STATE OF THE ART
MANAGEMENT of PARAGANGLIOMA

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PARAGANGLIOMAS

HEAD & NECK

• Neural Crest Origin
• Rare: 0.6% of H&N Tumours
• Multiple / Familial
• Sex Distribution
  \[ F>M \text{, } CBT : M>F \]
• Malignancy
  \[ Glomus <5\% \text{, } CBT <20\% \]
• Catecholamine Secretion
  \(<3\% \)
PARAGANGLIOMAS OF THE HEAD & NECK

DECISION MAKING POINTS in MANAGEMENT

➤ CLINICAL ASSESSMENT
➤ DETAILS OF TUMOUR
➤ CATECHOLAMINE SECRETING
➤ GENETIC TESTING
➤ SYNCHRONOUS TUMOURS
➤ TREATMENT
➤ COMPREHENSIVE FOLLOW-UP
PARAGANGLIOMAS OF THE HEAD & NECK

CLINICAL ASSESSMENT

PATIENT HISTORY
- Pre Referral Diagnosis & Rx
- Pulsatile Tinnitus - Causes
- Hearing Loss
- Cranial Nerve Symptoms
- Symptoms of Hormone Secretion
- Family History
- Associated Conditions / Syndromes
PARAGANGLIOMAS OF THE HEAD & NECK

SYMPTOMS

- Pulsatile Tinnitus
- Pain
- Vertigo / Imbalance
- Ear discharge
- Headache
- Hearing Loss
- Ear Pressure
- Cranial Nerve paresis
- Neck Mass
- Voice change
PARAGANGLIOMAS OF THE HEAD & NECK

CLINICAL EXAMINATION

- Vascular Mass in Ear
- Lateral Neck Mass
- Bruit over the tumour
- Cranial Nerve Palsies 7, 9, 10, 11, 12
- Blood Pressure
PARAGANGLIOMAS OF THE HEAD & NECK

TUMOUR DETAILS : IMAGING

- **CT SCAN** Ear, Head & Neck
  - Bone Detail
  - Contrast
  - CTA

- **MRI SCAN** Head and Neck
  - Standard Sequences
  - Contrast
  - MRA *Arterial supply*
  - MRV *Venous drainage*
  - MRV *Venous obstruction*
### Jugulo-Tympanic Glomus Tumour Classification

#### Jackson & Glasscock

<table>
<thead>
<tr>
<th>GLOMUS TYMPANICUM</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1</td>
<td>Promontory</td>
</tr>
<tr>
<td>Type 2</td>
<td>Middle Ear Space</td>
</tr>
<tr>
<td>Type 3</td>
<td>ME and Mastoid</td>
</tr>
<tr>
<td>Type 4</td>
<td>ME, Mastoid and EAC / Ant to ICA</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>GLOMUS JUGULARE</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1</td>
<td>Jug Bulb, ME and Mastoid</td>
</tr>
<tr>
<td>Type 2</td>
<td>Below IAC</td>
</tr>
<tr>
<td>Type 3</td>
<td>Petrous Apex</td>
</tr>
<tr>
<td>Type 4</td>
<td>Clivus or Infratemp Fossa</td>
</tr>
<tr>
<td>(Types 2 - 4 may have intracranial extension)</td>
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</table>

#### Fisch

<table>
<thead>
<tr>
<th>Type A</th>
<th>Confined to Promontory</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type B</td>
<td>ME and Mastoid (Jug Bulb &amp; Carotid Canal Intact)</td>
</tr>
<tr>
<td>Type C</td>
<td>Originate in Jugular Bulb:</td>
</tr>
<tr>
<td></td>
<td>In to Carotid Canal only</td>
</tr>
<tr>
<td></td>
<td>Vertical Carotid</td>
</tr>
<tr>
<td></td>
<td>Horizontal Carotid</td>
</tr>
<tr>
<td></td>
<td>Beyond Foramen Lacerum</td>
</tr>
<tr>
<td>Type D</td>
<td>De Extradural 1cm / 2cm</td>
</tr>
<tr>
<td></td>
<td>Di Intradural 1cm /2cm / Unresectable</td>
</tr>
</tbody>
</table>

