# STUDY OF INCIDENCE & MANAGEMENT OF PARA PHARYNGEAL TUMORS

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**ABSTRACT: INTRODUCTION:** Parapharyngeal tumors are rare, comprising approximately 0.5% of all head and neck tumours. Most of them are benign. These tumors present with difficulties in diagnosis - complementary MRI and CT scanning are necessary for diagnosis, and Fine Needle Aspiration Cytology (FNAC) is very specific in the histological diagnosis of these tumours. Open biopsy is not advisable due to bleeding, breaching of the capsule and seeding of the tumor. These tumors presents a challenge to the surgeon due to its anatomical complexities. This study deals with the incidence and management of various parapharyngeal tumors.

**OBJECTIVE OF THE STUDY:** This study deals with the incidence of various tumors in the parapharyngeal space in different age and sex groups, role of sophisticated diagnostic modalities like CT, MRI, MR Angio. Colour Doppler along with FNAC and various surgical approaches to this space. This study also deals with intra-operative and post operative complications. In this series, a total of 25 cases has been studied retrospectively in a time period of 2 years from 2012 to 2014, presenting in our ENT and Head and Neck Dept., Gandhi hospital.

**RESULTS:** According to this study, there is male preponderance (52%) and highest incidence is seen in 3rd and 5th decade (24% each). Most common presenting symptoms are difficulty in swallowing (36%) and swelling either intraoral or in the neck (28%). Least common symptoms being cranial nerve palsy (4%), difficulty in breathing/noisy breathing (4%), nasal regurgitation (4%) and hard of hearing (8%). FNAC was done in 21 cases, in which 13 were correlating with the biopsy report. CT scan was required in all cases. MR Angiography was done in 4 cases and colour Doppler in 2 cases. Surgery is the mainstay of the treatment. Most common tumor in PPS is neurogenic (schwannoma/neurofibroma).i.e 44%. Next commonly occurring tumor in our study is of salivary origin-pleomorphic adenoma (24%), paragangliomas (12%). Other less commonly seen tumors are AV malformation, epidermal cyst, rhabdomyosarcoma, mucoepidermoid ca., fibroangioma.

**KEYWORDS:** Parapharyngeal space, Prestyloid compartment, Post styloid compartment, Transcervical approach, Transparotid approach.

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**INTRODUCTION:** The parapharyngeal space (PPS) is defined as the deep space that forms an inverted triangular pyramid in the neck where the posterior belly of the digastric muscle and hyoid bone forms the apex of the pyramid, and the temporal bone, its base. Anteriorly, the space is bounded by the pterygoid muscles with their interpterygoid fascia and those fasciae directed laterally towards the buccinator muscle and angle of the mandible. Laterally, the space is limited by the ascending ramus of the mandible, while posterolaterally the deep lobe of the parotid gland and retromandibular fossa can be identified. Medially, there is the pharynx with the tonsillar fossa inferiorly and eustachian tube superiorly. The posterior border is limited by the cervical spine covered by prevertebral muscles and fascia.[1,2,3]

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The parapharyngeal space is known in the literature under the names of pterygomaxillary space, pharyngomaxillary space (Mosher, 1929),[4] lateral pharyngeal space and pterygopharyngeal space (Coller and Yglesias, 1935).[5] The term 'parapharyngeal space' seems most appropriate and is commonly used in the current literature.

The fascia stretching from the styloid process to the tensor veli palatini muscle divides the PPS into prestyloid and poststyloid compartments.[6,7]

The retropharyngeal space is connected with the parapharyngeal space in an area just medial to the carotid sheath and its contents. Situated at the junction of these two spaces is the superior lateral lymph node (node of Rouvière, 1927)[8] normally draining the nasopharynx, upper oropharynx and sinuses. The retropharyngeal space provides a pathway towards the mediastinum (Lincoln's highway), while anteriorly and laterally there are connections to other spaces located about the oral cavity and salivary glands.[9]

