CAROTID BODY TUMORS

Hasan Ashkanani (PGY-1 Vascular Surgery)

Division of Vascular & Endovascular Surgery

University of Ottawa

WVES Winnipeg Vascular & Endovascular Symposium

Presenter Disclosure

Presenter: Hasan Ashkanani (PGY-1 Vascular Surgery, uOttawa)

- I have no current relationships with commercial entities

OUTLINE

- Epidemiology
- Etiology
- Anatomy
- Physiology
- Pathology
- Clinical Presentation
- Diagnosis
- Treatment
- Preoperative Preparation
- Surgical Technique: Positioning & Exposure
- Surgical Technique: Resection of Tumor
- Postoperative Care

EPIDEMIOLOGY

- Usually Dx in the 3rd-5th decades of life (earlier if +ve FHx)

- Gender predominance has not been established
  - Slightly more prevalent in women

ETIOLOGY

- Classified as
  - Sporadic: 70-80% of cases
  - Familial: 10% of cases
    - Younger age 30% are bilateral AD inheritance
  - Hyperplastic:
    - Not true neoplasms
    - Chronic hypoxia (High altitude, COPD, Cyanotic heart disease)
- Genetics
  - Mice deficient in the SDHD (succinyl dehydrogenase – subunit D) gene
  - → continued activation of the carotid body cells in response to hypoxia

ANATOMY

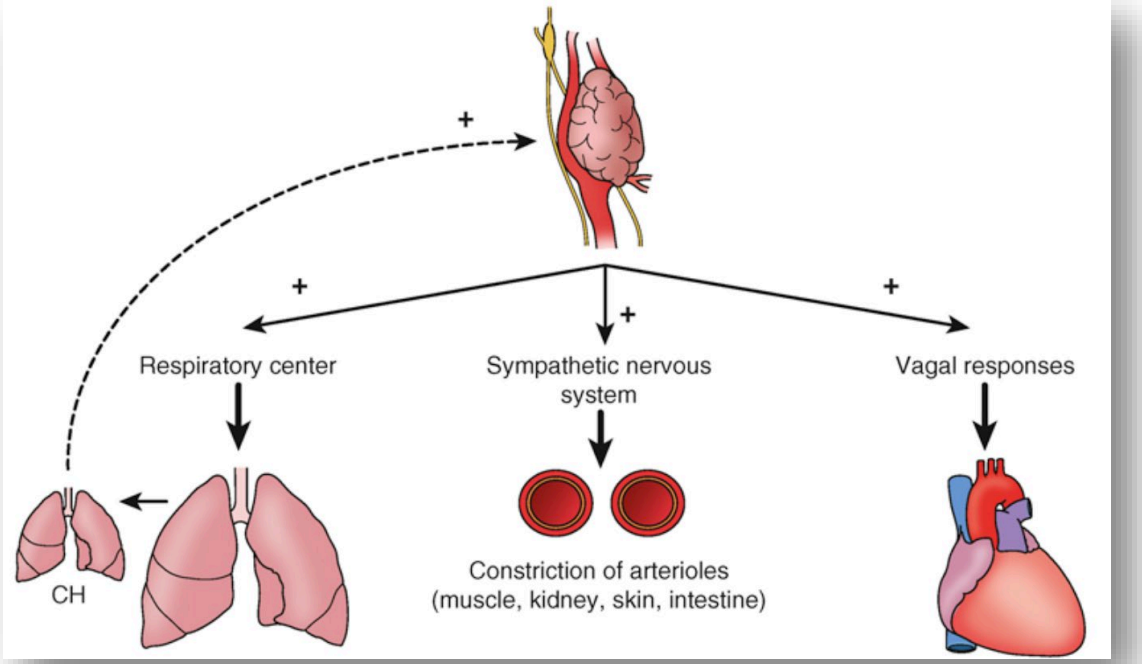
- The carotid body
  - Largest mass of chemoreceptor tissue in the body.
  - Located within the peri-adventitia of the posterior surface of the carotid bifurcation.
  - Ovoid in shape
  - ~ 5mm in its longest dimension
  - Arterial supply through = branches of the external carotid artery.
  - Venous drainage = lingual & laryngopharyngeal veins.
  - Innervation: sensory innervation from the glossopharyngeal nerve (CN 9)

EMBRYOLOGY

- Originates from both neural crest ectoderm and mesodermal tissue from the 3rd branchial arch.

- **The neural crest cells (ectoderm tissue)**
  - Migrate in close association with autonomic ganglion cells
  - Hence they are often referred to as paraganglioma cells.
  - These cells differentiate into the chemo-receptors, also known as type I glomus cells.

