Trigeminal Neuralgia and Other Cranial Neuralgias

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FINANCIAL DISCLOSURES

• Consulting
  – Allergan
  – Amgen
  – Amag
  – Analgesic Solutions
  – Avanir
  – Biomobie
  – Promius
  – Teva

OBJECTIVES:

• Participants will be able to accurately diagnose trigeminal neuralgia and other cranial neuralgias
• Participants will be able to describe the pathophysiology and natural history of trigeminal neuralgia and other cranial neuralgias
• Participants will be able to select the optimal medication and interventional techniques used for the treatment of trigeminal neuralgia and other cranial neuralgias
CLASSICAL TRIGEMINAL NEURALGIA DIAGNOSTIC CRITERIA

- A) At least three attacks of unilateral facial pain fulfilling criteria B and C
- B) Occurring in one or more divisions of the trigeminal nerve, with no radiation beyond the trigeminal distribution
- C) Pain has at least three of the following four characteristics:
  - Recurring in paroxysmal attacks lasting from a fraction of a second to two minutes
  - Severe intensity
  - Electric shock-like, shooting, stabbing or sharp in quality
  - At least three attacks precipitated by innocuous stimuli to the affected side of the face (some attacks may be, or appear to be, spontaneous)
- D) No clinically evident neurologic deficit
- E) Not better accounted for by another ICHD-3 diagnosis


CLASSICAL TRIGEMINAL NEURALGIA

- Previously known as Tic douloureux
  - When very severe, the pain often evokes ipsilateral facial muscle contraction
- Trigeminal neuralgia developing without apparent cause other than neurovascular compression.
  - Most frequently by the superior cerebellar artery


CLASSICAL TRIGEMINAL NEURALGIA

- Following a painful paroxysm there is usually a refractory period during which pain cannot be triggered.
- If there is prolonged background pain in the affected area
  - Subform 13.1.1.2 Classical trigeminal neuralgia with concomitant persistent facial pain

TRIGEMINAL NEURALGIA FEATURES

- Triggered by trivial stimuli including washing, shaving, smoking, talking and/or brushing the teeth (trigger factors) and frequently occurs spontaneously.
- Usually involves the second or third divisions with first division involvement in <5% of patients.


TRIGEMINAL NEURALGIA FEATURES

- In some cases a paroxysm can be triggered from somatosensory stimuli outside the trigeminal area, such as a limb, or by other sensory stimulation such as bright lights, loud noises or tastes.
- Attack periods can last for weeks to months followed by remissions, but the pain usually returns.
- Usually responsive, at least initially, to pharmacotherapy.


TRIGEMINAL NEURALGIA FEATURES

- TN does not typically involve unilateral autonomic features that can be seen with Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), short-lasting unilateral neuralgiform headache attacks with autonomic symptoms (SUNA).
- Based on this image President Vladimir Putin is more likely to have SUNCT than TN.

TRIGEMINAL NEURALGIA

EPIDEMIOLOGY

- Annual incidence of TN is 4-13 per 100,000 people
- Incidence increases with age, and most idiopathic cases occur to those over the age of 50
- Male to female ratio of TN is about 1:1.7
- Hypertension may be a risk factor
- Some estimates as high as 91% have vascular compression


TRIGEMINAL NEURALGIA

RED FLAGS

- According to the AAN and EFNS
  - Structural causes in up to 15% of patients
  - Features that increase risk of underlying lesion
    - Trigeminal sensory deficits (Trigeminal Neuropathy)
    - Bilateral involvement of the trigeminal nerve
    - Younger age


TRIGEMINAL NEURALGIA

- TN due to secondary causes = Painful Trigeminal Neuropathy
  - Acute herpes zoster/Postherpetic neuralgia
    - Most commonly affects V1
  - Post-traumatic trigeminal neuropathy
  - Multiple Sclerosis
  - Vestibular schwannoma/acoustic neuroma
  - Cerebellar pontine Meningioma
  - Epidermoid or other cyst
  - Saccular aneurysm
  - Arteriovenous malformation

TRIGEMINAL NEURALGIA

PHARMACOTHERAPY

• According to the AAN and EFNS
  – Carbamazepine (Level A, Established as effective)
    • 100mg daily → 600mg BID
    • Test for HLA-B*1502 allele in patients of Asian ancestry
  – Oxcarbazepine (Level B, Probably effective)
    • 300mg daily → 900mg BID
  – Baclofen (Level C, Possibly Effective)
    • 5 mg PO q8hr → 80 mg/day
  – Lamotrigine (Level C, Possibly Effective)
    • 50mg daily → 400mg daily
  – Phenytoin, Valproic acid, Gabapentin, Pregabalin, and Topiramate have small study support


