GRAND ROUNDS DECEMBER 3, 2009

STATE UNIVERSITY OF NEW YORK DOWNSTATE MEDICAL CENTER

Ramanath Bhandari, MD

CASE PRESENTATION

- 62 yo female presents with foreign body sensation and irritation of the right eye for the last 6 weeks presents as referral from primary care medicine.
 Pt. also reports subjective changes in hearing.
- Pt. reports her symptoms have been ongoing for the past 6 years, and has recently arrived from Jamaica.
- Pmhx: Hypertension, Hypercholesterolemia
- Pohx: denies
- Meds: recently started hydrochlorothiazide, zocor, aspirin.
- Allergies: NKDA

Patient Care

CASE PRESENTATION

o Dvasc: 20/20 ou

• Tapp: 16,16

- EOM: Full OU, No diplopia
- CVF: grossly full OU
- Pupils: 3-2mm OD, 5-3mm OS in room light
 4-2mm OD, 7-5mm OS in dark room

o SLE

- LLA: See picture CS: w/q ou K: clear ou
- AC: quiet ou
- IP: intact OU
- L: 1-2+NS OU



CASE PRESENTATION

• DFE:

V: Clear OU D: 0.35 c/d ratio OU M: Flat OU V: mild arteriole attenuation OU

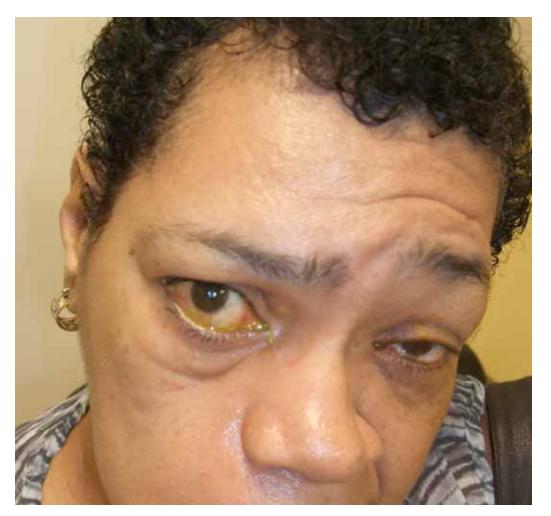
P: wnl ou.



Color Photograph



Color Photograph



DIFFERENTIAL DIAGNOSIS:

- Facial Nerve Paralysis
- Cerebello-Pontine Angle Mass
- Epstein Barr-Virus Infection
- Lyme Disease
- Herpes Virus
- o Syphilis
- Sarcoidosis
- Collagen Vascular Disease
- o HIV
- o Parotid Tumor
- o Trauma



FURTHER WORK UP OF OUR PATIENT?

- HIV testing.
- Lyme Titers.
- HSV titers
- ESR and CRP
- o RPR, VDRL
- ACE level, Lysozyme, Chest X-ray
- MRI of Brain with and without contrast focus on cerebello-pontine Angle



WORK UP COMPLETED

• HIV – negative

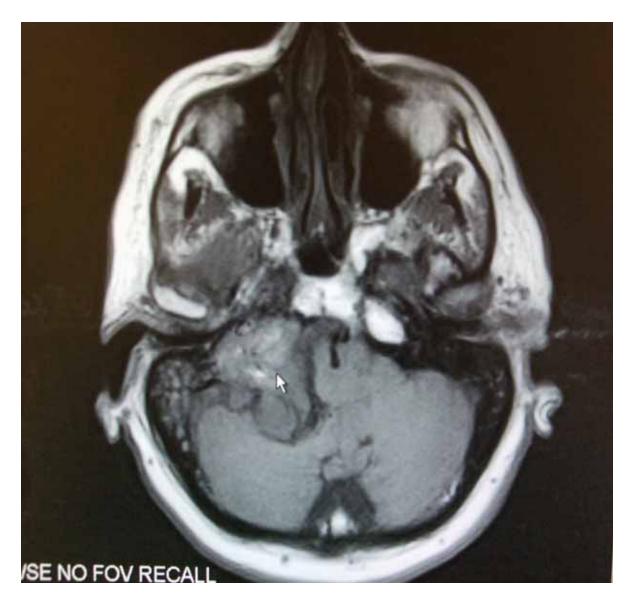
• Lyme Titers: WNL

• HSV titers, IgG positive, IgM negative

- RPR, VDRL non-reactive
- ESR: 26 (wnl), CRP: 1.5 (wnl)
- ACE, Lysozyme and CXR: wnl