- **The mesoderm tissue**
  - Type II glomus cells
  - Form the rich vascular stroma
  - Support the chemoreceptor cells.



PHYSIOLOGY

- Stimulation
  - Primarily by the pO_2 (Low pO_2)
  - Lesser degree the pCO_2 & arterial pH (High pCO_2 & Low pH)

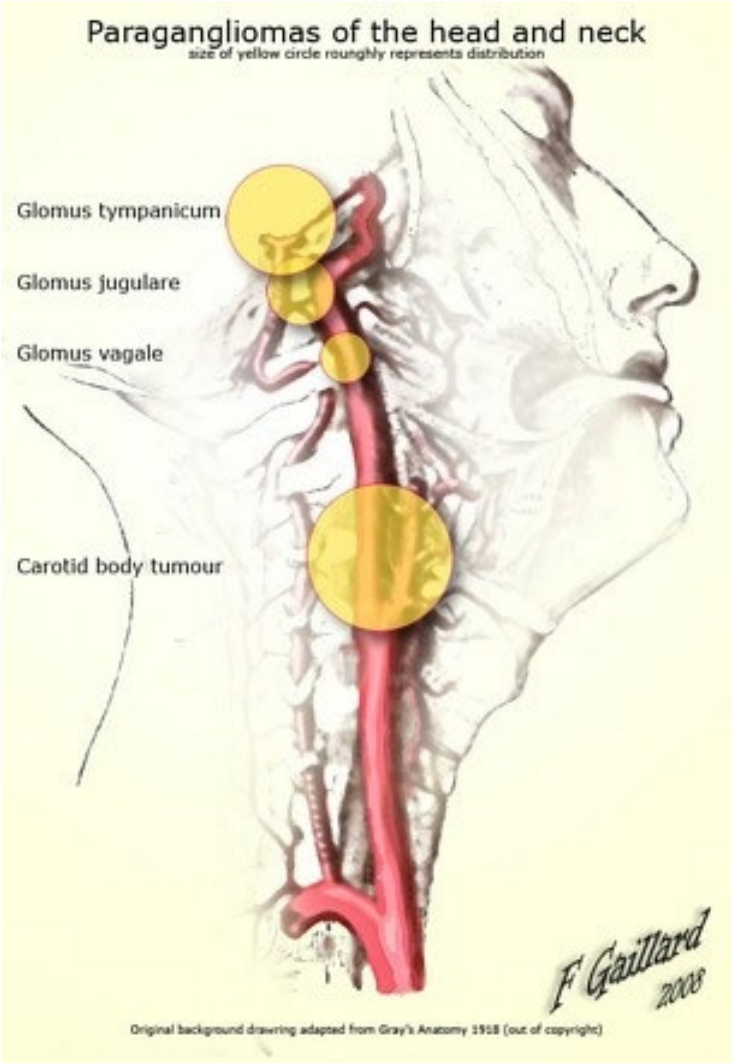
- Response
  - Type I glomus cells release neurotransmitters
  - → CN 9 nerve fibers.
  - → Medulla oblongata
  - → Cardiopulmonary centers in the brain
  - → increases in RR, TV, BP via vasoconstriction

PATHOLOGY

- CBTs AKA:
  - carotid chemodectomas
  - carotid paragangliomas
  - glomus tumors.

