Approach to Cranial Pain and Headache

Matthew Boyko

Reviewed by Dr. Farnaz Amoozegar

CLINICAL NEURO SCIENCES
CALGARY CANADA

Please view this presentation as a slideshow to listen to the accompanying audio. Use the arrow keys or mouse click to move to the next slide.

Objectives

- What is headache?
- Pain structures in the head
- Approach to headache (including history, physical, investigations)
- Primary headaches
- Secondary headaches

Headache

Noun | head•ache | \’hed-āk\

: an ache or pain in the head

: a difficult or annoying situation

merriam-webster.com

Epidemiology

- Very common!
- Actively affects nearly half of worldwide adult population
- Large socio-economic burden

Pain Sensitive Cranial Structures

Extracranial:

1. Skin
2. Fascia
3. Scalp vessels
4. Head and neck muscles
5. Skull and upper cervical vertebrae periosteum
6. Eyeball and orbital structures
7. Salivary glands
8. Teeth and gums
9. External auditory canal
10. Paranasal sinus mucus membranes
11. TMJ joints

Intracranial:

1. Dura mater
2. Dural arteries and large veins
3. Great venous sinuses
4. Pial arteries
5. Falx cerebri
6. Proximal sections of intracerebral arteries
7. Cranial nerves V, VII, IX, X

Classification

- Primary headaches

  Most common:

  – Tension type (31-69%)

  – Migraine (5-42%)

  – Chronic daily headache (3%)

- Secondary headaches

  Most common:

  – Medication overuse (3%)

Lebedeva et al. 2016

Approach

- Gold standard for diagnosis is history and general/neurological examination
- Most patients will have normal examination

Goal of your evaluation…

1. Rule out serious underlying pathology
2. Look for secondary causes
3. Then, determine type of primary headache

Some Serious Etiologies

INTRACRANIAL

- Subarachnoid hemorrhage (SAH)
- Ischemic stroke
- Cerebral Venous Sinus Thrombosis (CVST)
- Epidural hemorrhage
- Subdural hemorrhage (SDH)
- Mass lesion
- Idiopathic Intracranial Hypertension (IIH)
- Meningitis

EXTRACRANIAL

- Dissection
- Giant Cell Arteritis (GCA)
- Glaucoma
- Orbital cellulitis
- Pre-eclampsia
- HTH (dBP > 120 mmHg)
- CO poisoning

Secondary Headaches

- **Infection:** sinusitis, otitis media, meningitis, abscess, encephalitis, orbital cellulitis
- **Substance induced:** medications, substances induced/withdrawal, poisoning (CO)
- **Vascular:** GCA, SAH, SDH, ICH, CVST, TIA, RCVS, dissection
- **Homeostasis problems:** metabolic, hydrocephalus, exhaustion, HTN
- **Facial structures:** refractive errors, TMJ, trigeminal neuralgia
- **Trauma or degenerative:** cervical spondylosis, head injury, other trauma
- **Intracranial pressure disorders:** Idiopathic Intracranial Hypertension (high), Spontaneous Intracranial Hypotension (low), CSF leak (low)
- **Other:** psychiatric

Red Flags SNOOPPP

S – Systemic symptoms (fever, weight loss, travel, cancer, immunocompromised, pregnant, nuchal rigidity)

N – Neurological signs or symptoms

O – Older age (>50 yo)

O – Onset is sudden (thunderclap)

P – Prior headache history (now differs)

P – Progressive temporally or change in character

P – Pressure/postural (suspicious for raised ICP)

Thunderclap headache

- Severe head pain (worst headache of life)
- Abrupt onset
- Reaches maximal intensity in <1 min

Differential diagnosis for thunderclap

- Primary
  - Primary thunderclap headache
  - Primary exertional headache
  - Primary cough headache
  - Primary sexual headache
  - Migraine
- Secondary
  - Vascular
    - SAH
    - Sentinel headache
    - ICH
    - Ischemic stroke
    - Retroclival hematoma
    - Pituitary apoplexy
    - RCVS
    - CVST
    - Dissection
    - Hypertensive emergency
    - PRES
  - Non-vascular
    - Third ventricle colloid cyst
    - Spontaneous intracranial hypotension
    - Intracranial infection