TRIGEMINAL NEURALGIA

PHARMACOTHERAPY

• Medications with IV formulations may be useful in intractable cases and/or the ED
  – Levetiracetam
  – Phenytoin
  – Valproic Acid


TRIGEMINAL NEURALGIA

BOTULINUM TOXIN INJECTIONS

• Randomized controlled trial with 42 subjects with TN
  – 22 subjects received 75 units of BTX
  – 20 subjects received saline injections
  – Significant reduction in pain frequency at week 1 and intensity at week 2
  – More responders in BTX group (68.18%) than in the placebo group (15.00%).
  – BTX was well tolerated, with few treatment related adverse events at the end of 12 weeks

TRIGEMINAL NEURALGIA
INTERVENTIONAL PROCEDURES

- In cases resistant to pharmacotherapy, there are multiple procedures that can be used for the treatment of TN
  - Microvascular Decompression
  - Denervating/Destructive Procedures
    - Percutaneous Trigeminal Rhizotomy
    - Radiofrequency, Glycerol, or Balloon
    - Stereotactic Radiosurgery
    - Gamma Knife

MICROVASCULAR DECOMPRESSION

- 2 inch craniotomy exposes area posterior to ear
- Under microscope, the superior cerebellar artery is decompressed from nerve, and teflon felt is placed in between
- More invasive than other procedures, but no nerve destruction
- Faster results and longer lasting
- If no compression found, open denervation (microsurgical rhizotomy) could be performed
- Destructive procedures could be considered in MVD failure


RISKS OF NEUROSURGERY

- Highest rates of permanent cranial nerve deficit
- Meningitis/Encephalitis
- Intracranial Hemorrhage/Stroke
- Cranial Nerve Deficits/Neuralgias
- CSF Leaks
TRIGEMINAL NEURALGIA

- First denervating/destructive procedures were peripheral trigeminal neurectomies
  - Caused dense numbness
  - Earlier recurrence of pain
  - Treated focused, small, superficial branch of TN
- Proximal treatment (rhizotomy, root exit zone) has better results
  - Longer lasting
  - Less or no facial numbness
  - Worst case is anesthesia dolorosa

TRIGEMINAL NEURALGIA

- Percutaneous Trigeminal Rhizotomy
  - Needle inserted through cheek one inch from angle of the mouth
  - Needle advanced through foramen ovale using fluoroscopy

TRIGEMINAL NEURALGIA

- Percutaneous Trigeminal Rhizotomy Via
  - Radiofrequency Ablation (heat) → 6205 patients
    - Stimulation is performed prior to ablation to ensure correct target
    - Only selective technique
    - If V1 involved, caution to not over-numb corneal sensation, which risks keratopathy
    - Highest rates of initial pain relief and the lowest rates of pain recurrence
  - Glycerol (chemical) → 1217 patients
    - CSF coming from needle is a good finding before bathing nerve
  - Balloon (mechanical) → 759 patients
    - More likely to affect mastication

TRIGEMINAL NEURALGIA

- Stereotactic Radiosurgery
  - Used for treatment of tumors, vascular lesions, and functional disorders like TN
  - Highly focused beams of ionizing radiation with high precision
  - Useful for targets that are inaccessible for open surgery
  - Immediately outside of target there is a steep drop in radiation so surrounding tissues are relatively spared
  - Not useful for large targets

TRIGEMINAL NEURALGIA

- Stereotactic Radiosurgery
  - 497 patients presenting with TN underwent GKS
  - No clear vascular compression or history of multiple sclerosis
  - Results
    - 169 patients became pain free within the first 48 hours of GKS
      - Pain recurrence in 66 patients (39%)
      - Postoperative hypesthesia in 18 patients (13.7%)
    - 194 patients became pain free within post treatment Day 3-30
      - Pain recurrence in 71 patients (36.6%)
      - Postoperative hypesthesia in 30 patients (19%)
    - 91 patients became pain free 30 days post-GKS
      - Pain recurrence in 27 patients (29.7%)
      - Postoperative hypesthesia in 22 patients (30.6%)


GLOSSOPHARYNGEAL NEURALGIA DIAGNOSTIC CRITERIA

- A. At least three attacks of unilateral pain fulfilling criteria B and C
- B. Pain is located in the posterior part of the tongue, tonsillar fossa, pharynx, beneath the angle of the lower jaw and/or in the ear
- C. Pain has at least three of the following four characteristics:
  1. recurring in paroxysmal attacks lasting from a few seconds to 2 min
  2. severe intensity
  3. shooting, stabbing or sharp in quality
  4. precipitated by swallowing, coughing, talking or yawning
- D) No clinically evident neurologic deficit
- E) Not better accounted for by another ICHD-3 diagnosis

### Glossopharyngeal Neuralgia

- Previously used term: Vagoglossopharyngeal neuralgia.
- May remit and relapse in the fashion of classical trigeminal neuralgia.
- Less severe than classical trigeminal neuralgia but can be bad enough for patients to lose weight. These two disorders can occur together.
- Rare cases associated with vagal symptoms
  - Cough, hoarseness, syncope and/or bradycardia.
- Imaging may show neurovascular compression of the glossopharyngeal nerve.
- Usually responsive, at least initially, to antiepileptics
- Application of local anaesthetic to the tonsil and pharyngeal wall can prevent attacks for a few hours.


