the great vessels of the neck and other neurovascular structures. Surgery is the mainstay of treatment. The surgical approach chosen should facilitate complete tumor extirpation with minimal morbidity. Irradiation is administered as primary therapy in patients with unresectable tumors, poor surgical candidates, and selected other patients. Radiation therapy is also used after surgery for high-grade malignancies or when wide surgical margins cannot be achieved. [ONCOLOGY 11(5):633-640, 1997]

Introduction

Parapharyngeal Space
The parapharyngeal space is a potential space shaped like an inverted pyramid. The temporal bone comprises the base of the pyramid, represented by the greater cornu of the hyoid bone (Figure 1). Posteriorly, the space is bounded by the prevertebral fascia, prevertebral muscles, and vertebrae. Laterally, the space is defined by the posterior belly of the digastric muscle and the ascending ramus, coronoid process, and condylar neck of the mandible, covered by the pterygoid muscles and their fascia. These three boundaries are rigid, and therefore, limit the expansion of a tumor arising within the space to the anterior, inferior, and medial directions (Figure 2 and Figure 3). The medial boundary consists of the buccopharyngeal fascia and superior constrictor muscle. The pterygomandibular raphe and the fascia of the pterygoid muscles constitute the anterior boundary. The parapharyngeal space, thus, does not communicate with the masticator space.[1]

A layer of fascia extending from the levator veli palatine muscle to the styloid process divides the parapharyngeal space into an anterolateral compartment, the prestyloid compartment, and a posteromedial compartment, the post-styloid compartment (Figure 2 and Figure 3). The anterolateral compartment contains the stylomandibular process of the parotid gland, fat, and lymph nodes. The posteromedial compartment includes the contents of the carotid sheath; namely, the carotid artery, internal jugular vein, cranial nerves 9 through 12, and lymph nodes. The sympathetic nerve chain lies beneath the deep cervical fascia, and, although not part of the parapharyngeal space, it has nerve branches that travel in the post-styloid parapharyngeal space along the path of the internal arteries.

Benign and malignant tumors can arise from any of the structures contained within the parapharyngeal space. Malignant tumors from adjacent areas (eg, pharynx) can extend into the space by direct growth, or distant tumors may metastasize to the lymphatics within the space. Nonetheless, tumors arising in this area are extremely rare, comprising less than 1% of the neoplasms arising within the head and neck region.[2-4]

**Diagnosis**

**Clinical Evaluation**
Deviation of Oropharyngeal Structures

A parapharyngeal space tumor most commonly presents clinically as a painless mass in the neck or bulging of the oropharynx, which causes a lump-in-the-throat sensation. A significant number of parapharyngeal space tumors are discovered as an incidental finding when the patient is examined for an unrelated problem.[4] Rarely, patients may complain of having difficulty swallowing, pain, or trismus.[4,5] Lower cranial neuropathies, most commonly associated with paragangliomas or malignant tumors, compound the swallowing difficulty, and a vocal cord paralysis may present as hoarseness (Figure 4). FIGURE 6

Prestyloid Mass
FIGURE 7

Left Parotid Tumor

The physical examination should be thorough and should include a full assessment of the cranial nerves and flexible laryngoscopy. The latter ascertains the status of the airway and the motor and sensory innervation of the larynx and pharynx.

The clinical examination provides clues to the site of origin and nature of the tumor. The presence of a pulsatile, compressible mass or a thrill suggests a vascular tumor. Bimanual palpation may identify the origin of the mass as the deep lobe of the parotid gland. A history of rapid growth, the presence of a mass that extends outside of the parapharyngeal space, or a mass that is associated with severe pain all suggest the possibility of a malignancy. Nonetheless, the site of origin and extent of the mass, as well as its vascularity and relationship to the great vessels of the neck and other neurovascular structures, are better defined by imaging.