History

- Start with open ended questions
- Patients will usually talk for less than 2 minutes if not interrupted (Langewitz et al. 2002).
- Listen to the patient
  - Builds patient-physician rapport
  - Creates empathy, which is therapeutic
- Patients can have more than one type of headache so listen carefully to distinguish each

- Pain history: OPQRST
  - Location?
  - Duration?
  - Frequency and periodicity?
- Preceding symptoms: Aura?
  - Visual (positive or negative), somatosensory, language disturbances, motor, brainstem
- Associated symptoms
  - Nausea, vomiting, photophobia, phonophobia, vision changes, autonomic changes, Horner syndrome, jaw claudication, fever, seizure
- Precipitating factors
  - Effects of exercise, lights, sounds, smell, cough, bending forward, laying still, standing upright
- Family history
  - Especially migraine and tension headaches
- Prior treatments (both acutely and prophylaxis)
  - Dose and duration?
  - Response?
  - Rule out medication overuse

General Physical Exam

- Mental status
- Vitals
  - Temp, HR, BP, RR, O2 saturation, BMI
- HEENT
  - Scalp or temporal artery tenderness/thickening
  - Localized infection
  - Signs of trauma
  - Conjunctival injection or lacrimation
  - Ocular bruit
  - Cervical spine or muscle tenderness
  - Meningismus
- Cardiac
- Pulmonary
- Abdomen
- Skin
  - Meningococcal rash
  - Herpes Zoster rash
  - Malar rash
  - Osler's nodes, Janeway lesions, Splinter hemorrhages
  - Café-au-lait spots
  - Cutaneous angiomas

Neurological Exam

- Mental Status
  - Level of alertness, confusion, language disturbances
- Cranial Nerves
  - Pupils, fundoscopy, visual acuity and fields, EOM, ptosis
- Motor
  - Bulk, tone, strength, reflexes, plantar reflexes
- Sensory
  - Pinprick, vibration, proprioception, neglect
- Coordination
  - Dysmetria, RAM
- Gait
  - Ataxia, shuffling gait, up from chair, Romberg

Investigations

If signs/symptoms suggesting secondary cause:

- CRP, and platelets → elevated in GCA
- Carboxyhemoglobin → elevated in CO poisoning
- Alcohol and drug screen
- TSH → elevated in hypothyroidism
- Plasma/urine catecholamines and metanephrines → rule out pheochromocytoma
- CSF → rule out meningitis, SAH, encephalitis, leptomeningeal cancer, lymphoma

Imaging

If signs/symptoms suggesting secondary cause:

- MRI → encephalopathies, white matter disease, meninges, posterior fossa pathology, infarct, venous thrombosis, neoplasm, infection
- CT → fractures, acute hemorrhage, sinus or mastoid air cell disease
- X-rays (rarely done)

PRIMARY HEADACHES

Case 1

28 yo Female presents with right sided severe headaches lasting 1-2 days and during which she cannot work and prefers to lay still in a dark room. Pain builds up gradually over 1 hour.

Migraine

Migraine

- Affects 4 000 000 Canadians
- Prevalence 23-26% in women and 7.8-10% in men
- Results in loss of 7 000 000 workdays annually in Canada
- Subtypes: migraine with brainstem aura, hemiplegic migraine, retinal migraine, chronic migraine, vestibular migraine, menstrual migraine

Migraine without aura

A. At least five attacks fulfilling criteria B-D
B. Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)
C. At least two of the following:
   1. Unilateral
   2. Pulsating
   3. Moderate to severe intensity
   4. Aggravated by or causing avoidance of physical activity
D. At least one of the following:
   1. Nausea and/or vomiting
   2. Photophobia and phonophobia
E. Not better accounted for by another diagnosis

ICHD 3rd Ed. Beta. 2013

Clinical Features: Four Stages

1. Prodrome
   - Affective or vegetative symptoms for 24-48 hours prior to headache onset
   - Occurs in 60% of people
2. Aura
   - Gradual development of one or more positive or negative neurologic symptoms lasting no more than 1 hour
   - Occurs in approximately 1/3 of patients with migraine
3. Headache
   - Usually throbbing, unilateral, severe, intensity increasing over several hours with associated features
4. Postdrome
   - After the headache has resolved patients may feel exhaustion, fatigue, or pain upon sudden head movements

