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REVIEW ARTICLE

The Ophthalmologists' Headache

Shibal Bhartiya

Senior Consultant, Glaucoma Services, Fortis Memorial Hospital, Gurgaon, Haryana, India

Patients with headaches often present to the ophthalmologist or are referred by their primary care providers, with or without eye-related symptoms. Given that headaches may be portentous of a spectrum of clinical conditions, some of which life or vision threatening, a thorough history and examination is critical. This includes a cranial nerve and peripheral neurological examination, imaging studies, and a neurology opinion, whenever indicated. This is also true for headaches that present with ophthalmic signs and symptoms. The associated visual phenomenon may help in the formulation of a differential diagnosis and management plan.

Headache History

It is important to note the following, to distinguish the phenotype and underlying cause of headache, as well as formulate the management plans for the disease:

a. Severity of pain
b. Duration of pain;
c. Frequency of pain
d. Nature of pain (throbbing, pulsating, stabbing, and band-like constriction)
e. Anatomic location of pain (frontal, temporal, face, and occipital)
f. Prodromal features (visual, sensory or motor aura, and dysarthria)
g. Associated features (photophobia, phonophobia, osmophobia, nausea or vomiting, visual disturbances, and jaw claudication)
h. Aggravating factors (stress, sleep deprivation, prolonged near effort, physical activity, environmental triggers such as cold and bright lights, and menstruation).

Types of headaches

Headaches may be classified into primary or secondary headache disorders and facial pains or neuralgias. The four primary headaches are migraine, tension type headaches, trigeminal autonomic cephalgias (characterized by cluster headaches), and a diverse group of atypical headaches called "other primary headache disorders."[1] The International Classification of Headache Disorders, 3rd Edition (ICHD-3)[2,3] provides a systematic classification of headaches with explicit diagnostic criteria based on the documented longitudinal history. These headache phenotypes may or may not be exclusive for any disorder and often overlap. A brief summary of the most common phenotypes of primary headaches is discussed here. The characteristics of secondary headaches have been discussed in subsequent sections.

Migraine without aura

Diagnostic criteria:

A. Recurrent headaches, at least five attacks fulfilling criteria B-D
B. Headache episode lasting 4–72 h
C. With 2 or more out of the following four characteristics:
    1. Unilateral
    2. Pulsating quality
    3. Pain moderate or severe
    4. Aggravation by, or resulting in disruption of/avoiding routine activities
D. Headache associated with 1 or more the following:
    1. Nausea and/or vomiting
    2. Photophobia and phonophobia
E. Not better accounted for by another ICHD-3 diagnosis.

Migraine with aura

A. At least two attacks fulfilling criteria B and C
B. One or more of aura symptoms which are temporary. These may be visual, motor, sensory, retinal, related to speech or language, or the brainstem
C. At least three of the following six characteristics:
    1. One or more aura symptoms spread gradually over 5 min
    2. At least two aura symptoms occurring in succession
    3. Each aura symptom lasting for 5 min to an hour
    4. Minimum one unilateral aura symptom
    5. Minimum one positive aura symptom
    6. Aura accompanied by, or followed by headache within an hour
D. Not better accounted for by another ICHD-3 diagnosis.

Tension-type headache

A. Ten or more episodes of headache occurring for one day each month on average (<12 days a year) are classified as infrequent. If occurring for 1–14 days a month on average for more than 3 months (Between 12 and 180 days/year), the headaches are classified as Frequent Tension type Headaches,

if fulfilling criteria B–D
B. Lasting from 30 min to 7 days
C. Two or more of the following: bilaterality, pressing or tightening (non-pulsating), mild-to-moderate pain, not aggravated by routine physical activities
D. Absence of nausea/vomiting and no more than one of photophobia or phonophobia
E. Not better accounted for by another ICHD-3 diagnosis.

Cluster headache

Trigeminal autonomic cephalalgia
A. Five or more attacks fulfilling criteria B–D
B. Severe or very severe unilateral orbital, supraorbital, and/or temporal pain lasting from 15–180 min
C. One or more of the following characteristics:
   1. At least one of the following autonomic dysfunction on the same side as the headache: conjunctival injection and/or lacrimation, nasal congestion and/or rhinorrhea, eyelid swelling, forehead and facial sweating, miosis, and/or ptosis
   2. A sense of restlessness or agitation
D. From once every alternate day to as many as eight times per day
E. Not better accounted for by another ICHD-3 diagnosis.