#### My Simplified Version

<table>
<thead>
<tr>
<th>Type A</th>
<th>Air of Middle Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type B</td>
<td>Bone of Mastoid involved</td>
</tr>
<tr>
<td>Type C</td>
<td>Carotid Canal</td>
</tr>
<tr>
<td>Type D</td>
<td>Deep / Dural Penetration</td>
</tr>
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</table>
PARAGANGLIOMAS OF THE HEAD & NECK

TUMOUR DETAILS : IMAGING cont

➤ ANGIOGRAPHY (DSA)
  - Arterial supply
  - Venous drainage
  - Embolisation pre surgery
  - Carotid Occlusion

➤ NUCLEAR STUDIES
  - Dotatate / MIBG / FDG
  - Multiple Tumours / Recurrence
PARAGANGLIOMAS OF THE HEAD & NECK

ASSOCIATED PARAGANGLIOMAS

- TYMPANICUM
- JUGULARE
- NECK
  - Carotid Body
  - Glomus Vagale
  - Sympathetic Chain
- ABDOMEN
  - Adrenal: Phaeochromocytoma
  - Extra-Adrenal
- CHEST
# CAROTID BODY TUMOURS

**CLASSIFICATION** *(Shamblin 1971)*

- **GROUP 1** Just in contact with carotid  
  *Resection “straightforward”*

- **GROUP 2** Intimately attached to carotid  
  *Resection may involve vessel wall*

- **GROUP 3** Surrounding carotid  
  *Vessel definitely involved  
  May need replacement*
PARAGANGLIOMAS OF THE HEAD & NECK

CATECHOLAMINE SECRETING

• CATECHOLAMINES
  Urinary
  Plasma
• METANEPHRINES
  Urinary
  Plasma

Neck & other Extra-Adrenal PGL lack PNMT’ase
  --Secrete Normetanephrines & Dopamine only
PARAGANGLIOMAS OF THE HEAD & NECK

FAMILIAL SYNDROME

• Significant implications for Patient & Family
  
  *Present younger age*
  
  *More complex disease*
  
  *Multicentric in 80% vs 20% if Sporadic*

• Must be tested

• Informed Consent required

• Genetic Counselling
# Familial Syndromes in H&N Paraganglioma

<table>
<thead>
<tr>
<th>Mutation Gene</th>
<th>Syndrome</th>
<th>Phaeo</th>
<th>MultiFocal</th>
<th>Malignancy</th>
<th>Recurrence</th>
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<tbody>
<tr>
<td>SDHD 11q23</td>
<td>PGL1</td>
<td>+</td>
<td>++</td>
<td>+/-</td>
<td>+</td>
</tr>
<tr>
<td><strong>Autosomal Dominant with Maternal Imprinting - Carrier only if from mother</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SDHB 1p36</td>
<td>PGL4</td>
<td>++</td>
<td>+</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td><strong>Autosomal Dominant / More complex disease</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SDHC 1q21</td>
<td>PGL3</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>+ CBT</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SDHAF2 11q12.2</td>
<td>PGL2</td>
<td>-</td>
<td>++</td>
<td>+/-</td>
<td></td>
</tr>
</tbody>
</table>
PARAGANGLIOMAS OF THE HEAD & NECK

ASSOCIATED SYNDROMES

Renal Cell Carcinoma
Pituitary tumours
Gastro-Intestinal Stromal Tumours (GIST)
Multiple Endocrine Neoplasia
  Types 2a & 2b
  RET Proto-oncogene mutation
von Hippel Lindau Disease
  Some sub types
  VHL gene mutation
Neurofibromatosis Type I
  NF1 gene mutation
PARAGANGLIOMAS OF THE HEAD & NECK

MANAGEMENT

• CHANGE OF PRACTICE from 1980 to 2018
  - Natural History of Disease
  - Implications & Complications of Rx
  - Complex Algorithm