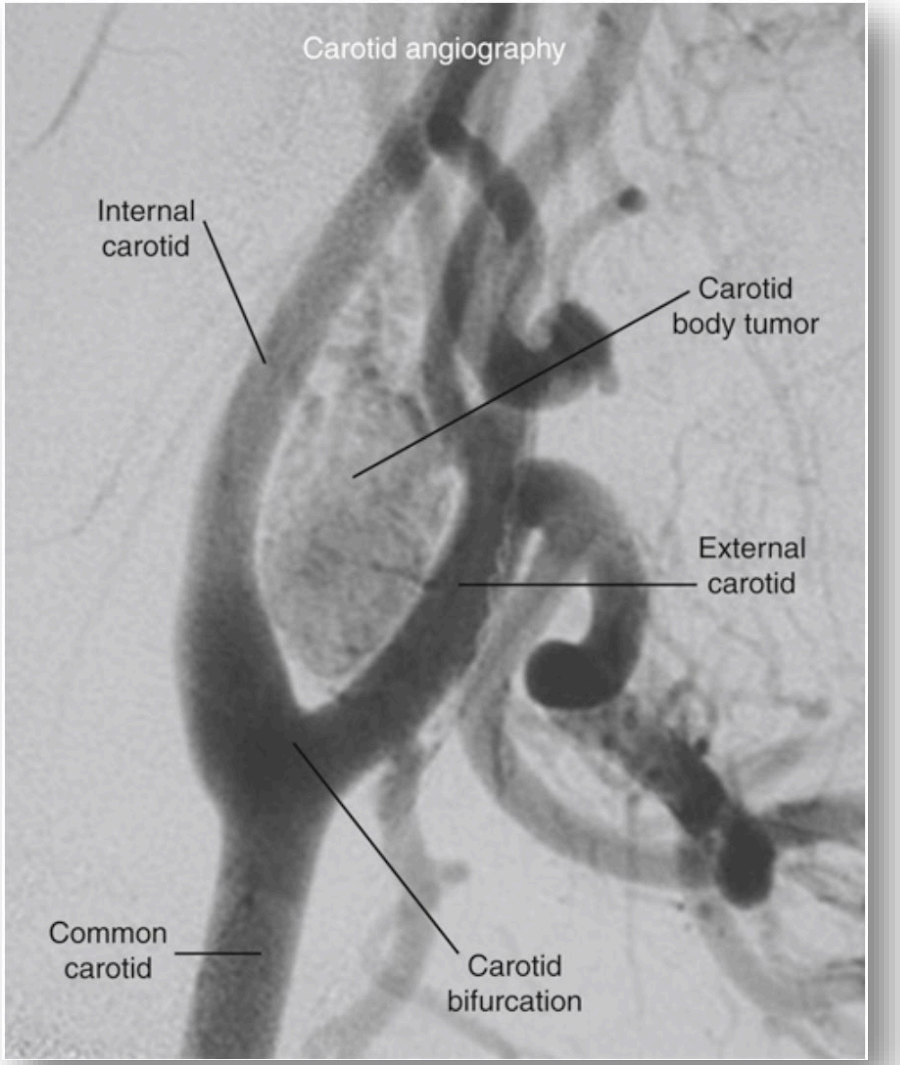
- Neoplastic growths of the chemoreceptive tissue.

- Belong to the family of paragangliomas
  - Tumors arising from the autonomic ganglion chain from the head - pelvis.
  - <u>Most common paraganglioma in the H&N.</u>



PATHOLOGY

- Macroscopically
  - CBTs resemble normal carotid body tissue.
  - Reddish brown, rubbery & well-circumscribed
  - Highly vascular
  - Can invade the adventitia of adjacent carotid vessels.
  - Splaying of ICA & ECA = "Lyre sign"



PATHOLOGY

- Microscopically:
  - CBTs tend to resemble normal carotid body architecture
  - Well-differentiated benign appearance.
  - Very rarely does histology demonstrate degenerative malignant characteristics
  - Although capable of neuro-endocrine secretion, it’s usually non-functional

- MCQ 1: What is the main sensory innervation of the carotid body:
    - Glossopharyngeal nerve
    - Main vagus nerve
    - Superior laryngeal nerve
    - Cervical sympathetic nerves
    - Hypoglossal nerve

- MCQ 1: What is the main sensory innervation of the carotid body:
  - Glossopharyngeal nerve
  - Main vagus nerve
  - Superior laryngeal nerve
  - Cervical sympathetic nerves
  - Hypoglossal nerve

PATHOLOGY

- Most CBTs are benign.
  - Malignancy is based on clinical behavior instead of histology.
  - Even benign tumors are capable of aggressive invasion into adjacent structures, as well as nodal and distal mets
  - Local growth may include adherence or encasement of neuro & vascular structures

CLINICAL PRESENTATION

- Asymptomatic neck masses (most common presentation)

