นิพนธ์ต้นฉบับ

Tumors of the parapharyngeal space : a 25-year review

Pimpet Sukumalpaiboon* Virachai Kerekhanjanarong*
Pakpoom Supiyaphun* Amnuay Cutchavaree*

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Objective : *To evaluate the incidence, clinical presentations and management of parapharyngeal space tumors*

Design : *Descriptive study*

Setting : *Department of Otolaryngology, Faculty of Medicine, Chulalongkorn University*

Material and Method : *The medical records of 20 patients with the diagnostic of parapharyngeal space tumors during 1971 to 1996 were reviewed. The data on clinical presentations, diagnosis imaging techniques, type of treatments, pathological outcomes were evaluated.*

Results : *Sixty percent of lesions were benign, 40% were malignant. Fifty five percent (11/20) were neurogenic in origin, 15% (3/20) were salivary in origin, 15% (3/20) were connective tissue in origin, 15% were miscellaneous origin. Four cases of rare lesion were found; i.e., synovial sarcoma, ectomesenchymona, mixed tumor of neurilemmoma and hemangioma, and metastatic tumor of papillary cell cancer of thyroid. The most common presenting symptom and sign were neck mass and pharyngeal mass, respectively. The most common surgical approach was cervical approach.*

*Department of Otolaryngology, Faculty of Medicine, Chulalongkorn University

Conclusion : *Parapharyngeal space tumors are rare. Diagnosis of the lesion requires high degree of suspicion, thorough physical examination, and either CT scan or MRI. Most of the lesions are benign and of neurogenic origin. Almost all of the tumors are surgically excised through various surgical approaches either cervical, combined parotid cervical or intraoral approach.*

Key words : *Parapharyngeal space tumors, Neurilemmoma, Synovial sarcoma.*

Repring request : Sukumalpaiboon P, Department of Otolaryngology, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.

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วัตถุประสงค์ : *เพื่อศึกษาอุบัติการณ์การเกิดเนื้องอกข้างฟาริงซ์ ลักษณะอาการและอาการแสดง แนวทางการวินิจฉัย และรักษา*

รูปแบบการศึกษา : *การศึกษาเชิงพรรณา*

สถานที่ที่ทำการศึกษา : *ภาควิชาโสต นาสิก ลาริงซ์วิทยา คณะแพทยศาสตร์ จุฬาลงกรณ์-มหาวิทยาลัย*

วิธีการ : *ศึกษาย้อนหลังจากทะเบียนประวัติผู้ป่วยนอกและในที่ได้รับการวินิจฉัยว่าเป็นเนื้องอกข้างฟาริงซ์ระหว่างปี พ.ศ. 2524 ถึง พ.ศ. 2539 จำนวนทั้งหมด 20 ราย ข้อมูลที่นำมาศึกษา ได้แก่ ลักษณะอาการ และอาการแสดงชนิดของรังสีวินิจฉัยที่ใช้ประกอบการวินิจฉัย และรักษา วิธีการรักษา และการผ่าตัดผลทางพยาธิวิทยาของเนื้องอก*

ผลการศึกษา : *เนื้องอกส่วนใหญ่เป็นชนิดไม่ร้ายแรง (ร้อยละ 60) และพบเนื้องอกที่เกิดจากเซลล์ประสาทร้อยละ 55 เกิดจากเซลล์ของต่อมน้ำลายร้อยละ 15 นอกจากนั้นพบเนื้องอกที่เกิดจากเนื้อเยื่อประสาน ต่อมน้ำเหลือง เป็นต้น ก้อนเนื้องอกบางชนิดพบได้ยาก ได้แก่ synovial sarcoma, ectomesen chymoma ก้อนเนื้องอกผสมระหว่างนิวริเลมโมมากับแองจิโอมา ก้อนเนื้องอกข้างฟาริงซ์ที่เกิดจากมะเร็งต่อมไทรอยด์ อาการและอาการแสดงที่พบบ่อย คือ ผู้ป่วยมาด้วยก้อนเนื้อที่บริเวณข้างลำคอ และตรวจพบก้อนอยู่ข้างฟาริงซ์ วิธีการผ่าตัดที่ใช้มากที่สุด คือ transcervical*

