Paragangliomas of the Head and Neck

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http://mingaonline.uach.cl/fbpe/img/cuadcir/v23n1/art06-figura01.jpg

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Objectives

• Paragangliomas in general
  – Features, radiology, histology, treatment

• A focus on carotid body tumors

• Discussion of other head and neck paragangliomas
Differential Diagnosis of Neck Mass

- Regional node metastasis (SCC) vs distant metastasis
- Lymphoma
- Skin cancer
- Congenital → thyroglossal duct/branchial cleft cyst
- Salivary gland tumor → parotid, submandibular
- Thyroid carcinoma and goiter
- Vascular → Glomus tumor, aneurysm
- Infection → Strep/Staph, EBV, TB, HIV, Bartonella
- Benign neoplasm → lipoma, hemangioma, fibroma
What is a Paraganglioma?

- Neuroendocrine neoplasm in the Autonomic Nervous System
  - neural crest cell tumor outside of the adrenal medulla
  - NC cells function as chemoreceptors

- Majority of paragangliomas found in body are from the Sympathetic Nervous System (e.g. pheochromocytoma)
  - Those found in head and neck are from the Parasympathetic Nervous System

- Head and neck locations: vagus, middle ear, carotid body, nose, orbit, thyroid and larynx (rare)

- Adrenal medulla has highest concentration of NC cells, thus called “para” ganglioma b/c outside of adrenal
- Location: Along the track of the Vagus nerve, Middle Ear (glomus jugulare, glomus tympanicum)
Epidemiology of Paragangliomas

- 90% arise from the adrenals (pheochromocytoma)
  - 8.5% in abdomen
- 0.3% found in the head and neck
- Tends to occur in middle age adults
- More common in females

Pellitteria et al 2003
Head and Neck Paragangliomas

- Carotid body tumors $\rightarrow$ 60%
- Jugulotympanic tumors $\rightarrow$ 2nd most common
- Glomus vagale $\rightarrow$ only 5% (nodose ganglion)
- Malignancy
  - 6-16% of carotid body tumors
  - 4% of jugulotympanic tumors
- Other locations
  - Larynx, sinonasal, orbit

Pellitteria et al 2003
Familial Paragangliomas

- 5-10% of paragangliomas are familial with **autosomal dominant** inheritance pattern
- Tend to be:
  - Multiplicity (78-87% of familial cases, 10% sporadic)
  - Bilateral
  - Earlier age of onset
- Gene ➔ PGL1 at 11q23 locus; also PGL2 and 3 (may test for these; also known as SDH gene)
- Indium pentetretotide testing ➔ labels somatostatin receptors
  - Helpful when suspecting multiplicity in familial case
- May be found in MEN syndrome (Type II)

Pellitteria et al 2003 - Pg 565
Types of Head & Neck Paraganglioma

Carotid body

Glomus jugulare

Glomus tympanicum

Glomus vagale
Gross appearance of Paragangliomas

• Well-circumscribed
• Firm to rubbery consistency
• Highly vascular → readily bleed in surgery
• Color
  – Jugulotympanic → deep red when viewing through tympanic membrane
  – Carotid body → deep red to brown-grey

Pellitteria et al 2003 – page 565
Histology of Paragangliomas

- Chief cells = primary cell (a.k.a Type I cells) → may contain catecholamine bound granules
- Type II cells are structural support (fibrovascular)
  - a.k.a sustenacular cells
  - Zellballen = nest of cells

*Cannot use histology to determine malignancy → Why

- Nuclear polymorphism, neurovascular invasion, high rate of mitosis, and necrosis often present in both benign and malignant paragangliomas

- Cell balls = nest of cells
- Malignancy only detected once tumor metastasizes to regional nodes or distantly
Zellballen - Nest of Cells

- Cell balls or nests demonstrated here
- Close association with blood vessel
Chief cells
Support cells
Fibrovascular septae and granular cytoplasm

Immunohistochemical Staining

• Chief cells (Type I)
  – (+) for chromogranin, synaptophysin, neuron specific enolase, serotonin, & neurofilament
  – (-) for S100

• Sustenacular cells (Type II)
  – (+) for S100
A. H and E stain → Nest-like pattern

B. Neuroendocrine-type chromatin and pale-to-pink granular cytoplasm

C. (+) immunostaining for chromogranin

D. Sustenacular (II) cells (+) for S100 stain
Labs- Paragangliomas

• May have (+) urinary catecholamines if functional

Note: most paragangliomas in the head and neck are non-functional
- Only 2-4% present with hypertension or tachycardia

• Unlike paragangliomas in the rest of the body, H and N paragangliomas unlikely to be functional
• If pt asymptomatic, screening for these is only indicated if the tumor is suspected to be familial or if pt had previous carotid body tumor

http://www.cvpharmacology.com/norepinephrine.htm
Radiology of Paragangliomas

• Preferred diagnostic modality

• Combination of multiple modalities
  – Ultrasound
  – CT with contrast
  – MRI with contrast
  – Angiography
Other Workup

Biopsy?

