Headache and Facial Pain

Sharon Ramos, MD
Faculty Advisor and Discussant:
Farrah Siddiqui, MD
The University of Texas Medical Branch
Department of Otolaryngology
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Archivist: Melinda Stoner Quinn, MSICS
Headache and facial pain are common complaints otolaryngologist evaluate in practice

- Migraine headache
- Tension headache
- Cluster headache
- Rhinogenic headache
- Trigeminal neuralgia
- Herpes zoster
- Tolosa-Hunt syndrome
- First bite syndrome
- Eagle Syndrome
- Giant cell arteritis
- Carotidynia
Migraine Headaches

- Second most common form of headache
- Prevalence 10% of population
  - 18% in women, 6% in men
  - Peak age onset 20’s-30’s
- Recurrent episodes of severe, throbbing, unilateral headaches
- Associated Symptoms
  - Nausea, vomiting, photophobia, phonophobia
- Precipitating factors
  - Stress, lack of sleep, hormonal changes, diet, etc.
- Pathophysiology (Theories)
  - Vasospasm
  - Cortical Spreading Depression
This theory asserts an explanation why people experience pain and auras with migraines. A trigger event causes neurons to fire which control the vascular tone round the cerebral blood vessels, decreasing blood to the cortex. This hypoxic event is thought to be the cause for the aura event prior to the headache. After vessel constriction the blood vessels relax and dilate and distend from blood reperfusion. This distension stretch the neuron fibers innervating the meningeal vasculature found in a trigeminal nerve distribution producing the headache pain.
Cortical Spreading Depression

- A triggering event depolarizes hyperexcitable neurons in a wave like fashion across the brain followed by a prolonged inhibition of cortical activity.
- “Spreading Depression” - elicits the sensation of an aura -- Causes release of NO, ATP, glutamate, etc.
- These molecules diffuse toward the surface of the cortex triggering vessel dilation/inflammation resulting in migraine pain.

- A triggering event depolarizes hyperexcitable neurons in a wave like fashion across the brain (2-6mm/min) followed by a prolonged inhibition of cortical activity (15-30 min).
- **Cortical spreading** is what provokes an aura. It also involves local release of ATP, NO, H+ ions, etc. by neurons/vascular cells.
- These molecules diffuse toward the surface of the cortex, triggering a consequential neurogenic inflammation (vasodilation, plasma protein extravasation) causing pain.

Migraine without Aura

Table 1. ICHD III 1.1 Migraine without Aura

<p>| | |</p>
<table>
<thead>
<tr>
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<tbody>
<tr>
<td>A.</td>
<td>At least five attacks fulfilling criteria B-D</td>
</tr>
<tr>
<td>B.</td>
<td>Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)</td>
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<tr>
<td>C.</td>
<td>Headache has at least <strong>two</strong> of the following four characteristics:</td>
</tr>
<tr>
<td></td>
<td>1. unilateral location</td>
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<tr>
<td></td>
<td>2. pulsating quality</td>
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<tr>
<td></td>
<td>3. moderate or severe pain intensity</td>
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<td></td>
<td>4. aggravation by or causing avoidance of routine physical activity (eg, walking or climbing stairs)</td>
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<tr>
<td>D.</td>
<td>During headache at least one of the following:</td>
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<tr>
<td></td>
<td>1. nausea and/or vomiting</td>
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<td></td>
<td>2. photophobia and phonophobia</td>
</tr>
<tr>
<td>E.</td>
<td>Not better accounted for by another ICHD-III diagnosis.</td>
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</tbody>
</table>

The International Classification of Headache Disorders, 3rd Ed.
Migraine with Typical Aura

Table 2. ICHD III 1.2.1 Migraine with Typical Aura

| A. At least two attacks fulfilling criteria B and C |
| B. Aura consisting of visual, sensory and/or speech/language symptoms, each fully reversible, but no motor, brainstem or retinal symptoms |
| C. At least two of the following four characteristics: |
|   1. at least one aura symptom spreads gradually over ≥5 minutes, and/or two or more symptoms occur in succession |
|   2. each individual aura symptom lasts 5-60 minutes |
|   3. at least one aura symptom is unilateral |
|   4. the aura is accompanied, or followed within 60 minutes, by headache |
| D. Not better accounted for by another ICHD-III diagnosis, and transient ischemic attack has been excluded. |
## Vestibular Migraine

**Table 3. ICHD III Appendix criteria for A1.6.5 Vestibular Migraine**

<table>
<thead>
<tr>
<th>A.</th>
<th>At least five episodes fulfilling criteria C and D</th>
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<tbody>
<tr>
<td>B.</td>
<td>A current or past history of 1.1 <em>Migraine without aura</em> or 1.2 <em>Migraine with aura</em></td>
</tr>
<tr>
<td>C.</td>
<td>Vestibular symptoms of <em>moderate or severe intensity</em>, lasting between 5 minutes and 72 hours</td>
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<tr>
<td>D.</td>
<td>At least 50% of episodes are associated with at least one of the following three migrainous features:</td>
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<tr>
<td></td>
<td>1. headache with at least two of the following four characteristics:</td>
</tr>
<tr>
<td></td>
<td>1. a) unilateral location</td>
</tr>
<tr>
<td></td>
<td>2. b) pulsating quality</td>
</tr>
<tr>
<td></td>
<td>3. c) moderate or severe intensity</td>
</tr>
<tr>
<td></td>
<td>4. d) aggravation by routine physical activity</td>
</tr>
<tr>
<td></td>
<td>2. photophobia and phonophobia</td>
</tr>
<tr>
<td></td>
<td>3. visual aura</td>
</tr>
<tr>
<td>E.</td>
<td>Not better accounted for by another ICHD-III diagnosis or by another vestibular disorder.</td>
</tr>
</tbody>
</table>
A section in Vestibular Migraine was found in the Appendix on page 795, book has a total of 807 pages.

The purpose of the appendix is to present novel entities that have not been sufficiently validated by research conducted thus far.

BUT in 2013 the migraine work group did include an appendix definition of Vestibular Migraine, which allows vertigo as the sole accompaniment to headache.

Moderate

**Visual auras** are characterized by bright scintillating lights or zigzag lines, often with a scotoma that interferes with reading. Visual auras typically expand over 5–20 minutes and last for less than 60 minutes.
Migraine induced vasospasm leads to a decrease in regional blood flow to the inner ear via the internal auditory artery from the anterior inferior cerebellar artery (AICA) causing transient ischemia and resulting in possible transient or maybe permanent peripheral dysfunction.
# Vestibular Migraine

## Table 1. Diagnostic criteria for definite migrainous vertigo

<table>
<thead>
<tr>
<th>Definite migrainous vertigo</th>
<th>Neuhauser et al.</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Episodic vestibular symptoms of at least moderate severity</td>
<td></td>
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<tr>
<td>B. Current or previous history of migraine according to the 2004 criteria of the HIS</td>
<td></td>
</tr>
<tr>
<td>C. One of the following migrainous symptoms during ≥ 2 attacks of vertigo: migrainous headache, photophobia, other auras</td>
<td></td>
</tr>
<tr>
<td>D. Other causes ruled out by appropriate investigations</td>
<td></td>
</tr>
</tbody>
</table>

**Comment:**

Vestibular symptoms are rotational vertigo or another illusory self or object motion. They may be spontaneous or positional. Vestibular symptoms are moderate if they interfere with but do not prohibit daily activities and ‘severe’ if patients cannot continue daily activities.

## Probable migrainous vertigo

A. Episodic vestibular symptoms of at least moderate severity

B. One of the following:
   1. Current or previous history of migraine according to the 2004 criteria of the HIS
   2. Migrainous symptoms during vestibular symptoms
   3. Migraine precipitants of vertigo in more than 50% of attacks: food triggers, sleep irregularities, hormonal change
   4. Response to migraine medications in more than 50% of attacks

C. Other causes ruled out by appropriate investigations

In a study conducted by Neuhauser he found that only 94% of patients diagnosed with vestibular migraine actually had a migraine headache associated with episodes of vertigo, the most common duration of vertigo was 5-60 minutes (33%) and the second most common associated symptoms was photophobia (70%) followed by phonophobia (64%)
Diagnosis

- Radtke et al. assessed 75 patients with prior diagnosis of definitive vestibular migraine and probable vestibular migraine. They were re-assessed after a mean followed up of 8.75 years. DVM was confirmed in 40 of 47 patients with prior diagnosis of dMV (85%). 14 of the 28 initially classified as pVM met criteria for dVM (50%), nine for pVM (32%). Six additional patients with dVM and two with pVM had developed mild sensorineural hearing loss, formally fulfilling criteria for bilateral Meniere’s disease (MD), but had clinical features atypical of MD. Seven of these also met criteria for dVM at follow-up. The initial diagnosis was completely revised for 4 patients.