• MRI see Images:











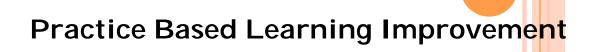
MRI REPORT:

- Extra-axial mass demonstrating heterogeneous high signal on T2 weighted images and intermediate signal on T1 weighted images extends from the right cerebellopontine angle, internal auditory canal, middle ear, and jugular foramen.
- The right internal carotid artery is encased by this mass in its high cervical segment.
- There is mild mass effect on the pons and medulla.
- Findings as above suggest glomus jugulotympanicum tumor.
 - Differential diagnosis also includes meningioma and other vascular extra-axial tumors.

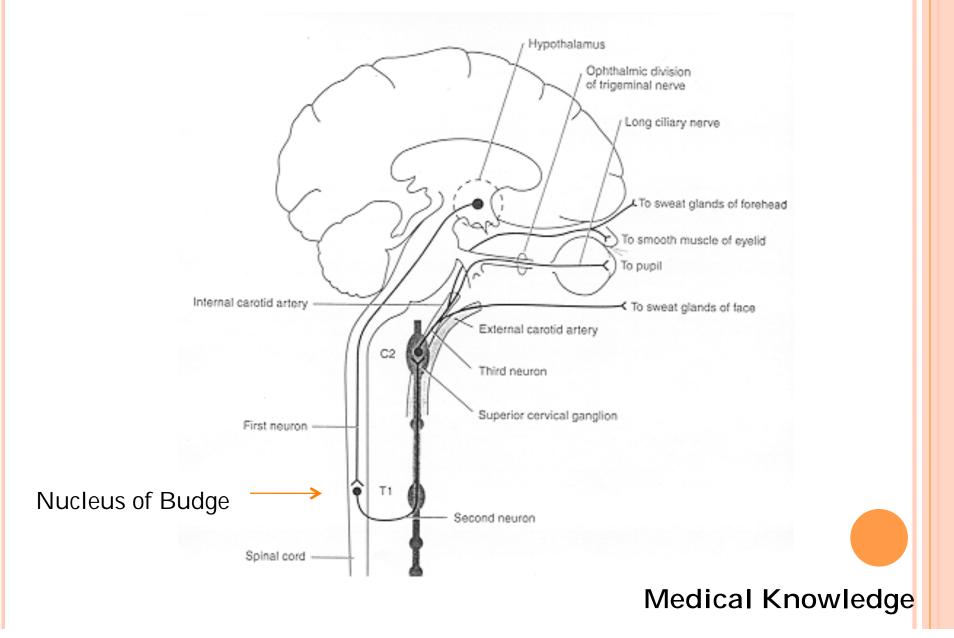
Medical Knowledge

PUPIL FINDINGS:

