STATE OF THE ART
MANAGEMENT of PARAGANGLIOMA

IFOS, Lima, 2018

VINCENT C COUSINS

ENT-Otoneurology Unit, The Alfred Hospital
&
Department of Surgery, Monash University
MELBOURNE, AUSTRALIA
PARAGANGLIOMAS

HEAD & NECK

• Neural Crest Origin
• Rare: 0.6% of H&N Tumours
• Multiple / Familial
• Sex Distribution
  \[ F>M , \quad CBT : M>F \]
• Malignancy
  \[ Glomus <5% , \quad CBT <20\% \]
• 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*  
  *Venous obstruction*
### Jugulo-Tympanic Glomus Tumour Classification

#### Jackson & Glasscock

**GLOMUS TYMPANICUM**
- **Type 1**: Promontory
- **Type 2**: Middle Ear Space
- **Type 3**: ME and Mastoid
- **Type 4**: ME, Mastoid and EAC / Ant to ICA

**GLOMUS JUGULARE**
- **Type 1**: Jug Bulb, ME and Mastoid
- **Type 2**: Below IAC
- **Type 3**: Petrous Apex
- **Type 4**: Clivus or Infratemp Fossa
  (Types 2 - 4 may have intracranial extension)

#### Fisch

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type A</td>
<td>Confined to Promontory</td>
</tr>
<tr>
<td>Type B</td>
<td>ME and Mastoid</td>
</tr>
<tr>
<td></td>
<td>(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

- **Type A**: Air of Middle Ear
- **Type B**: Bone of Mastoid involved
- **Type C**: Carotid Canal
- **Type D**: Deep / Dural Penetration
PARAGANGLIOMAS OF THE HEAD & NECK

TUMOUR DETAILS : IMAGING cont

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

➢ NUCLEAR STUDIES
  Dotate / MIBG / FDG
  Multiple Tumours / Recurrence
PARAGANGLIOMAS OF THE HEAD & NECK

ASSOCIATED PARAGANGLIOMAS

• TYMPANICUM
• JUGULARE
  \[Ipsi. / Contralateral\]
• 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>
</tr>
</thead>
<tbody>
<tr>
<td>SDHD 11q23</td>
<td>PGL1</td>
<td>+</td>
<td>++</td>
<td>+/-</td>
<td>+</td>
</tr>
<tr>
<td></td>
<td><strong>Autosomal Dominant with Maternal Imprinting -Carrier only if from mother</strong></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></td>
<td><strong>Autosomal Dominant / More complex disease</strong></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>
</tr>
<tr>
<td></td>
<td>+ CBT</td>
<td></td>
<td></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