Indications for Neuroimaging

Experts believe that all patients with headaches do not require neuroimaging, since only a minority of these patients has an underlying secondary etiology.[4-8] Moreover, incidental findings, atypical features, and benign pathologies are commonly seen on imaging studies, and may cause significant anxiety for both the patient and the doctor.

In case of any of these red flags noted while evaluation of the patient with a headache, immediate or urgent investigations, including a neurology opinion, are warranted, to rule out an underlying pathology. The earlier SNOOPS mnemonic of 2003 (where S stood for secondary illnesses, HIV or history of neoplasia) was modified in 2018 into the currently used SNNOOP10 [Table 1].[9]

Table 1: SNNOOP10

- Systemic symptoms (e.g. fever, weight loss)
- Neoplasm history
- Neurological signs/symptoms
- Onset (sudden)
- Older age (>50 years)
- 10 "P"s—
  1. Pattern change/recent onset of headache,
  2. Positional headache,
  3. Precipitated by coughing/sneezing,
  4. Papilledema,
  5. Progressive headache,
  6. Pregnancy/Puerperium,
  7. Painful eye with autonomic features,
  8. Posttraumatic onset,
  9. Pathology of immune system,
  10. Painkiller overuse or new drug at onset of headache

Ocular Causes of Headache

According to the ICHD-3,[2] eye-related causes can be broadly classified into:

1. Headache attributed to acute angle-closure glaucoma: Angle closure disease can cause episodic headaches[10-12] and acute angle closure can mimic an acute attack of migraine with autonomic symptoms.
2. Trochlear headache: Peritrochlear inflammation or dysfunction results in a frontal and/or periorbital headache, with or without associated eye pain. The pain worsens with movements of the eye, especially supraduction,[13,14] and may radiate to the ipsilateral cranium.
3. Headache attributed to ocular inflammatory disorder: Ocular inflammatory disorders such as uveitis,[15] scleritis, and even conjunctivitis may present similarly, with pain and photophobia. The clinical course of the headache parallels that of the inflammatory pathology, and is usually ipsilateral. Any improvement in the latter makes the headache better, and vice versa. Meibomian gland disease and dry eyes may exacerbate migraines. Optic neuritis may also be associated with headaches, in addition to painful ocular movements.
4. Headache attributed to refractive error, especially important with increasing use of electronic devices and associated eye strain.[17-19] These include headaches associated with refractive error, convergence insufficiency (CI), and accommodative spasm (AS). The intensity of the headache is more severe with higher refractive errors and usually occurs toward the end of the day, and with increasing near effort. Patients with CI and AS may present with diplopia and strabismus, as also a sudden onset diminution of vision (induced myopia).[19]

Eye Findings in Patients with Headache

Visual disturbances associated with headache

Visual disturbances may include photophobia, amaurosis fugax, Uhthoff's phenomenon, transient visual obscuration (TVO), palinopsia, "visual snow", and aura.[20]

a. Monocular transient visual loss (TVL) or amaurosis fugax is caused due to the interruption of blood flow to the central retinal/ophthalmic artery, resulting in a blackout. This may affect the entire, or half the field of vision, and is often described a curtain sweeping across the visual field. This is usually transient, lasting from less than a minute, to up to an hour. This may be caused by either an embolic thrombus originating from an atherosclerotic plaque in the internal carotid artery, from the heart or aorta, or from carotid artery dissection. These patients are at a higher risk of stroke, and therefore, must undergo a complete cardiac evaluation. Giant

cell arteritis (GCA) may also cause of TVL, in association with headache. It is also important to differentiate between TVL caused by migraine aura or transient ischemic attack.

