



PARAPHARYNGEAL TUMOURS OUR EXPERIENCE

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ABSTRACT:

This study is conducted to evaluate the clinicopathological features and the management of parapharyngeal space tumours. This study is conducted as a retrospective analysis of patients from 2008 to 2010 at Madras Medical College, Rajiv Gandhi General Hospital, Chennai. All the patients were diagnosed on the basis of clinical examination, FNAC, radiological imaging. Total number of patients was 13 out of which 6 were women and 7 males. Age group varied from 25 to 55 years average 35 years. All thirteen patients underwent surgical excision of tumour by transcervical approach. There was no major post operative complication. All patients are followed over a period of three years.

Introduction:

The parapharyngeal space is a complex anatomical area. Primary parapharyngeal tumors are rare tumors and only account for 0.5% of all the head and neck tumors. About 80% of these tumors are benign and the other 20% are malignant. The differential diagnosis of the primary parapharyngeal tumor includes salivary gland neoplasm, neurogenic tumor and many miscellaneous tumors. Most common tumour of PPS is pleomorphic adenoma followed by neurilemmoma and paraganglioma. Clinical examination of the PPS is difficult and therefore CT, MRI are essential to delineate the tumour extent, spread, intracranial involvement, relationship to the adjacent vital structures. Imaging also helps in planning the surgical procedure. FNAC of the tumour is done first.

Materials and methods:

The clinical features, histopathology and radiological features, surgical management, outcome of all the thirteen patients were analysed in a retrospective manner. Depending on the location of the tumour in PPS the cases were classified as prestyloid and post styloid space tumours. Among the thirteen patients twelve patients underwent transcervical approach; one patient underwent transcervical approach with mandibulotomy. A horizontal incision at the level of hyoid bone or a vertical incision along the anterior border of the sternocleidomastoid made, submandibular gland retracted, apex of the prestyloid space gained between the digastric muscle and mandible. Post styloid region accessed medial to posterior belly of digastric muscle. Tumours are divided without damaging the capsule or the nerve of origin. Post operatively patients were followed up with regular ultrasonography and imaging.

Results:

Total of thirteen patients where 7 males and 6 females included in the study (table: 1, chart 1). All the patients had neck swelling, change in voice, difficulty in swallowing, with pushing of lateral oropharyngeal wall and soft palate. One of the patients presented late with slow growing tumour on the right side of the neck as a firm non pulsatile swelling in the sub mandibular region.

All the thirteen patients underwent all the preliminary investigations and the imaging. Among them in eight patients the tumour was in the post styloid compartment and in the remaining five patients the tumour was in pre styloid compartment. No adjacent bony erosion was found in all the cases.

All the patients underwent surgical excision through transcervical approach. One patient with tumour in the pre styloid compartment was approached with additional mandibulotomy. Histopathological examination of the pre styloid compartment mass showed myxomatous stroma mixed with myo epithelial cells, epithelial cells and glandular elements suggestive of pleomorphic adenoma. In all other tumours the HPE showed Antoni A cells, Verocay bodies' interspersed with Antoni B cells suggestive of neurilemmomas. Post operative period of all the patients were uneventful and all the patients are being followed up with no recurrence (Table 2).

Discussion:

Review of Anatomy:

The parapharyngeal space is often described to be a deep potential neck space shaped as an inverted pyramid. The base of the pyramid is at the skull base and the apex is at the greater cornu of the hyoid bone. The boundaries of the parapharyngeal space are as following:

- 1) Superior border: small portion of the temporal bone.
- 2) Inferior border: junction of the posterior belly of the digastric muscle and the greater cornu of the hyoid bone
- 3) Medial border: pharyngobasilar fascia and pharyngeal wall.
- 4) Lateral border: medial pterygoid muscle fascia, the ramus of the mandible, retromandibular portion of the deep lobe of the parotid gland and posterior belly of the digastric muscle.

5) Posterior border: vertebral fascia and paravertebral muscles.

6) Anterior border: pterygomandibular raphe and medial pterygoid fascia.

Clinically, the parapharyngeal space should be considered in two spaces: pre-styloid space and post-styloid space. Fascia from the styloid process to the tensor veli palatini muscle divides the parapharyngeal space into these two compartments. The pre-styloid space is anterolateral and contains retromandibular portion of the deep lobe of the parotid gland, minor or ectopic salivary gland, a small branch of the CN V to the tensor veli palatini muscle, ascending pharyngeal artery and pharyngeal venous plexus. The majority of the pre-styloid space is actually fat. The post-styloid compartment is posteromedial and contains internal carotid artery, internal jugular vein, CN IX to XII, cervical sympathetic chain, lymph nodes and glomus bodies.