Migraine Pathophysiology

1. Cortical Spreading Depression
   - Self-propagating wave of neuronal depolarization thought to cause the aura, activate trigeminal nerve afferents, and alter BBB permeability
2. Trigeminovascular System
   - Neurons relay information from intracranial blood vessels and meninges to the Trigeminal nucleus caudalis, which leads to central sensitization and release of vasoactive peptides

Treatment Approach

1. Stratified
2. Step care within attacks
3. Step care across attacks
4. Hybrid

Treatment

Non-pharmacological:

1. Relaxation training, Biofeedback, CBT, physiotherapy, massage therapy
2. Avoid triggers
3. Behavioral/lifestyle modifications

Pharmacological:

1. Migraine specific – triptans and dihydroergotamine
2. Non-specific agents – ASA, ibuprofen, naproxen, diclofenac, acetaminophen
3. Adjuncts – metoclopramide, domperidone

Case 2

48 yo Male presents with bilateral squeezing headache 1-2 times per day over the past year that lasts 2-4 hours. Tightness also in neck. Headaches benefit from massages.

Tension Headache

Infrequent episodic tension-type headache

A. At least 10 episodes of headache on <1 day per month on average and fulfilling criteria B-D
B. Lasting 30 min to 7 days
C. At least 2 of the following:
   1. Bilateral
   2. Pressing or tightening
   3. Mild to moderate
   4. Not aggravated by physical activity
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 diagnosis

ICHD 3rd Ed. Beta. 2013

Tension Treatment

Non-pharmacological:

- Relaxation training, Biofeedback, CBT, physiotherapy, massage therapy

Pharmacological:

- Abortive: Ibuprofen 800 mg, Naproxen 500-825 mg, Tylenol 1g
- Prophylactic: TCAs @ 10 mg qhs and titrated as needed, Gabapentin and Pregabalin are used off-label
- Mixed results for muscle relaxants and botox

Case 3

52 yo Male presents with excruciating episodes of pain behind his right eye that lasts 2 hours and feels like stabbing. Pain is so severe that he will cry out then pace around his room. Occasionally associated runny nose.

Cluster Headache

Cluster headache

A. At least 5 attacks fulfilling criteria B-D
B. Severe unilateral orbital, supraorbital, temporal pain lasting 15-180 min (when untreated)
C. Either or both of the following:
   1. ≥1 of the following ipsilateral to the headache:
      a) Conjuctival injection or lacrimation
      b) Nasal congestion or rhinorrhea
      c) Eyelid edema
      d) Facial sweating
      e) Facial flushing
      f) Fullness in the ear
      g) Miosis or ptosis
   2. Restlessness or agitation
D. Attacks occur every other day and 8 per day for more than half of the time
E. Not better accounted for by another diagnosis

ICHD 3rd Ed. Beta. 2013

Cluster Treatment

Abortive Treatment:

- Oxygen via face mask @ 12-15L/min for >15 min (high flow rate)
- Sumatriptan 6 mg SC, then can repeat in 2 hours (max 2 doses in 24h)
- Zolmitriptan 5 mg IN, then can repeat in 2 hours (max 2 doses in 24h)
- DHE 0.5-1.0 mg SC
- Lidocaine solution IN on cotton swab

Prophylactic Treatment: (need at least 1-2 weeks of treatment)

- Verapamil 160-480 mg per day divided into tid dosing (monitor with ECG)
- Lithium (monitor with regular blood-work for CBC, lytes, Cr)
- Topiramate 25-200 mg per day
- Valproic acid 600-2000 mg per day

Bridging Treatment while starting Prophylaxis:

- Prednisone 80 mg per day x 2 days then taper

Trigeminal Autonomic Cephalalgias

CLUSTER HEADACHE
- 1-8 per day
- 15-180 min

PAROXYSMAL HEMICRANIA
- 1-40 per day
- 2-30 min
- Indomethacin sensitive

SUNCT and SUNA
- 1-200 per day
- 1-600 sec

HEMICRANIA CONTINUA
- Continuous pain
- Months to years
- Indomethacin sensitive

SECONDARY HEADACHES

Case 4

35 yo Female with previous headaches now presents with mild headache that occurs daily. Headache dull, bilateral, and 6-8/10. Has been alternating Tylenol and Advil every day with minimal effect.

