Evaluation and Management of Headaches in Children

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2 cases

• 14 year old with headaches once weekly
• 13 year old with daily headache now worsening

Patient 1

• 14 year old left handed girl with headaches
• Occur once weekly for one year
• More severe with menses
• Pain throbbing and frontal in location
• Associated with nausea, phono- and photophobia
• Worse with exercise
• Lasts 3-4 hours and often resolves with sleep
• History of motion sickness and sleep walking
• Mother and grandmother have “sick” headaches
• Examination normal
• Based on the history, diagnosis is______?
Migraine

- Stereotyped paroxysmal headache associated with:
  - Lasting 4-72hrs
  - Unilateral
  - Pulsating quality
  - Moderate to severe pain severity
  - Aggravated by physical exertion
  - Associated with phono-, osmo- and/or photophobia
  - Nausea and vomiting
  - +/- aura

- Differences in children
  - Not always >4hrs
  - Not always unilateral
  - Pain not always well described
  - Children will often seek sleep

Epidemiology

- Bille (1962)
  - 8993 children
  - 75% children will have notable headache by 15 yrs
  - Migraine 3.9% overall
    - 1.7% in 7 year olds
    - 5.3% in 15 year olds
- Isik (2009)
  - 2669 children
  - 5-13 yrs
  - 40% with debilitating headache
- Lateef et al (2009)
  - US survey
  - 10,816 individuals 4-18 yrs
  - 17% report frequent or severe headache in prior 12 mos
  - Prevalence similar in pre-pubertal children
  - Highest prevalence – adolescent girls – 27%
Migraine and Severe headaches in Children: Prevalence

Lateef TM et al. J Child Neurol 2009

Co-Morbidities

- Asthma
- Hayfever
- Frequent otitis media
- ADD
- Learning disability
- Stuttering

Not associated with hypertension or diabetes

Lateef TM et al. J Child Neurol, 2009
Migraine and variants

- Common
- Classic
- Basilar
- Ophthalmoplegic*
- Retinal*

- Alice-in-Wonderland syndrome
- Acute confusional migraine
- Migraine equivalents
  - Cyclic vomiting
  - Benign paroxysmal vertigo
  - Benign paroxysmal torticollis

*rare in children

Pathophysiology

Migraine trigger leads to cascade of events through the trigeminal-vascular system. Release of various peptides CGRP, substance P leads to pain, increased blood flow and neurogenic inflammation. Aura is associated with a spreading wave of cortical depression leading to “neuronal silence.”

Headache Evaluation

- History
  - Including co-morbidities
  - Medication history
  - Sleep history
- Physical examination
- Neurological examination
  - Look at fundus

- Neuroimaging not indicated unless:
  - Worrisome history
  - Focal deficits
  - Papilledema
  - MRI
- Lab studies
  - Only as clinically indicated
Other headaches syndromes

- Tension type
- Chronic daily headache
- Trigeminal autonomic cephalgias*
  - Cluster headache
  - Paroxysmal hemicrania
  - Short lasting unilateral neuralgiform headache attacks with conjunctival tearing and injection (SUNCT)
- Secondary headache
  *rare in children

Migraine Treatment

- 3 modalities
  - Lifestyle changes
  - Rescue therapies
  - Preventative therapies

Lifestyle changes

- Good sleep hygiene
- Appropriate diet
- Maintain hydration
- Exercise
- Smoking cessation
Lifestyle changes - Evidence

- Smoking, obesity, low physical activity associated with chronic headaches in adolescents*

*Robberstad et al. Neurology 2010

Rescue therapy

- NSAIDs
- Triptans
- Infusion therapy

NSAIDs
- Ibuprofen*
- Naproxen sodium
- Diclofenac sodium
- Diclofenac potassium
- Ketorolac

Triptans
- Sumatriptan* - inj, NS, tab
- Rizatriptan* - tab
- Zolmitriptan* - NS, tab
- Almotriptan* - tab
- Eletriptan* - tab
- Naratriptan - tab
- Frovatriptan - tab

*studied in children

Number of studies indicate that ibuprofen, sumatriptan, rizatriptan, zolmitriptan, almotriptan are effective and safe in children
Rescue Therapy: Infusion/injection

- Dihydroergotamine/metoclopramide
- Sodium valproate
- Keterolac IM/IV
- Dexamethasone

Preventative therapy

- Anti-depressants
- Beta-blockers
- Anti-epileptic drugs
- Anti-histamines
- Calcium channel blockers
- Nutraceuticals

Preventative

<table>
<thead>
<tr>
<th>Anti-depressants</th>
<th>Betablockers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amtriptyline</td>
<td>Propranolol</td>
</tr>
<tr>
<td>Mirtazepine</td>
<td>Atenolol</td>
</tr>
<tr>
<td>Trazodone</td>
<td></td>
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<tr>
<td>SSRIs</td>
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</tbody>
</table>
Preventative

Anti-epileptic drugs
- Topiramate
- Valproate
- Zonisamide
- Levetiracetam

Others
- Anti-histamines
  - Cyproheptadine
- Calcium channel blockers
  - Verapamil
  - Nimodipine
  - Flunarazine
- Nutraceuticals
  - CoQ
  - Riboflavin
  - Magnesium Oxide
- Biobehavioral

Outcome – generally good

Brna (2005)
- 20 year follow-up
  - 27% headache free
  - 33% had tension headache
  - 17% had migraine
  - 23% had both
  - 66% felt they were improved
  - 88% continued to have moderate to severe headache

