STATE OF THE ART
MANAGEMENT of PARAGANGLIOMA

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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
  \[ F > M,\ CBT : M > F \]
- Malignancy
  \[ \text{Glomus} < 5\%,\ CBT < 20\% \]
- 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

**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

<table>
<thead>
<tr>
<th><strong>GLOMUS TYMPANICUM</strong></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><strong>GLOMUS JUGULARE</strong></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></td>
<td>(Types 2 - 4 may have intracranial extension)</td>
</tr>
</tbody>
</table>

### Fisch

<table>
<thead>
<tr>
<th><strong>Type A</strong></th>
<th>Confined to Promontory</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Type B</strong></td>
<td>ME and Mastoid</td>
</tr>
<tr>
<td></td>
<td>(Jug Bulb &amp; Carotid Canal Intact)</td>
</tr>
<tr>
<td><strong>Type C</strong></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><strong>Type D</strong></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><strong>Type A</strong></th>
<th><strong>Air of Middle Ear</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Type B</strong></td>
<td><strong>Bone of Mastoid involved</strong></td>
</tr>
<tr>
<td><strong>Type C</strong></td>
<td><strong>Carotid Canal</strong></td>
</tr>
<tr>
<td><strong>Type D</strong></td>
<td><strong>Deep / Dural Penetration</strong></td>
</tr>
</tbody>
</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
  - Extra-Adrenal
• CHEST

Ipsi. / Contralateral
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><em>Autosomal Dominant with Maternal Imprinting - Carrier only if from mother</em></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><em>Autosomal Dominant / More complex disease</em></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>
</tr>
<tr>
<td>+ CBT</td>
<td></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>
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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 2019
  - 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*
  *Vessel stenting*
  *Pre-Op Balloon Occlusion*

NERVE MANAGEMENT

  Facial Nerve
  *Re-Route or not*
  Lower Cranial Nerves
PARAGANGLIOMAS OF THE HEAD & NECK

MANAGEMENT OF IMPLICATIONS & COMPLICATIONS

• HEARING LOSS & TINNIUS
  Hearing Aids - *Conventional*
  *Bone Conduction Implants*

• 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 Study
  1 -3 yearly Abdomen & Chest - CT or MRI or Nuc.
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