- 47 patients had a prior diagnosis of dVM and 28 were previously diagnosed with probable VM. These patients followed up after a mean of 8.75 years.
Diagnosis (cont’d)

- International Classification of headache disorders, 2013
  - Migraine without Aura
  - Migraine with Aura
  - Migraine with Brainstem Aura

- Vestibular Migraine
  - Challenging to diagnose-no set criteria?
    - 2013, International Classification of Headache Disorders (Appendix)
      - “Migraine with Brainstem Aura” is the only one to include vertigo as a symptom
    - 2005, Neuhauser and Lambert published a set of guidelines
      - Must experience “Moderate –Severe” symptoms
        - Definite VM
        - Probable VM
      - 2011, Radtke et al showed high validity of the proposed clinical criteria by Neuhauser for detecting patients with vertigo related to migraine
        - Must **rule out** other causes of vertigo
          - Stroke, BPPV, Menierre’s

- Family history of migraines
Diagnosis (cont’d)

- **Vestibular Migraines**
  - Neurological exam
    - Romberg’s Test
    - Vestibulo-ocular reflex test
  - Dix-Hall pike
  - Audiogram
  - VNG/ENG
  - MRI/CT

- Diagnosis, perform full ENT exam, neurological exam should include Romberg test which evaluates the body’s ability to sense positioning (proprioception), the vestibulo-ocular reflex test assess the visual compensations the brain makes for head rotation.
- Dix-hall pike is a positioning maneuver which elicits vertigo and nystagmus and is diagnostic for BPPV. Asses hearing with an Audiogram, presence of hearing loss associated with vertigo may suggest Meniere’s disease. VNG is used to evaluate for nystagmus.
- MRI to rule any masses or lesions, like an acoustic neuroma. Differential diagnosis for vestibular migraine are the following, Migraine with Brainstem Aura, Meniere’s Disease and BPPV.
Differential Diagnosis for VM

- **Migraine with Brainstem Aura**
  - Previously, known as Basilar Migraine
  - Requires at least 2 “brainstem symptoms” therefore excludes patients with typical migraine accompanied only by vertigo.

- **Meniere’s Disease**
  - Vertigo lasting between 20 min-24 hours, tinnitus, unilateral, low frequency fluctuating hearing loss, not typical for VM

- **BPPV**
  - Dix-hallpike (+)
    - VM can have positional vertigo, nystagmus is not fatigable w
Migraine with Brainstem Aura

**Table 4. ICHD III Migraine with Brainstem Aura**

A. At least two attacks fulfilling criteria B-D

B. Aura consisting of visual, sensory and/or speech/language symptoms, each fully reversible, but **no motor or retinal symptoms**

C. At least **two of the following brainstem symptoms:**
   1. dysarthria
   2. vertigo
   3. tinnitus
   4. hypoacusis
   5. diplopia
   6. ataxia
   7. decreased level of consciousness

D. At least **two** of the following four characteristics:
   1. at least one aura symptom spreads gradually over ≥5 minutes, and/or two or more symptoms occur in succession
   2. each individual aura symptom lasts 5-60 minutes
   3. at least one aura symptom is unilateral
   4. the aura is accompanied, or followed within 60 minutes, by headache

E. Not better accounted for by another ICHD-III diagnosis, and transient ischemic attack has been excluded.

The International Classification of Headache Disorders, 3rd Ed.
Migraine with Brainstem Aura (cont’d)

- Previously known as “Basilar Type Migraine”. These patients should supposedly have symptoms and/or signs suggestive of the posterior cerebral circulation such as bilateral visual symptoms, dysarthria, vertigo, hearing loss, diplopia, or ataxia (Table 7), but the new label may be another misnomer as the evidence for brainstem dysfunction in migraine is scant.
- This category also requires at least 2 “brainstem symptoms” therefore excluding patients with typical migraine accompanied only by vertigo.
Treatment

- Nonpharmacologic
  - Avoid triggers
  - Symptom Diary
  - Dietary modifications
  - Regularity in exercise, eating, sleeping
- Photophobia/Phonophobia
  - Lay down in a dark/quiet room

Avoid

* Ripened cheeses (such as cheddar, Emmentaler, Stilton, Brie, and Camembert)
  - Chocolate
  - Marinated, pickled, or fermented food
  - Foods that contain nitrites or nitrates (bacon, hot dogs) or MSG (soy sauce, meat tenderizers, seasoned salt)
  - Sour cream
  - Nuts, peanut butter
  - Sourdough bread
  - Broad beans, lima beans, fava beans, snow peas
  - Figs, raisins, papayas, avocados, red plums
  - Citrus fruits
  - Excessive amounts (more than 2 cups total) of caffeinated beverages such as tea, coffee, or cola
  - Alcohol (including red wine and beer)
Treatment (cont’d)

- **Abortive**
  - Triptans/Ergot derivatives
    - Sumatriptan, rizatriptan, ergotamine tartrate
    - If used >2d/wk can cause ergot-induced headache
  - S/E-nausea, angina
  - Fioricet
    - Butalbital, acetaminophen, caffeine
  - Fiorinal
    - Butalbital, ASA, caffeine

- **Prophylactic**
  - Episodes >5/mo
  - Antihypertensives
    - BB-Metoprolol, atenolol, propranolol
    - CCB- verapamil
  - Antidepressants
    - TCA-amitriptyline
  - Anticonvulsants
    - Gabapentin, valproic acid, topiramate
  - NSAIDs
  - BOTOX
    - onabotulinumtoxinA
    - Chronic migraines
    - 31 injection sites in the H&N (155-195 U) every 12 weeks

***Propranolol is used in children***
Treatment (cont’d)

- Nausea and Vomiting/Suppression of Vestibular System
  - Antiemetics - Promethazine, ondansteron
  - Antihistamines - Dimenhydrinate, meclizine
  - Metoclopramide
  - Benzodiazepines

- Physical Therapy
  - Effective for patients with VM
Tension Headaches

- Most common headache
  - Affects 80% of population
    - more common in women
    - Triggered by stress or anxiety
  - Headaches are bilateral, with a tightening/band-like sensation, in the frontotemporal region, radiates to occipital region and trapezius muscles.
  - Onset is gradual, pain is non-throbbing and constant.

Persistent muscle contraction is no longer considered the cause of the pain
Tension Headache

- **Subdivisions**
  - **Infrequent Episodic**
    - At least 10 episodes occurring <1 day/month on average (12 days/year)
  - **Frequent Episodic**
    - At least 10 episodes occurring on >1 but < 15 days/month for at least 3 months (≥12 and <180 days/year)
  - **Chronic**
    - Headache occurring on >15 days/month on average for > 3 months (≥180 days/year)
Tension Headaches

Table 5. ICHD III 2.1 Infrequent Episodic Tension-type Headache

A. At least 10 episodes of headache occurring on <1 day per month on average (<12 days per year) and fulfilling criteria B-D
B. Lasting from 30 minutes to 7 days
C. At least two of the following four characteristics:
   1. bilateral location
   2. pressing or tightening (non-pulsating) quality
   3. mild or moderate intensity
   4. not aggravated by routine physical activity such as walking or climbing stairs
D. Both of the following:
   1. no nausea or vomiting
   2. no more than one of photophobia or phonophobia
E. Not better accounted for by another ICHD-III diagnosis.

The International Classification of Headache Disorders, 3rd Ed.
Treatment

- Nonpharmacologic
  - Reassurance, muscle relaxation, stress management, biofeedback, physical therapy with thermal modulation or electrical stimulation.

- Pharmacological
  - Abortive
    - Acetaminophen, ASA, caffeine, NSAIDs
      - Should not be taken >2 days/week
  - Prophylactic
    - Reserved for patients with frequent headaches ≥2/wk
    - Amitriptyline-first line
    - Topiramate, valproate, venlafaxine

Botox was speculated to be effective in the treatment of tension type headaches, which had been previously attributed to increased muscle tone. Recent studies have failed to show a significant effect of the toxin in headache.
Cluster Headaches

- Less common than migraine or tension headaches
- Men>Women (3:1)
  - Middle age
- Headaches are unilateral, excruciating, and located around the eyes or in the maxilla.
- Associated with unilateral lacrimation, rhinorrhea, and injected conjunctiva, +/- ptosis and miosis.
- No aura or nausea.
- Pain lasts minutes - ~2-3 hours
Cluster Headaches

**Diagnostic criteria:**

A. At least 5 attacks fulfilling criteria B-D
B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15-180 minutes if untreated
C. Headache is accompanied by at least one of the following:
   1. ipsilateral conjunctival injection and/or lacrimation
   2. ipsilateral nasal congestion and/or rhinorrhoea
   3. ipsilateral eyelid oedema
   4. ipsilateral forehead and facial sweating
   5. ipsilateral miosis and/or ptosis
   6. a sense of restlessness or agitation
D. Attacks have a frequency from one every other day to 8 per day
E. Not attributed to another disorder
Cluster Headaches

- **A cluster period**
  - Segment of time during which attacks tend to occur several times per day for several weeks.
  - Remission can last months to years.