### Classical Nervus Intermedius Neuralgia Diagnostic Criteria

- A. At least three attacks of unilateral pain fulfilling criteria B and C
- B. Pain is located in the auditory canal, sometimes radiating to the parieto-occipital region
- C. Pain has at least three of the following four characteristics
  1. recurring in paroxysmal attacks lasting from a few seconds to minutes
  2. severe intensity
  3. shooting, stabbing or sharp in quality
  4. precipitated by stimulation of a trigger area in the posterior wall of the auditory canal and/or periauricular region
- D) No clinically evident neurologic deficit
- E) Not better accounted for by another ICHD-3 diagnosis


### Classical Nervus Intermedius Neuralgia

- Can involve lacrimation, salivation and/or taste alteration
- Neurovascular compression can be a cause
- Nervus intermedius neuropathy attributed to Herpes zoster
  - Ramsay Hunt syndrome
  - Herpetic eruption has occurred in the ear and/or oral mucosa, in the territory of the nervus intermedius
  - Peripheral facial paresis
- Sensory innervation of the ear is complicated

OCCIPITAL NEURALGIA

DIAGNOSTIC CRITERIA

• A. Unilateral or bilateral pain fulfilling criteria B-E

• B. Pain is located in the distribution of the greater, lesser and/or third occipital nerves

• C. Pain has two of the following three characteristics:
  – 1. recurring in paroxysmal attacks lasting from a few seconds to minutes
  – 2. severe intensity
  – 3. shooting, stabbing or sharp in quality

• D. Pain is associated with both of the following:
  – 1. dysaesthesia and/or allodynia apparent during innocuous stimulation of the scalp and/or
    hair
  – 2. either or both of the following:
    a) tenderness over the affected nerve branches
    b) trigger points at the emergence of the greater occipital nerve or in the area of
    distribution of C2

• E. Pain is eased temporarily by local anaesthetic block of the affected nerve

• F. Not better accounted for by another ICHD-3 diagnosis.

Headache Classification Committee of the International Headache Society (IHS). The International Classification of

OCCIPITAL NEURALGIA EXAM

• Exam maneuvers to perform…
  – Cranial tinel’s sign demonstrating pain/paresthesias along nerve distribution
  – Neck passive range of motion elicits pain
  – Best results: Lancinating pain occurs with tinel’s and PROM when patient
    denies any significant headache otherwise

NERVE BLOCKS

DIAGNOSTIC AND THERAPEUTIC

• Generally safe, well tolerated
  office based procedures

• Can be performed for the acute
  treatment of numerous
  headache disorders.

• Can have prolonged effects
  beyond the duration of the
  injected anesthetic at times
  lasting weeks to months

Afridi SK, Shields KG, Bhola R, Goadsby PJ. Greater occipital nerve injection in primary headache syndromes—
NERVE BLOCK COMPOSITION

- Nerve blocks are performed with an anesthetic with or without a steroid
- Anesthetic is usually lidocaine, bupivacaine, or a combination.
  - 0.75% bupivacaine is my preference
- Steroids added can include methylprednisolone and triamcinolone
- Steroid alone proven to be useful, but lack of immediate relief makes this less successful;


NERVE BLOCK CENTRAL EFFECTS

- Peripheral nerve blocks may modulate central pain structures
- In one study, occipital nerve blocks were performed in the setting of an acute migraine with improvement of
  - Migraine pain
  - Brush allodynia in the trigeminal nerve distribution
  - Photophobia


Occipital Nerve Block

Prone, 6cc per side

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TRIGEMINAL NERVE BRANCHES...

