

**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
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

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

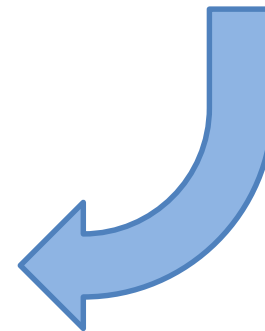
(Types 2 - 4 may have intracranial extension)

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	<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
 - 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	+	++	+/-	+
<i>Autosomal Dominant with Maternal Imprinting -Carrier only if from mother</i>					
SDHB 1p36	PGL4	++	+	++	+
<i>Autosomal Dominant / More complex disease</i>					
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 2019**
 - Natural History of Disease
 - Implications & Complications of Rx
 - Complex Algorithm
- **OPTIONS**
 - **OBSERVATION**
 - **RADIATION**
 - **SURGERY**

MANAGEMENT of HNPG

- **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