- **Triggers**
  - Alcohol, histamine, nitroglycerin, REM sleep, low oxygen saturation, OSA
    - Patients with nocturnal attacks should get a sleep study to r/o OSA
Cluster Headaches

- Etiology unknown
  - Circadian hormonal fluctuation
    - Hypothalamic dysfunction
  - Excitation of a nerve plexus in the carotid sheath and adventitia may increase trigeminal nerve discharge → facial pain
Cluster Headaches

- Treatment
  - Abortive
    - Inhalation of 100% $O_2$ x 10 minutes
    - Triptans
      - Sumitriptan 6 mg subcutaneously, relief in 15 min
      - Zolmitriptan PO, relief in 30 min
    - Dihydroergotamine IM or IV, relief in 30 min and 10 min respectfully
    - All of the above work by central vasoconstriction
  - Intranasal 4% lidocaine may also be effective
    - SPG
Cluster Headaches

● Prophylactic
  ◦ To be used early in the cluster period until headache free for 2 weeks
  ◦ Transitional prophylaxis
    • Used during a cluster period to suppress attacks
    • Prednisone, dexamethasone, ergotamine tartrate
  ◦ Maintenance prophylaxis
    • Used before and throughout the duration of the cluster period or in anticipation of a cluster season
    • CCB-nifedipine, verapamil
    • Low-dose ergotamine
    • Lithium carbonate
    • Methysergide

● Refractory cases
  • Trigeminal nerve block and sphenopalatine ganglion block
    • Most effective is Radiofrequency ablation of trigeminal ganglion
Trigeminal Neuralgia

- Most common in adults >50 y/o
- Women>Men (1.5:1)
- Incidence 4/100,000
- Etiology
  - Ignition Hypothesis of Devor
    - A trigeminal nerve injury (vascular compression, tumors, demyelinating plaques) induces physiological changes that lead to a population of hyperexcitable and functionally linked trigeminal sensory neurons.
    - The discharge of any individual neuron in this group can quickly spread to activate the entire population, resulting in a sudden synchronous discharge and a sudden jolt of pain.
Trigeminal Neuralgia (tic-douloureux)

- Recurrent episodes of unilateral, excruciating, stabbing pain occurring most often along V1 and V2.
  - Ipsilateral twitching may occur hence the name “painful-tic”.
- Episodes occur without warning, and can recur several to many times a day.
  - Lasting seconds to about 2 minutes.
- Light touching of the face, chewing, talking, shaving, smiling can precipitate an attack.
- Trigger zones
  - Nasolabial fold, lips, or gums.
- Pain attacks are stereotyped, recurring with the same distribution and intensity.
- There are no other neurological deficits, no numbness/paresthesias.
  - Sensation is intact
Trigeminal Neuralgia (tic-douloureux)

Diagnostic criteria from International Headache Society

<table>
<thead>
<tr>
<th>Trigeminal neuralgia</th>
</tr>
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<tbody>
<tr>
<td>A. Paroxysmal attacks of pain lasting from a fraction of a second to 2 minutes, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C</td>
</tr>
<tr>
<td>B. Pain has at least one of the following characteristics:</td>
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<tr>
<td>1. intense, sharp, superficial or stabbing</td>
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<td>2. precipitated from trigger areas or by trigger factors</td>
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<tr>
<td>C. Attacks are stereotyped in the individual patient</td>
</tr>
<tr>
<td>D. There is no clinically evident neurological deficit</td>
</tr>
<tr>
<td>E. Not attributed to another disorder</td>
</tr>
</tbody>
</table>
Trigeminal Neuralgia (tic-douloureux) – *(speaker notes for previous slide)*

- Recurrent episodes of unilateral, excruciating, stabbing pain occurring most often along V1 and V2. During an episode, ipsilateral twitching may occur hence the name “painful-tic”.

- Episodes occur without warning, and can recur several to many times a day. Each episodes lasting seconds to about 2 minutes. Light touching of the face, chewing, talking, shaving, smiling can precipitate an attack. Trigger zones are most common in the nasolabial fold, lips, or gums. Pain attacks stereotyped, recurring with the same distribution and intensity. There are no other neurological deficits, no numbness/paresthesias. Sensation is intact.
Trigeminal Neuralgia

- **Diagnosis**
  - **History**
  - **MRI of head**
    - Part of the initial evaluation
      - Vascular anomalies, aneurysm, tumors, MS
    - 3D MRI reconstruction and MR cisternography
      - Allows better identification of neurovascular compression and can even measure the volume of neurovascular compression at CPA and can predict prognosis of initial treatment
  - **LP**
    - Patients with MS are predisposed to TGN and 2% of patients with TGN have MS
    - Infectious process-neurosyphilis, meningitis
Trigeminal Neuralgia

- A good history on pain characteristics. In the majority of patients with TN clinical examination, imaging and labs are unremarkable but in a small group TN can be attributed to another disease process. Therefore MRI studies should be part of the initial disease process. One must pay special attention to MS plaques, tumors and vascular anomalies that may be the source of root compression.
Trigeminal Neuralgia

(B) A 69-year-old man with secondary progressive MS since age 43 and first symptoms of TN at age 63. At the time of MRI he had frequent episodes of severe electric shock-like pain in the area of the right eye and cheek quality. An oval-shaped brainstem lesion follows the course of the pontine fifth nerve fibers (arrow). Another small hyperintense pontine lesion can be seen anteriorly on the right.
Trigeminal Neuralgia

- (C) A 28-year-old man with first sensory symptoms of MS at age 27. Hypoesthesia in the distribution of the maxillary branch of the right trigeminal nerve gradually increased over 4 months. This further progressed and, besides resulting in permanent hypoesthesia in that area touching the upper lip and gingiva, caused electric shock-like shooting pains to this area. A pontine lesion following the course of the fifth nerve fascicular fibers is demonstrated on the trigeminal nuclei close to the floor of the fourth ventricle (long arrow). The plaque reaches to the junction of central and peripheral myelin in the nerve as it enters the pons (short arrow).
Trigeminal Neuralgia

- **Treatment**
  - Education and Reassurance
  - Pharmacological treatment
    - Must be used for **at least 2 weeks**
    - **Carbamazepine** (First line treatment)
      - 80% of pts experience initial sx control
      - 75% have long-term control
    - Baclofen can be just as affective
    - Oxcarbazepine is effective for those unresponsive to carbamazepine
    - Phenytoin, clonazepam, valproic acid, gabapentin, NSAIDs
    - Amitriptyline
    - Opioids are ineffective and should be avoided
Trigeminal Neuralgia

Carbamazepine will provide symptomatic relief in the majority of the patients. Other agents have shown to be effective and can be used in combination. Any drug should be employed for at least 2 weeks
Trigeminal Neuralgia

Surgical treatment

- Microvascular decompression
  - Posterior cranial fossa approach
  - Artery is separated of the nerve (teflon)
    - Superior cerebellar artery most common

- Trigeminal radiofrequency rhizotomy
  - A radiofrequency needle is inserted through the foramen ovale under the guidance of fluoroscopy.

- Stereotactic radiosurgery/Gamma knife
  - Stereotactic methods are used to concentrate ionizing radiation on the trigeminal root entry zone
  - Currently recommended as 1st line noninvasive surgical technique
  - Median time for pain relief is 3 weeks

- Percutaneous injection-nerve block
  - Local anesthetic + steroid (0.25% Bupivacaine + triamcinolone)
  - ultrasound guided
  - Immediate relief
Trigeminal Neuralgia

- **Microvascular decompression-** This operation involves opening the skull behind the ear, and exposing the Cerebello-pontine Angle using microsurgical techniques. The trigeminal nerve is exposed, and the offending artery is separated off the nerve. To stop the artery from falling back on the nerve, a small piece of teflon is placed around the artery.
Trigeminal Neuralgia

- **Stereotactic radiosurgery** is a form of single fraction radiation therapy in which a stereotactic coordinate system is used for precise targeting. Gamma Knife is an accurate and precise method for delivering stereotactic radiosurgery. An MR image showing the trigeminal nerve. The inset shows targeting used in Gamma Knife radiosurgery for trigeminal neuralgia (the green line is the 20% isodose line and the yellow line is the 50% isodose line).
Sluder’s Neuralgia

- In 1908, Sluder described a symptom complex consisting of neuralgic, motor, sensory and gustatory manifestations attributed to the sphenopalatine ganglion.
- He believed that irritation of this ganglion resulted from the extension of inflammation from the sphenoid and the posterior ethmoid sinuses.
Sluder’s Neuralgia