....Usually not necessary
Treatment Considerations

- **Surgical resection** is generally preferred treatment modality

- If (+) urine catecholamines ➔ look for pheochromocytoma too
Preoperative embolization

• Goal = reduce intra-operative bleeding

• Controversial as being beneficial but commonly used

• Prefer surgical resection within 2 days of embolization ➔ prevent regrowth of vessels

• May lead to scarring obscuring tissue planes

Notes: If tumor is invading the carotid wall and lots of operative bleeding expected, can work with vascular surgery to place a balloon shunt in the internal carotid.
Non-surgical treatment

Radiotherapy
- Primarily use when not a surgical candidate
  - Elderly patients with multiple comorbidities
- Controversial
- Also in patients with multiple paragangliomas
  - E.g patient w/ carotid body and glomus vagale tumor
  - Recurrent tumors

Notes: Also, observation is a viable option if the patient is not a surgical candidate and tumor appears to be stable
Radiation vs Surgery

• Depends on...
  – who you ask
    • surgeon vs radiation oncologist
  – tumor characteristics
    • Size, location, multicentricity
  – Patient’s health status
    • RT for the sicker patients

NOTES: Many of the articles written by rad/onc strongly advocate considering radiation over surgery
Role of Radiation

• “fractionated RT offers high probability of tumor control with minimal risk to patients...” - Hinerman et al. 2008
  – Studied RT on 121 paragangliomas; control rates of 96%
  – Mean follow-up time 10.6 yrs and median f/u of 8.5 yrs
  – Philosophy is absence of tumor progression = cure
  – Recommend surgery mainly for healthier patients with smaller tumors that have low risk for cranial nerve sacrifice

• Sheehan et al. (Laryngoscope 2010) demonstrated “Gamma Knife Radiosurgery is an effective treatment for patients with glomus jugulare tumors...”
  – Tumor control in 12/15 patients (mean follow up of approx 4 yrs)

Notes: Hinerman ➔ Local control defined as absence of tumor progression or partial/complete regression
Malignancy of Paragangliomas

- Offergeld et al 2012
  - “only occurrence of lymph node and/or distant metastases are the ultimate criteria for malignancy”
  - “tumor invasion into adjacent structures including bone does not indicate biologic malignancy”

**Lymph node/distant organ mets** = overall consensus on malignancy definer

- 4-15% head & neck paragangliomas believed to be malignant
  - Highest rate in *vagal tumors (16-19%)*

- Moskovic 2010
  - Somewhat frustrating b/c how will you know a tumor is malignant if still defined to the neck
Other Malignancy Predictors?

- Chapman et al. (2010) studied clinical, histopathologic and radiologic features
- 84 patients; 77 benign, 7 malignant (8%)
- Of 7 malignant, 6 were considered based on lymph node metastasis
  - 1 based on very aggressive local features
  - 6/7 were carotid body tumors, 1/7 was glomus vagale
### Factors Determining Malignancy

**Table 3**

Pathologic characteristics of malignant paragangliomas

<table>
<thead>
<tr>
<th>Patient</th>
<th>Metastasis</th>
<th>Vascular invasion</th>
<th>Lymphatic invasion</th>
<th>Perineural invasion</th>
<th>Poorly circumscribed/capsular invasion</th>
<th>Dense sheet-like growth</th>
<th>Diffuse nuclear atypia</th>
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<tr>
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<td>No</td>
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</tbody>
</table>

N/A, pathologic data not available for review.

*Unable to obtain and review original slides; based on pathology report only.

†Aggressive malignant features, but no metastasis.
Lymph node invasion

Perineural invasion & tumor encircling nerve

Figure 1  Lymph node with metastatic tumor within the subcapsular space. (Hematoxylin-eosin; original magnification: $\times200$.)

