

Headaches in ophthalmology (part 2)

Ophthalmologists see a large number of patients with headaches or facial pain in the ophthalmic outpatient clinics or in emergency clinics. Over two articles, I will discuss several causes of headaches, ocular manifestations and proposed management and referral options. It is beyond the scope of these articles to discuss the details of each case and its treatment protocols.

As these patients are presenting to ophthalmic departments, it would be useful to classify these headaches as headaches with red eye or headaches with white eye, then further sub group each in order to find a likely diagnosis.

This article will discuss headaches with a white eye. Most of these patients would have primary neurological causes with possible ophthalmic manifestations.

Primary headaches

Tension-type headaches: Usually tend to be frontal or generalised type of headache with no neurological manifestations and more often in afternoons or after work or visual display units (VDU) tasks, very common and usually responds to simple analgesia.

Migraine: One of the most common types of headaches. It is usually throbbing, one sided, with or without aura. Patients tend to be photophobic and sonophobic, they could feel nauseous or vomit. They tend to avoid physical activity and sit in a dark quiet room. Aura usually lasts around 20 minutes but anything between five and 60 minutes and precedes the headaches which could last a full day. A good detailed history usually gives up the diagnosis but atypical cases might need neuro-imaging.

Other primary headaches trigeminal autonomic cephalalgia (TACs) were discussed under the headaches with red eye section in the previous article due to the usual presence of conjunctival congestion and redness.

Secondary headaches

It is important to identify secondary causes of headaches and facial pain and arrange necessary treatment or appropriate referrals.

Trauma: It is important to have a detailed clear history of headaches and arrange necessary neuro-imaging to rule out any treatable causes that might be associated with trauma.

Vascular causes: Examples would include:

- **Giant cell arteritis (GCA):** Patients usually elderly with recent onset temporal headaches, jaw claudication, temporal tenderness with or without general symptoms such as weight loss or night sweats. Patients could present to eye emergency clinics with loss of vision due to anterior ischaemic optic neuropathy and blood tests usually reveal raised inflammatory markers. It is a medical emergency and patients need to be assessed and started on treatment (steroids) urgently to save the other eye.
- **Subarachnoid haemorrhage (SAH):** Very acute presentation with severe headaches (thunderclap headache) and possible loss of consciousness. The diagnosis should be suspected in patients with risk factors or known aneurysms and should have emergency admission and neuro-imaging and possible neuro-surgical intervention.
- **Cerebrovascular accident (CVA):** Patients with recent strokes can develop persistent headaches.
- **Carotid dissection:** This tends to affect patients with known vascular risk factors or following major trauma. Patients will need neuro-imaging to confirm diagnosis (CTA or MRA) and would need vascular referral. Depending on the location of the dissection, presentation can be an ipsilateral Horner's syndrome or a VI nerve palsy.
- **Cerebral sinus thrombosis (CVS):** It is also a medical emergency. Patients tend to present with severe headaches, symptoms of raised intracranial pressure such as tinnitus. Examination would probably reveal bilateral optic nerve swelling (papilloedema) and VI nerve palsy.
- **Pituitary apoplexy:** Patients usually have a known pituitary adenoma. Patients present with sudden onset headaches with various endocrine manifestations. Be aware of this diagnosis following major surgery or following pregnancy. It is important to recognise the diagnosis urgently, arrange neuro-imaging and refer for treatment.
- Other rarer vascular causes of headaches include genetic causes such as cerebral autosomal recessive arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL), and mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS).

Non-vascular causes: Examples would include:

- **Idiopathic intracranial hypertension (IIH):** Patients are usually young females who are overweight. They usually present with headaches associated with tinnitus and transient visual symptoms or diplopia. Ophthalmic examination could reveal optic nerve swelling and possible VI nerve palsy. Neuro-imaging is necessary to rule out brain tumours or sinus thrombosis followed by lumbar puncture to measure intracranial pressure (ICP). It is important to rule out secondary causes of raised ICP such as brain tumours, effects of certain medications and several medical conditions that could cause raised ICP.

Neurological inflammatory conditions such as meningitis, neuro sarcoid and Tolosa Hunt can also present with headaches and cranial nerve palsies.