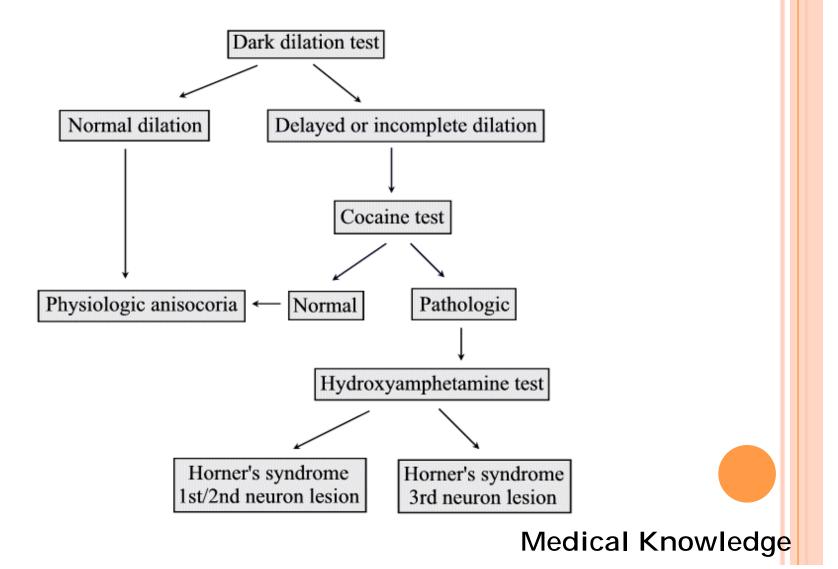
- What is the reason for the Pupil findings in this patient?
- 3-2mm OD and 5-3mm OS in room light
 4-2mm OD and 7-5mm OS in dark room



Sympathetic Pathway:



ANISOCORIA AND PHARMACOLOGIC TEST IN HORNER'S SYNDROME:



GLOMUS TUMOR:

• Estimated Incidence of 1/30,000.

- Benign Tumor of the paraganglionic system also known as chemodectomas or paragangliomas
- 80% are sporadic, 20% may be inherited as an autosomal dominant trait.
- Familial forms tend to be bilateral.
- Malignant glomus tumors are rare.
- Early tumors may present with symptoms related to the middle ear cleft (i.e. pulsatile tinnitus or conductive hearing loss).
- Head and Neck glomus tumors tend to be derived from the parasympathetic system as opposed to the sympathetic paragangliomas seen in NF I, MEN 2A and VHL.

Medical Knowledge

GLOMUS TUMOR

- 2% of Glomus Tumor secrete clinically significant levels of catecholamines.
- These patients may have facial flushing, tachycardia, labile hypertension.
- These tumors are highly vascular as demonstrated on MRI and CT angiography.
- Tumors have a propensity to grow and must be proactively treated even though the majority are benign.

Medical Knowledge

• Endovascular Embolization is commonly used prior to resection or radiation therapy.

OUR PATIENT'S COURSE:

- Our patient was referred to ENT and Neurosurgery
- ENT surgeons were able to visualize a pulsatile mass behind the tympanic membrane.
- The patient was diagnosed with a glomus tumor.
- It was decided that our patient would first undergo a neuro-interventional procedure prior to resection of the tumor.

Dr. Sundeep Mangla

- Director of Interventional Neuroradiology
- Associate Professor of Radiology, Neurosurgery and Neurology
- Diplomate, American Board of Radiology and National Board of Medical Examiners
- He has served on the faculty of Columbia University, Cornell University and Yale University School of Medicine
- He completed fellowships in Interventional Neuroradiology at Yale-New Haven Medical Center and University of Iowa Hospital and Clinics.

Professionalism

PATIENT S.N.