Differential diagnosis of primary parapharyngeal tumors:

Most of the tumors of the parapharyngeal space are metastatic disease or direct extension from adjacent spaces. Primary parapharyngeal tumor is uncommon. As per literature, 80% tumors of the parapharyngeal space are benign and 20% are malignant. The location of the tumor can be helpful for differential diagnosis. Tumors in the pre-styloid space are most likely to be salivary gland tumor, lipoma, or rare neurogenic tumors.

tumors. And all the structures in the post-styloid compartment are potential sources for post-styloid tumors. Hughes reviewed 172 patients with parapharyngeal space neoplasm and found that pleomorphic adenoma was the most common neoplasm (40%), followed by paraganglioma (20%), neurogenic tumor (14%), malignant salivary gland tumor (13%), miscellaneous malignant tumors (7%), and miscellaneous benign tumors (6%). This result is consistent with most reports. However, there are few literatures reported neurogenic tumors as the most frequent entities.

Pleomorphic adenoma is the most common salivary gland tumor in the parapharyngeal space. It can originate in the deep lobe of the parotid gland as well as from the minor salivary gland tissue. The frequency of malignant parapharyngeal salivary neoplasm varied greatly in the literature. The reported malignant tumors include mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, malignant mixed carcinoma, squamous cell carcinoma, adenocarcinoma, and a case of malignant Warthin's tumor.

In most of the reports neurogenic tumor is the second most common tumor in the parapharyngeal space. Schwannoma is the most common type of neurogenic tumor. The vagus nerve has been reported to be the nerve of origin in 50% of parapharyngeal schwannoma. Schwannomas in general are slow growing and usually do not affect the nerve of origin. The symptoms are mainly compressive.

Paraganglioma is the second most common neurogenic tumor in the parapharyngeal space. They are either vagal paragangliomas or carotid body tumors. Vagal paragangliomas are parapharyngeal in location in 2/3 of the cases.

Neurofibroma is the third most common neurogenic tumor in the parapharyngeal space. Malignant neurogenic tumors have been reported in the parapharyngeal space include malignant schwannoma, malignant paraganglioma, malignant neuroblastoma, or sympatheticoblastoma.

Clinical presentation and Evaluation:

Clinical detection of parapharyngeal tumor is difficult. The tumor has to grow to 2.5 to 3.0 cm to be detected clinically. The presentation can be very subtle. Generally it presents as an asymptomatic mass causing mild bulging in the soft palate or tonsillar region, or fullness near the angle of the mandible. With tumors enlarging superiorly, they can cause soft palate and nasopharyngeal swelling. And inferior growth of tumors result palpable masses at the angle of the mandible. The ensuing symptoms depend on the affected site. The pre-styloid lesion can present as serous otitis media, voice change, nasal obstruction, aspiration or dyspnea. The post-styloid lesion can compress the 9th, 10th, 11th and 12th nerve and cause hoarseness, dysphagia, dysarthria, or Horner's syndrome by tumor pressure on the superior cervical sympathetic ganglia.

Cranial nerve palsy, pain, and trismus often suggest malignancy. Other than a complete head and neck exam, imaging studies such as CT, MRI and angiogram are important modalities to support the diagnosis of parapharyngeal tumors and distinguish pre-styloid tumors from those in the post-styloid space.

On CT scan, salivary gland tumor is in pre-styloid space and displaces the carotid artery posteriorly. Also if there is a fat plane between the lesion and the deep lobe of the parotid gland, the mass is extraparotid. The most common enhancing extraparotid lesion on CT scan is the schwannoma. The schwannoma on the CT scan is in the post-styloid space and usually displaces the carotid artery anteromedially. Lesions that show enhancement on CT scan include paragangliomas, hemangiomas, hemangiopericytomas, aneurysms, and schwannomas.

MRI provides the most useful preoperative information about the extent of the tumor and its relationship to surrounding structures. If carotid involvement is suspected, an angiogram should be done. It can be used to accurately define the vascular anatomy and collateral circulation, to perform the carotid occlusion study and possibly to embolize the tumor preoperatively. Carotid occlusion test must always be done whenever a malignant tumor involves the post-styloid space or for extensive vascular tumors that surround the carotid artery at the level of the skull base.

Surgical approaches:

The goal of parapharyngeal surgery is to provide adequate tumor visualization to achieve complete tumor removal, while preserving the surrounding nerves and vessels and control of any hemorrhage. Many surgical approaches have been reported in the literature.

