Tumours of Parapharyngeal space

- The parapharyngeal spaces (PPS), as the name implies, lie laterally on either side of the pharynx. They are potential spaces, filled with fat and areolar tissue containing branches of the maxillary nerve and maxillary vessels.

**PPS boundaries**

- Superiorly – Skull base
- Inferiorly – Greater cornu of hyoid and Submandibular gland fascia
- Laterally – Ascending ramus of mandible
- Medially – Superior constrictor muscle and pharyngobasilar fascia with tonsillar fossa
- Anteriorly – Interpterygoids and buccinators fascia, tail of Submandibular gland
- Posteriorly – Vertebral column, paravertebral muscles.
- Posterolaterally – Parotid fascia, posterior belly of diagastric muscle

**Prestyloid Compartment**

- Tonsillar fossa inferiorly and fossa of rosenmuller superiorly.

**Contents** – Internal Maxillary Artery, Inferior Alveolar Nerve, Auriculotemporal Nerve and Lingual Nerve

- Poststyloid compartment – Internal carotid artery, internal jugular vein, 9th, 10th, 11th, 12th Nerves, lymph nodes and globus system

- The **Styloid process** with its muscles and condensations of fascia divide the PPS into a prestyloid and a poststyloid compartment.
- While the prestyloid compartment has its apex at the hyoid bone, the poststyloid compartment continues to low in the neck
- More accurately, the poststyloid compartment is separated from the prestyloid compartment by the **tensor palati fascial layer**:
- The main structures contained in the anterior compartment include the pterygoid and tensor palati muscles, fat and the deep lobe of the parotid gland.
- Anatomical perspective – The parotid deep lobe is not within the PPS but separated from it by the deep parotid fascia.
- Surgical importance is the relation of the anterior part of the PPS to the masticator space

**MASTICATOR SPACE**

- This space is formed by the splitting of the deep cervical fascia as it reaches the inferior margin of the mandible, and there envelops the pterygoid, temporalis and masseter muscles.
- Masticator space also contains the **mandibular branch of the trigeminal nerve**.
- Masticator space attaches to a much larger area of the skull base and has the **foramen ovale** in its roof, allowing potential tumour spread.
- Inferiorly, the **masticator space terminates at the lower margin of the mandible**.
- The **poststyloid compartment** contains the carotid sheath, with the carotid artery, jugular vein and vagus nerve. The compartment also contains the sympathetic trunk, the 9,10 (in carotid sheath), 11,12 cranial nerves and the major part of the internal maxillary artery.
Nearly all tumours of the prestyloid compartment are deep lobe parotid neoplasms, whereas most tumours of the poststyloid compartment are either paragangliomas or schwannomas.

## HISTOPATHOLOGY AND SPREAD OF TUMOURS IN THE PARAPHARYNGEAL SPACE

- One of the commonest tumours to involve the PPS is nasopharyngeal carcinoma
- Histologically and anatomically, the most common tumour is the parotid pleomorphic adenoma, located in the prestyloid compartment.

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### Patients with PPS lesions of Liverpool (over a 30-year period).

**Tumour type**

- Benign
  - Pleomorphic adenoma
  - Carotid paraganglioma
  - Vagal paraganglioma
  - Schwannoma
  - Other benign salivary tumours
  - Other benign tumours
  - Meningioma
  - Neurofibroma

- Malignant
  - Adenoid cystic carcinoma
  - Mucoepidermoid carcinoma
  - Other malignant salivary tumours
  - Lymphoma
  - Carotid body paraganglioma
  - Vagal paraganglioma
  - Other malignant tumours

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### SALIVARY GLAND TUMOURS

- These are the most common neoplasms to involve the PPS and invariably are located in the prestyloid compartment and almost always arise from the deep lobe of the parotid gland

### NEUROGENIC TUMOURS

- Paragangliomas arising from the carotid bodies were previously known as carotid body tumours or chemodectomas.
- The paragangliomas cell is sensitive to changes in the blood gas partial pressure, particularly of carbon dioxide but also of oxygen, and is also sensitive to pH.
The cells are positive for Neuron Specific Enolase (NSE) and are classed as neuroectodermal tissue of ANS.

Cervical paragangliomas rarely secrete catecholamines. Familial paragangliomas are notorious in that they secrete catecholamines, and the patients manifest with fluctuating hypertension.

Relatively common Neoplastic change in such a small body of cells is related to their very high metabolic rate.

They also have the highest blood flow of any organ relative to their size.

High altitude paragangliomas have high F:M ratio of 5:1

Low altitude paragangliomas have a F:M ratio of 2:1

Paragangliomas have a brownish colour and are encapsulated. Histologically, they demonstrate a typical appearance of epithelial cell clusters in an extremely vascular and fibrous stroma. The clusters have been given the name zellballen.