Right Neck/Skull Base Neoplasm Paraganglioma/Glomus

Professionalism

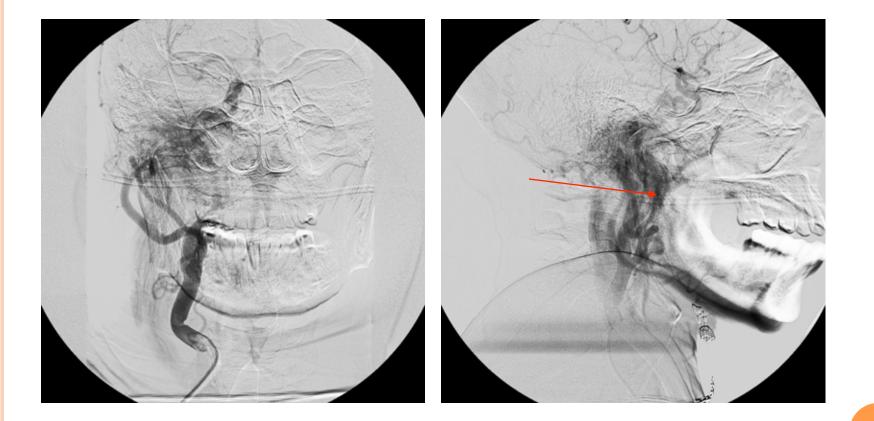
ARTERIAL FEEDERS

• Multiple arterial feeders

- Right ECA
 - Middle Meningeal Branches/IMAX branches
 - Superficial Temporal
 - o Occipital
 - Posterior Auricular
 - Ascending Pharyngeal
- Right Vertebral



VERY HYPERVASCULAR RAPID A-V SHUNTING



Filling defect suggests tumor within or compressing right IJ Tumor found within vein at surgery Medical Knowledge

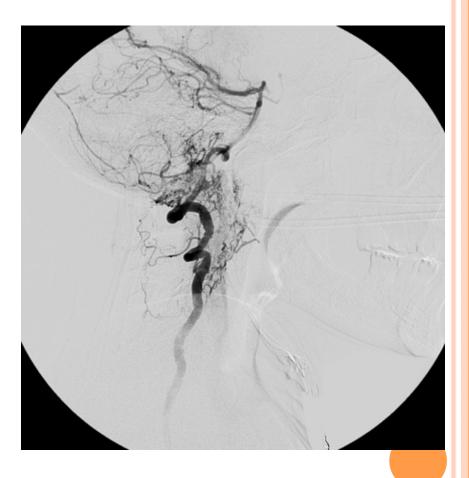
RIGHT VERTEBRAL

o Right vertebral

- Muscular branches
- PICA/vert branches

ODO NOT TOUCH

- Brainstem Strokes
- Spinal Cord Strokes



Medical Knowledge

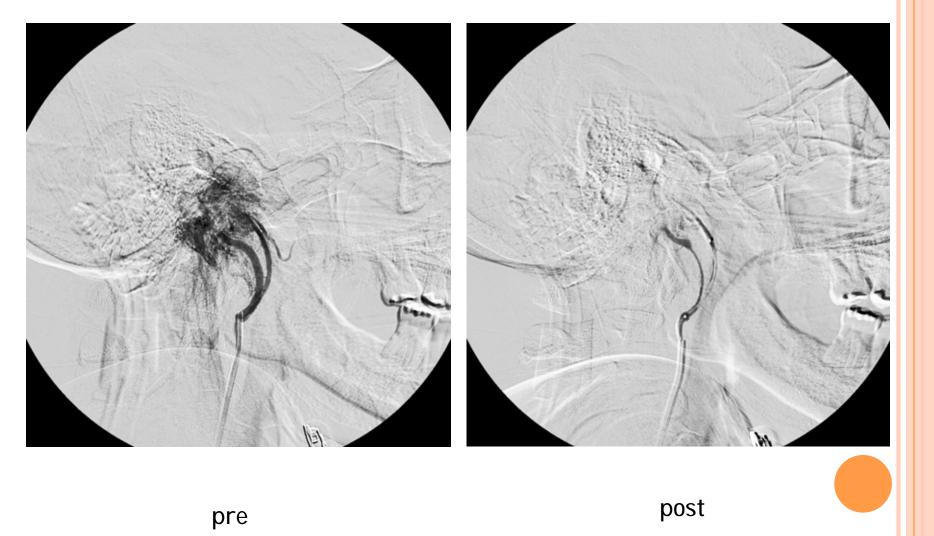
Right Ascending Pharyngeal (pre, AP)