• OPTIONS
  ➢ OBSERVATION
  ➢ RADIATION
  ➢ SURGERY
MANAGEMENT of HNPGL

• Patient Factors

• Tumour Factors

• Treatment Factors
  – What is available
MANAGEMENT of HNPGL

• **Patient Factors**
  – Symptoms
    • Hearing Loss / Tinnitus
    • Dysphagia / Dysphonia – Lower CN Palsies
    • Facial Palsy
    • Raised Intracranial Pressure
    • Hypertension

  – General Health
    Co-Morbidities / Age
MANAGEMENT of HNPGL

• Tumour Factors
  – Mass Effect : size / site(s)
  – Single / Multiple
  – H & N / Other
  – Intracranial extension
  – Progressive growth
  – Hormone secretion
  – Metastasis / Malignancy

Relative / Absolute
PARAGANGLIOMAS OF THE HEAD & NECK

➤ OBSERVATION

• WHOLE DISEASE PROCESS
  Sporadic or Familial

• MONITORING of PRESENTING TUMOUR
  MRI Repeat at 6 months
  Then at 12 – 18 months

• SURVEILLANCE for ASSOCIATED TUMOURS
  MRI  H&N, Abdomen, Chest
  3 yearly
  Catecholamine Secretion
PARAGANGLIOMAS OF THE HEAD & NECK

RADIATION

• Mode of Delivery
  External Beam
  Stereotactic
  *Single Treatment vs Fractionated*

• Control Rates
  90% +

• Complications
  Radionecrosis
  Cranial Nerve Palsy
MANAGEMENT of HNPGL

➢ SURGERY

• Benefits vs Risks

• Implications & Complications

• Aims
  • Tumour resection
    Total vs Sub-total vs Partial
  • Vital structure preservation
  • Relief of symptoms
  • Eradication of hormone secretion
PARAGANGLIOMAS OF THE HEAD & NECK

➢ SURGERY

• JUGULO-TYMPANIC
  TYPE A  Tympanotomy
  TYPE B  CWU / CWD Mastoidectomy
  TYPE C  Infratemporal Approach
  TYPE D  Infratemporal + PCF approach

• NECK  Complete Resection
  (Intercarotid / Intravagal / Other)

➢ Vessel Management
➢ Nerve Management
INFRATEMPORAL FOSSA SURGERY

• VESSEL MANAGEMENT
  
  Jugular Bulb Resected

  *Ligation of Sigmoid & IJV

  Carotid Artery

  *Preservation

  *Pre-Op Balloon Occlusion

NERVE MANAGEMENT

  Facial Nerve

  *Re-Route or not

  *Lwr Cranial Nerves
PARAGANGLIOMAS OF THE HEAD & NECK

MANAGEMENT OF IMPLICATIONS & COMPLICATIONS

• HEARING LOSS & TINNIUS
  Hearing Aids - Conventional / Bone anchored

• FACIAL WEAKNESS
  Physical Therapy / Eyelid / Repair / Replace

• DYSPHAGIA & DYSPHONIA
  Speech & swallowing therapy
  Vocal cord augmentation

• VERTIGO / IMBALANCE
  Physical therapy
PARAGANGLIOMAS OF THE HEAD & NECK

LONG TERM SURVEILLANCE

• SPORADIC
  Annual Clinical
  Annual MRI + Gad / Nuclear
  Catechol. secretion: 1 to 3 yearly

• FAMILIAL
  Annual Clinical + BP
  Annual Catechol. secretion
  Annual H+N MRI / Nuclear
  1-3 yearly Abdomen & Chest - CT or MRI
PARAGANGLIOMAS OF THE HEAD & NECK

SUMMARY

• FULL ASSESSMENT
  Clinical
  Imaging
  Catechol. Secretion
  Genetic

• MANAGEMENT
  Obs. / Rad. / Surgery / Combined
  Natural History is relevant
  Treatment is individualised to patient