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

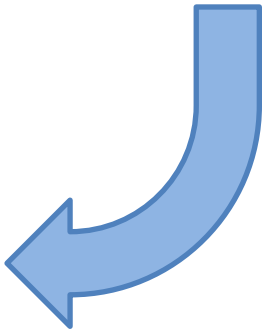
(Types 2 - 4 may have intracranial extension)

Fisch

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

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**
  - 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

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

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*