สรุป : *เนื้องอกข้างฟาริงซ์พบได้น้อยมาก การวินิจฉัยต้องอาศัยประวัติที่บ่งชี้ การตรวจร่างกายที่ละเอียดและข้อมูลจากจากภาพรังสีทางคอมพิวเตอร์ (CT scan, MRI) ส่วนใหญ่เนื้องอกที่พบจะเป็นชนิดไม่ร้ายแรง และมาจาก เซลล์ประสาท การรักษาส่วนใหญ่คือการผ่าตัดทางด้านข้างของคอ*

Parapharyngeal space tumors are very uncommon, comprising only 0.5 - 1 % of all head and neck tumors.[1-3] The complexity of the anatomy of the parapharyngeal space renders it difficult in assessment of tumors during physical examination, especially in the extension of the tumors in this space. With the aid of recently advanced imaging techniques, i.e., computered tomography scanning (CT) and magnetic resonance imaging (MRI) , an otolaryn- gologist can better evaluate the parapharyngeal tumor. However, a thorough understanding of anatomy and some knowledge of parapharyngeal tumors will tremendously help in making the diagnosis , choosing the most appropriate imaging technique, and planning surgical approaches. At present, a preoperative diagnosis can be made in 90-95% of patients through the use of clinical and imaging information.[4] In this paper, we present details of clinical presentation, diagnostic studies, surgical approaches and pathological outcome.

Materials and methods

A retrospective study of parapharyngeal space tumors treated at the Department of Otolaryngology, Faculty of Medicine, Chulalongkorn University from January 1971 to January 1996 was conducted. Patients with the tumors primarily located in the parapharyngeal space were included, and patients with the tumors extending to or impinging into the space were excluded from this study. The outpatient office files, admission charts and pathological reports were reviewed to evaluate the clinical findings, diagnosis imaging techniques, types of treatment and pathological outcome.

Results

Clinical findings

Sixteen males (80 %) and four females (20 %) with a mean age of 44.7 years (age range of 4-77 years) were studied. Presenting symptoms and signs are shown in tables 1, 2 and figures 1, 2.

Table 1. Presenting symptoms.

Clinical Presentation	Number of Patients
Asymptomatic neck mass	6
symptomatic neck mass	5
neck mass with fever	(1)
neck mass with fever and dysphagia	(1)
neck mass with rapid growth	(1)
neck mass with rapid growth and dyspnea	(1)
neck mass with hoarseness of voice and history of thyroid cancer	(1)
Foreign body sensation in the throat	5
with dysphagia and dyspnea	(3)
with dysphagia	(2)
Swelling of cheek	1
Swelling of cheek with aural fullness	1
Pre-auricular mass with dyspnea	1
Syncope and somnolence	1

Table 2. Presenting signs.

Clinical Presentation	Number of Patients
Pharyngeal mass	10
Pharyngeal mass	(7)
Pharyngeal mass and cervical node enlargement	(1)
Pharyngeal mass and recurrent laryngeal nerve paralysis	(1)
Pharyngeal mass and tonsil an enlargement	(1)
Submaxillary mass	5
Parotid mass	5
Parotid mass	(3)
Parotid mass and serous otitis media	(1)
Parotid mass which extend to carotid trangle	(1)

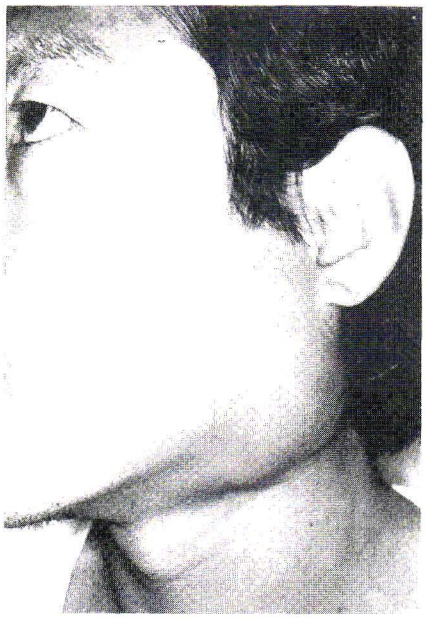

Figure 1. Patient presented with left neck mass.