- **Neurological Features**
  - “Lower half headache”
  - Ipsilateral pain
    - constant with exacerbations, cyclical or episodic
    - Duration: hours to days, daily
    - begins at the root of the nose, spreads ipsilaterally in and around the eye, frontotemporal area, cheek and teeth, beneath the zygoma to the ear and mastoid
    - Pain is most severe at a point 5cm posterior to the mastoid
    - It can extend to the occiput, neck, shoulder, shoulder blade and breast
Sluder’s Neuralgia

- **Sensory Signs**
  - Anesthesia of soft palate, oropharynx, tonsils, anterior lower part of the nose
  - Hyperesthesia along trigeminal nerve
  - Aura
    - Metallic or peculiar acid sense of taste before or during an attack

- **Motor Signs**
  - Higher palatine arch on the affected side
  - Uvula deviated to unaffected side
Sluder’s Neuralgia

- Parasympathetic Signs
  - Ipsilateral Lacrimation
  - Injected Conjunctiva
  - Nasal obstruction, inflamed nasal mucosa
  - Rhinorrhea
  - Increased salivation
Anatomy and Physiology of the Sphenopalatine ganglion

- Sluder’s description
  - Small triangular body placed deep within the sphenomaxillary fossa
  - Behind the posterior tip of the middle turbinate
  - 1-9mm from the lateral nasal wall and separated from the nasal cavity by mucous membrane and fatty tissue
Sluder’s Neuralgia

- Possible theories
  - Neurological mechanism
    - Trigeminal Nerve
      - There is an overlap between cervical and trigeminal root afferents fibers in the most caudal part of the spinal nucleus of trigeminal nerve
      - Thus, irritation of the pterygoplatine nerves can cause stimulation of trigeminal nerve and pain along the distribution of the maxillary nerve
    - Medial pterygoid nerve
      - Branch of mandibular nerve
      - Innervates tensor veli palatini
      - Stimulation of this nerve → tension of soft palate/higher palatine arch
    - Parasympathetic fibers
      - Lacrimal Nucleus (parasympathetic nucleus of facial nerve), receives afferent fibers from sensory nuclei of trigeminal nerve for reflex lacrimation secondary to irritation of the cornea
Sluder’s Neuralgia

- Lacrimal Nucleus is used to refer to a portion of the superior salivary nucleus, a parasympathetic nucleus of the facial nerve, which is situated in the lower part of the pons, it receives afferent fibers from sensory nuclei of trigeminal nerve for reflex lacrimation 2/2 irritation of the cornea or conjunctiva.
Sluder’s Neuralgia

- Vascular mechanism
  - Vasodilators
    - Vasoactive intestinal peptide (VIP)
    - Nicotinamide-adenine dinucleotide phosphate (NADP) diaphorase
    - Nitric Oxide (NO)
  - Zygomatic N.
    - Branch of maxillary n
    - Conveys sensory fibers and post-ganglionic parasympathetic fibers from SPG via the zygomaticotemporal n. to the lacrimal gland
    - Fibers abundant in NADP-diaaphorase
      - These fibers originate from SPG
      - Parasympathetic fibers are responsible for the secretory and vasodilatory changes in nasal cavity and migraine like vascular pain
Sluder’s Neuralgia

- The sphenopalatine ganglion neurons are rich in the following compounds: VIP, NADP and NO, these are known vasodilators in cerebral arteries.

- Relatively abundant NAPD-diaphorase positive fibers were found in the zygomatic nerve, a branch of the maxillary nerve, that conveys sensory fibers as well as post ganglionic parasympathetic fibers from the SPG via the zygomaticotemporal nerve to the lacrimal gland.

- NADP-diasphorase positive and NO containing nerves are known to induce non-androgenic, non-cholinergic vasodilation.
Sluder’s, classic migraine and cluster headaches share a neurovascular origin

- Sluder’s Headache/Classic migraine
  - He described the pain to be migraine-like and identified patients who complained of an aura preceding the development of a headache.
  - Intranasal lidocaine treatment during an aura in a patient suffering from migraines prevented a headache in 73 out of the 75 episodes in an 18 month period.
# Sluder’s Neuralgia

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Sluder’s syndrome$^3$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain quality and intensity</td>
<td>Burning or aching pain which was either a constant pain with exacerbations or a pain that stopped and reappeared cyclically or stopped and reappeared with stabbing sharpness.</td>
</tr>
<tr>
<td>Site</td>
<td>Typically unilateral pain starting at the root of nose, involving the cheek, eye, teeth, frontotemporal region, mastoid region. Maximum pain intensity was experienced 5 cm posterior to the mastoid. Occasionally bilateral pain.</td>
</tr>
<tr>
<td>Frequency</td>
<td>Attacks could last hours to several days. Attacks could occur daily.</td>
</tr>
</tbody>
</table>
Sluder’s Neuralgia

Classic migraine
[IASP (V-1)]
Throbbing, pulsating pain, mild to severe in intensity.

Typically unilateral pain beginning most commonly in fronto-temporal area, may involve whole hemicranium, alternating sides between or during an attack.

Attacks last 4–72 hours if unmodified by drugs. Attacks most commonly occur 1–4 times a month.
## Sluder’s Neuralgia

<table>
<thead>
<tr>
<th>Associated symptoms and signs</th>
<th>Sensory signs:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Anaesthesia of soft palate, pharynx, tonsils, nose.</td>
</tr>
<tr>
<td></td>
<td>Hyperesthesia along distribution of trigeminal nerve.</td>
</tr>
<tr>
<td></td>
<td>Motor signs:</td>
</tr>
<tr>
<td></td>
<td>Palatine arch higher on affected side, the deviation of uvula to normal side.</td>
</tr>
<tr>
<td></td>
<td>Parasympathetic signs:</td>
</tr>
<tr>
<td></td>
<td>Ipsilateral lacrimation, conjunctival injections, nasal obstruction, rhinorrhoea, serous nasal discharge, inflamed mucosa.</td>
</tr>
<tr>
<td></td>
<td>Gustatory signs:</td>
</tr>
<tr>
<td></td>
<td>Delayed or diminished perception of taste.</td>
</tr>
</tbody>
</table>

| Aura                          | Distorted sense of taste described as ‘metallic’ or ‘peculiar acid’ before or during an attack. |
Sluder’s Neuralgia

Anorexia, nausea, vomiting, photophobia and phonophobia. Redness and swelling of the mucous membrane of the nose and conjunctival injection may also occur with migraine.

Visual disturbances; unilateral paresthesia of hand and mouth or mild paresis; dysarthria and aphasic disturbance occurred before or during an attack.
**Sluder’s/Cluster headache**

- **Similarities**
  - Pain distribution
  - Autonomic manifestations
  - Precipitating factors

- **Differences**
  - Pain in cluster headaches are generally mediated by ophthalmic division of trigeminal nerve, in Sluder’s pain is usually Maxillary
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain quality and intensity</td>
<td>Burning or aching pain which was either a constant pain with exacerbations or a pain that stopped and reappeared cyclically or stopped and reappeared with stabbing sharpness.</td>
<td>Excruciating severe attacks of constant stabbing, burning or even throbbing pain.</td>
</tr>
<tr>
<td>Site</td>
<td>Frequency</td>
<td></td>
</tr>
<tr>
<td>-------------------------------------------</td>
<td>------------------------------------------</td>
<td></td>
</tr>
<tr>
<td>Typically unilateral pain starting at the root of nose, involving the cheek, eye, teeth, frontotemporal region, mastoid region. Maximum pain intensity was experienced 5 cm posterior to the mastoid. Occasionally bilateral pain.</td>
<td>Attacks could last hours to several days. Attacks could occur daily.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Attacks occur during cluster periods of 4–12 weeks. Attacks last 30 minutes to 2 hours and occur 1–3 times a day. Nocturnal attacks were typical. No attacks occur during the remission period, which last 6–18 months.</td>
<td></td>
</tr>
</tbody>
</table>
Sluder’s/Cluster headache

Associated symptoms and signs

Sensory signs:
- Anaesthesia of soft palate, pharynx, tonsils, nose.
- Hyperesthesia along distribution of trigeminal nerve.

Motor signs:
- Palatine arch higher on affected side, the deviation of uvula to normal side.

Parasympathetic signs:
- Ipsilateral lacrimation, conjunctival injections, nasal obstruction, rhinorrhoea, serous nasal discharge, inflamed mucosa.

Gustatory signs:
- Delayed or diminished perception of taste.

Aura
- Distorted sense of taste described as ‘metallic’ or ‘peculiar acid’ before or during an attack.

Sensory signs:
- Dysaesthesia on touching scalp hairs in ophthalmic division of trigeminal nerve, Photophobia.

Motor signs:
- Ipsilateral miosis or ptosis.

