Trigeminal Autonomic Cephalalgias
Learning Objectives

• At the conclusion of this case, participants should
  – Know how to perform a differential diagnosis for trigeminal autonomic cephalalgias
  – Know the current therapeutic options for treatment of trigeminal autonomic cephalalgias
Medical History

- 35 yo female
- Pain in face and temple- right side also around eye and maxilla
- Daily pain for 20 minutes
- Early morning in timing
- Throbbing, stabbing, severity 5-8 (out of 10)
- Lacrimation on side of pain
- 2 years in duration
- Photophobia
Overview of Medical History

• No known aggravating factors
• Alleviating factors: high dose of antiinflammatory (12-16 tablets of ibuprofen daily) for partial relief of pain
• No other medical conditions
• Regular menses
• Negative history of surgeries or trauma
• Laboratory evaluations all normal
Family/Social History

Family history
• Married for 6 years, 2 children (4 yr, 5 yr)
• No known headache history in the family
• Parents and sister alive and healthy

Social history
• Works 40-hour weeks
• Administrative assistant
Review of Systems

• Review of systems:
  • GENERAL: SKIN: Normal
  • HEAD AND NECK: Normal
  • HEMATOLOGIC: Normal
  • CARDIOPULMONARY: Normal
  • GASTROINTESTINAL: Normal, denied abdominal pain
  • GENITOURINARY: Normal
  • MUSCULOSKELETAL: Normal
  • NEUROLOGIC: Normal, denied trauma
  • INFECTIOUS: Past history of measles and chickenpox, denied shingles
Physical Exam

- **General:** Patient Ht 64; Weight 140bs; BP 120/80; afebrile. Current pain level 0 /10.
- **Head/Neck:** minor conjunctival inflammation
- Temporomandibular joint examination – within normal limits (no clicking, normal ROM)
- Cervical spine examination – good range of motion, NT
- Lymph nodes: no lymphadenopathy, within normal limits
- **Heart:** Regular rhythm.
- **Lungs:** Clear
- **Abdomen:** Clear
- **Neuro:** Cranial nerve examination II – XII within normal limits with normal motor and sensory reflexes
Approaching the Patient with Daily Headache

Headache ≥15 d/mo

Excluding secondary headache

Classify based on duration

- **Short duration**
  - Cluster headache
  - Paroxysmal hemicranias
  - Hypnic headache
  - Trigeminal neuralgia
  - Other

- **Long duration**
  - Chronic daily headache
  - Chronic migraine
  - Chronic tension-type headache
  - Hemicrania continua
  - New persistent daily headache
  - Other

Approaching the Patient with Daily Headache

**Headache ≥15 d/mo**

Exclude secondary headache

*Secondary headache identified* → Diagnose

*Secondary headache excluded*

Classify primary headache based on duration

**Short duration**
- Cluster headache
- Paroxysmal hemicrania
- Hypnic headache
- Trigeminal neuralgia
- Other

**Long duration**
- Chronic daily headache
- Chronic migraine
- Chronic tension-type headache
- Hemicrania continua
- New persistent daily headache
- Other

Trigeminal Autonomic Cephalalgias

- Cluster headache
  - Episodic cluster headache
  - Chronic cluster headache
- Paroxysmal hemicrania
  - Episodic paroxysmal hemicrania
  - Chronic paroxysmal hemicrania (CPH)
- Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
- Probable trigeminal autonomic cephalalgia
  - Probable cluster headache
  - Probable paroxysmal hemicrania
  - Probable SUNCT
Question 1: Which of the following would be a possible diagnosis for this patient?

- Migraine
- Cluster headache
- Chronic paroxysmal hemicrania
Question 1: Which of the following would be a possible diagnosis for this patient?

- Migraine
- Cluster headache
- Chronic paroxysmal hemicrania
Clinical Features of Paroxysmal Hemicrania

• Severe, unilateral, orbital, supraorbital or temporal pain that lasts from 2 to 20 minutes in duration
• Headaches accompanied by:
  1. Ipsilateral conjunctival injection and or lacrimation
  2. Ipsilateral nasal congestion and or rhinorrhea
  3. Ipsilateral eyelid edema
  4. Ipsilateral forehead and facial sweating
  5. Ipsilateral miosis or ptosis.
• Frequency of about >5 per day for more than half the time
• Responds absolutely to indomethacin
Clinical Features of Cluster Headache

- Severe, unilateral, orbital, supraorbital or temporal pain that lasts from 15-180 minutes in duration
  1. Conjunctival injection, lacrimation
  2. Nasal congestion, rhinorrhea
  3. Eyelid edema
  4. Forehead and facial sweating
  5. Mitosis, ptosis
- Frequency of every other day to 8/day
Other Features of Cluster

- 75% males; 25% females
- Onset 20-40 yrs
- 5% of cases may be inherited
  - Autosomal dominant link
- 10-15% have chronic cluster
  - No remission
- Common triggers
  - Alcohol
  - Histamine
  - Nitroglycerine
- May occur during sleep
  - Up to 80% have obstructive sleep apnea
Question 2: Which of the following headache conditions may neuroimaging be considered for a differential diagnosis?

- Migraine
- Tension-type headache
- Cluster
- Paroxysmal hemicrania
- SUNCT
Question 2: Which of the following headache conditions may neuroimaging be considered for a differential diagnosis?

- Migraine
- Tension-type headache
- ? Cluster
- ✓ Paroxysmal hemicrania
- ✓ SUNCT
Question 3: Which treatments are effective in chronic paroxysmal hemicrania?

- Indomethacin
- Ibuprofen
- Topiramate
- Triptans
Question 3: Which treatments are effective in chronic paroxysmal hemicrania?

- Indomethacin
- Ibuprofen
- Topiramate
- Triptans
### Question 4: Which treatments are effective for cluster headache?

- Nonpharmacologic acute therapy
- Pharmacologic acute therapy
- Nonpharmacologic prophylactic therapy
- Pharmacologic prophylactic therapy
Question 4: Which treatments are effective for cluster headache?

- ✓ Nonpharmacologic acute therapy
- ✓ Pharmacologic acute therapy
- ❑ Nonpharmacologic prophylactic therapy
- ✓ Pharmacologic prophylactic therapy
Clinical Course

- Indomethacin 25 mg/d tid
  - Dose escalated 25 mg/wk to max daily dose of 150 mg (after 3-5 days)
- Record attack symptoms on diary
  - Frequency of attacks
  - Signs and symptoms associated with each attack
  - Treatment taken
  - Time to relief
  - Other important triggers or factors that she noticed.
Follow-up

3 months return to office with diary
- Diary was well completed for first 2 weeks, then relatively unpopulated due to good control
- Achieved pain-free within 48 hours of starting treatment
  - Some gastrointestinal upset
    - Prescribed proton pump inhibitor
    - Dose of indomethacin reduced to 125 mg/d

6 months - indomethacin discontinued due to gastrointestinal side effects
- Asked to return to office if headaches returned