Primary tumours of the PPS are very rare, comprising approximately 0.5% of all head and neck tumours.[10,11] They often present asymptomatic growth and can stay undetected for long periods of time or may be detected as

an incidental mass during screening for another reason. These tumours frequently manifest via medial displacement of the lateral wall of the oropharynx or via a growth on the upper neck, and nearly 50% of patients present with a neck mass.[12]

Of special interest is the deep lobe of the parotid gland between the mandible laterally, the medial pterygoid muscle medially and the masseter muscle anteriorly. The sternocleidomastoid and posterior belly of the digastric muscles as well as those structures arising from the styloid process are in a posterior position.[13] Patey and Thackray (1956) coined the term 'stylomandibular tunnel' formed by the posterior margin of the ascending ramus of the mandible anteriorly, the styloid process and stylomandibular ligament behind and the base of the skull above. This tunnel resists pressure so that tumours of the deep lobe of the parotid gland assume a dumb-bell shape as they grow.[14]

Tumours of the parapharyngeal space originate from the various types of cells and tissues present within this space, by direct extension from neighbouring structures or through lymphogenous or haematogenous spread. Numerous primary benign and malignant tumours have been described, the group of salivary gland tumours being most prevalent (50%) followed by the neurogenic tumours (30%).[15,16] The parapharyngeal space is richly endowed with lymphatics that drain the sinuses, pharynx and thyroid gland (Robbins and Woodson, 1985).[17]

**PATHOLOGY:** Tumors of the parapharyngeal space originate from the various types of cells and tissues present within this space, by direct extension from neighbouring structures, or through lymphagenous or hematogenous spread. Numerous primary benign tumors have been described.

1. **Salivary gland tumours:** Two types of salivary gland tumours occur.
    1. The first type arises de novo from salivary gland tissue in the parapharyngeal space and has no connection with the parotid gland.
    2. The second type is the 5% of parotid tumours that arise within the deep lobe of the parotid gland (Conley and Clairmont, 1978) and it is this type that has an intimate relationship with the gland.[18]

Most are benign pleomorphic adenoma while the most common malignant tumour is adenoid cystic carcinoma (Hanna et al, 1968; Work and Gates,[19] 1969; Johns, 1977).

2. **Neurogenic tumours:**
    - Nerve sheath tumours.
    - Schwannomata.
    - Neurofibromata.
    - Malignant Schwannoma.

3. Paraganglioma.
4. Hemangioma.
5. Lymphangioma.
6. Branchial Cyst.
7. Benign Lymphoepithelial lesions.
8. Dermoid tumors.
9. Amyloid tumors.
10. Teratoma.
11. Meningioma.

**MALIGNANT:**

1. Lymphoma.
2. Fibrosarcoma.
3. Fibrous histiocytoma.
4. Hemanangiopericytoma.
5. Plasmacytoma.
6. Chordoma.
7. Rhabdomyosarcoma.

The commonest tumors are those arising in the deep lobe of the parotid gland, and they compromise 50% of parapharyngeal tumors.[20] They may be dumb-bell shaped because of constriction in the stylomandibular tunnel, and may present either as a pharyngeal or external swelling.

Neurogenic tumors are the next commonest type of tumor. These are schwannomas, neurofibroma, ganglioneuroma or neuroblastoma. Meningiomas may extend into the space through the jugular foramen.

Paragangliomas is the most common sarcoma described at this site. Lymphomas may arise in the parapharyngeal nodes other lesions include teratomas, developmental cysts, lipomas and less commonly hemangiomas and rhabdomyosarcomas.[21]

**Neurilemmoma (Schwannoma):** All peripheral motor and sensory axons are covered by Schwann cells throughout their length. Tumors arising from these cells are called schwannomata and are the most common neurogenic tumours in the parapharyngeal space. They are also more common in the head and neck than elsewhere in the body.

Microscopic examination reveals that the tumour is composed of alternating areas, some organized and compact (Antoni type A) and some of loosely arranged, relatively acellular tissue (Antoni type B).[22]

**Malignant Schwannomata:** A malignant schwannoma is a malignant neoplasm of nerve sheath origin that infiltrates locally and also metastasizes. There is pleomorphism, hyperchromatism, increased cellularity and frequent mitoses.