Overall, transcervical and transparotid approach are the two main approaches. They have been reported to be used alone or in combination of each other. They have also been used with mandibulotomy to increase exposure. Transcervical approach is usually used for post-styloid tumors and transparotid approach for pre-styloid tumors. Transparotid approach is commonly used for deep lobe parotid tumors.

Transcervical approach starts with a transverse incision at the level of the hyoid bone. The submandibular gland is often removed or retracted anteriorly. An incision through the fascia deep to the submandibular space allowed for entry into the parapharyngeal space and blunt dissection of the tumor. Many modifications have reported. Some surgeons divide the digastric, stylohyoid, and styloglossus muscles from the hyoid bone to improve exposure. The styloid process and the stylo-mandibular ligament can also be divided to elevate the mandible anteriorly to improve access. This approach frequently involves blind finger dissection in the parapharyngeal space and does not provide enough exposure for larger benign lesions extending cranially or those with a more aggressive growth pattern.

Transcervical approach can be combined with mandibulotomy. Various locations for osteotomy have been reported, including mandibular body, angle, ramus, and parasymphiseal. The key is to try to limit injury to the inferior alveolar nerve while providing access to the parapharyngeal space. The risk of mandibulotomy includes inferior alveolar nerve anesthesia, loss of dentition, malocclusion, mandibular malunion or nonunion, and possibly requires a tracheostomy. It is thought to be necessary in less than 10% of patients.

Many surgeons prefer the transcervical-transparotid approach. A standard parotidectomy incision is made and carried into the lateral neck. The main trunk and lower division of the facial nerve are identified. The posterior belly of the digastric and stylohyoid muscle are divided, allowing for visualization of the internal and external carotid arteries, internal jugular vein, CN IX, X, XI, XII, and sympathetic chain. The styloid process and stylo-mandibular ligament may be transected to give a wide opening into the parapharyngeal space. This approach is recommended for all deep-lobe parotid tumors, extraparotid salivary tumors and most of the post-styloid neurogenic tumors.

For vascular tumors that extend into the superior portion of the parapharyngeal space, Olsen recommended cervical-parotid approach with midline mandibulotomy. Midline lip splitting is used to expose anterior mandible. Then the mandible is divided in the midline. An intraoral incision is then made in the floor of the mouth extending back to the anterior tonsillar pillar and up to the level of the hard palate. The hypopharyngeal nerve and lingual nerve are preserved. The styloglossus and stylopharyngeus muscles are divided. The mandible is retracted laterally and superiorly to give wide exposure to the parapharyngeal space. Tracheostomy is necessary for this approach.

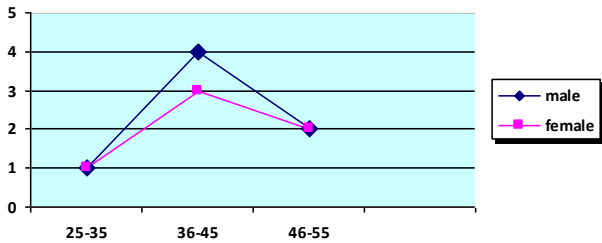
Conclusion:

Primary parapharyngeal tumors are rare and are located in a complex anatomical region. The clinical presentation of these tumors can be subtle.

Therefore radiographic study provides important information for diagnosis and surgical planning. Majority of the tumors are benign with salivary gland neoplasm being the most common tumor. Surgical resection is the mainstay of treatment. Transcervical approach with or without mandibulotomy is the most preferred method.

s. no	Age in years	Number of patients operated (2008 to 2010)	
		Male	Female
1	25-35	1	1
2	36-45	4	3
3	46-55	2	2

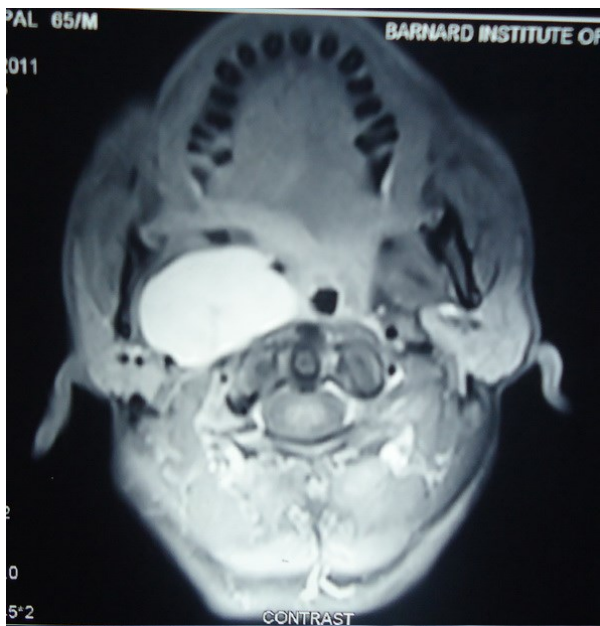
Age distribution of the patients Table 1