Chapman et al 2010
Predictors of Malignancy

Found mainly clinical factors help predict malignancy

- Pain
  - 5/7 (71%) malignant; 5/77 (8%) benign → p<0.0001
- Enlarging cervical mass in past 12 months
  - 100% of malignant, 33% of benign
- Younger age of presentation (p<0.02)
  - Benign → 54 ± 16 y.o
  - Malignant → 40 ± 12 y.o

Chapman et al 2010
Carotid Body Tumor
Carotid Body Anatomy

• Reddish brown-tan ovoid structure
• Location ➔ Posterior & medial at carotid bifurcation
• Found in adventitia of common carotid
• Fed primarily by small feeder blood vessels from the ascending pharyngeal artery off the external carotid
• 3-5 mm in diameter
• Innervation = Herring’s nerve (branch of IX)

CB is on average larger in people of higher altitudes; related to lower O2 content?
Sensory afferent to medulla
Physiology

Carotid body = chemoreceptor
- Glomus cells →
- Senses pH, CO2, and O2 levels
- Stimulators → acidemia, hypercarbia, hypoxia
  - Result is increased HR, BP, RR, and TV*

- This sympathetic nervous system stimulation may relate to why people w/ OSA have systemic hypertension
- Hypoxia thought to be the most potent stimulant of CB
Parapharyngeal space

**Carotid body tumor:**

Most common tumor of the post-styloid parapharyngeal space

http://www.advancedonc.com/parapharyngeal-space-tumors/
Carotid Body Tumor - Signs and Symptoms

- Usually asymptomatic (painless swelling)
- 10% have cranial nerve symptoms
  - IX, X, XI, XII, and sympathetic chain
    - can result in pain, hoarseness, dysphagia, shoulder drop, tongue paresis & Horner’s syndrome
  * Vagus most commonly affected
- Could be functional → HTN, diaphoresis, palpitations

• Asymptomatic → may grow for years before being noticed (doubling time is about 7yrs at 0.83 cm/yr)
• If functional, sx’s of pheochromocytoma (palpitations, sweating, paroxysmal hypertension)
• Vagus affected → in the carotid sheath
Signs and Symptoms

• Slowly enlarging, non-tender neck mass
  – Anterior to sternocleidomastoid mm at level of hyoid
  – Compression may cause dysphagia itself

• May produce a carotid bruit

• Laterally mobile but fixed in cephalic-caudal direction
  – Known as (+) Fontaine’s sign

Offergeld et al 2012
Duplex U/S
- hypervascular mass next to carotids (also see carotid plaques)

https://www.vascularweb.org/APDVS/Pages/CerebrovascularModule.aspx
Radiology- CT with contrast

http://radiopaedia.org/images/392640

- CT description same as the US
- R arrows are two different carotids

http://ispub.com/IJRA/10/2/3794
MRI (gadolinium)

Said to have a “salt and pepper” appearance on T2
MRI Features

T1: isointense to muscle
T2: hyperintense

- Avid enhancement on post-contrast imaging

“Salt and pepper”
- Pepper: low signal contrast flow voids
- Salt: high signal foci of hemorrhage and/or slow flow

Wienke 2009
Angiography

• Used for definitive diagnosis

• Often not necessary for initial diagnosis due to use of MRI detection
  – More historical

• Helps delineates feeder blood vessels and means of embolization

• Best means to show Lyre’s sign
Lyre’s Sign = splaying of external and internal carotid arteries by tumor

http://www.ghorayeb.com/carotidbodytumor.html
Angiography

Carotid angiography

Other → MRA

Bilateral tumors

- much higher incidence in inherited form

Other Imaging Modalities

MRA 3D reconstruction

http://dualibra.com/wp-content/uploads/2012/04/037800~1/Part%2015.%20Endocrinology%20and%20Metabolism/Section%201.%20Endocrinology/337.htm
Intra-op

http://www.ghorayeb.com/carotidbodytumor.html
Intra-op
Intraop

Steps of the procedure - Meyers Ch 76

1. Incision → place transversely hidden in skin crease
   - Alternatively vertical incision over SCM (carotid endarterectomy type)
2. Dissect through subcutaneous tissue with skin flaps elevated down to SCM
3. Find the sternocleidomastoid muscle, retract posteriorly to expose carotid sheath
4. Dissect IJV off the carotid artery
5. Identify and dissect out CN X and CN XII and place vessel loops around them
6. Identify the common carotid; skeletonize it and place vessel loop around it
7. Start subadventitial dissection on the CC, directed superiorly up to bifurcation
   - This begins just superior to commonly seen venous plexus overlying CC
8. Carry subadventitial dissection superiorly onto External Carotid
   - Recommend clamping, cutting and suture ligating ECA to aid in deeper dissection, especially for larger tumors extending to prevertebral fascia
9. Dissect tumor off the Internal Carotid, remove, and then close