**Nerve Cell Tumours:** The terms neuroblastoma and ganglioneuroma indicate the cell of origin. Although these tumours usually occur in the adrenal medulla, they occasionally develop along peripheral nerves elsewhere in the body including the head and neck. They may produce catecholamines.

**Paragangliomata:** The parapharyngeal space contains vagal bodies closely associated with the ganglion nodosum of the vagus nerve. These bodies contain clusters of

chemoreceptor cells and make up a part of the chemoreceptor or glomus system of the body. Tumours originating from glomus bodies are called glomus tumours, chemodectomata or paragangliomata.[23]

**Diagnosis and Treament:** PPS tumors are difficult to diagnose early due to their location and plethora of presentation. CT scan and FNAC have a great role in making the diagnosis and deciding the suitable surgical approach. CT scan can demonstrate the exact size and extent of the tumor and its relationship to surrounding structure. Magnetic resonance imaging (MRI) has also become a very useful diagnostic tool.

The new generation of CT scanners with better resolution power not only accurately delineate the extent of the tumour, but also give information as to whether it arises from the deep lobe of the parotid or de novo. In the latter case there may be a lucent line representing the compressed layer of fibroadipose tissue between the tumour and the deep lobe of the parotid (Som, Biller and Lawson, 1981).[24,25] Computerized tomographic scanning may indicate the degree and extent of bone erosion.

If the radiological picture is non vascular then a FNAC is done to get the diagnosis. Open biopsy is not advised, as it increases the risk of bleeding, breakage of the capsule and, accordingly, the seeding of the lesion.[26,27]

The surgical approach best applied to the parapharyngeal space tumours is an external one, which affords adequate visualization, control of bleeding, and identification of major vessels and nerves.[28] Internal approaches are to be discouraged, except perhaps in the rare circumstance of an extremely small lesion localized to the medial aspect of the space that can clearly be defined as such. Hughes et al.[29] and Malone et al.[30] in their study mostly used the transcervical approach, though Hughes et al used the transparotid approach also but trancervical was the most common one. McElroth[31] and Ehrilch[32] described the transoral approach but this was limited for small non vascular tumours.

The success of parapharyngeal tumor surgery depends on two conditions: correct identification and proper exposure of the lesion, allowing for complete removal; and minimum functional and aesthetic morbidity as a consequence of the surgery.

**AIMS AND OBJECTIVES:** This is a retrospective study of parapharyngeal space tumors, their incidence, mode of presentation and management. Management includes diagnosis with use of investigations like fine needle aspiration, CT scan,MRI Neck, Angiography and colour Doppler. The study also emphasizes on the approach used in accessing the tumor and its complications.

**SELECTION OF THE PATIENTS:** Only those patients who presented with mass in parapharyngeal space were included from 2012 to 2014. Any swelling mimicking or pushing the oropharyngeal contents medially eg quinsy or tumors of tonsil were excluded. Careful history noted down with emphasis on each symptom. And cases which were fully evaluated with thorough history, clinical examination and investigations are taken into the study.

**MATERIALS AND METHODS:** As this is a retrospective study, the history sheets and operative notes were retrieved from the record room of the years 2012 to 2014. 25 patients are included in this study attending our OPD at Gandhi Hospital, Secunderabad.

**OBSERVATIONS AND RESULTS:** Parapharyngeal tumors are rare. Observations done on retrospective study of 25 patients for the past 2 years 6 months making 0.023% of the total patients attending the ENT Dept at Govt ENT Hospital and Gandhi Hospital.

**ASSOCIATED SYSTEMIC DISEASES:**
This was seen in few cases.
Hypertension- 2 cases
Diabetes Mellitus- 1 case.

**INVESTIGATIONS:** Routine investigations like CBP, CUE, RBS, Blood urea, Sr.creatinine, ECG, 2DECHO, HIV1 and 2,HBsAg were done for all 25 cases.

FNAC was done for 21 cases, 13 were correlating with the biopsy report, remaining were inconclusive.

CT scan was performing for almost all cases, CT scan helped to delineate extent of the lesion with its relationship with the vital structures in the neck and the nature of the tumor.