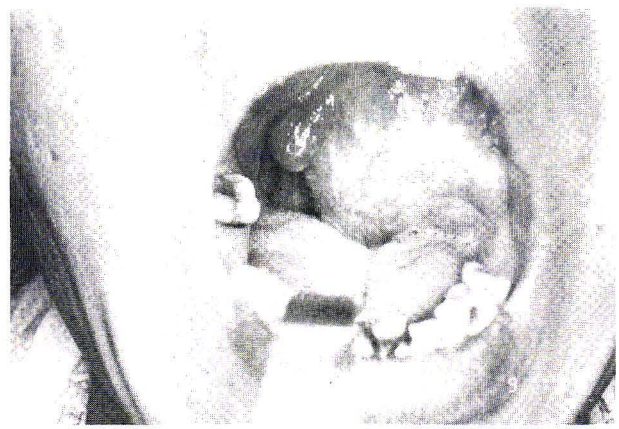

Figure 2. Upon intraoral examination, there was a bulging at left parapharyngeal wall medially displaced left tonsil. The pathological report was malignant schwanoma

Half of the cases were presented as pharyngeal masses (50%), some were either submaxillary or parotid mass. Regardless of their presentation, on intraoral examination there was medial bulging of the lateral pharyngeal wall and tonsil, and a fullness. Bimanual palpation revealed the continuity between the external mass and internal mass with ballottedment.

Table 3 shows the histopathology of the parapharyngeal tumors, and most were are benign (60%). The most common benign tumor found was neulilemmoma (50%). In the six patients with neurilemmoma there was an asymetrical sex distribution with a male to female ratio of 4 to 1. The majority of cases (83%) were in their sixth decade of life or older. Tumors occurred more commonly on the left side (83%) than the right side. The chief complaints were asymptomatic neck or submandibular mass, and foreign body sensation with dyspnea and dysphagia in most of the cases (71%). And interest-ingly, in one case the chief complaint was somnolence and syncope. This particular patient had a history of recurrent syncope temporarily relieved by surgical removal of the parapharyngeal neurilemmoma.

Table 3. Parapharyngeal space pathology.

Histologic Type	Number of Cases
Neurogenic Tumor	
Benign	
Neurilemoma*	6
Mixed neurilemoma with hemangioma[17]	1
Neurofibroma	1
Malignant	
Malignant schwanoma	2
Ectomesenchymoma[16]	1
Soft tissue	
Rhabdomyosarcoma	1
Synovial sarcoma	1
Inflamatory retention cyst	1
Parotid	
Pleomorphic adenoma	3
Lymphnode	
Malignant lymphoma	1
Lymph node hyperplasia	1
Metastastatic Cancer	
Papillary Cancer from Thyroid	1

* 2 cases were previously reported by our senior author.

Diagnostic imaging techniques

Due to the non-availability of imaging devices (CT scan and MRI) before 1988 at our hospital, the diagnosis of cases then relied mainly on clinical suspicion, physical examination and preliminary pathological reports from biopsies. Parotid sialography and angiography were occasionally performed in indicated cases.

After 1988, CT scans were more routinely used as an aid in diagnosis. CT helps make distinctions between extraparotid and intraparotid masses by observation of parapharyngeal space fat. CT also provides information whether the lesion is solid or cystic. MRI are ordered occasionally in selected cases in which the CT scan cannot provide adequate information. MRI enables a more reliable distinction between intraparotid and extraparotid lesions and allows most schwannoma and extraparotid salivary gland tumors to be differentiated. The contour of the tumor is better identified by MRI.