- Auriculotemporal Neuralgia
- Supraorbital Neuralgia
- Supratrochlear Neuralgia

Auriculotemporal Nerve Block
Supine, 2cc per side

Supraorbital/Supratrochlear Nerve Block
Supine, 0.5-1cc per foramen
## OCCIPITAL NEURALGIA AND MIGRAINE

- 35 consecutive occipital neuralgia cases, 15 had both occipital neuralgia and migraines
- Chances are good that many patients with migraines and focal neuralgias are only being diagnosed with migraines
- Patients being treated with decompression procedures for migraines may be responding because they actually have a cranial neuralgia

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## MIGRAINE SURGERY

### Surgical Deactivation of Potential Trigger Sites
- **Frontal Trigger Site**
  - Supraorbital and supratrochlear nerves
  - Resection of corrugator supercili, depressor supercili muscles, lateral procerus

- **Temporal Trigger Site**
  - Zygomatico-temporal branch of Trigeminal Nerve through the temporalis muscle
  - Avulsion of the nerve

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MIGRAINE SURGERY

• Surgical Deactivation of Potential Trigger Sites
  – Occipital Trigger Site
  • Greater occipital nerve
  • Resection of small portion of semispinalis capitis muscle and shielding of the nerve with a subcutaneous flap (fat pad)
  • If there is contact between the occipital artery and occipital nerves, the artery is at times also resected


CRITIC OF THE CRITIC

• Insult to study neurologists
• Two separate diagnoses cannot co-exist at the same time in the same patient.
  – That would be like having carpal tunnel syndrome and cervical radiculopathy at the same time


MIGRAINE PRE-SURGERY EVALUATION

• Doppler Evaluation
  – Headache point of origin identified with 1 finger by patient
  – Site is explored with Doppler.
  – If an arterial Doppler signal is identified at the site, it is considered an active arterial trigger site.
FRONTAL, TEMPORAL, OCCIPITAL TRIGGER SITES

- If nerve compression is serving as a trigger for migraines, why are branches of the trigeminal nerve being resected rather than decompressed in the temporal region.
  - Based on the trigeminal neuralgia literature, damaging or destroying a peripheral nerve can lead to numbness, paresthesias, dysesthesias, and even worsening of preoperative pain
- If nerve compression is thought to be occurring, why do these patients not have numbness, paresthesias, or neuralgiform pain in the distribution of the suspected nerve compression.
  - Supraorbital, Supratrochlear, Auriculotemporal, and Greater/Lesser Occipital Neuralgia may have existed in these patients in addition to migraine.
  - Decompression of the nerve improved/resolved the neuralgia, which has a tendency to improve, but not CURE migraine.


CRANIAL NEURALGIA OR PRIMARY STABBING HEADACHE?

- DIAGNOSTIC CRITERIA
  - A. Head pain occurring spontaneously as a single stab or series of stabs and fulfilling criteria B-D.
  - B. Each stab lasts for up to a few seconds.
  - C. Stabs recur with irregular frequency, from one to many per day.
  - D. No cranial autonomic symptoms.
  - E. Not better accounted for by another ICHD-3 diagnosis.


CRANIAL NEURALGIA OR PRIMARY STABBING HEADACHE?

- Transient and localized stabs of pain in the head that occur spontaneously in the absence of organic disease.
- When stabs are strictly localized to one area, structural changes at this site and in the distribution of the affected cranial nerve must be excluded.
- Involves extratrigeminal regions in 70% of cases.
- If cranial autonomic symptoms are present, think Short-lasting unilateral neuralgiform headache attacks (SUNCT).
- Primary stabbing headache is more common in migraineurs.
  - Stabs tend to be localized around areas of frequent migraine headache pain.

CRANIAL NEURALGIA OR PRIMARY STABBING HEADACHE?

• My take…
  – Reproducible physical exam findings + single location = neuralgia
  – No significant exam findings + multiple locations = primary stabbing headache
  – Little risk and high potential yield with nerve blocks

CONCLUSIONS REGARDING CRANIAL NEURALGIAS

• There are many treatments for these conditions
• Medication trials should start at a low dose, and titrations should be fast/slow based on patient preference and side effects
• Combination therapies should be considered
• Do not hesitate to refer patients to another provider for treatments that you may not provide
• AHS has issued a position statement on migraine surgery for a reason

If you enjoyed the clinical content…

Principles and Practice of Pain Medicine

Dr. Mathew (4 chapters), Dr. Scrivani
If you enjoyed the humor...

Harvard Health Publications
HARVARD MEDICAL SCHOOL
Trusted advice for a healthier life

http://www.health.harvard.edu/blog/author/pgmathew
Sinus headache or sign-us up for a migraine consultation
Spinning out of control: Vertigo
Snored to death: The symptoms and dangers of untreated sleep apnea
Unlocking the lock jaw: Temporomandibular Joint (TMJ) dysfunction
White coat syndrome or white coat logo syndrome: The pitfalls of doctor shopping by brand

THANK YOU!!!

JOHN R. GRAHAM HEADACHE CENTER
FELLOWS AND STAFF
2017-2018