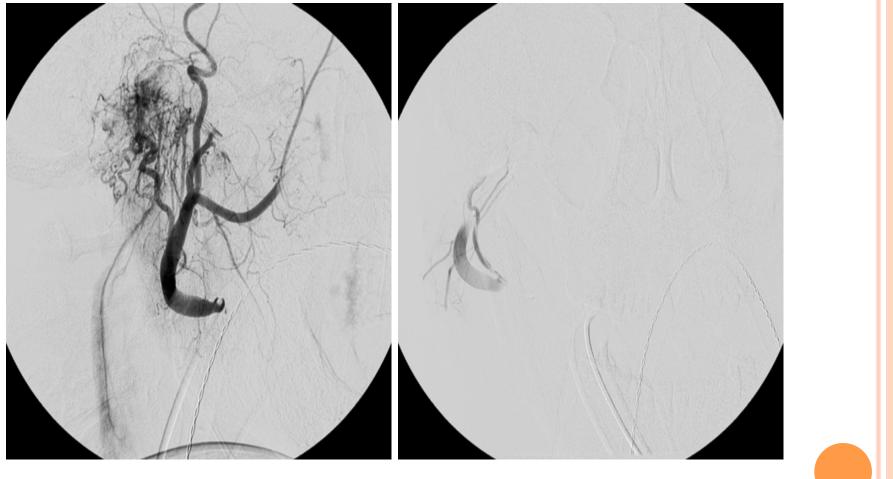
Right Ascending Pharyngeal (Pre, LAT)



RIGHT ASCENDING PHARYNGEAL (EMBOLIZATION)



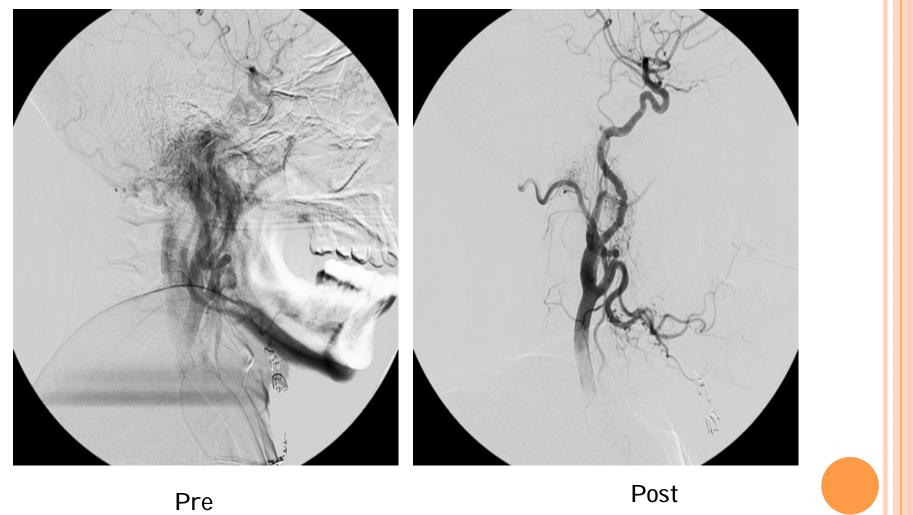
RIGHT POSTERIOR AURICULAR







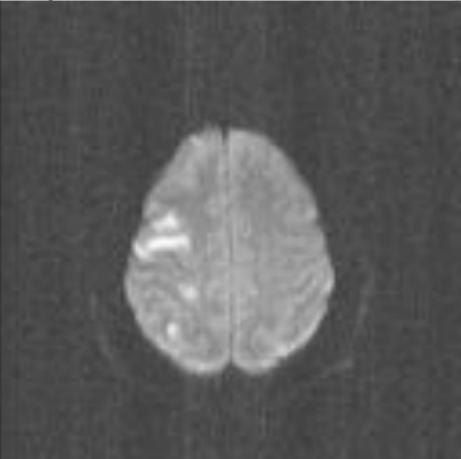
FINAL





CLINICAL COURSE (POST EMBOLIZATION)