Parasympathetic signs:
- Ipsilateral lacrimation, conjunctival injection, nasal obstruction, rhinorrhoea.

Reduced heart rate, irregular in severe attacks. Nausea and vomiting may occur.
Sluder’s neuralgia as a cluster headache

- Current Classification categorized Sluder’s neuralgia as a cluster headache.
  - Several studies have shown that the SPG is involved in cluster headaches
    - The effect of intranasal cocaine and lidocaine on nitroglycerin induced cluster headache attacks were measured in a double blinded placebo study.
      - All patients treated with cocaine or lidocaine had complete resolution of symptoms
      - Pain intensity increased in those patients treated with saline
Sluder’s neuralgia as a cluster headache

- The following are studies support the hypothesis that the SPG is involved in cluster headaches
  - In a double blinded placebo study, a cluster headache attack was induced on 15 patients with history of cluster headaches using nitroglycerin.
  - Once pain intensity measured at least 5/10 after ~10 minutes, either a 10% cocaine HCL solution, a 10% lidocaine solution or saline was applied to area corresponding to sphenopalatine fossa.
Sluder’s neuralgia as a cluster headache

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Pain relieved by anaesthetization of SPG, ganglionectomy, clonazepam</th>
<th>Pain improved with triptans, pizotifen, ergot preparations, oxygen, 5HT₁ agonists, steroids, verapamil, lithium methysergide.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Precipitating factors</td>
<td>Alcohol, tobacco, changes in climate or exposure to drafts</td>
<td>Alcohol or smoking. Long lasting stress may predispose to bouts.</td>
</tr>
<tr>
<td>Prevalence</td>
<td>Rare</td>
<td>7 per 10 000 population</td>
</tr>
<tr>
<td>Sex ratio</td>
<td>Female : Male 2:1</td>
<td>Male : Female 4:1</td>
</tr>
<tr>
<td>Age of onset</td>
<td>30–50 years</td>
<td>18–40 years</td>
</tr>
</tbody>
</table>
Sluder’s neuralgia as a cluster headache

- In another study, the efficacy of SPG blockade in 66 patients who suffered from cluster headaches that did not respond to pharmacological management, showed significant therapeutic effect following radiofrequency treatment of SPG via an infratemporal zygomatic approach.
  - 60% of pts with episodic cluster headaches and 30% of pts with chronic cluster headaches experienced complete relief over a 12-70 month period.
Rhinogenic Headache

- Headache or facial pain secondary to mucosal contact points in the nasal cavity in the absence of inflammatory sinonasal disease, purulent discharge, nasal polyps, nasal mass, or hyperplastic mucosa
  - septal deviation contacting nasal wall
  - septum to middle turbinate
  - septum to inferior turbinate
  - concha bullosa
  - superior turbinate pneumatization
  - any other visualized mucosal contact point
Rhinogenic Headache

ICHDIll. Appendix 11.5.1 Mucosal Contact Point Headache

Diagnostic criteria:
A. Intermittent pain localized to the periorbital and medial canthal or temporozygomatic regions and fulfilling criteria C and D
B. Clinical, nasal endoscopic and/or CT imaging evidence of mucosal contact points without acute rhinosinusitis
C. Evidence that the pain can be attributed to mucosal contact based on at least one of the following:
   1. Pain corresponds to gravitational variations in mucosal congestion as the patient moves between upright and recumbent postures
   2. Abolition of pain within 5 minutes after diagnostic topical application of local anesthesia to the middle turbinate using placebo- or other controls\(^1\)
D. Pain resolves within 7 days, and does not recur, after surgical removal of mucosal contact points

Note:
1. Abolition of pain means complete relief of pain, indicated by a score of zero on a visual analogue scale (VAS).

Comment:
Der A11.5.1 Mucosal contact point headache is a new entry to the classification for which evidence is limited. Controlled trials are recommended to validate it, using the listed criteria for patient selection.
Rhinogenic Headache

- Most common diagnostic method used to identify possible surgical candidates has been application of topical anesthetics and decongestants to intranasal contact areas during a headache. Improvement of headache after decongestion test may predict the surgical success (FESS) in patients with rhinogenic headaches.
Rhinogenic Headache

- **Contact point pain**
  - Is mediated by the stimulation of intranasal receptors that are innervated by afferent C fibers of $V_1$ and $V_2$
  - **Substance P**
    - Neuropeptide found in nasal mucosa causes vasodilation, plasma extravasation and perivascular inflammation, resulting in pain. This mechanism is similar to that found in migraines.
    - can be released by multiple etiologies including pressure
Rhinogenic Headache

- Contact point pains are mediated by the stimulation of intranasal polymodal receptors that are innervated by afferent C fibers of ophthalmic and maxiallry branches of the trigeminal nerve. The pain is projected and felt in the cutaneous distributions of the corresponding dermatomes. Substance P is a neuropeptide found in nasal mucosa and is known to cause referred pain and local reflexes. These reflexes cause vasodilation, plasma extravasation, and perivascular inflammation similar to mechanism in migraine headaches.
Rhinogenic Headache

- **Surgical Outcomes**
  - Ramadan.
    - 23 patients with refractory headaches and contact points on CT and nasal endoscopy.
    - 15 patients underwent ESS and 8 refused surgery.
    - 9 (60%) of those patients that underwent surgery, reported improvement of their symptoms.
  - Bektas et al.
    - 36 patients with rhinogenic contact point headaches underwent ESS.
    - All patients reported a decrease in the intensity of pain post-operatively.
    - 19 (52.7%) experienced complete relief.
  - Several studies have reported successful treatment of rhinogenic headaches with surgical management of contact points.
First Bite Syndrome

- Netterville et al. 1998
  - Facial pain characterized by a severe cramping or spasm in the parotid region with the first bite of each meal that diminishes over the next several bites.
  - Pathogenesis
    - Loss of *sympathetic* input resulting in hypersensitivity of sympathetic receptors on the myoepithelial cells of the parotid gland. Cross stimulation by parasympathetic release of Ach is believed to cause hyperintense contraction of myoepithelial cell contraction throughout the gland, causing pain.
First Bite Syndrome

- FBS is debilitating syndrome that can have a significant impact on a patients quality of life. Pain can be so severe that patients are afraid to eat and end up with significant weight loss.
First Bite Syndrome

- **Pain**
  - Onset: 5-7 days after surgery
  - Duration: a few seconds
  - So severe some patients are afraid to eat and result with significant weight loss
  - Worse with first meal of the day, but present with every meal or when salivating/thinking about food

- **Potential sequela of surgery involving**
  - Infratemporal fossa
  - Parapharyngeal space
  - Deep lobe of the parotid
# Treatment

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Efficacy</th>
<th>Safety</th>
<th>Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dietary modification</td>
<td>Completely ineffective</td>
<td>Safe</td>
<td>Nettervile et al, 1998&lt;sup&gt;5&lt;/sup&gt;; Sims et al, 2012 (present article)</td>
</tr>
<tr>
<td>Pharmacological treatments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>NSAIDs</td>
<td>Completely ineffective</td>
<td>Relatively safe</td>
<td>Casserley et al, 2009&lt;sup&gt;11&lt;/sup&gt;; Cernea et al, 2006&lt;sup&gt;12&lt;/sup&gt;; Chiu et al, 2002&lt;sup&gt;13&lt;/sup&gt; (7 patients)</td>
</tr>
<tr>
<td>Gabapentin</td>
<td>Decreased duration but same intensity</td>
<td>Relatively safe</td>
<td>Philips et al, 2009&lt;sup&gt;14&lt;/sup&gt; (1 patient)</td>
</tr>
<tr>
<td>Pregabalin</td>
<td>Improvement in 2 wks, Asymptomatic at 18 mos</td>
<td>Relatively safe</td>
<td>Casserley et al, 2009&lt;sup&gt;11&lt;/sup&gt; (1 patient)</td>
</tr>
<tr>
<td>Carbamazepine</td>
<td>Conflicting: complete resolution in 1 report, ineffective in 3 reports</td>
<td>Relatively safe</td>
<td>Effective: Cernea et al, 2006&lt;sup&gt;12&lt;/sup&gt; (1 patient) Ineffective: Philips et al, 2009&lt;sup&gt;14&lt;/sup&gt;; Casserley et al, 2009&lt;sup&gt;11&lt;/sup&gt;; Chiu et al, 2002&lt;sup&gt;13&lt;/sup&gt; (3 patients)</td>
</tr>
<tr>
<td>Radiation therapy</td>
<td>Complete resolution after 7–10 mos</td>
<td>High morbidity</td>
<td>Costa et al, 2011&lt;sup&gt;15&lt;/sup&gt;, Chiu et al, 2002&lt;sup&gt;13&lt;/sup&gt; (3 patients)</td>
</tr>
<tr>
<td>Surgical treatments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tympanic neurectomy</td>
<td>Completely ineffective</td>
<td>Relatively high risk</td>
<td>Ali et al, 2008&lt;sup&gt;9&lt;/sup&gt;; Chiu et al, 2002&lt;sup&gt;13&lt;/sup&gt; (4 patients)</td>
</tr>
<tr>
<td>Auriculotemporal nerve resection</td>
<td>Effective initially, long term efficacy unknown</td>
<td>Relatively high risk</td>
<td>Nettervile et al, 1998&lt;sup&gt;5&lt;/sup&gt; (1 patient)</td>
</tr>
<tr>
<td>Total parotidectomy</td>
<td>Complete resolution</td>
<td>Relatively high risk</td>
<td>Deganello et al, 2011&lt;sup&gt;3&lt;/sup&gt;; Diercks et al, 2011&lt;sup&gt;2&lt;/sup&gt;; Lieberman et al, 2011&lt;sup&gt;4&lt;/sup&gt; (3 patients)</td>
</tr>
<tr>
<td>Botulinum toxin injections</td>
<td></td>
<td>Relatively safe: currently no reported complications</td>
<td>Lee et al, 2009&lt;sup&gt;10&lt;/sup&gt; (5 patients) Ali et al, 2008&lt;sup&gt;9&lt;/sup&gt;; Sims et al, 2012, (present article) (4 patients)</td>
</tr>
</tbody>
</table>