**MR Angiography was done for 4 cases.**

- Splaying of carotids was observed in two cases of paraganglioma.
- 1 case of schwannoma with pushing the carotids anteriorly and to the right side.
- 1 case of rhabdomyosarcoma.

Colour Doppler was done for 2 cases of paraganglioma to study the status of carotid vessels.

**TREATMENT:** Mainly surgical treatment.

Surgery was done for 21 cases, in 2 cases procedure was abandoned because of AV malformation in one case and carotid body tumor in the other, remaining two cases left against medical advice.

All operated cases were approached externally.

- Transcervical approach – 12.
- Transcervical Transparotid – 4.
- Transcervical submandibular – 5.

**POST OPERTIVE CARE:** All the patients were put on higher antibiotics for 7 days and Ryles tube feeding for the first 48 hours. Ipsilateral facial paresis was recognized in 1 case.

Vagus nerve was sacrificed in one case as the tumor was neurogenic, of vagal origin.

**FOLLOW UP:** All the patients were followed up for 2 months to 1 year, no recurrences were found. There were no intraoperative or postoperative deaths.

**DISCUSSION:** Total number of patients attending ENT Department during the period Jan 1999 to July 2001 was 1,00,500. The incidence of parapharyngeal tumors is 0.023%.

Other parapharyngeal tumors like Teratoma, Malignant Lymphoma, Lymphangioma, Lipoma were not encountered in our study.

**AGE INCIDENCE:** Dr.Mangal Singh[33] reported highest age incidence in the 3rd and 4th decade, with minimum age as 2 years and maximum age 60 years.

In our study the highest age incidence was in 3rd and 5th decade, with minimum age at 5 months and maximum at 70 years.

**SEX INCIDENCE:** Male preponderance 13 of 25 cases as it was in Dr Mangal Singh study.

**MODE OF PRESENTATION:** Commonest mode of presentation was difficulty in swallowing, intraoral swelling followed by swelling in the neck in the present study.

Swelling in the oropharynx was the commonest mode of presentation in Dr. Mangal Singh study. Cranial nerve palsies was also observed in the present study where as Dr Mangal Singh reported 36.8% of his patients had cranial nerve palsies at the time of presentation.

**SURGERY:** 12 of 25 cases were exclusively transcervical approach, 5 cases by transcervical approach submandibular approach and 4 cases by transcervical transparotid approach.

**INCISION:** In 12 cases a transverse skin crease incision was given on the swelling depanding on the extent of the swelling.

In 6 cases, a transverse skin crease incision was given at the level of the hyoid bone extending from the greater horn of the hyoid anteriorly to the mastoid process posteriorly.

In 4 cases, a superficial parotidectomy incision was given. Emergency tracheostomy was done in one case.

Surgery was abandoned in 2 cases. In one there was an AV malformation and in the other cleavage between the tumor and the carotid vessel was not possible.

Mandibular branch of the facial nerve was severed in 2 cases in transcervical transparotid approach.

**PERIOD OF FOLLOW UP AND RESULTS:** In the present study no recurrence of the growth was observed. Residual facial paresis improved over a period of 6 months.

In Dr Mangal Singh study too there was no recurrence of growth during follow up.

**CONCLUSIONS:** This is a retrospective study of parapharyngeal space tumors and their management done during the period Jan 1999 to June 2001 at Govt ENT Hospital and Gandhi Hospital, Secunderabad.

1. Parapharyngeal tumors are rare tumors with an incidence of 0.023% in our study.
2. Tumors are common in 3rd and 5th decade.
3. There is a a male preponderance.
4. Most common mode of presentation is difficulty in swallowing.
5. Intra oral biopsy should be condemned as it may lead to an uncontrollable hemorrhage.
6. FNAC plays a major role in provisional diagnosis of parapharyngeal space tumors.
7. we have found that HRCT (High Resolution Computerised Tomography) is now the best initial diagnostic study tool as it helps to determine the extent of tumor, differentiate tumors of parotid and de novo parapharyngeal space tumors, determines the degree of vascularity and separates benign from malignant.
8. MR Angiography, Carotid Doppler also helps in identification of soft tissue involvement, status of carotid vessels and degree of adherence of the growth to the vessels and the recognition of the feeding vessels.
9. Most of the parapharyngeal space tumors are benign in nature, surgery with transcervical approach that was used most of the cases in our study allowed better exposure of all vital structures.
10. Majority of the parapharyngeal space tumors are benign in nature, neurogenic followed bt salivary gland tumors.
11. Two cases were sent for radiotherapy.
12. Very few post operative complications were encountered like facial paresis, X nerve palsy.
13. Recurrences was not noticed in the operated cases during the follow up period.