- Initially mild left arm/leg hemiparesis
- Multiple small embolic strokes right hemisphere during embolization
- Complete recovery in
 < 1 week.
- Surgical Resection
 planned 1 week later

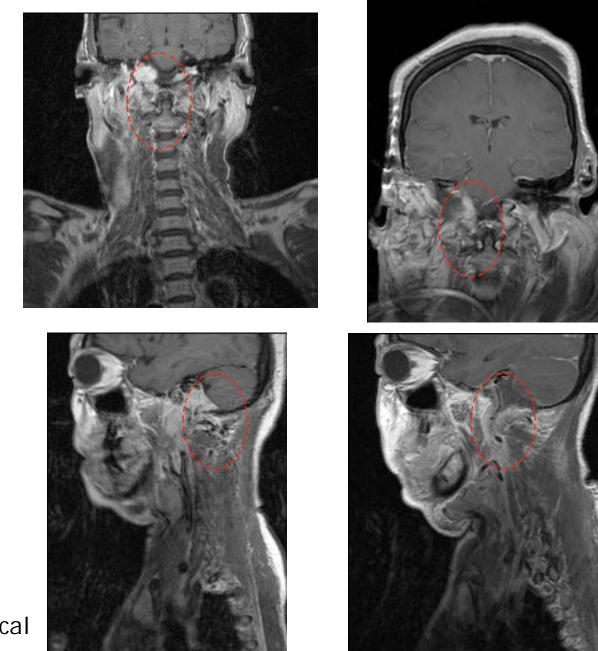


POST NEURO-INTERVENTIONAL PROCEDURE

- Our Patient underwent surgical resection of the mass lesion in a combined procedure with ENT and Neurosurgery.
 - Dr. Matthew Hanson ENT
 - Dr. Ethan Benardete Neurosurgery
- The operation was successful, a large amount of the tumor burden was resected.
- The patient continues to have a peripheral facial nerve paralysis and conductive hearing loss.
- Our Patient is scheduled to follow up in oculoplastics clinic later this month at KCH.



Patient Care



Post surgical

Pre surgical

SELF REFLECTION SLIDE

- The care of this patient was appropriate. She obtained the correct referrals in a timely organized fashion and was taken care of with a multidisciplinary approach among several surgical subspecialties.
- Ophthalmologists must routinely collaborate with other specialists to deliver the highest quality patient care and must advocate for their patients.



REFERENCES:

- Martin TP. What we call them: the nomenclature of head and neck paragangliomas. Clin Otolaryngol 2006; 31: 185-186.
- Sobol SM, Dailey JC. Familial multiple cervical paragangliomas: report of a kindred and review of the literature. Otolaryngol Head and Neck 1990; 102: 382-390.
- Sniezek JC, Netterville JL, Sabri AN. Vagal Paragangliomas. Otolaryngol. Clin North AM 2001; 34:925-939.
- Niemann S, Steinberger D, Muller U. PGL3, a third, not maternally imprinted locus in autosomal dominant paraganglioma. Neurogenetics 1999; 2: 167-170.
- Tikkakoski T, Luotonen J, Leinonen S, et al. Preoperative Embolization in the management of neck paragangliomas. Laryngoscope 1997; 107:821-826.
- Sanna M, Jain Y, De Donato G, et al. Management of Jugular paragangliomas: the Gruppo Otologico Experience. Otol Neurotol 2004; 25:797-804.
- Willen SN, Einstein DB, Maciunas RJ, et al. Treatment of Glomus Jugulare Tumors in patients with advanced age: planned limited surgical resection, followed by staged gamma-knife radiosurgery: a prelim report. Otol Neurotol 2005; 26:1229-1234.
- Semaan MT, Megerian CA. Current Assessment and Management of glomus tumors.Current Opinion in Otolaryngology and Head and Neck Surgery 2008, 16:420-426.

THANK YOU

- o Dr. Purewal
- o Dr. Sundeep Mangla
- o Dr. E.C. Lazzaro
- o Dr. Gabriel Schaab
- KCH residents