Abbreviation: NSAIDs, nonsteroidal anti-inflammatory drugs.

- Sims J and Suen J
Treatment

- A case report of 3 patients treated with botulin toxin and a literature review of other treatment modalities for FBS was done at the university of Arkansas.
- **Dietary modification** (avoid sour foods) ineffective,
- **Permanent solutions** Total Parotidectomy is the most effective but is also the most radical and most morbid as it can lead to facial nerve paralysis.
Treatment

- Radiation therapy also shows complete resolution but who would want to unnecessary side effects given other more tolerable/safer treatments.
- They believe local botox injection is a reasonable 1st line treatment for FBS. In their study, 2 of the 3 patients experienced complete relief when injecting 75 units of botox. The third patient had near-complete resolution.
Botox Treatment

- **Botox**
  - Inhibits the release of Ach from the presynaptic terminal at the **neuromuscular junction** by degrading SNAP-25, a membrane bound protein that mediates presynaptic vesicle fusion with plasma membrane.
  - In Sims et al they found that symptoms begin to return within 3-5 months, symptoms return gradually and most patients do not request another injection until 5-4 months have passed.
  - **Concerns?**
    - Risk of Facial Nerve paralysis is very small
      - Only the axons of the Facial Nerve travel through the parotid gland and no NMJ are present
    - Dry mouth
      - Botox may inhibit salivation from the injected gland but enough saliva can be produced by contralateral Parotid, SMGs and Sublingual glands.
Acute Herpes Zoster

- Most common nerves affected
  - Trigeminal N
    - Herpes zoster ophthalmicus ($V_1$) most common
  - Facial Nerve
    - Ramsey Hunt Syndrome
      - Acute facial paralysis
      - Vesicular lesions – external ear, meatus, preauricular skin, EAC
  - Cervical rootlets

- Presentation
  - Intense burning, stabbing pain in the involved nerve distribution
  - Followed within 1 week by herpetic eruption
  - Motor division involvement
Acute Herpes Zoster

- **Treatment**
  - Goal is minimize duration of the attack, decrease severity of pain, and prevent post-herpetic neuralgia
    - Acyclovir 800 mg 5 times/day x 7-10 days (Cheaper), Valacyclovir
      - Prevents post-herpetic neuralgia
    - Prednisone 40 mg and taper x ~7 days
      - Risk of virus dissemination in treatments >1 month or in immunocompromised
    - Pain medications

- **Prevention**
  - Vaccine
    - Zostavax for people >60 y/o
Chronic Post-herpetic Neuralgia

- Pain persists for >3 months
- Pain remains in the same distribution of the originally afflicted nerve
- More common and less likely to resolve spontaneously in patients >60 years of age
- Treatment:
  - Topical anesthetics
    - Lidocaine patches
    - EMLA-lidocaine/prilocaine topical
  - Gabapentin
  - Tricyclic antidepressants (Amitriptyline)

Exists when post-herpetic pain persists for >3 months, it remains in the distribution of the originally affected nerve.
Tolosa-Hunt Syndrome

- Tolosa, 1954
  - Severe continuous retro-orbital pain preceding or coinciding with ipsilateral ophthalmoplegia and spontaneous remission with recurrences without evidence of neurological findings outside the cavernous sinus and the orbit.

- Hunt, 1961
  - Reported the therapeutic effect of systemic steroids with prompt improvement of signs and symptoms.

- International Headache Society, 2005
  - Idiopathic granulomatous process involving the cavernous sinus/superior orbital fissure or orbit diagnosed by MRI.
Tolosa-Hunt Syndrome

- Episodic unilateral orbital pain along V₁ with palsies in cranial nerves III, IV, VI and may also involve sympathetic fibers and optic nerve (II), if orbital apex is involved.
  - Pain is “severe”, “intense”, “boring”, “lancinating” or “stabbing”
  - Location: Periorbital, retro-orbital, frontal and temporal
  - Spontaneous Remission/recurrence
  - Episodes last about 8 weeks untreated and resolve within 3 days with steroids
    - Positive response to steroids have also been seen with chondromas, giant cell tumor, lymphoma, vasculitis etc.
Tolosa-Hunt Syndrome

- **Diagnosis**
  - MRI with contrast
    - Multiple views, coronal *
    - Isointense on T1 and Hypointense on T2
    - Enhancement is seen with IV contrast
  - High resolution CT scan
    - Less sensitive than MRI
  - Labs:
    - CBC, ACE, ANA, pANCA, anti-dsDNA Ab, etc.
    - ESR, C-reactive protein (usually increased)

- **Treatment**
  - Prednisone
Tolosa-Hunt Syndrome

An 18-year-old girl with a 2-week history of severe right-sided headache, diplopia and sluggish right papillary reaction.

A) Heterogeneous, hyperintense lesion expanding the right cavernous sinus and Meckel’s cave
B) Lesion extends into pituitary fossa
C) Mass showing dural enhancement, central area of necrosis
D) Follow-up MRI after 9 weeks of steroid therapy, shows complete resolution
Tolosa-Hunt Syndrome

- An 18-year-old girl with a 2-week history of severe right-sided headache, diplopia and sluggish right papillary reaction.
- (a) Coronal T2-weighted image shows a heterogeneous, predominantly hyperintense lesion (arrowheads) expanding the right cavernous sinus and Meckel’s cave.
- (b) Contrast-enhanced fat-saturated T1-weighted coronal image shows an enhancing lesion (arrowheads) expanding the right cavernous sinus, and extending into the pituitary fossa.
- (c) Contrast-enhanced fat-saturated T1-weighted transverse image shows a large mass extending from the right cavernous sinus anteriorly, into the Meckel’s cave and showing associated dural enhancement. Note the central necrotic area within the mass (*).
- (d) Follow-up MRI after 9 weeks of steroid therapy. Coronal T2-weighted image shows complete resolution of the lesion.
Eagle Syndrome

- Elongated styloid process
  - Impingement of carotid plexus or branches of CN IX
- Recurrent throat and neck pain exacerbated by swallowing
  - Associated symptoms
    - Otalgia, dysphagia, foreign body sensation
- Palpation of styloid process in the tonsillar fossa causes pain
- Common after tonsillectomy because of inflammatory changes
Eagle Syndrome

- **Diagnosis**
  - Lateral plain x-ray of the head
    - Styloid process >2.5 cm long
    - 4% of population have an elongated process, ~10% are Symptomatic
  - Injection of local anesthetic into tonsillar fossa provides relief and is diagnostic

- **Treatment**
  - NSAIDs, corticosteroid injection around styloid ligament
  - Shortening of calcified styloid ligament through tonsillar fossa.
Giant Cell Arteritis

- Presents as new-onset, constant localized temporal headache.
  - Pain is moderate to severe, burning, throbbing
  - Pain can be unilateral or bilateral
  - Associated symptoms: jaw claudication, weight loss, generalized fatigue, low grade fevers, malaise, and extremity pain.
- Visual symptoms
  - Involvement of ophthalmic artery causes anterior ischemic optic neuropathy
  - Blurring, scotomata, and sudden blindness
    - Blindness ~20% of patients

Vision loss can be unilateral or bilateral (less common), vision loss can be transient or permanent, partial or complete. Vision loss that lasts more than a few hours will not reverse.
Giant Cell Arteritis

- Most commonly in patients >50 y/o, (average age is 79)
  - women:men 2:1
  - Highest incidence in Scandinavians or Americans of Scandinavian descent
  - Associated with Polymyalgia rheumatica
- Physical Findings
  - Palpable thickened and tender scalp arteries with diminished or absent pulse.
  - Fundoscopic examination
    - Ischemic optic neuritis –slight pallor and edema optic disc, with scattered cotton-wool patches
Giant cell arteritis

- **Pathogenesis**
  - Segmental Inflammation of medium sized muscular arteries
    - “Skip” lesions
    - Superficial temporal > vertebral > ophthalmic > posterior ciliary arteries
  - Early
    - Internal or external elastic lamina or adventitia
  - Advanced disease
    - Intimal thickening with prominent cell infiltration
    - Transmural inflammation
Giant cell arteritis

Inflammation is found most often in medium sized muscular vessels that originate from the arch of the aorta. Inflammation affects arteries in a segmental fashion, leading to “skip lesions” within arteries.