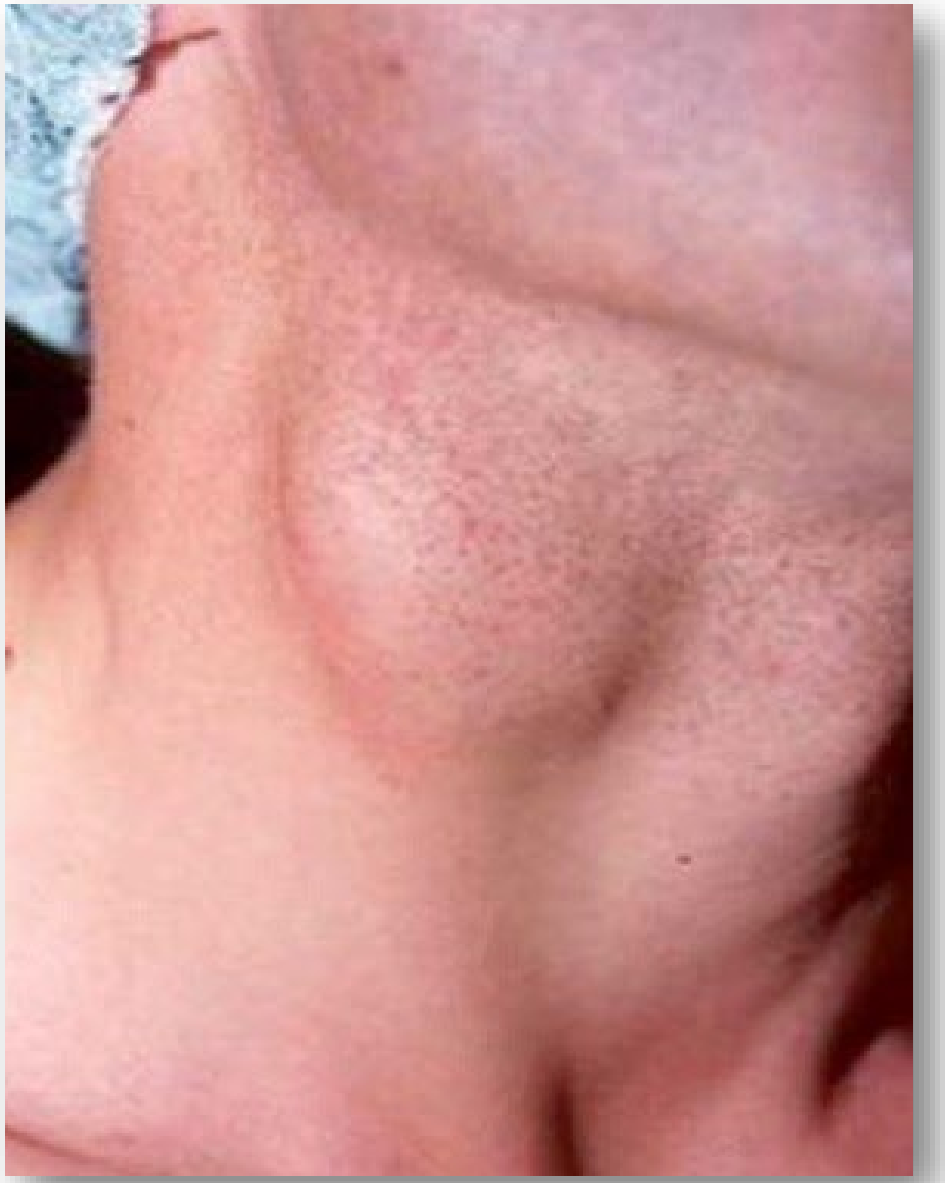
- Compression or local invasion symptoms (Large tumors)
  - Localized tenderness, fullness, numbness
  - Dysphagia, hoarseness, chronic cough, and tinnitus.

- Family Hx

- On exam:
  - Firm, smooth, and lobulated
  - Mobile laterally but NOT up & down (Fontaine sign)
  - Audible bruit over the tumor (30-40% of pts)

- Tumors rarely produce cranial nerve dysfunction,
  - Vagal (CN 10), hypoglossal (CN 12), & cervical sympathetics (Horner's)

- Almost never functional



DIAGNOSIS

- Hx & PE

- Neck mass DDx
  - Congenital lesions (vascular malformations, branchial cleft cysts, hygromas),
  - Inflammatory disorders (chronic lymphadenitis, reactive lymphadenopathy),
  - Infectious lymphadenopathy
  - Benign lesions (lipomas, cysts, parotid & salivary tumors),
  - Malignancies (metastatic head & neck cancer, lymphoma).

Diagnosis - Tests

- Duplex US: 1st line.
  - Excellent anatomic detail (vascularity, size, vessel encasement, & atherosclerosis)
  - Less expensive, no radiation.
  - Downsides
    - Inability to clearly visualize the proximal (chest) & distal extent (intracranial) of the carotid artery
    - Not as sensitive for small lesions.
    - Highly operator dependent.