- Venous plexus located about half the distance between clavicle and carotid bifurcation
- Subadventitial dissection need because carotid body naturally lies in this plane at bifurcation
Intraop Considerations

• Greater nerve/arterial injury risk with increased size
  - do no paralyze patient ➔ monitor cranial nerves

• ID of cranial nerves ➔ IX, X, XI, XII
  - Superior laryngeal N. (X) often on posterior edge of tumor (most commonly injured/sacrifice)

• Bilateral ➔ only excise one at a time

  - Superior laryngeal nerve the most commonly injured nerve during dissection
  - Don’t do bilateral at same time b/c of potential hemodynamic instability after both carotid sinuses are disrupted (baroreflex failure syndrome) and to ensure preservation of at least one vagus nerve
Surgical Classification- Shamblin

- **Purpose**: Predict morbidity in highly vascular lesions

- **Class I**: minimally attached to vessels and easily removed → complete resection, minimal risk

- **Class II**: partially surround vessels, more adherent to adventia → more risk

- **Class III**: adherent to entire surface of carotid bifurcation → resection impossible

Wieneke 2009
Offergeld 2012
Shamblin

Type I

Type II

Type III

ICA
ECA
Sup. laryn. n
Sup. laryngeal n.
N. X
N. XII
N. X
Shamblin Class I

- T2-weighted axial MRI
- Splaying but not encasing
- Complete resection with minimized risk

Offergeld et al. 2012
Class II

- T2-weighted axial MRI
- Partially encasing carotid branches
- Significantly more risk in resection

Offergeld et al 2012
Shamblin Class III

- CT angiography
- Complete encasement of carotids
- Highly risky resection → requires blood flow interruption for vascular reconstruction *

Offergeld et al. 2012

* May require vascular surg to place intra-op carotid balloon/stent
Surgery complications

1. Bleeding → Carotids, IJV
2. CN injury → Mainly X, XI, XII
   - X → aspiration, hoarseness
   - XI → shoulder pain/weakness
   - XII → speech and swallowing problems
3. Cervical sympathetic chain
   - Ipsilateral Horner’s
   - First bite syndrome (parotid pain)
4. Stroke → due to embolism or decreased blood flow

- CN X → Superior laryngeal nerve dysfunction will cause aspiration (decreased sensation of supraglottic larynx) and vocal dysfunction (cricothyroid mm)
- Horners → ptosis, miosis, anhidrosis; sympathetic cervical chain lies posterior to carotid sheath
- First Bite → lack of SNS leads overwhelming myoepithelial stimulation by Parasympathetic NS
Other Paragangliomas

- Jugulotympanic tumors
  - Glomus tympanicum
  - Glomus jugulare

- Glomus vagale
Jugulotympanic tumors

Glomus Tympanicum

http://american-hearing.org/disorders/glomus-tumors/
Glomus tympanicum - deep red

Brown’s sign → blanching of the TM on pneumatic otoscopy

- Owing to the factor that it is a hypervascular tumor
Glomus tympanicum

• Confined to middle ear
  – Arise along Jacobson’s nerve (inferior tympanic nervebranch of CN IX)
  – Fisch Type A
• Most common tumor of the middle ear
• Presenting symptoms
  – 80% ➔ pulsatile tinnitus
  – 60% ➔ unilateral hearing loss
• Treatment ➔ tympanotomy (smaller lesions) or mastoidectomy (larger lesions)

Pellitteria et al 2003- 566
Alaani 2009-
Glomus tympanicum

CT Temporal Bone
- round mass on cochlear promontory
Glomus Jugulare

- Arise from paraganglia in/around jugular bulb
  - Along Jacobson’s (CN IX) or Arnold’s nerve (X)
- Most common symptoms → tinnitus/hearing loss
- CT distinguishes tympanicum from jugulare
  - MRI defines extent of disease
- Tendency towards multiplicity of tumors
  - 10% in non-familial and 50% in familial cases
- 30% have CN deficits

Cummings 1916
## Glassock-Jackson Classification

<table>
<thead>
<tr>
<th>Table 1</th>
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<tbody>
<tr>
<td>Glassock-Jackson Classification of Glomus Jugulare Tumors</td>
</tr>
</tbody>
</table>

- Small tumor involving jugular bulb, middle ear, and mastoid (Class I)
- Tumor extending under internal auditory canal; may have intracranial extension (Class II)
- Tumor extending into petrous apex; may have intracranial extension (Class III)
- Tumor extending beyond petrous apex into clivus or infratemporal fossa; may have intracranial extension (Class IV)

- Classified by location in temporal bone
- Less commonly used

http://www.physicianspractice.com/review-article/paraganglioma-potentially-challenging-tumor
Fisch Classification of Temporal Bone Paraganglioma

- Based on extension of the tumor to surrounding anatomic structures and is closely related to mortality and morbidity.