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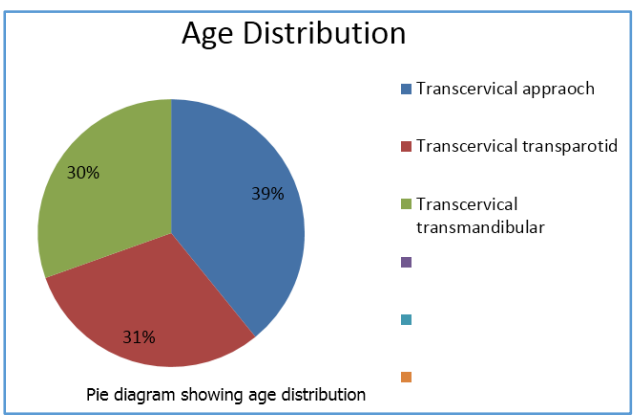
| | Pre styloid | Post styloid |
|---|---|---|
| Arteries | Maxillary | Internal Carotid |
| Veins | - | Internal Jugular |
| Nerves | Inferior Alveolar | 9, 10, 11, 12 |
| | Lingual | |
| | Auriculotemporal | Cervical sympathetic chain |
| Lymph Nodes | - | ++ |
| Glomus Bodies | - | ++ |

***Structures located in the compartments of the parapharyngeal space and in the retropharyngeal space***

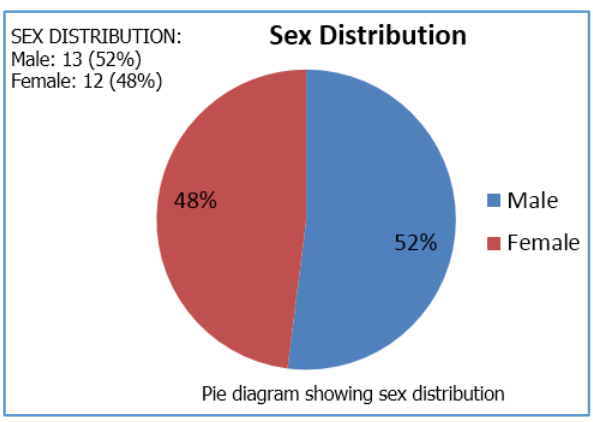
Source: Hans Heeneman, anatomy, chap 21; Parapharyngeal space tumors.

| Age Group | No. of cases | % |
|---|---|---|
| Infancy | 1 | 4 |
| 1-10 | 2 | 8 |
| 11-20 | 4 | 16 |
| 21-30 | 6 | 24 |
| 31-40 | 4 | 16 |
| 41-50 | 6 | 24 |
| 51-60 | 2 | 8 |