**Imaging Studies**

FIGURE 8

Undifferentiated Sarcoma
Computed Tomography (CT)--A CT scan with contrast ascertains the site of origin, extent, and vascularity of tumors of the parapharyngeal space and their relationship to adjacent neurovascular structures (Figure 5). The presence or absence of fat planes between the tumor and adjacent structures and the direction in which the tumor displaces the fat planes help determine whether the tumor is located in the prestyloid or post-styloid compartment and whether the site of origin is the parotid gland[1,4,5] (Figure 6 and Figure 7). In addition, a CT scan provides a precise definition of the skeletal framework, demonstrating bone remodeling, expansion, or destruction (Figure 8).

Magnetic resonance imaging (MRI) provides exquisite definition of the soft-tissue planes and allows for multiplanar imaging (Figure 9).[6] Magnetic resonance imaging is preferred over CT when soft-tissue invasion or perineural invasion is suspected, when the use of intravenous contrast is contraindicated due to a previous major allergic reaction, and in patients who should avoid exposure to radiation (ie, pregnant woman, small children). However, MRI is more expensive than CT, takes longer to complete, is more prone to motion artifact, and cannot be tolerated by some patients due to claustrophobia. The CT or MR scan often suggests the histologic diagnosis (Figure 10 and Figure 11). The presence of a malignancy is strongly suggested by a mass that extends into adjacent spaces, (eg, masticator space), infiltrates soft tissue, destroys bone, or is associated with lymphadenopathy (Figure 8).
Glomus Vagale

**Angiography** is still considered the test of choice to define the blood supply of vascular tumors and establish their site of origin. Widening of the carotid bifurcation (lyre sign) by a vascular tumor strongly points to the presence of a carotid body tumor. Anterolateral displacement of the internal carotid artery indicates a glomus vagale (Figure 12).[7,8] In addition, angiography provides the opportunity to embolize tumors in preparation for surgical extirpation.

**Magnetic resonance angiography (MRA)** delineates the relationship of malignant or vascular tumors to the great vessels of the neck. It may be used in lieu of angiography to ascertain the presence of multiple paragangliomas or to map the blood supply of tumors of the parapharyngeal space. It is most useful for the evaluation of compression of a major vessel (Figure 5), such as the internal carotid artery or internal jugular vein; when intraluminal extension of the tumor is suspected; or for the assessment of collateral circulation and venous drainage (eg, large glomus jugular with sigmoid sinus obstruction). Magnetic resonance angiography does not offer the possibility of tumor embolization, however.[9]

**Cerebral Blood Flow Supply**

**Cerebral blood flow** studies, such as transcranial Doppler and balloon occlusion studies (eg, single-photon emission CT [SPECT], xenon CT), are indicated when the patient is at risk for sacrifice of or major injury to the internal carotid artery. These studies predict whether the blood flow provided by the contralateral internal carotid artery is adequate to supply both cerebral hemispheres (Figure 13).[10]

**Other Diagnostic Tests**

Patients presenting with symptoms that suggest the presence of a catecholamine-secreting paraganglioma, such as tachycardia, high blood pressure, diarrhea, and facial flushing, are tested with a 24-hour urine collection for catecholamine analysis. If the catecholamine levels are elevated, the patient is advised to undergo an abdominal CT scan to rule out a pheochromocytoma. If the source of abnormal catecholamine production is unknown, a metaiodinated benzylguanidine (MIBG) radioisotope scan may help elucidate the location of the secreting tumor.[11]

**Differential Diagnosis**

**TABLE 1**

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<th>Most Common Origins of Tumors of the Parapharyngeal Space</th>
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**TABLE 2**
Histologic Diagnosis of Parapharyngeal Space Tumors

Tumors of the parapharyngeal space are most commonly benign, although malignant tumors comprise approximately 20% to 30% of reported series. Most commonly, the tumors are of salivary or neurogenic origin (Table 1).[12-14] Other histologies include sarcomas, lymphoreticular malignancies, and metastasis from distant tumors (Table 2).