Medication Overuse Headache

Medication Overuse Headache

A. Headache occurring ≥15 days per month in a patient with a pre-existing headache disorder
B. Regular overuse for >3 months of ≥1 drugs taken to treat headache
C. Not better accounted for by another diagnosis

ICHD 3rd Ed. Beta. 2013

MOH Treatment

- Should resolve after the overuse is stopped!
- Bridge Therapy:
  - Long acting NSAIDs
  - Prednisone
- Prophylactic Therapy:
  - Tailored to the underlying type of headache

Case 5

79 yo Male with HTN, T2DM, and dyslipidemia presents with a new right sided headache. Pain affecting his chewing. No nausea. No photophobia. No weakness. No vision changes.

Giant Cell Arteritis

Giant Cell Arteritis

- Inflammation of medium and large arteries
- Incidence ranges from 10-29 per 100 000 per year
- Incidence significantly increases after 50 yo
- Over half associated with Polymyalgia rheumatica
- 50% of patients with Amaurosis fugax will become partially or totally blind if left untreated

Presentation

Symptoms:

- Headache
- Scalp tenderness
- Jaw claudication
- Tenderness over temporal artery
- Amaurosis fugax
- PMR

Signs:

- Temporal artery inflammation (49% of patients)
- Large artery bruits
- Diminished pulses
- Infarct from vasculitis of other vessels

Diagnosis and Treatment

- Check for ↑CRP, ↓hemoglobin, ↑platelets
- Diagnosis made with Temporal artery biopsy for histology
- Start corticosteroids!
  - Prednisone 40-60 mg daily x 1 month then taper
  - Symptoms and laboratory abnormalities should normalize in days
  - Continue to monitor CRP for relapses; titrate prednisone accordingly
  - May start immunomodulatory medication in consult with Rheumatology
  - Complications such as neuropathies or stroke may still occur months after starting steroids

Case 6

32 yo Female with recent weight gain presents with severe bilateral headache for the past 6 months. Worse when she lays down or coughs. Upon further questioning, she endorses that her vision will become transiently blurry.

Idiopathic Intracranial Hypertension

IIH

Often affects overweight females with menstrual irregularities

Symptoms: headache (consistent with ↑ICP), transient visual obscurations, pulsatile tinnitus, diplopia, sustained vision loss.

Diagnosis: rule out CVST with imaging, elevated CSF opening pressure, otherwise normal CSF studies

Treatment:

1. Weight loss
2. Carbonic anhydrase inhibitors: acetazolamide 500 mg bid and titrate up as tolerated
3. Large volume LP taps
4. VP shunting or optic nerve sheath fenestration for refractory cases or progressive vision loss

Conclusion

- Evaluation of headache relies on the history
- Headaches are largely classified as primary vs. secondary
- The clinician must rule out potential serious underlying causes
- Pursue further investigations or imaging if warranted

References

- Bradley's Neurology in Clinical Practice 6th Edition
- Uptodate.com
- Lebedeva ER, Kobzeva NR, Gilev D, Olesen J. Prevalence of primary headache disorders diagnosed according to ICHD-3 beta in three different social groups. *Cephalalgia.* 2016; 36(6): 579-588.
- Worthington I, Pringsheim T, Gawel MJ, Gladstone J, Cooper P, Dilli E, Aube M, Leroux E, Becker WJ. Canadian headache society guideline: Acute drug therapy for migraine headache. *Can J Neurol Sci.* 2013; 40.
- International Classification of Headache Disorders (3rd Edition Beta).
- Benoliel R and Eliav E. Primary headache disorders. *Dent Clin N Am.* 2013; 57: 513-539.

- Thank you Theo Mobach for providing previous slides