***Age Distribution***



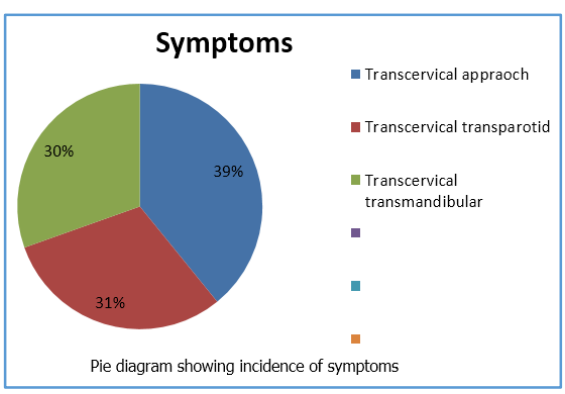
Pie diagram showing age distribution



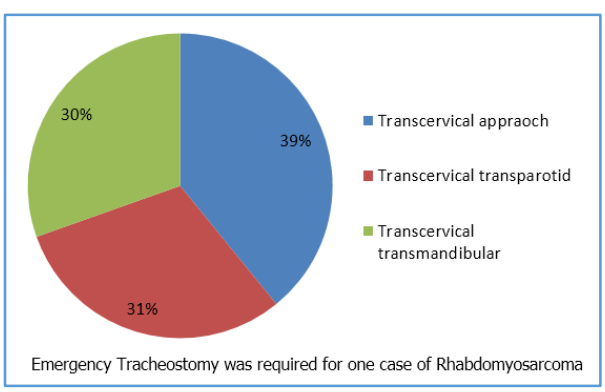
Pie diagram showing sex distribution

| Symptom | No. of cases | % |
|---|---|---|
| Difficulty in swallowing | 9 | 36 |
| Intraoral swelling | 7 | 28 |
| Swelling in the neck | 7 | 28 |
| Change in Voice | 5 | 20 |
| Noisy breathing | 1 | 4 |
| Difficulty in breathing | 1 | 4 |
| Cranial nerve palsy | 1 | 4 |
| Hard of hearing | 2 | 8 |
| Nasal Regurgitation | 1 | 4 |

***Symptoms***

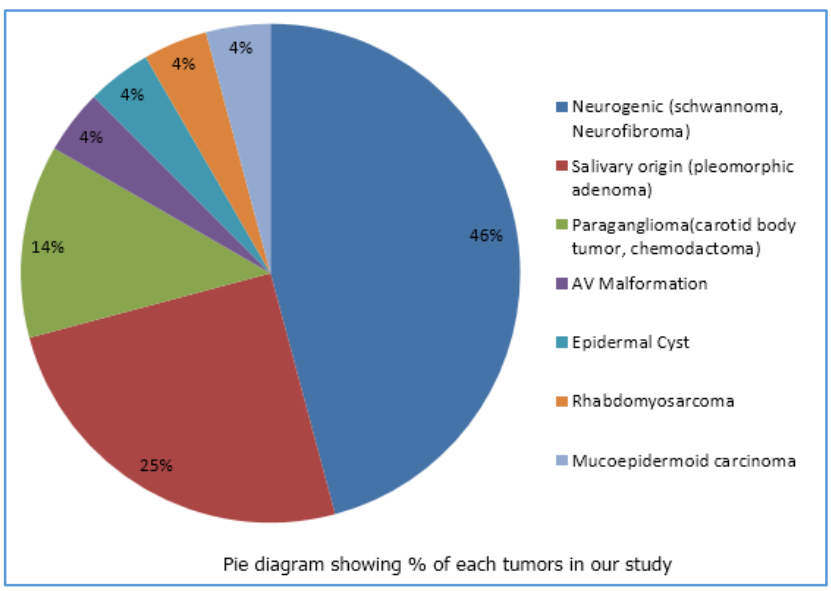


Pie diagram showing incidence of symptoms



Emergency Tracheostomy was required for one case of Rhabdomyosarcoma

| Tumor | Work et al(1969) 18 cases | Mayo's clinic 101 cases | Mangal Singh 26 cases | Our study 25 cases |
|---|---|---|---|---|
| Neurogenic (schwannoma, Neurofibroma) | 27.7% | 16% | 3.8% | 44% |
| Salivary origin (pleomorphic adenoma) | 33.3% | 43% | 0.8% | 24% |
| Paraganglioma(carotid body tumor, chemodactoma) | - | 12% | 27% | 12% |
| AV Malformation | - | - | - | 4% |
| Epidermal Cyst | - | - | - | 4% |
| Rhabdomyosarcoma | - | - | - | 4% |
| Mucoepidermoid carcinoma | -- | - | - | 4% |
| Fibroangioma | - | - | - | 4% |
| ***Specific incidence of parapharyngeal tumors*** | | | | |



Pie diagram showing % of each tumors in our study