Case-Based Diagnosis and Management of Headache Disorders

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Case – 1

- 34-year-old teacher
- Referred for recurrent headaches for the last five years taking only occasional ibuprofen to treat the headaches
- Headaches occurred on average twice a month, lasting a day or two at a time
- Bilateral, over her forehead, and sometimes accompanied by neck pain
- Her job was stressful and wondered whether that might be causing the headaches Physical and neurologic examinations were normal except for mild tenderness on palpation over the posterior neck and upper trapezius muscles.
- Told that she probably had “tension headaches” and had been referred for physical therapy but did not find that treatment effective.

A.

- **How can migraine be reliably distinguished from tension-type headache in this patient?**

Case – 1

- Careful evaluation of the full headache history
  - Characteristic features of migraine
    - Nausea
    - Vomiting
    - Photophobia
    - Phonophobia or
    - Worsening with physical activity

Diagnostic features of migraine without aura: common pitfalls

"Silver platter" migraine features	"Not so obvious" migraine history
Duration of 4-72 hours	Duration uncertain because patient treats early or falls asleep. Shorter headaches often seen in children.
Unilateral (often over the temple)*	Bilateral, posterior location of pain or prominent complaints of neck pain often lead to a diagnosis of tension-type headache, but neck pain occurs in almost three-quarters of migraine attacks
Thrombbing*	Not all patients who otherwise have clear-cut migraine report throbbing pain; for many patients the throbbing quality of the pain is obvious only in fully developed, longer duration headaches, so patients who treat early or fall asleep may not experience this. Be alert for synonymous descriptions such as "pounding" or "with my heartbeat"
Moderate to severe pain intensity*	In migraine attacks that are treated or do not progress, pain may never reach severe intensity. Differences in pain reporting behaviours and pain perception among patients may affect patient ratings of pain intensity
Aggravated by or causing avoidance of physical activity*	Sedentary patients may not have noticed this feature of their headaches
Nausea and/or vomiting#	Vomiting is prominent in children with migraine, but often lessens as patients get older or headache frequency increases. Decreased appetite may be present instead
Photophobia and phonophobia#	Sensitivity to light or sound may become apparent only in headaches that have a chance to develop fully; these symptoms may not develop in milder "forme fruste" or treated attacks

* Only two of these four features are required for a diagnosis of migraine.
# Only one of these two features is required for a diagnosis of migraine. At least five attacks meeting criteria are required before migraine without aura can be diagnosed.

Case – 1

B. Most predictive of migraine as opposed to tension-type headache are nausea, photo and phonophobia.

◦ Further questioning revealed

- Loss of appetite, photophobia which support a diagnosis of migraine.

C. Discussion

- Both Migraine and Tension type headache have muscle pain or tenderness in the neck and shoulder.
- Both have lower thresholds for experiencing pain with pressure on muscles than do people without headache.
- Emotional stress - headache trigger in both.
- Headache diary can distinguish between the two.

Case – 1

D. Diagnosis

- Migraine without aura

E. Tip

Migraine often presents with features assumed to be highly characteristic of tension-type headache. Most patients consulting physicians for troublesome headaches have migraine and not tension-type headache.

Case – 2

- 27-year-old woman

- Frequent headaches characterized by retro-orbital pain, always on the right, up to four hours in duration

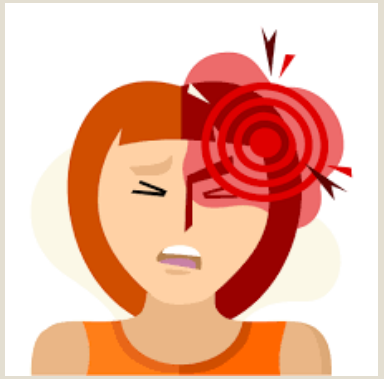
- With the most severe pain she also noticed tearing and redness of the right eye. When the pain was milder she preferred to be lying down

- Associated photophobia and some nausea but no aura

- Headaches occur very frequently, up to twice a day, and other times when she would have several weeks without a headache.

- Had been told she had "cluster migraines." One of her other physicians thought she had cluster headache and started verapamil, which had improved the frequency of headaches only mildly.

A.



- **Are this patient's autonomic feactures helpful in making a diagnosis?**

Case – 2

- Autonomic features (lacrimation,conjunctival injection, forehead or facial sweating, miosis, ptosis, eyelid edema, rhinorrhea, or nasal congestion) are defining characteristic of the headaches classified as trigeminal autonomic cephalagias (TACs), including cluster headache.
  - ICHD-3 beta does not require autonomic feactures to diagnose cluster headache if the patient has the characteristic restlessness or physical agitation that are seen in cluster headache.
  - 56% of patients with migraine had cranial autonomic symptoms. (One prospective study comparing migraine and clustert headache)

B.

Is “cluster migraine” an appropriate diagnosis in this patient?

Case – 2

- Cluster migraine is not a recognized term in any edition of the ICHD, including ICHD-3 beta.