- **Tolosa Hunt syndrome:** Patients usually have inflammatory lesions in the orbital apex. They tend to present with headaches, pain behind their eye along with possible cranial nerve palsies such as III or VI or trigeminal neuralgias. Patients usually require steroid treatment with possible long-term immunosuppression.
- **Transient headache and neurologic deficits with cerebrospinal fluid lymphocytosis (HaNDL):** It is a headache disorder where patients experience moderate to severe headache attacks, neurological symptoms, and an increase in the amount of a type of white blood cell in the cerebrospinal fluid (CSF). Neurological symptoms include stroke-like symptoms (hemiparesis), altered sensation (hemisensory disturbances), and aphasia. These symptoms usually last up to two hours.
- **Substance use or withdrawal:** Examples include exposure to nitrous oxide (NO) and carbon monoxide (CO). Withdrawal headaches can happen also after exposure to codeine and caffeine. Medication overuse headaches can affect patients who are on multiple analgesics. Key feature is good detailed history with medications history, the patients need to come off their analgesia, reduce or stop caffeinated drinks and tolerate the period of increased headaches before improvement of symptoms.
- **Homeostasis disruption headaches:** These tend to happen in the setting of significant hypoxia, uncontrolled hypertension (especially malignant hypertension) and fasting (due to dehydration).

- **Malignant hypertension:** Presentation is usually with severe headaches, reduced vision (due to optic nerve swelling and possible macular oedema) and various other general symptoms such as altered mental status. Most patients would need acute medical intervention to reduce their blood pressure and improve their symptoms.
- **Headaches due to head or neck disorder:** Examples of these headaches include cases of cervicogenic headaches due to neck problems, ocular related headaches (examples below), ear problems (infections), nasal and paranasal disease, temporomandibular joint disorders (TMJ) and teeth abnormalities.
- **Refractive errors / presbyopia:** It is important to have a recent refraction and correct any uncorrected refractive errors and address presbyopia with appropriate reading adds.
- **Optic neuritis:** In typical optic neuritis, patients usually present with acute deterioration of vision with ocular pain especially on eye movement. Examination would show reduced vision and colour vision with an relative afferent pupillary defect (RAPD). Fundus examination is usually normal and patients would need MRI to prognosticate their condition and rule out other possible causes.
- **Posterior scleritis:** The pain is usually ocular or retrobulbar and could be associated with reduced vision. Most patients would have general inflammatory background such as rheumatoid arthritis. Orbital ultrasound could help with the diagnosis.
- **Painful cranial nerves:** Several cranial nerve palsies can present with associated pain, facial pain or headaches. Examples include: trigeminal neuralgia, painful trigeminal neuropathy (such as HZO), glossopharyngeal neuralgia, occipital neuralgia, Raeder syndrome (severe boring headaches with ipsilateral Horner's syndrome), III cranial nerve palsy (possible aneurysm) and Tolosa Hunt syndrome. It is important to rule out urgent cases such as III nerve palsy which could be cause be an aneurysm. Most of these patients would need neuro-imaging (ideally MRI with contrast, possible CT angiogram if aneurysm is suspected) and referral to neurology might be warranted.

Conclusion

Headaches are very common presentation in ophthalmic clinics. It is important to take a clear detailed history of the headaches / ocular or facial pain, focusing on its frequency, location, duration, intensity, course, triggers and family history of headaches. It is also important to perform a

HEADACHES IN OPHTHALMOLOGY – FLOW CHART

1. Red eye – headaches:

- Is there any skin rash?
 - Consider HZO
- Are there eye surface symptoms?
 - Dry eyes: FB sensation, irritation, no discharge
 - Conjunctivitis: discharge
 - Keratitis: blurring of vision, hazy cornea
- Is there sectorial / nodular redness with ++ pain?
 - Episcleritis: young patient, sectorial redness with moderate pain
 - Scleritis: sever pain, history of RA, etc.
- Is there irregular pupil, hazy vision?
 - Iritis: recurrent episodes, floaters, blurring of vision, perilimbal flush
- Are there halos on light, mid dilated pupil?
 - Acute angle closure glaucoma: sever pain, nausea, hard globe, hazy cornea
- Is there any unilateral or bilateral proptosis?
 - Consider thyroid eye disease, CCF or orbital inflammation.

2. White eye – headaches:

- Optician review to rule out refractive errors, astigmatism or presbyopia
- Loss of vision or severe blurring in elderly patient
 - Consider anterior ischaemic optic neuropathy, rule out GCA
- Young patient with a loss of colour vision, RAPD and blurring
 - Consider optic neuritis
- Diplopia
 - Consider cranial nerve palsies III, IV, VI, VII
- Pupil abnormality
 - Consider Horner syndrome
- Bilateral optic disc swelling
 - Consider papilloedema, IHH
 - Consider malignant hypertension
- Dull unexplained ache
 - Consider posterior scleritis
 - Consider Tolosa Hunt syndrome
- Remember migraines and cluster headaches
 - Visual aura (bilateral)
 - Watery eyes, mid-night episodes: consider cluster.

detailed ophthalmic examination to identify any ocular causes. Most patients would need neuro-imaging and might need further referrals to appropriate medical specialties.

Further reading

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- Medscape http://www.medscape.com/viewarticle/488345_9
- The International Classification of Headache Disorders 3rd edition www.ichd-3.org

(All links last accessed October 2018)

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