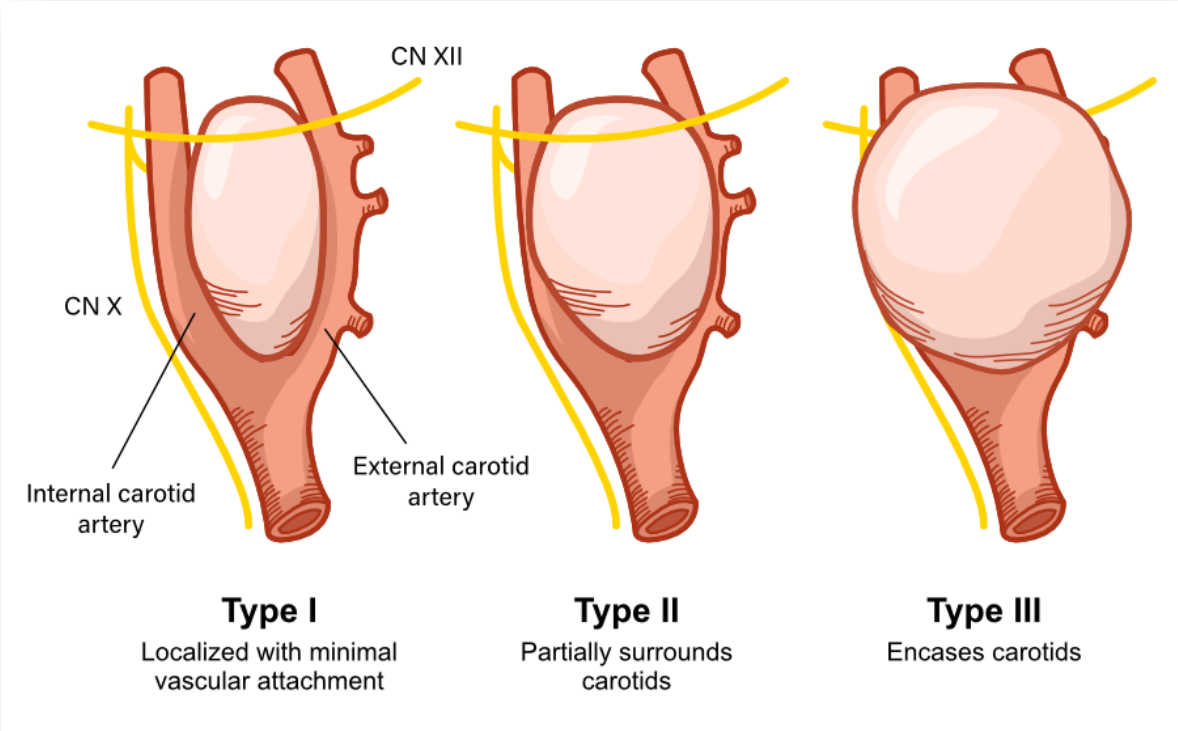
- CTA/MRA
  - Shows proximal and distal extents of tumor
  - Radiation & contrast exposure in CTA
  - Time consuming & contrast exposure in MRA

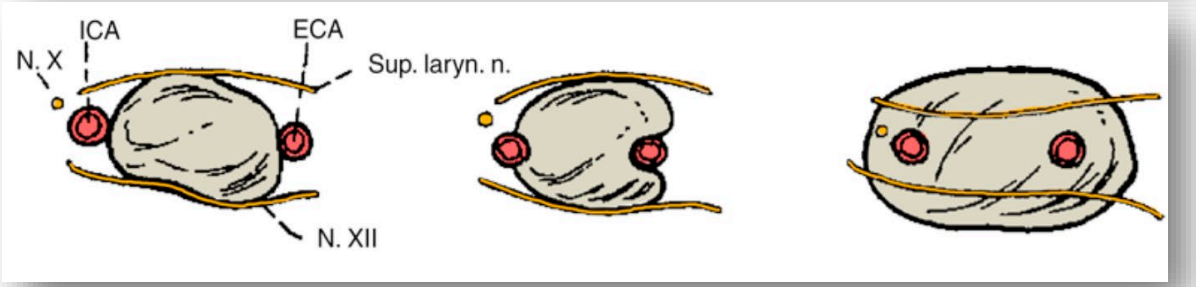
- Conventional angiography
  - Traditionally the gold standard
  - Now largely replaced by US Duplex & CTA/MRA
  - Main use is to check for feeding vessels & pre-op embolization (if > 5cm OR Shamblin 2-3)
  - Risks: high cost, access complications, wound complications, distal embolization w/ stroke , contrast

- Percutaneous needle biopsies and incisional biopsies contraindicated ! (highly vascular tumors)

DIAGNOSIS – SHAMBLIN CLASSIFICATION

- Shamblin 1:
  - Localized to carotid bifurcarion
- Shamblin 2:
  - Partially surrounding ICA/ECA
- Shamblin 3:
  - Completely surrounding ICA/ECA





TREATMENT

- Surgical resection is the mainstay of treatment for CBTs.
  - In the absence of prohibitive comorbidities, perioperative risks, or limited life expectancy.
  - Early resection is better (as soon as the diagnosis is made)
    - Smaller tumors are easier to remove
    - Future local invasion will complicate resection

- Radiation therapy
  - CBTs are traditionally NOT radiosensitive
  - Radiation is used for suppression.
  - Role is controversial
  - May render surgery more difficult due to fibrosis.
  - Usually reserved for
    - Poor operative candidates
    - Bulky, unresectable tumors
    - Recurrent tumors.