Kabbouche (2005)
- 1.2.5 year follow-up at multidisciplinary clinic
- Improvements reported by 94%, 85% and 94% at the 1,2 and 5 year intervals
- Reduction in frequency, severity, and school absences

Case 2
- 12 year old right handed boy with headache
  - 3 weeks ago – Headache, fever
  - Diagnosed with sinusitis and ABX started
    - Parents deny symptoms URI
  - Pain 8/10 and persistent
  - Intermittent vomiting without pattern
  - Re-evaluated at one week and changed to Augmentin
  - One week prior to eval, seen in ER because of diplopia and left facial weakness – CT, CBC, ESR, monospot - normal; Lyme titer sent
  - Doxycycline and Prednisone started
  - Day of eval, persistent headache, intermittent horizontal diplopia
  - Headache worse in am
  - Parents report no one looked at his fundus
Case 2

- Physical examination
  - Afebrile, obese
  - No nuchal rigidity
  - Otherwise normal

- Neurological examination
  - Bilateral VI nerve palsy
  - Minimal left facial weakness
  - Fundi – next slide
  - No other abnormalities

Case 2

- What is your concern?
- What is your differential?
  - Chronic or partially treated meningitis
  - Mass lesion – abscess vs. tumor
  - Sinus thrombosis
  - Benign intracranial hypertension
Case 2: Evaluation

- MRI/MRV: normal
- LP:
  - Opening pressure 550mm
  - Fluid clear
  - 1 wbc, 0 rbc
  - Glucose 67 mg/dl; Protein 18mg/dl
  - Cytology normal
  - Lyme PCR normal

And the diagnosis is?

Benign Intracranial hypertension
Pseudotumor cerebrii

Pseudotumor cerebri

- Syndrome of disturbed CSF hydrodynamics without identifiable cause
  - Normal neurological examination (except for abducens palsy)
  - No evidence of hydrocephalus, cerebral mass lesion, vascular abnormality such as sinus thrombosis
  - Elevated ICP by lumbar puncture with normal indices
    - Neonates >70mm
    - <8 years with papilledema >180mm
    - > 8yrs or less than 8 yr w/o papilledema >250mm
  - Primary or Secondary
Secondary Pseudotumor cerebri – Associated conditions

- Female gender
- Obesity
- Drugs:
  - Minocycline, tetracycline
  - Hormonal – GH, OCPs, Corticosteroid treatment and withdrawal
  - Hypervitaminosis A
  - Lithium
- Otitis media and craniofacial infections
  - Usually with sinus thrombosis
- Anemia
- Endocrine conditions
  - Thyroid replacement
  - Addison disease
  - Menarche
- Renourishing of malnourished children

Pseudotumor cerebri

- Epidemiology
  - More common in women
  - 1/100,000 to 13/100,000 (women 20-44yrs who are overweight)
  - Female to male ratio: 4-8:1
  - Prepubertal: no gender difference
  - More common in obese individuals
  - Can be familial

Pseudotumor cerebri - Clinical features

- Headache
  - Chronic, acute or paroxysmal
- Strabismus secondary to VIth nerve palsy
- Nuchal rigidity
- Irritability
- Emesis
- Visual loss (late)

- Atypical
  - Cranial nerve palsy
  - Apathy
  - Seizures
  - Parathesias
  - Facial and limb numbness
  - INO
  - Nystagmus
  - Pulsatile tinnitus

Patient may be asymptomatic – papilledema identified on routine eye exam
Pseudotumor cerebri – Diagnosis/Evaluation

- History including medications
- Lumbar puncture with opening pressure
  - Normal indices
- MRI with angiography (venous)
- Ophthalmologic evaluation
  - Visual acuity
  - Formal visual fields
  - If necessary, orbital ultrasound or fluorescein angiography

Mimickers of Papilledema

- Optic nerve drusen
- Myelinated optic nerve fibers

Useful clues suggesting pseudopapilledema:
- Presence of SVPs
- Absence of optic cup in mild to moderately “swollen” disk
- Presence of anomalous vasculature at the disk
- Non-enlargement of the blind spot on formal perimetry

Pseudotumor Treatment

- Weight loss if overweight
- Discontinue offending agents
- Mediations
  - Acetazolamide - starting with 25mg/kg/day divided t.i.d. to q.i.d
  - Topiramate, Furosemide
  - Corticosteroids
- Surgical
  - Lumboperitoneal shunt, VP shunt
  - Optic nerve sheath fenestration
  - Subtemporal decompression (rare)

INDICATION FOR SURGERY: PROGRESSIVE VISUAL LOSS!!
Pseudotumor - outcome

- Self limited in most pediatric patients
  - Relapse rate similar children ~20%
  - Can occur months after treatment discontinued
  - 24% can have permanent visual loss

Patient 2

- Patient started on Acetozolamide
- Ophtho evaluation
  - Significant constriction of visual fields
  - OD 20/70, OS 20/60
  - Acetozolamide increased
  - F/U progressive visual loss
  - Referred for ONSF – no improvement
  - LP shunt placed

Patient 2

- Following shunt
  - Vision stabilized and improved
  - Headache resolved
  - Did not lose weight
  - One year later, develops recurrent headache
    - No papilledema or vision changes
    - Headaches have migrainous quality
    - Responds to migraine prophylaxis
Keys to headache evaluation

- History, history history
- Complete examination
  - Evaluation of disks is key
- Tailor therapy to diagnosis
  - Multimodal
- Remind parents: Be patient!
  - Response to therapy is not immediate

QUESTIONS?