The greatest frequency of severe involvement are Superficial temporal>vertebral>ophthalmic>posterior cilliary arteries. Early in disease, Collections of lymphocytes are confined to the region of the internal or external elastic lamina or adventitia. In more advanced disease you will see intimal thickening with prominent cellular infiltration possible transmural thickening.
Giant Cell Arteritis

- **Diagnosis**
  - ESR \( \geq 50 \text{mm/hr} \)
  - Superficial Temporal Artery biopsy
    - Segment should be at least 5 cm long
    - Granulomatous inflammation with multinucleated giant cells
  - Color Duplex ultrasonography
    - Clear Halo around the lumen
      - Sensitivity of 93%

- **Treatment**
  - Prednisone 40-60 mg/day (initial)
  - Then, 10-20 mg/day x several months while checking ESR
Giant Cell Arteritis

- An ESR of >50 mm/hr is almost always elevated with GCA, if the diagnosis is suspected, a temporal artery biopsy is essential to conform diagnosis. Inflammation of the artery is segmental therefore a 5 cm long segment of artery should be excised. If you get a negative biopsy result but suspicion is high consider a second biopsy of temporal artery or occipital artery.

- **Figure**: Giant cell arteritis. **A**, Cross-section of a temporal artery showing transmural inflammation with mononuclear cells and giant cells (hematoxylin and eosin, ×10). **B**, Higher-power (×100) view demonstrating giant cells infiltrating the media.
Giant Cell Arteritis

- Color duplex ultrasound examination of a swollen, tender temporal artery in a patient with giant cell arteritis. The variably thickened artery wall is visible as a clear “halo” (*solid arrows*) around the lumen in the center (*open arrow*).

- Treatment, initial dose should be 4-60 mg/day and should be continued until all reversible symptoms, signs, and labs abnormalities have reverted to normal. Gradually Reduce prednisone to about 10-20 md/day for several months, this will allow identification of the minimal suppression dse. If there is a flare up just increase dose by 10 mg/day.
Carotidynia

- International Headache Society Classification for Idiopathic Carotidynia, 1988

<table>
<thead>
<tr>
<th>A. At least one of the following overlying the carotid artery:</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Tenderness</td>
</tr>
<tr>
<td>2. Swelling</td>
</tr>
<tr>
<td>3. Increased pulsations</td>
</tr>
<tr>
<td>B. Appropriate investigations not revealing structural abnormality</td>
</tr>
<tr>
<td>C. Pain over the affected side of the neck; may project to the ipsilateral side of the head</td>
</tr>
<tr>
<td>D. A self-limiting syndrome of less than 2 weeks’ duration</td>
</tr>
</tbody>
</table>

- 2004, IHS removed it as a distinct pathological entity
  - Controversy still exists
    - A symptom of various heterogeneous causes of neck pain
    - Recent studies, have demonstrated it is not only a symptom but also a pathological entity with structural abnormalities and characteristic radiological findings
      - Inflammation of carotid fascia or adventitia

- Diagnosis
  - History
  - Response to treatment
  - MRI

- Treatment
  - NSAIDs
  - Self limited
Carotidynia

48-year-old man with left-sided carotidynia.

- **A**, Axial T1-weighted image (700/16/2 [TR/TE/excitations]) at the level of the distal common carotid artery. A skin marker has been placed over the area of swelling and tenderness. Note the abnormal soft-tissue signal surrounding the left carotid (arrow).

- **B**, Axial contrast-enhanced T1-weighted image (550/16/2) shows striking enhancement of the tissue surrounding the artery.

- **C**, Coronal contrast-enhanced T1-weighted image (550/16/2) shows the enhancement extending to the level of the carotid bifurcation (arrows). A lymph node is seen above the bifurcation (arrowhead).

- **D**, Axial contrast-enhanced T1-weighted image (500/12/2) obtained several months later, while the patient remained asymptomatic, shows resolution of the thick rim of enhancement.
Vestibular Migraine

- Vestibular Symptoms, as defined by Barany Society’s of Vestibular Symptoms
  - Spontaneous vertigo
    - Internal vertigo - false sensation of self motion
    - External vertigo - false sensation that the visual surround is spinning
  - Positional vertigo after change in head position
  - Head motion-induced vertigo, occurring during head motion
  - Head motion induced dizziness with nausea
    - (dizziness is characterized by a sensation of disturbed spatial orientation; other forms of dizziness are currently not included in the classification of vestibular migraine)
Anatomy and Physiology of the Sphenopalatine ganglion

- Parasympathetic ganglion
- Receives fibers from
  - Maxillary Nerve
    - Sensation to nose, palate, tonsils, gingivae
  - Greater Petrosal Nerve (branch CN VII)
    - Taste fibers → palate
    - Parasympathetic fibers
      - Post-synaptic fibers supply the lacrimal gland and mucosa of the palate, nasopharynx, nasal cavity
  - Deep Petrosal Nerve (branch of internal carotid plexus)
    - Carries post ganglionic sympathetic fibers from superior cervical sympathetic ganglion
      - these fibers do NOT synapse at SPG
      - Joins Maxillary N. → NC, palate, superior pharynx
    - Vidian Nerve
      - Greater Petrosal + Deep Petrosal join at the foramen lacerum → pterygoid canal → SPG
Anatomy and Physiology of the Sphenopalatine ganglion

- Lies just below the maxillary nerve near the sphenopalatine foramen

- **Greater petrosal** nerve a branch of the facial nerve carries taste and parasympathetic fibers. Taste fibers pass thru the sphenopalatine ganglion to the palate.

- **Taste fibers** pass through the sphenopalatine ganglion to the palate

- **Parasympathetic fibers** synapse in the ganglion and postsynaptic fibers supply the lacrimal gland and mucosa of the palate, Nasopharynx and nasal cavity

- **Deep petrosal nerve** is a branch of the internal carotid plexus and carries postganglionic fibers from nerve cell bodies in the superior cervical sympathetic ganglion. The sympathetic nerve fibers pass through the SPG without synapsing and join branches of the maxillary nerve where they are distributed to the nasal cavity, palate and superior part of the pharynx. The greater petrosal N and deep petrosal N join at the foramen lacerum to form the vidian nerve, which travels thru pterygoid canal to the SPG.
Tolosa-Hunt Syndrome

- 17/o F with new onset of diplopia, right sided headache and facial pain along V₁ and V₂. Patient was afebrile with right serous OM and Right abducens palsy. No mastoid tenderness.

- Labs: elevated ESR, CRP, CBC, ANA, ANCA, ACE, PPD-wnl

- Patient taken to OR for tympanocentesis and placement of tympanostomy tube.

- MEE Culture-no growth, no neutrophils.

- Patient started on Vanc and meropenum as there was a concern for Gradenigo Syndrome.
Tolosa-Hunt Syndrome
Tolosa-Hunt Syndrome

Contrast-enhanced T1-weighted images (A, B, C) and FLAIR images (D) obtained at time of initial presentation. (A) Enhancing soft tissue infiltrating the right cavernous sinus. (B) Enhancing soft tissue extending into Meckel's cave on the right. (C) Largest deposit of enhancing soft tissue (white open arrow) centered in the right carotid space, causing severe narrowing of the right internal carotid artery (black arrow) and internal jugular vein. (D) Right mastoid effusion secondary to obstruction of the right eustachian tube.
Tolosa-Hunt Syndrome

- Patient returned 10 days later after being discharged on IV antibiotics. MEE was gone but diplopia, facial pain and headache were unchanged.

- Patient was started on Prednisone. Within 1 week, patient’s eye pain, facial pain, headache and diplopia improved, completely resolution within 2 months.