Treatment

Surgery

The mainstay of treatment of tumors of the parapharyngeal space is surgical extirpation. Complete excision of the tumor is recommended to corroborate the diagnosis suggested by the clinical examination and imaging and, in most cases, as final treatment. Fine-needle aspiration biopsy is recommended for patients who are poor surgical candidates or for those who are suspected of having a lymphoma or metastatic tumor. An incisional biopsy is indicated when the fine-needle biopsy fails to establish a definitive diagnosis.

Surgical Approaches--The ideal surgical approach should facilitate the complete extirpation of the tumor with minimal morbidity. The choice of the surgical approach is dictated by the index of suspicion for malignancy; histology, site of origin, and size of the tumor; relationship of the tumor to neurovascular structures; and experience of the surgeon.

Transoral Approach--The transoral approach offers a direct route for the extirpation of small tumors of the prestyloid compartment that present as a bulge in the oropharynx and do not extend into the post-styloid compartment. However, this approach affords only limited exposure of the tumor and does not allow for control of the great vessels.

Transcervico-Submaxillary Approach--The transcervico-submaxillary approach provides a direct route to the prestyloid compartment, as well as access to the great vessels of the neck. The exposure provided by this approach may be enhanced by medial displacement of the tumor toward the pharynx. This maneuver facilitates visualization of the tumor and control of the distal extracranial internal carotid artery. If necessary, transection of the stylomandibular ligament and prognathic dislocation of the mandible widens the exposure by more than 50%. This approach is ideally suited for benign prestyloid tumors of minor salivary gland origin.[4]

Transcervical Approach--This approach is reserved for post-styloid masses. It shares all of the advantages and disadvantages of the transcervico-submaxillary approach. It does not violate the submandibular triangle, however.

Transmandibular Approaches--Both the lateral mandibulotomy approach and the median mandibu- otomy-transpharyngeal approach provide vessel control and exposure for vascular tumors extending into the skull base or for squamous cell carcinomas of the oropharynx extending into the parapharyngeal space.[15,16] A tracheotomy and primary repair of the mandible are required for all transmandibular approaches. A mandibulectomy is indicated when the tumor has invaded the mandible.

Transparotid Approach--Some surgeons have advocated the use of this approach for the surgical extirpation of all parapharyngeal space tumors. Although the transparotid approach provides good exposure of tumors of the post-styloid space, it involves a total parotidectomy and extensive
dissection of the facial nerve. We consider this approach to be indicated only for those tumors that originate in the deep lobe of the parotid gland and extend into the parapharyngeal space. Figure 14

CT Scan

**Infratemporal Fossa Dissection/Craniofacial Approach**—These approaches are reserved for malignant tumors, lesions involving the skull base, or tumors extending into the cranial cavity (Figure 14).

**Complications**—Surgical morbidity varies according to the approach used, histologic diagnosis, and tumor differentiation. Benign tumors that can be excised transorally or transcervically are associated with minimal morbidity; the most common complication in this setting is temporary weakness of the marginal mandibular nerve induced by traction, pressure, or dissection of the nerve. Transparotid approaches are associated with paresis or temporary paralysis secondary to similar mechanisms. It is important to counsel the patients about the neurologic defect expected after resection of a neurilemmoma of the parapharyngeal space (eg, Horner’s syndrome after removal of a tumor arising from the sympathetic plexus).

Surgery for paragangliomas and malignant tumors is associated with a greater incidence of neurovascular complications, including cranial neuropathies and cerebrovascular accidents. Thus, the current trend is to observe asymptomatic paragangliomas for which surgical extirpation would require extensive dissection or sacrifice of the internal carotid artery or injury to multiple cranial nerves.

**Radiation**

Irradiation, as primary treatment, is reserved for patients who have unresectable tumors, metastatic tumors from distant sites, or lymphoreticular malignancies and for patients who are poor surgical candidates. Postoperative irradiation is recommended for high-grade malignancies or when wide surgical margins cannot be obtained.

**Other Treatments**

It is possible that in the near future, vascular tumors of the parapharyngeal space, such as paragangliomas and hemangiomas, may be controlled with antiangiogenic agents, obviating the need for surgery.

**References:**


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