b. Note: TVO is the graying or blacking out in the vision, which lasts typically for a few seconds, especially when a patient moves or bends forward. This is usually seen in patients with papilledema.
c. The Uhthoff phenomenon is the blurring of the vision after vigorous exercise or activities that increase body temperature, like a hot water bath. This is usually seen in patients of optic neuritis, with or without an underlying demyelinating disorder such as multiple sclerosis and neuromyelitis optica.
d. Visual Aura typically develops gradually from the periphery, over 15 min, and may last up to an hour. Symptoms include positive scotomas, phosphenes, and visual hallucinations such ase flashes of light, zigzags, scintillating scotomas, teleopsia, and metamorphopsia. These may be seen in migraine, and non-convulsive epilepsy, and are remarkably short lived in the latter, lasting for a few seconds only. Retinal migraine is characterized by recurrent attacks of monocular visual disturbances, including scintillating scotomata and TVL along with migrainous headaches. These patients often report a diplopia and third nerve involvement. The disorder has now been renamed Recurrent Painful Ophthalmic Neuropathy and is considered a demyelinating disorder.[20]

Ophthalmoplegia, papilledema, and ocular ischemia

a. Ophthalmoplegic migraine is a rare disease, characterized by childhood onset, recurrent headaches, and ophthalmoplegia, usually involving the third cranial nerve.[21]
b. Papilledema may be noted in case of Idiopathic Intracranial Hypertension (IIH), as well as ICSOLs. Cavernous sinus thrombosis and superior orbital fissure syndrome with ophthalmoplegia may also be associated with the papilledema.

Note:

I. Idiopathic Intracranial Hypertension: Or pseudotumor cerebri is caused by increase in intracranial pressure with no identifiable organic lesion or cerebrospinal fluid composition abnormality and chronic headache. Almost all patients have papilledema, which may result in progressive visual deterioration. It typically affects young women of working age, and the headache is worse on waking up and on physical activity. It may be associated with visual field loss, TVO, optic atrophy, photophobia, phonophobia, nausea and vomiting, cranial nerve palsies and diplopia, cognitive deficits, olfactory deficits, and tinnitus.[22,23]
II. Cavernous Sinus thrombosis is also associated with ophthalmoplegia, since the third, fourth, and sixth cranial nerves pass through the cavernous sinus. Associated symptoms include corneal and facial sensory loss, conjunctival chemosis; Horner's syndrome and proptosis Causes include neoplasia-like meningioma; inflammatory pathologies such as sarcoidosis; infections (especially fungal); trauma, and vascular lesions like carotid-cavernous fistula. The latter may be associated with an orbital bruit, increased eye pressure, engorged conjunctival vessels, and relative afferent pupillary defect.
III. Pituitary apoplexy is caused by infarction or hemorrhage of the pituitary gland, usually in a pre-existing adenoma, leading to localized edema or bleed. It is potentially life-threatening endocrinological emergency. It typically presents with headache, vomiting, visual impairment, and loss of consciousness. It may be associated with loss of vision, field loss, and third, fourth, and sixth cranial nerve palsies. There may be an associated pituitary insufficiency, especially corticotropic deficiency.

c. GCA: It is a systemic granulomatous vasculitis, affecting medium and large arteries, and can potentially cause irreversible loss of vision, and stroke. GCA usually presents with new onset headache (~67% patients), jaw claudication (>50% patients), scalp tenderness, neck pain, temporal allodynia, polymyalgia rheumatica and flu-like symptoms including fever, malaise, night sweats, and weight loss. Ocular manifestations are usually a result of ischemia: they include diplopia, eye pain, amaurosis fugax, choroidal infarcts, acute ischemic optic neuropathy (anterior and posterior), as well as cilioretinal artery and central retinal artery occlusions.[24-26]

Conclusion

Headaches are a complex group of disorders with varied presentation, etiologies, and management protocols. Their management requires a through history taking and examination, as well as diagnostic tools such as the ICHD-3 criteria to identify the etiopathogenetic. Even though most headaches are primary, secondary causes of headaches can often be vision, and even, life threatening. An urgent neurology consult, along with neuroimaging, therefore, is essential in case of any red flags to avoid any diagnostic dilemmas and therapeutic pitfalls.

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Address for correspondence:
Shibal Bhartiya
Senior Consultant, Glaucoma Services,
Fortis Memorial Hospital, Gurgaon, Haryana, India.
E-mail: shibalbhartiya@gmail.com

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