

**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 , CBT : M>F*
- Malignancy  
*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

## *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 |

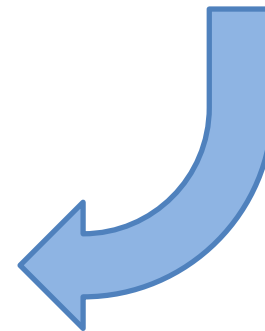
(Types 2 - 4 may have intracranial extension)

## Fisch

|        |  |
|--------|--|
| Type A | Confined to Promontory   |
| Type B | ME and Mastoid<br>(Jug Bulb & Carotid Canal Intact)  |
| Type C | Originate in Jugular Bulb:<br><br>In to Carotid Canal only      C1<br><br>Vertical Carotid                      C2<br><br>Horizontal Carotid                      C3<br><br>Beyond Foramen Lacerum      C4 |
| Type D | De Extradural 1cm / 2cm<br>Di Intradural 1cm / 2cm / Unresectable  |

## My Simplified Version

|        |  |
|--------|--|
| Type A | <u>A</u> ir of Middle Ear                |
| Type B | <u>B</u> one of Mastoid involved         |
| Type C | <u>C</u> arotid Canal                    |
| Type D | <u>D</u> eep / <u>D</u> ural Penetration |





# 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
- 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

| <b>Mutation Gene</b>  | <b>Syndrome</b> | <b>Phaeo</b>       | <b>MultiFocal</b> | <b>Malignancy</b> | <b>Recurrence</b> |
|---|-----------------|--------------------|-------------------|-------------------|-------------------|
| <b>SDHD<br/>11q23</b>   | <b>PGL1</b>     | <b>+</b>           | <b>++</b>         | <b>+/-</b>        | <b>+</b>          |
| <i>Autosomal Dominant with Maternal Imprinting -Carrier only if from mother</i> |                 |                    |                   |                   |                   |
| <b>SDHB<br/>1p36</b>  | <b>PGL4</b>     | <b>++</b>          | <b>+</b>          | <b>++</b>         | <b>+</b>          |
| <i>Autosomal Dominant / More complex disease</i>                                |                 |                    |                   |                   |                   |
| <b>SDHC<br/>1q21</b>  | <b>PGL3</b>     | <b>-<br/>+ CBT</b> | <b>-</b>          | <b>-</b>          |                   |
| <b>SDHAF2<br/>11q12.2</b>   | <b>PGL2</b>     | <b>-</b>           | <b>++</b>         | <b>+/-</b>        |                   |

# 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*