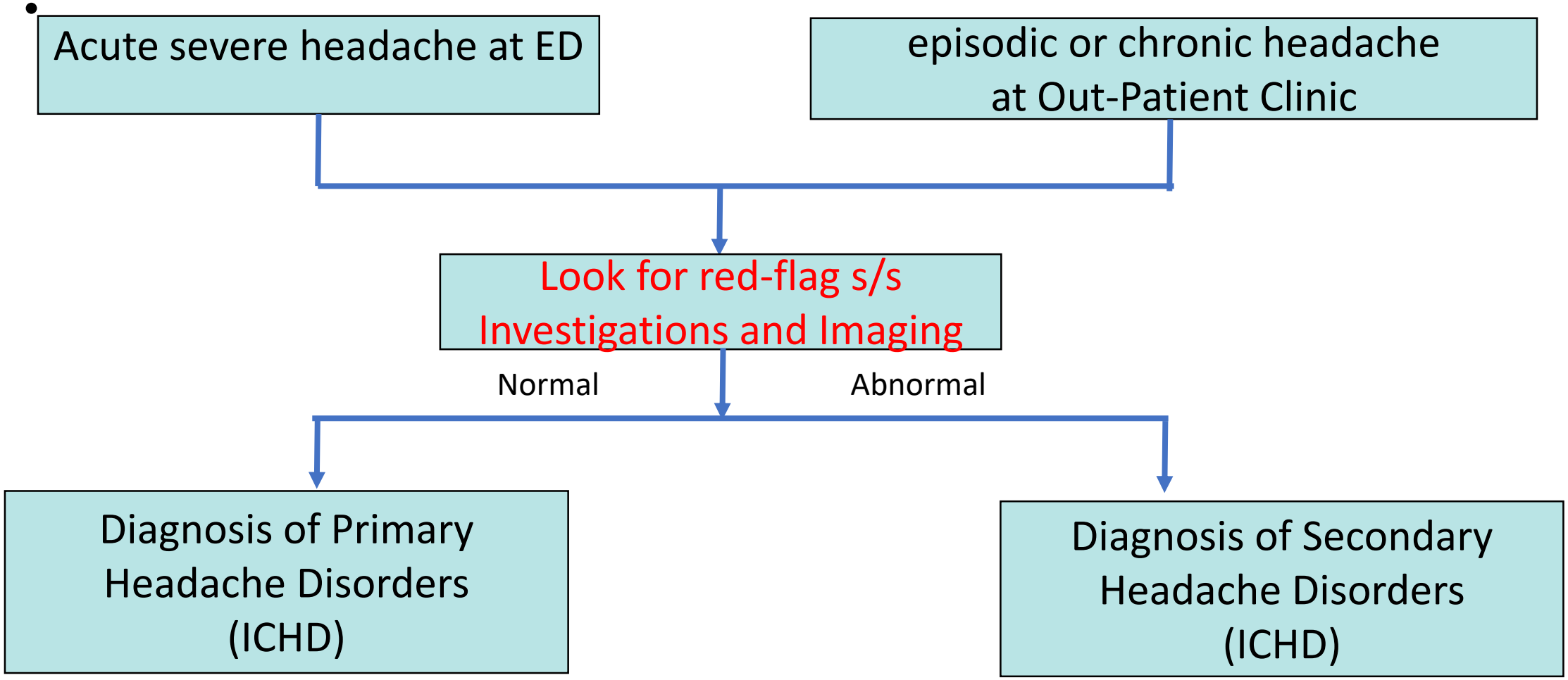
Headache attributed to Non-vascular Intracranial Disorder

Dr Seinn Mya Mya Aye
Senior Consultant Neurologist

9th July,2023

Diagnosis of Headache



Non-vascular Intracranial Disorder

Non-vascular Intracranial Disorder

A. Headache attributed to cerebrospinal fluid (CSF) pressure

B. Headache attributed to non-infectious inflammatory intracranial disease

C. Headache attributed to intracranial neoplasia

D. Headache attributed to intrathecal injection

E. Headache attributed to epileptic seizure

F. Headache attributed to Chiari malformation type I(CM1)

G. Headache attributed to other non-vascular intracranial disorder

A. Headache attributed to increased cerebrospinal fluid (CSF) pressure

- New Headache, or a significant worsening of a pre-existing headache
- Early morning headache , or worsening headache by lying
- Headache is relieved by reducing the intracranial hypertension
- Papilloedema
- Cerebrospinal fluid (CSF) pressure exceeds 250 mm CSF