Treatment

Almost all of the tumors (90%) were surgically excised through various approaches. In cases of malignant lymphoma, the patient underwent chemotherapy, and in cases of metastatic papillary cancer no curative treatmant was offered. Table 4 shows various surgical approaches aim at extirpation of the tumor masses. The most common surgical approach employed was the cervical approach, either alone or in combination with a parotid approach (75%). Intraoral approach were used for small avascular tumors which were not palpable in the neck and parotid region. The cervical-parotid approach with a step-wise mandibulectomy, an approach that provide better tumor visualization and preservation of nerves and vessels,[12] was used in cases of recurrent neurilemmoma and malignant schwannoma. In three cases a tracheostomy were required prior to definite surgical treatment due to a compromised airway from the large intraoral component of masses.

Table 4 . Surgical Approaches.

Surgical Approach	Number of Cases
Cervical approach (Submandibular)	10
Cervical - Parotid approach	5
with mandibular osteotomy	(2)
without mandibular osteotomy	(3)
Intraoral approach	3

Discussion

The parapharyngeal space is a potential space located deep in the neck with the boundaries forming an inverted pyramid within the base of skull and the greater cornu of the hyoid bone. Superiorly, its base is formed by the temporal and sphenoid bones. The lateral boundary is composed of pterygoid muscle , the ramus of the mandible , and the posterior belly of the digastric muscle . The anterior boundary is composed of interpterygoid and buccinator fascia and raphe. The posterior boundary is made up of the vertebral column and paravertebral muscles. The similarity among the aforementioned boundaries is that they are rigid. However, the medial and inferior boundaries are soft and more pliable and are made up of superior pharyngeal constrictor muscle, tonsillar fossa, and submadibular gland fascia, respectively. Hence, a parapharyngeal tumor will tend to enlarge medially and inferiorly presenting as a bulging in the pharyngeal wall with a submandibular mass and /or a neck mass.[12]

The parapharyngeal compartment is further divided into *the prestyloid* and *poststyloid* compartments. The *prestyloid* compartment is adjacent to the tonsillar fossa and nasopharynx. It contains the internal maxillary artery and inferior alveolar, lingual and auriculotemporal nerves. Thus, tumors within this space can cause symptoms of cranial nerve V_3 involvement. The *poststyloid* compartment contains the internal jugular vein, internal carotid artery, cranial nerves IX, X, XI, XII, cervical sympathetic chain, and numerous lymph nodes. A tumor within this space will cause symptoms pertaining to nerve structures involvement.

Cumulative results of several previous large series studies with a total of 336 cases reviewed by Stanford et al. in 1985 revealed that the most common tumor presenting in the parapharyngeal space were salivary gland tumors (40.7%), followed by neurogenic tumors (30.6%) and lymphoma (10.7%).[6] In our study, we found that 55% of the tumors were of neurogenic origin and 15% salivary in origin. This result is similar to Myer's, et al. Study. They found that 57 % of the tumors were of neurogenic origin and 30% salivary.[6] The discrepency may be to the differences in defining the papharyngeal space tumor which excludes tumors that impinge upon the space and also may be due to the population studied. Even though in both our study and Myer's study neurogenic origin tumors comprised the majority of the tumors, the difference is that 63 % were neurilemmomas in our study and 86% were paragangliomas in Myer's study.