Age distribution of patients Chart 1

s.no	Approaches	No. of cases operated		Success	Failures	Recurrences over 3 year follow up
		Males	Females			
1	Transcervical app.	6	6	12	Nil	Nil
2	Transcervical with mandibulotomy	1	-	1	Nil	Nil

Results Table 2



Intraoral view of the mass

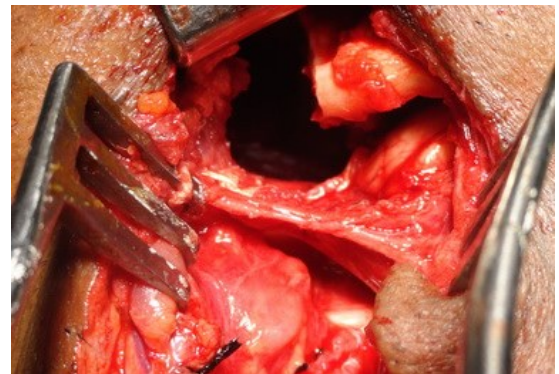


Image showing transcervical approach



Imaging in parapharyngeal tumors

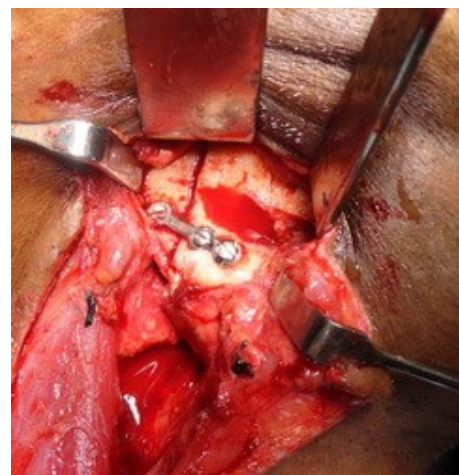
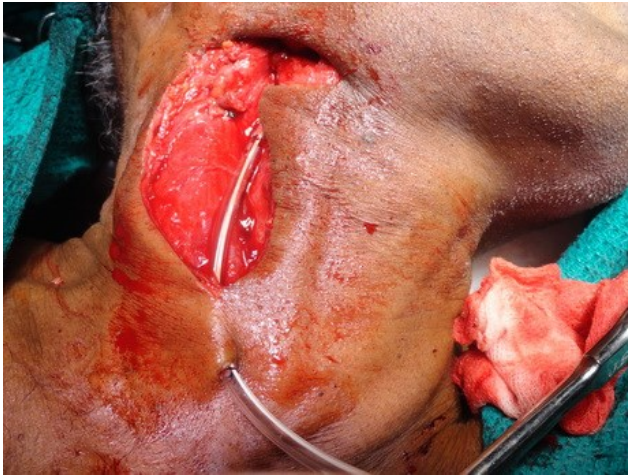


Image showing mandibular fixation after mandibulotomy approach



Closure after transcervical approach



Dumb bell shaped mass after removal

References:

Hughes KV., Olsen KD., McCaffrey TV. Parapharyngeal space neoplasms, Head Neck 1995; 17; 124-130.

Luna-Ortiz K, Navarrete-Aleman JE., Granados-Garcia M., Herrera-Gomez A. Primary parapharyngeal space tumors in a Mexican cancer center. Otolaryngology and Head and Neck Surgery 2005; 132(4); 587-591.

Olsen KD. Tumors and surgery of the parapharyngeal space. Laryngoscope 1994; 104: 1-28.

Shahab R., Heliwell T., Jones AS. How we do it: A series of 114 primary pharyngeal space neoplasms. Clinical Otolaryngology 2005; 30; 364-367.

Batsaki JG, sneige N(1989) parapharyngeal and retro pharyngeal space disease.

Shahinan H Dirinier c, Fisch U (1995) parapharyngeal space tumors , skull base surgery 5 (2):73-81

Betka J Chovenac M, Klozer(2010) transoral combined transoral - transcervical approach to parapharyngeal tumours . Eur Arch otorhinolaryngological 267: 765 -772