The tumours are histologically similar to the adrenal medullary phaeochromocytoma and cases of catecholamine-secreting carotid body tumours have been reported.

**VAGAL PARAGANGLIOMAS**

- These are rare tumours of neural crest origin and less than 200 cases have ever been reported
  - Can arise anywhere along the course of the vagus nerve and its branches, including the middle ear.

  - Microscopically, they have a fusiform appearance and are usually sited close to or involving the jugular foramen, and tumours are well encapsulated.
  - It is frequently intracranial rather than involving the PPS, doing so only as it expands.
  - While carotid body paragangliomas only very rarely secrete catecholamines, it is more common in vagal paragangliomas.

  - A 24-hour urinary Vanillylmandelic acid (VMA) collection should be performed. If the results are suspicious the consensus view seems to be to proceed to a MIBG (metaiodobenzylguanidine) scan.

**MALIGNANT PARAGANGLIOMAS**

- Up to 10 percent of paragangliomas are malignant. In the absence of regional or distant metastasis it is not possible to distinguish benign from malignant lesions on histological features alone.

  - Post operative radiotherapy is mandatory

**FAMILIAL AND MULTIPLE PARAGANGLIOMAS AND PHAEOCHROMOCYTOMAS**

- Multiple paragangliomas, 50% in familial and 10% in non familial.

  - Familial tumours mode of inheritance is AD.

  - The commonest synchronous tumours are carotid body and jugulotympanic tumours.

  - With paragangliomas thus phaeochromocytoma screening should also be done & this should include blood pressure monitoring, preferably by continuous ambulatory electronic monitoring, measurement of plasma catecholamine levels and a 24-hour urinary VMA estimation.

  - Other Neuroendocrine and endocrine tumours can happen in association with paragangliomas. These include parathyroid adenomas, thyroid carcinomas and certain other rare neural crest tumours ie MEN syndromes.
- Medullary thyroid carcinoma + phaeochromocytoma and hyperparathyroidism was termed MEN 2A (Sipple's Syndrome).
- MEN-2 category with medullary thyroid carcinoma and phaeochromocytoma also had multiple mucosal neuromas, sometimes with marfanoid features, puffy lips, prominent jaw, pes cavus and certain other occasional abnormalities with absence of parathyroid disease.
- In MEN gene is located on Chromosome 10.
- Paragangliomas may be involved in the MEN-2B syndrome.

- Histologically, the neurofibromas does not have a capsule and the nerve fibres are within the tumour. Typical cafe au lait spots associated with multiple neurofibromas are pathognomonic of von Recklinghausen's disease.

**VON RECKLINGHAUSEN'S DISEASE**
- Diagnostic criterion of five or more spots, characterized by a light-brown cutaneous macule 1.5 cm or more in diameter, is diagnostic.
- Gliomas may be seen
- Usually localized to skin but cranial nerves may also be affected. MC being 2nd & 8th > 10th.

**RARE TUMOURS OF THE PARA PHARYNGEAL SPACE**
- Lipomas, Sacomas, Rhabdomyosarcomas, leiomyosarcomas, Masson’s tumour, Meningiomas
- Masson's tumour, otherwise known as intravascular papillary endothelial hyperplasia.
- Extracranial meningiomas are rare and MC site is Orbit > PPS.

**METASTASES TO THE PARAPHARYNGEAL SPACE**
- PPS LN usually not cleared during standard procedures and are major cause of recurrence.
- BUT morbidity of removal is very high
- Thus irradiation preferred modality.
- A more common metastasis to the PPS is from nasopharyngeal carcinoma, Thyroid cancer and among distant tumours is Ca Breast.

**THE SPREAD OF TUMOUR WITHIN THE PARAPHARYNGEAL SPACE**
- 80% tumours of PPS benign.
- Expansion of such a tumour, in the plane of least resistance, tends to result in displacement of the lateral oropharyngeal wall and tonsil medially. For such a mass to be apparent it must be at least 3 cm in diameter.
- Obstructive sleep apnoea has been described when medial displacement of the lateral pharyngeal wall is pronounced.
- More superficially placed lesions may expand laterally to produce a lump in the neck, behind the angle of the mandible.
- Submandibular gland and may be displaced downwards.
- Lesions may enter the cranial cavity via one of the foramina, in particular, the carotid canal or jugular foramen. Anteriorly, the foramen ovale can theoretically be entered.
- Pterygoid muscle involvement can lead to trismus.
- High poststyloid lesions may compress the cartilaginous part of the Eustachian tube, causing **middle ear effusion and deafness**.
- **Jugular foramen syndromes** can occur. **Palsies** of 9, 10, 11, 12 nerves can be seen

- Significant spread into the PPS by the tumours like oral, oropharyngeal, nasopharyngeal Ca as well as maxillary cancers is a relative indication of inoperability.