- No effective chemotherapy treatment !

PREOPERATIVE PREPARATION

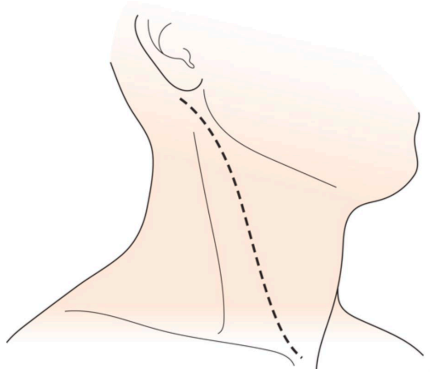
- Optimize general medical conditions
- Imaging (tumour size, proximal and distal extents, other cervical tumors)
- Document pre-existing CN involvement & neuro deficits
- If endocrine dysfunction or hormone imbalance is suspected (B/L or familial disease)
  - 24 hrs urinary catecholamines
- Pre-op ECA embolization: Selective cath of ECA and embolizing feeding branches to CBT
  - Usually for tumors > 5cm or Shamblin 2-3
  - Evidence is controversial. Mostly for reduction of total blood loss NOT operative time, stroke, death
- +ve workup for other active synchronous lesions (e.g. adrenal pheochromocytomas) → Tx FIRST

SURGICAL TECHNIQUE - MAIN CONSIDERATIONS

- Tumor volume & distance from skull base
- Massive hemorrhage risk (highly vascular lesion)
- Adherence to neurovascular structures (esp cranial nerves)
  - Meticulous dissection
  - Use of bipolar forceps
- Choice of anesthesia: GA preferred
  - Better airway management
  - Longer OR than carotid endart
  - Less movement esp with higher lesions
- Cerebral monitoring
  - EEG
  - Shunting
    - Selective: if ischemic changes on EEG
    - ? Routine: idea is to divert blood away from ECA (making dissection easier)

POSITIONING, EXPOSURE, & MARKING – LIKE A CEA

- Supine
- Head rotated to the contralateral side.
- +/- Shoulder roll may be placed to extend the neck.



- The head of the table can be elevated 10 to 15 degrees
  - reduce venous pressures
  - reduce incisional blood loss.
- Longitudinal incision along the anterior border of the SCM (variations exist)
  - Centered over the carotid bifurcation
  - +/- proximally toward the sternal notch and distally toward the mastoid process
  - Planning for distal exposure technique (11 techniques in Rutherford !)
    - Nasotracheal intubation
    - Curve the incision behind the ear to the mastoid process
    - Detachment of the posterior belly of the digastric muscle from the mastoid process
    - Division of the stylohyoid muscle with or without the styloid process
    - Subluxation or division of the mandible
    - Etc.

Large aneurysms and aneurysms extending to the distal ICA can be technically challenging. Nasotracheal intubation should be considered because it allows complete closure of the jaw which opens the space around the distal ICA. Several additional techniques can be used to improve distal operative exposure, including:

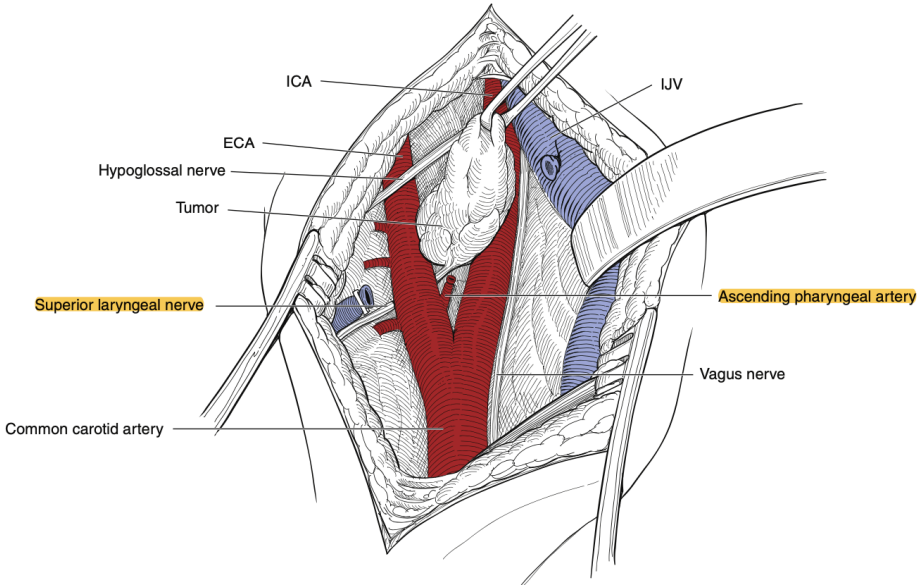
1. The first step in obtaining distal exposure should always be extension of the incision to curve in a posterior fashion behind the ear to the mastoid process.
2. Divide the ansa cervicalis to allow gentle retraction on the hypoglossal nerve.
3. Divide the posterior belly of the digastric muscle.
4. Divide the occipital artery and adjacent venous branches.
5. Divide the ascending pharyngeal artery.
6. Divide the sternocleidomastoid muscle from its mastoid attachment and elevate or resect the parotid gland. Careful dissection of the facial nerve and its branches is mandatory.
7. Remove the styloid process and its attached muscles.
8. Subluxate the mandible to increase the width of exposure at the skull base by approximately 1 cm. This maneuver is in general performed by an oral surgery team after nasotracheal intubation and prior to carotid exposure.
9. Drill and remove portions of the inferior surface of the petrous portion of the temporal bone. This process usually requires a multidisciplinary approach, including the participation of nonvascular surgeons with experience in skull base surgery.
10. Use of intraluminal balloons, usually as part of a shunt (e.g., the Pruitt–Inahara shunt), to control distal internal carotid back-bleeding can be a useful adjunct when distal control is difficult.