- Diagnosis: Tolosa-Hunt Syndrome

- Repeat MRI showed near complete resolution of the enhancing mass
Tolosa-Hunt Syndrome
Tolosa-Hunt Syndrome

Contrast-enhanced T1-weighted images demonstrating temporal changes in the largest focus of enhancing soft tissue involving the right carotid space just lateral to the right longus coli muscle. (A) Initial presentation. Large enhancing soft tissue lesion centered in right carotid space causing narrowing of the distal cervical portion of the internal carotid artery and obliteration of the internal jugular vein. (B) 2 months later. Near complete resolution of the enhancing mass with mild residual enhancement and narrowing of the right cervical internal carotid artery. (C) 11 months after initial presentation (8–9 months after discontinuation of prednisone). Recurrent soft tissue mass centered in the right carotid space with recurrent narrowing of the cervical internal carotid artery. (D) Nearly two years after initial presentation. Complete interval resolution of the soft tissue mass in the right carotid space.
Other conditions that need to be considered in patients with herpes zoster

<table>
<thead>
<tr>
<th>Age</th>
<th>Possible Cause</th>
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<tr>
<td>0–18 years</td>
<td>AIDS/HIV, leukemia, Hodgkin's disease, tuberculosis</td>
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<tr>
<td>20–40 years</td>
<td>Steroid therapy, AIDS/HIV, diabetes mellitus, major operations (organ transplant), infection (viral, bacterial, fungal, or parasitic)</td>
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<tr>
<td>60–80 years</td>
<td>Malignant conditions should be the first possibility, and most of the above-mentioned factors could also be present</td>
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First Bite Syndrome

A retrospective cohort study by Morris et al.
- The cumulative incidence of FBS was 9.6%, patients undergoing sympathetic chain sacrifice, the incidence was 48.6% (P<.001); patients undergoing deep parotid lobe resection incidence was 38.4% (P<.001); patients undergoing PPS dissection incidence was 22.4% (P<.001)
- Incidence of FBS in Total parotidectomy was 0.8% (P<.001)
- Patients that received radiation therapy before or after surgery to the H&N had a 0.9% incidence of FBS
  - No radiation to H&N 17.5%
First Bite Syndrome

- Patients with exposure to radiation of the H&N were included. Patients that had previous surgery in these sites prior to referral to Memorial Sloan Kettering, procedures limited to biopsy and patients seen in clinic in a follow-up visit < 3 months were excluded.
First Bite Syndrome
First Bite Syndrome

The sympathetic pathway is a three-neuron chain, with origins in the posterolateral hypothalamus. First-order neurons descend the brainstem to synapse with second-order neurons located between the eighth cervical and second thoracic vertebrae. The second-order neurons exit the spinal cord and ascend along the cervical sympathetic chain to synapse with third-order (postganglionic) neurons within the superior cervical ganglion. The superior cervical ganglion is roughly 3 cm long, lies at the level of the second and third cervical vertebrae and posterior to the carotid sheath, between the internal carotid artery and the longus capitis muscle. Postganglionic sympathetics to the sweat glands and skin of the face/scalp leave the superior cervical ganglion to travel along the external carotid artery as a nerve plexus. The sympathetic supply to the parotid gland itself branches from the external carotid artery plexus to travel along the middle meningeal artery. Postganglionic nerves to the eyelid and orbit run in the adventitia of the internal carotid artery.
First Bite Syndrome

- Thus, sympathectomy to the parotid can exist even in the absence of Horner's syndrome.

- The parotid gland receives innervation from both sympathetics and parasympathetics. Although these nerves travel to the parotid gland by way of different nerves, once in the gland, individual axons pass together in the same Schwann-axon bundles to reach their destinations. Contrary to popular general belief, sympathetic and parasympathetics do not act antagonistically but rather in a collaborative association. Electron microscopy shows that salivary gland myoepithelial cells receive a dual innervation by sympathetic and parasympathetic nerves, and both impulses cause myoepithelial cell contraction.

- It is therefore reasonable to theorize that the absence of sympathetic input to the myoepithelial cells may result in a supersensitivity of these cells to parasympathetic innervation, especially when the two nerves travel in the same nerve bundle and innervate the same individual cells. This supersensitivity may result in a supramaximal contraction of the myoepithelial cells, resulting in the first bite pain. Subsequent bites may then desensitize this effect and cause an amelioration of symptoms, only to recur with the first bite of the next meal.
First Bite Syndrome

- All the patients with FBS had residual parotid gland tissue. Although three underwent excision of deep lobe tumors, residual parotid tissue was left in the superficial lobe and/or anterior periphery in deference to the preservation of the facial nerve.

- The exact amount of residual parotid tissue needed to elicit first bite symptoms is unknown but may be small, given that one of our neck dissection patients also underwent a superficial parotidectomy.
Sluder’s Headache/ Classic migraine

- He described the pain to be migraine-like and identified patients who complained of an aura preceding the development of a headache.

- Intranasal lidocaine treatment during an aura in a patient suffering from migraines prevented a headache in 73 out of the 75 episodes in an 18 month period
### Sluder’s Headache / Classic migraine

<table>
<thead>
<tr>
<th>Precipitating factors</th>
<th>酒,烟草,气候变化或暴露于 drafts</th>
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<tr>
<th>Treatment</th>
<th>痛缓解的麻醉、SPG 解剖学切除术、clonazepam</th>
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<th>频率</th>
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<th>性别比例</th>
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<tr>
<th>发病年龄</th>
<th>30–50 years</th>
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Sluder’s Headache/ Classic migraine

- Pain relieved by triptans, pizotifen, ergotamines, beta-blocking agents, calcium channel blocking agents, NSAIDs, 5HT\textsubscript{1} agonists.

- Numerous, including changes in stress, mood changes, relaxation, dietary factors.

- High
- Female > Male
- Childhood to 35 years
Carotid artery pain

- Carotid dissection
  - Head & neck pain on side of condition following trauma or can be spontaneous
  - Otalgia, Horner’s syndrome, Cranial Nerve palsies, pulsatile tinnitus.
  - Cerebral or retinal ischemia may develop
  - Diagnosis
    - MRI/MRA>U/S, intra-arterial angiography
  - Treatment
    - ASA or Warfarin x 6 months, allows recanalization of carotid artery and prevents ischemic complications.
    - +/- Surgery- balloon angioplasty, distal thrombectomy, ligation of the distal carotid artery, and external – internal carotid bypass

Surgery is for patient with recurrent ischemic strokes or are non responsive to conservative therapy
That was a very vast topic and I think you covered it well. I think that for us as otolaryngologists headache will always be a major symptom that we see and there are so many rhinologic causes and there are so many vestibular causes that can affect headache and once the patient has seen the neurologist and had other testing done, sometimes even before they see a neurologist, they are referred to us for evaluation. Sometimes headache and vertigo come together and it’s good to recognize that they are migraines as an entity because after ruling everything out the patients don’t fit with ischemic pictures; they don’t fit with Meniere’s disease; they don’t really have true aura’s as in migraine headache as such but they do respond to treatment and I actually have treated vestibular migraine on my own without neurological help. Minor to moderate complaints of headache with or without vertigo I have successfully treated with amitryptiline and dopimerate. But when you’re using such medication you want to be sure that you know about the side effects you counsel patients and as for me I generally don’t feel safe going beyond 50 mg. of amitryptiline ideally based on 75 mg. of timerate. In very very persistent headache I get a neurologist involved and with amitryptiline it’s important to remember that in older patients that it can cause urinary retention so that’s something patient should be counselled about. With Tifimerate, you don’t have as many autonomic side effects, but it causes weight gain and a lot of women don’t like that.
In cluster headache Dr. Ramos brought up a very important point that oxygen deprivation is a major causative factor in this and one classic question that you tend to get asked about in cluster headache is although we don’t usually use this in treatment of cluster headache as a first line is indomethacine works really well to stop or abort a cluster headache and to help with the inflammatory symptoms that you’re having. Of course, indomethacine has a lot of side effects so we really don’t use it as a long term medicine if we can avoid it. These days you can actually get ergotamine nasal spray and you can even get a compounded lidocaine nasal spray that tends to work well for cluster headache.

In trigeminal neuralgia, the effects of nitrous oxide on the pathogenesis of this condition. Nitrous oxide is coming up in otolaryngologic literature as a factor in the normal ventilation of the sinuses because it’s a big vasodilator. This has led to some recommendations that the ostia of the sinuses be kept to a certain size to maintain sinus health following surgery, and possibly to prevent or modify the occurrence of headache. There really isn’t any evidence-based data on this as yet.
Sluder headache is a much more involved syndrome, so we should not use it to denote rhinogenic headache. Identifying contact points in the office can be useful to isolate areas responsible to rhinogenic headache, which then can be remedied in the course of endoscopic endonasal sinus surgery although the basis for this is largely anecdotal and I have not seen any big studies which might tend to confirm this theory of the pathogenesis of rhinogenic headache.
8. Levine et al. An Otolaryngology, Neurology, Allergy and Primary Care Consensus on Diagnosis and Treatment of Sinus Headache. Otolaryngology Head and Neck surg 2006 134:516.