Neurilemmoma

The neurilemmoma are typically benign, slow growing, solitary tumors of Schwann cell origin and were first described by Verocay in 1908. It is also known under various terminologies as schwannoma, neurilemmoma and neurinomas; the first two terms are more commonly used.[7,8] They are not commonly found in the head and

neck region (25%-45% of all reported neurilem moma), and even are more rare in the parapharyngeal space.[7] In the previous study by Cutchavaree, et al., of 63 cases of neurilemmoma of the head and neck region, only two cases were found in the parapharyngeal space.[9] They usually arise from Schwann cell of the vagus nerve or the cervical sympathetic chain[10] and are easily separated from the nerve because they are encapsulated tumors that are attached to, or surrounded by the nerve pushing the axon aside. These tumors have distinctive histological characteristics. Neurilem moma has a centrifugally compacted cellularity arranged with palisading nuclei (Antoni A pattern) alternating with a more loosely arranged myxoid cellularity (Antoni B pattern). Verocay body may also be seen in the Antoni A pattern tumors.[11]

The most frequent initial complaints found in our study were an asymptomatic neck mass and an awareness of a mass in the throats with some degree of dysphagia and dyspnea. The mean duration of suffering of the second most common complaint (3 months) was much shorter than the first complaint (22 months). Dyspnea,requiring a tracheostomy was observed in two patients in whom the tumors had grown intraorally to large size with the greatest tumor dimension of 6 centimeters or larger. One patient had a history of recurrent syncope which was relieved by sugical removal of the tumor mass. The symptom may occur as a result of pressure exerted on the baroreceptor in carotid sinus created by large parapharyngeal mass. None of our subjects had symptoms of cranial nerve dysfunction or Horner's syndrome. Pain and characteristic radicular pain were not found in our study.[11]

Synovial Sarcoma

Synovial sarcoma accounts for 5-15% of all soft tissue malignancies.[13] This tumors is usually found in the extremities where synovial tissue is present and it is rarely found in the head and neck region.

Lesions occurring in the head and neck have been found most commonly in young adult males. The most common presenting symptoms are either a painless mass lesion, airway obstruction, dysphagia or hoarseness.[14] Our one such patient was thirty years old with a four month history of cheek swelling and aural fullness. She did not complain of dysphagia or dyspnea. Examination revealed a swelling at the right parotid area, bulging at the right parapharyngeal wall and medially placed right tonsil, and right serous otitis media. The mass was excised transparotid and gross pathologic examination revealed a firm gray-white irregularly lobulated mass. The histopathologic examination revealed synovial sarcoma. Histologically "classic" synovial sarcoma is characterized by a biphasic cellular pattern consisting of epithelioid cells arranged in nests and acini surounded by a fibrosarcoma-like area.[13] This patient was followed-up regularly with CT scan and MR imaging for recurrence of the lesion. However,

six years after surgery no recurrence had been observed.

The usual method of treatment is a combination of surgical excision, radiation, and chemotherapy. The prognosis of patients with head and neck synovial sarcoma is less than 40 %. Pulmonary spread is the most common problem.[14]

Papillary CA

There are few cases of lymph node metastases to the parapharyngeal space as revealed in the literature.[2,6] The parapharyngeal space contains lymphatic which drain the nasal cavities, paranasal sinus, nasopharynx, oropharynx and a portion of the thyroid gland.[16] Ferrario, et al had purposed a mechanism of parapharyngeal lymph node metastases of papillary cancer of the thyroid. Papillary CA tends to develope lymphatic metastasis. The neoplastic emboli will first spread to pretracheal and paratracheal nodes and the superior and inferior jugular nodes before reaching the parapharyngeal space. A retrograde spread via a reversal of lymphatic stream may explain a possibility of parapharyngeal metastases and a slow progressive nature of papillary cancer.[16]

Our patient was a known papillary thyroid cancer case. He was on iodine ablasion therapy when he was presented at the ENT department with symptoms of hoarseness and a feeling of a lump in the throat. Incisional biopsy was made and the pathological results revealed papillary CA of thyroid origin. No surgical treatment was offered.

Summary

Parapharyngeal space tumors are rare. In our 25-year review, neurogenic origin tumor was the most common. Many rare tumors such as ectomesenchymoma, synovial sarcoma, mixed tumors of neurilemmoma and hemangioma, and metastatic tumor of the papillary carcinoma of thyroid were found. Removal of the tumors through a cervical approach was our method of choice for most tumors.

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