C. Discussion

- Autonomic activation during headache is a function of the trigeminal autonomic reflex. Trigeminal autonomic reflex occurs when nociceptive input from trigeminal system, as occurs during headache, stimulate trigeminocervical complex. This inturn stimulates the superior salivatory nuclus, which give rise to autonomic fibers providing innervation to the head.

- Autonomic symptoms are most commonly seen in the TACs, but can also occur in migraine.

- Autonomic symptoms in migraine:
  - bilateral,
  - mild or moderate in severity, and
  - occur inconsistently with headache attacks
  - Lacrimation is the autonomic symptom most commonly associated with migraines
- Autonomic symptoms associated with cluster headache:
  - unilateral 80% of the time and
  - almost always ipsilateral to the headache when they are unilateral
- Dishinguishing between "runs" of migraines and a bout of true cluster headache can be challenging. A very careful history taken with attention to headache duration, location of pain, other associated feactures such as phono or photophobia and frequency.

Distinguishing migraine from cluster headache

Pain features	Migraine	Cluster headache
Location	Often unilateral over the temple or forehead area but may be bilateral	Strictly unilateral; typically highly localized to behind one eye
Duration of attack	4-72 hours (adults)	15 minutes to 3 hours*
Frequency of attacks	Sporadic. Can "cluster" in bunches but rarely follow the distinctive pattern of true cluster headache	Attacks can occur once every other day up to eight times a day (for more than half of the time during an active cluster bout – headache frequency may increase or taper slowly at the beginning or end of a bout)*
Associated features	Nausea, vomiting, photo and phonophobia	Agitation or restlessness OR one of the following seven symptoms or signs must occur on the side of the headache: (1) eye redness or tearing; (2) nasal congestion or runny nose; (3) edema of the eyelid; (4) sweating of the forehead and face; (5) flushing of the forehead and face; (6) a feeling of ear fullness; (7) decreased pipil size or ptosis*
Sex ratio	Females > males	Males > females
Behaviour during attack	Quiet; prefer to lie quietly in a dark room	Agitated, restless
Temporal features	Attacks typically occur at ramdom and are not easily predictable	Attacks commonly occur at specific times of the day or night. Named for the way "cluster" together occuring daily or almost daily for 2- to 3-month bouts. In episodic cluster headache these bouts are separated by peroids of remission lasting at least a month; in chronic cluster headache remissions do not occur or are shorter than a month

*According to ICHD-3 beta, all of these criteria must be met in at least five attacks in order to make a diagnosis of cluster headache

Case – 2

- It can also be helpful to determine if the patient has headaches outside of the identified "cluster" periods.
- Episodic cluster headache – rarely has any headaches at all outside of their periods
- Migraine – usually has occasional isolated headaches even during their "good periods"
- The true cluster headache pattern, with peroids of headaches occuring from one every other day to eight per day followed by complete remission peroids, is almost never seen in migraine.

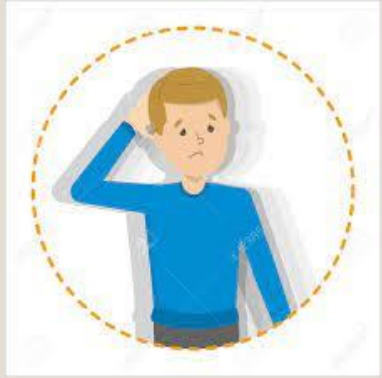
D. Diagnosis

Migraine without aura

E. Tip

Migraine headaches can present with autonomic features in up to half of patients. “Cluster migraine” is a confusing term which should be avoided.

Case – 3



- 28-year-old man
- Prior history of hypertension with a complaint of sudden visual changes
- Described a “flashing” in his peripheral vision that started in one eye and spread to both over a period of about ten minutes. This gradually evolved into as a "flashing floater" along the edges of his vision bilaterally and was followed by blurred vision in the originally affected eye.
- Lasted a total of 20 minutes and gradually disappeared
- Concerned about the possibility of a transient ischemic attack, and underwent an evaluation that included a carotid ultrasound, a CT scan of his head, and consultation by an ophthalmology specialist
- Results of these investigations were normal, and he was discharged home with instructions to take 81 mg of aspirin daily and follow up in the outpatient neurology clinic.
- Elicited a history of several similar episodes of visual disturbance when the patient was in his teens described as periods of “fluttery” peripheral vision lasting 5-25 minutes and followed by a period of mild head “soreness”
- Occurred sporadically over a period of several years and then disappeared. The patient had sought an ophthalmologic examination at the time, but no abnormalities were found.

A.

What is the differential diagnosis of these visual symptoms?