- **Type A** - Tumor limited to the **middle ear cleft (glomus tympanicum)**

- **Type B** - Tumor limited to the **tympanomastoid area** with no infralabyrinthine compartment involvement

- **Type C** - Tumor involving the **infralabyrinthine compartment** of the temporal bone and **extending into the petrous apex including portions of carotid canal**

- **Type D1** - Tumor with an **intracranial extension less than 2 cm** in diameter
- **Type D2** - Tumor with an **intracranial extension greater than 2 cm** in diameter

C- subclasses C1-3 based on involvement in carotid canal
### De La Cruz Glomus Tumor Classification with Associated Surgical Approach

<table>
<thead>
<tr>
<th>Classification</th>
<th>Surgical Approach</th>
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<tbody>
<tr>
<td>Tympanic</td>
<td>Transcanal</td>
</tr>
<tr>
<td>Tympanomastoid</td>
<td>Mastoid—extended facial ridge</td>
</tr>
<tr>
<td>Jugular bulb</td>
<td>Mastoid-neck (possible limited facial nerve rerouting)</td>
</tr>
<tr>
<td>Carotid artery</td>
<td>Infratemporal fossa ± subtemporal</td>
</tr>
<tr>
<td>Transdural</td>
<td>Infratemporal fossa/intracranial</td>
</tr>
<tr>
<td>Craniocervical</td>
<td>Transcondylar</td>
</tr>
<tr>
<td>Vagal</td>
<td>Cervical</td>
</tr>
</tbody>
</table>

Ballenger’s Otorhinolaryngology (2003) Ch 25
Jugular Bulb relation to Middle Ear

Middle Ear Viewed from Within
(1) Sigmoid Sinus
(2) Jugular Bulb *
(3) Hypotympanum
(4) Internal Carotid Artery *
(5) Eustachian Tube
(6) Descending Portion of Facial Nerve
(7) Aditus Ad Antrum
(8) Chorda Tympani Nerve
(9) Malleus Handle
(10) Epitympanic Space
(11) Head of Malleus
(12) Inner Aspect of Tympanic Membrane
(13) Tensor Tympanic Muscle

http://www.rcsullivan.com/www/wyeth-ayerst/02f.htm
Netter- Skull base from above

- Foramen spinosum
  - Middle meningeal artery and vein
  - Meningeal branch of mandible
- Sphenoid emissary foramen (Vesalius) (inconstant)
- Foramen lacerum
- Carotid canal for
  - Internal carotid artery
  - Internal carotid nerve plexus
- Hiatus for
  - Lesser petrosal nerve
- Hiatus for
  - Greater petrosal nerve
- Internal acoustic meatus
  - Facial nerve (VII)
  - Vestibulocochlear nerve (VIII)
  - Labyrinthine artery
- External opening of vestibular aqueduct
  - Endolymphatic duct
- Mastoid foramen (inconstant)
  - Emissary vein (and occasional branch of occipital artery)
- Jugular foramen
  - Inferior petrosal sinus
  - Glossopharyngeal nerve (IX)
  - Vagus nerve (X)
  - Accessory nerve (XI)
  - Sigmoid sinus
  - Posterior meningeal artery
Netter: Skull base from below

Carotid Canal

Jugular Foramen (IX, X, XI, IJV)

Close relationship of two structures
Glomus tumor

Glomus Jugulare

Glomus Tympanicum

Aural polyp
Glomus Vagale

- Arise from the inferior (nodose), middle, or superior vagal ganglia
- Least common type $< 5\%$ of head and neck paragangliomas
- Present as asymptomatic neck mass behind angle of mandible
  - Displaces internal carotid anteromedially
- Most likely of the head and neck paragangliomas to be malignant

Majority originate in the nodose ganglia, approx 2 cm below jugular foramen
Glomus vagale