**SYMPTOMATOLOGY**

- Late presentation
- Medial displacement of the oropharyngeal wall or a lump behind the angle of the mandible are relatively common
- In the masticator space, trismus.
- Nerve palsies
- Pain, Otalgia, Middle ear effusions, Deafness.
- The syncope may be associated with glossopharyngeal neuralgia.
- Finally, the syndrome of inappropriate antidiuretic hormone secretion or Schwartz-Bartter syndrome, is well recognized in head and neck cancer.

**DIAGNOSIS, INVESTIGATIONS AND STAGING**

- Examination of all signs symptoms mentioned above.
- **FNAC** (very useful but in few tumours like carotid body tumours, large amount of blood aspirated might lead to difficulty in diagnosis)
- The main problem with FNAC is in patients who have been previously treated and in whom recurrence is suspected.
- PET scanning better for recurrences.
- CT, MRI, Chest radiographs.
- In vascular tumours angiography should also subsequently be carried out.
- **Without angiography, vagal paragangliomas can be difficult to distinguish radiologically from a carotid body paragangliomas**
- Glomus lesions generally are vascular showing flow voids in Angiography.
- DSA (digital subtraction angiography) better in most cases. Major indication is planning for surgery.
- **Neither AJCC nor International Union Against Cancer have given any staging for PPS tumours.**

**THE TREATMENT OF PARAPHARYNGEAL SPACE TUMOURS AND OTHER LESIONS**

- In the case of a carotid body paraganglioma with a stable natural history in an elderly patient, who perhaps is suffering **few symptoms**, the best management is almost certainly **conservative**.
- Equally, a symptomatic distant metastasis to the PPS from another site, such as the **breast**, would only be suitable for palliative treatment, usually by a **short course of irradiation**.
Surgical Treatment

- Surgery may be necessary to obtain a histological diagnosis and this should be excisional rather than incisional.
- Although most of these tumours can be removed relatively easily, on a few occasions major internal carotid artery surgery including excision and grafting has proved necessary.
- Radical radiotherapy given postoperatively as recurrence in this region is extremely difficult to deal with.

Definitive Surgery

- 2 approaches → Transparotid & Transcervical.

Transparotid Approach

- Commonly, the standard transparotid approach may be extended as a transparotid Submandibular approach, which is indicated in larger deep lobe parotid tumours. All that is required is that the standard Modified Bailey Incision for a superficial parotidectomy is extended beneath the mandible anteriorly over the Submandibular space; the submandibular gland may then be displaced or more often excised.

The Transcervical Approach

- A reasonably extensive excision is made from a point 2 cm below the mandibular ramus, starting some 2 cm in front of the mandibular angle and extended backwards and upwards. The size and accessibility of the tumour will dictate how large the incision needs to be.
- Care is obviously required to preserve the mandibular and cervical branches of the facial nerve.
- Division of the digastric tendon then allows excellent exposure with direct visualization and proper oncological removal of the tumour.

Extended Transmandibular Approaches

- With the median mandibulotomy, open transpharyngeal resections can also be carried out.
- While the technique is very rarely necessarily for benign tumours, very extensive malignant tumours may be satisfactorily resected using this method.
- The mylohyoid muscle is divided and an anterior mandibulotomy performed, and the lateral floor of the mouth incised as far as the anterior tonsillar pillar. The contents of the carotid sheath can then be exposed.

Other Exposures Of The Skull Base

- Typically, massive tumours within the Infratemporal fossa and pterygopalatine fossae are approached via a Fisch type C procedure.
- The Fisch type D approach, which uses an infratemporal preauricular incision, provides more limited access but preserves hearing.
**RADIOTherapy**

- Recurrent, secondary’s, Carotid body tumours, tumour extensions etc.
- Not proved too useful in Vagal Body tumours.

**CHEMOTHERAPy**

- Chemoradiation protocols are used primarily in the treatment of rhabdomyosarcomas and other sarcomas of the PPS.
- NPC (nasopharyngeal carcinoma) → Cisplatin + 5 fluorouracil.

**TREATMENT OF THE NECK**

- Reduce the risk of neck node metastasis happening by employing postoperative irradiation to the primary site and the regional nodes.
- A final difficulty is in dealing with patients with bilateral vagal paragangliomas or neurofibromas. For successful removal of these tumours the vagus nerve will, almost certainly, have to be sacrificed. Therefore, it is best to operate on the side with the most advanced lesion and to administer radiotherapy to the opposite side.

- As the vast majority of tumours are benign, the tumour-specific five year survival was excellent at 97 percent.
- For Malignant tumours 5 yr survival was 85%.

**COMPLICATIONS OF SURGERy**

- 9,10,11,12 nerve palsies
- Marginal Mandibular nerve damage
- Haematomas
- Major bleed
- Radiotherapy effects like → Temporal lobe necrosis, xerostomia