RESECTION OF TUMOR – SHAMBLIN 1

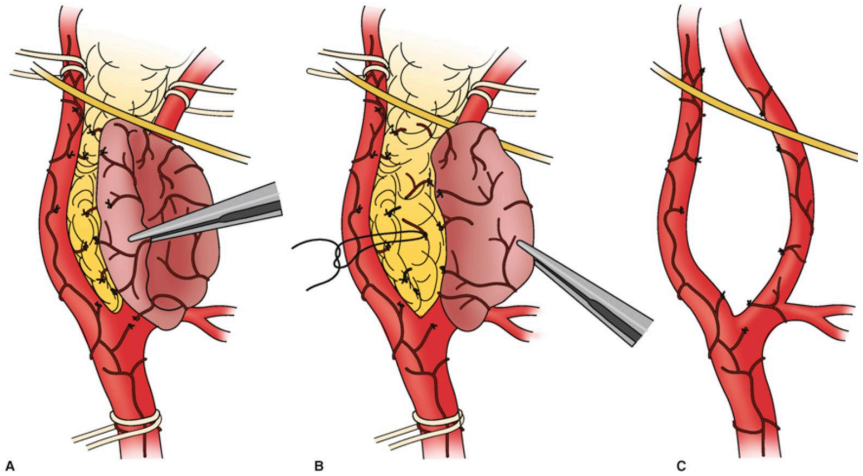
- "No touch" technique

- Preservation of nerve structures (esp in Shamblin 2-3)

Nerve	Location
CN 12	2-4 cm above the bifurcation, usually anterior to CBT
CN 10	Posterior to the ICA, (10% anterior)
Superior Laryngeal Nerve (CN 10)	Fascia posterior to the tumor / posterior to ICA
Marginal Mandibular Nerve (CN7)	Parallel to the mandibular ramus (retraction!)
Spinal accessory of CN 11	Anterior to most distal portion of ICA, post to stylohyoid near base of skull
CN9	crosses anterior to IJV and ICA near base of skull



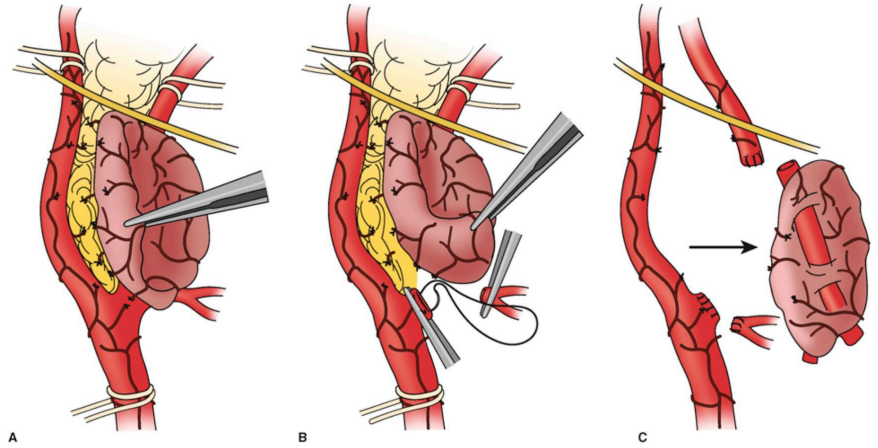
- Periadventitial plane: →
  - "white line" via counter-tension between CBT & ICA / ECA
  - Circumferential dissection: laterally, posteriorly, medially
  - Control & ligation of deep attachments
  - Single U-Stitch for a feeding vessel caudally