Case – 3

- D/Dx of isolated visual symptoms
  - Intraocular pathology
  - Retinal detachment
  - TIAs
  - Migraine aura
  - Seizures

} visual symptoms that resolves spontaneously

Visual phenomena

- ‘Positive visual phenomena’ - false visual images or distortions of normal vision (sparkling lights, lines or geometric shapes)
  - spots of colour
  - shimmering vision
- ‘Negative visual phenomena’ - visual deficits (dark or blind spots)

Selected feactures that distinguish the visual aura of migraine from ischaemic events

Visual aura of migraine	Ischaemic events
• Visual phenomena begin in the periphery and develop gradually	• Sudden onset of visual loss
• Both positive and negative visual phenomena are present	• Negative visual symptoms predominate
• Reversible, with duration of event no longer than an hour	• Short duration (typically no more than 15 minutes)
• If other aura symptoms are present, they occur in sequence	• If other neurologic symptoms are present, they occur simultaneously with visual phenomena
• Similar events on multiple occasions	• No prior history of similar events
• Symptoms are followed by headache (usually within 15 minutes)	• Headache is not a common associated feacture

Case – 3

- **Patient experienced a mixture of positive and negative visual features that developed gradually and were followed by headache → characteristic of visual aura**

◦ Patients often find it difficult to describe visual aura.

◦ Obtaining a previous history of such attacks can be very helpful. (H/O similar episodes at a younger age)

B.

What bedside test may have helped clarily the diagnosis?

The Visual Aura Rating Scale (VARS)

Visual symptom characteristic	Risk score
Duration 5 – 60 minutes	3
Develops gradually over 5 or more minutes	2
Scotoma	2
Zig-zag line (fortification spectrum)	2
Unilateral (homonymous)	1
Maximum VARS score	10
Migraine with aura diagnosis	≥ 5

Adapted frm: Eriken *et al*. The Visual Aura Rating Scale (VARS) for migraine aura diagnosis. *Cephalagia*. 2005;10:801-10, with permission.

C. Discussion

- Aura occurs in about a third of patients with migraine.
  - Visual aura – 99%
  - Sensory aura – 31%
  - Aura involving language symptoms (S/A aphasia) – 18%
  - Motor aura – rare (recognised as hemiplegic migraine)
- Due to the phenomenon of cortical spreading depression, which is an orderly wave of depolarisation that spreads anteriorly across the cerebral cortex.
- Followed by a prolonged peroid during which neurons struggle to repolarise and is followed by a small reduction in regional cerebral blood flow
- The wave of neuronal depolarisation spreads at a rate of 2-5 mm per minute, which correlates closely with the evolution of visual or sensory symptoms during an aura

Case – 3

D. Diagnosis

- Typical aura with headache

E. Tip

- Failure to elicit historical feactures consistent with migraine aura can lead to an erroneous diagnosis of cerebral ischaemia in patients with aura

Summary of level of evidence and special considerations for the acute treatment for migraine

Acute treatment for migraine	AHS evidence statement	Strength of evidence from meta-analysis	Special considerations
Acetaminophen (for non-incapacitating attacks)	Level A Established efficacy	Moderate	Limit use in patients with impaired hepatic function
NSAIDs Aspirin, celecoxib, diclofenac, ibuprofen, naproxen	Level A Established efficacy	Moderate	Limit use in patients with impaired renal function, history of gastrointestinal bleeding or concurrent antiplatelet therapy
Combination analgesics Acetaminophen/aspirin/caffeine	Level A Established efficacy	N/A	See above
Antiemetics Prochlorperazine, metoclopramide, promethazine, droperidol	Level B (Intravenous)	Low	Potential development of extrapyramidal symptoms and QTc prolomgation
Triptans (migraine-specific) Sumatriptan, rizatriptan, naratriptan, eletriptan, almotriptan, zolmitriptan, frovatriptan (oral, nasal spray, subcutaneous injection)	Level A Established efficacy	High	Contraindicated in patients with ischaemic stroke, coronary artery disease, uncontrolled hypertension
Ergots DHE, ergotamine	Level A (nasal spray) Level B, C (other forms of DHE and oral ergotamine) Established efficacy	High Moderate	Contraindicated in patients with peripheral arterial disease, uncontrolled hypertension, ischemic heart disease, or coronary artery vasospasms Cannot be used within 24 h of a triptan intake
Gepants (migraine-specific) Ubrogepant, rimegepant (zevegepant)	Established efficacy	Moderate to high	Not contraindicated in patients with the above-listed contraindications for triptans and ergots Not associated with medication overuse headache
Ditan (migraine-specific) Lasmiditan	Established efficacy	High	Not contraindicated in patients with the above-listed contraindications for triptans and ergots Does not cause vasoconstriction Should not drive within 8 h of medication intake
Opioid and butalbital	Level C	Insufficient	Not recommended

References

- Ailani J, Burch RC, Robbins MS; Board of Directors of the American Headache Society. The American Headache Society Consensus Statement: Update on integrating new migraine treatments into clinical practice. *Headache*. 2021 Jul;61(7):1021-1039. doi: 10.1111/head.14153. Epub 2021 Jun 23. PMID: 34160823.
- Loder, E., Burch, R.C. and Rizzoli, P. (2014) *Common pitfalls in the Evaluation and management of headache: Case-based learning*. Cambridge, United Kingdom: Cambridge University Press.