- tend to be higher than CB tumors
- Situated between IJV and ICA

Figure 3: Vascular Involvement—(A) Diagrammatic illustration of the relationship between a carotid body tumor and vagal paraganglioma and the large vessels of the neck. (B) Left carotid angiogram in a 50-year-old woman with a left vagal paraganglioma and carotid body tumor following embolization of the external carotid branches (asterisk). Note the displacement of the vessels related to the vagal (arrows) and carotid (arrowheads) tumors.
Glomus Vagale

- Intracranial extension in 22% of cases
  - Primary cause of death
- Up to 50% manifest with CN deficit
  - IX → dysphagia
  - X → hoarseness, aspiration
  - XI → shoulder drop
  - XII → aspiration and hemiatrophy of tongue
Dale et al. 1993

Type III is the classic “Dumbbell” tumor
Vagale Surgical Treatment

• Technique often requires lateral cervical incision with possible mandibulotomy for better exposure
  – Neurosurgery involved for intracranial extension tumors

• Involved vagus nerve usually sacrificed with associated morbidity
  – Dysphagia and aspiration expected complication

• Unilateral paralyzed cord
  – Vocal medialization via silastic thyroplasty commonly used post op
  – Some advocate thyroplasty at time of initial resection

• Greatest risk to aspiration found with concurrent CN X and XII injury
Glomus tympanicum & jugulare

Glomus jugulare

Glomus vagale

Carotid body
Summary of Paragangliomas

• Slow growing and usually benign
• Carotid body most common, then jugulotympanic, then vagal least common
  – Vagal most likely to be malignant with most cranial nerve morbidity
• Surgery preferred in smaller masses and healthier patients
  – some role for RT as alternative or adjunct
Dr. Susan McCammon: Thank you, Dr. Yantis. That was a terrific talk, a great summary of paragangliomata. Dr. Resto has asked me to comment on my experience and training at Memorial Sloan-Kettering where typically we did not embolize these with the thought that there was a transient inflammatory response that would make it more difficult to resect them. I think the incidence and morbidity of the cranial neuropathies that can result after surgery are really significant, and most of the people that I worked with are tending more toward radiating these now.

The larger ones are more invasive ones really only in early stage carotid body tumor - a one or two cm. one I would really resect. But it depends a lot on the patient’s age, functional status, but to take a young person and to give them a high vagus injury, especially if there’s a risk of twelfth nerve injury, as well virtually guarantees almost chronic and almost insurmountable aspiration, not just because of the cord paralysis, but also because of the loss of sensation plus the loss of tongue mobility. So that’s the thing I would add: the loss of sensation along with the loss of mobility really exacerbates the cord paralysis. Now I’ll turn it over to Dr. Resto, your Mentor.

Dr. Vicente Resto: Thank you, Dr. McCammon. Dr. Yantis, that was a great talk. So I guess over all, at a high level, I would echo what Dr. McCammon just said. I think that the experience with the often serious deficits that result from surgical intervention really has driven folks to observe these. Many times one can follow them with imaging, and if there is a tumor that is producing minimal symptoms, which is often the case, often they are found incidentally. You can follow them prospectively and show that there is little or no growth, observation is really at the top of the list. Traditionally resection was rooted in the fact that there has been this reported low rate of malignant transformation, but the reality is that the rate is so low that when put in context with the adverse effects of resection really have made over the history, now a strong argument for observation and or radiation - specifically if there is an elderly patient. With a growing tumor, radiation can be useful in arresting growth. It does not necessarily get rid of the tumor. (continued – next page)
Dr. Vicente Resto (continued):

Two points: if you choose engage in surgery of these tumors, one is to recognize that you will have significant cranial nerve defects particularly in the vagal version of these paragangliomas. I just don’t know a whole lot of people who will go up there and not have at least transient and very often permanent cranial nerve injury. The other is that the surgically approach is really all about management of vascular structures. That’s where certain maneuvers such as early take of the external carotid at its root when handling a carotid body tumor is a common technique that comes in very useful. Recognition that the ascending pharyngeal branch will not uncommonly come out of the bifurcation itself. So, one of the more common injuries is when you’re trying to dissect out the carotid body tumor from the base of the bifurcation is to end up with a rather sizeable rent in that area. Having vascular surgery, or some expertise in patching or repairing the carotid at that site, it might be mandatory when handling some of these tumors.

When talking about the vagal tumors again, it’s not rare at all that they engage the jugular foramen, and at that the jugular vein tends to be collapsed, and then it’s not uncommon to have injury of the internal jugular at a very high location which makes it technically challenging to repair. These are all things that need to be thought of ahead of time and having a definitive plan of how to manage them. These have always been a very tantalizing set of lesions.
Sources