RESECTION OF TUMOR – SHAMBLIN 2 & 3

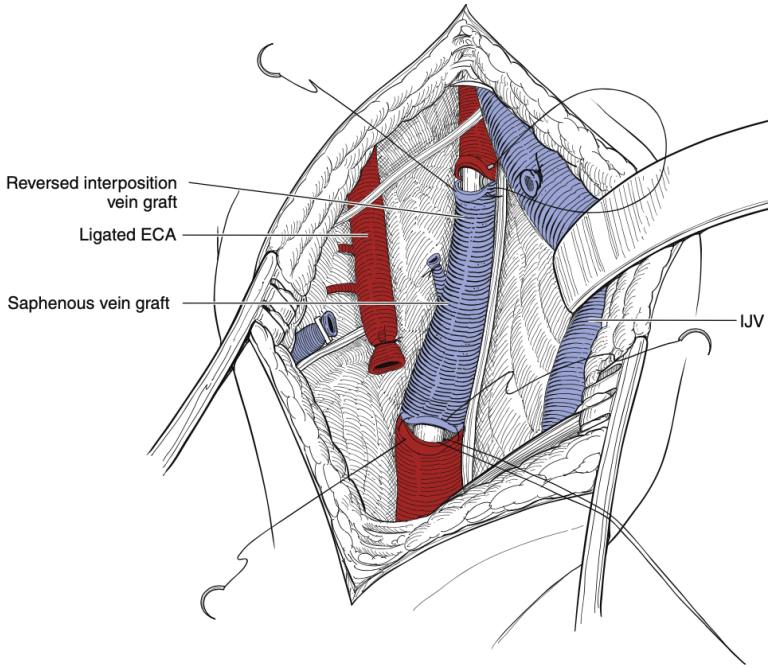
- Sacrifice of cranial nerves often in-avoidable

- ECA early proximal ligation & en-block resection with CBT tumor
  - Used as a ”handle” to facilitate retraction
  - Small en-block → re-anastomose the ECA
  - Large en-block → reconstruct ECA



- Maintain peri-adventitial dissection esp around the ICA
  - Ambitious dissection towards media of ICA = risk of rupture & blood loss

  - Larger ICA resection = interposition graft
    - 6mm PTFE (Rutherford: well documented long-term patency)
    - GSV (Chaikof)
    - W/ indwelling shunt



- MCQ 2: Techniques to help with distal ICA exposure include all except:
  - Nasotracheal intubation
  - Argyle shunt
  - Dissection of the posterior belly of the digastric muscles
  - Division of the styloid process and its muscle attachements
  - Division of the occipital artery

- MCQ 2: Techniques to help with distal ICA exposure include all except:
  - Nasotracheal intubation
  - Argyle shunt
  - Dissection of the posterior belly of the digastric muscles
  - Division of the styloid process and its muscle attachements
  - Division of the occipital artery

POSTOPERATIVE CARE

- Main consideration:
  - Monitor airway (neck hematoma)
  - Monior BP and neuro deficits
  - DAT and ambulate POD#0
  - DC home POD#1-2

- Complications:
  - Post op mortality < 0.5%
  - CN injury (24% in one series)
  - Horner's syndrome (ipsilateral): carotid sympathetic chain
  - First bite syndrome (ipsilateral): cervical sympathetic chain innervating parotid gland
  - Stroke
  - Neck hematoma
  - Baro-reflex failure:
    - Damage to nerve of Hering (CN 9)
    - Esp after bilateral CBT resection
    - Disruption of negative feedback of baroceptor tissue → life-threatining HTN crisis
    - Mx= Arterial line ICU-PSCU setting Aggressive HTN management
    - Some patient will have long term BP lability

MCQs

- MCQ 3: CBT resection is usually associated with nerve damage to all except:
    - Vagus nerve
    - Superior laryngeal nerve
    - Recurrent laryngeal nerve
    - Spinal accessory nerve
    - Marginal mandibular nerve

MCQs

- MCQ 3: CBT resection is usually associated with nerve damage to all except:
  - Vagus nerve
  - Superior laryngeal nerve
  - Recurrent laryngeal nerve
  - Spinal accessory nerve
  - Marginal mandibular nerve

REFERENCES

- Rutherford's Vascular Surgery & Endovascular Therapy. 10th Ed

- Chaikof's Atlas of Vascular Surgery & Endovascular Therapy: Anatomy & Technique

THANKS FOR LISTENING!

- Questions !