A. Headache attributed to increased cerebrospinal fluid (CSF) pressure

1.Headache attributed to idiopathic intracranial hypertension (IIH) -

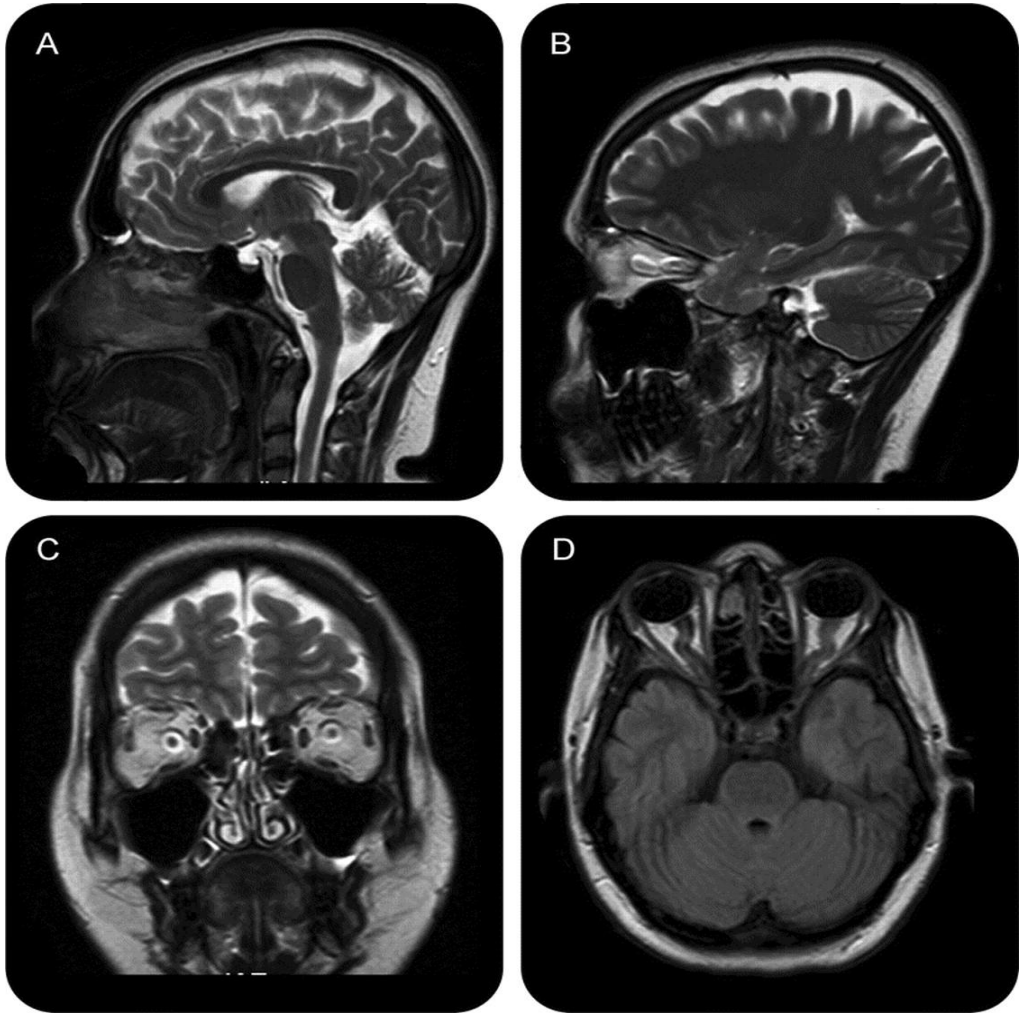
- Headache is accompanied by either or both of the following:

  a) pulsatile tinnitus

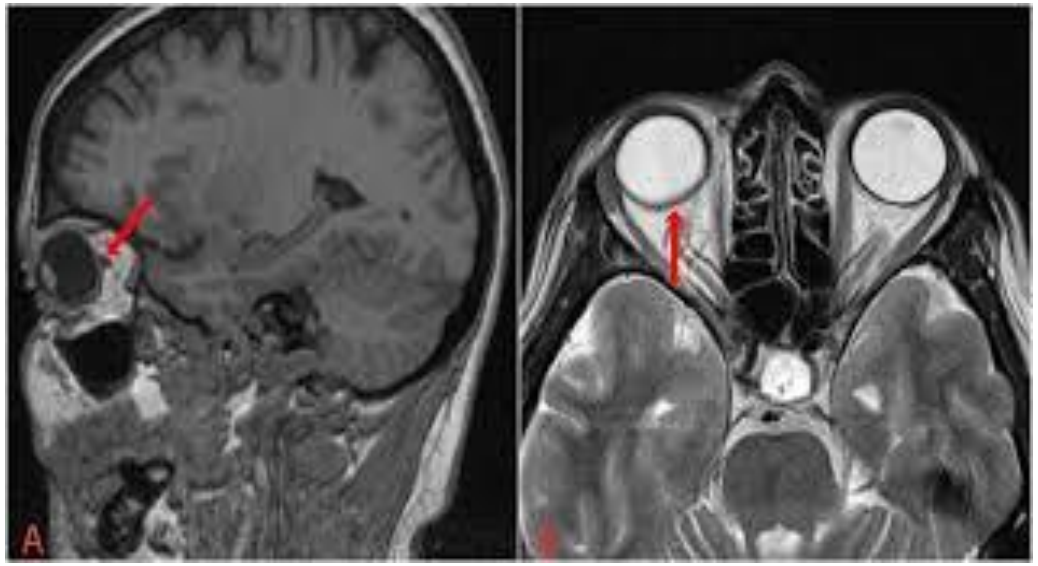
  b) papilloedema

- Imaging features of IIH

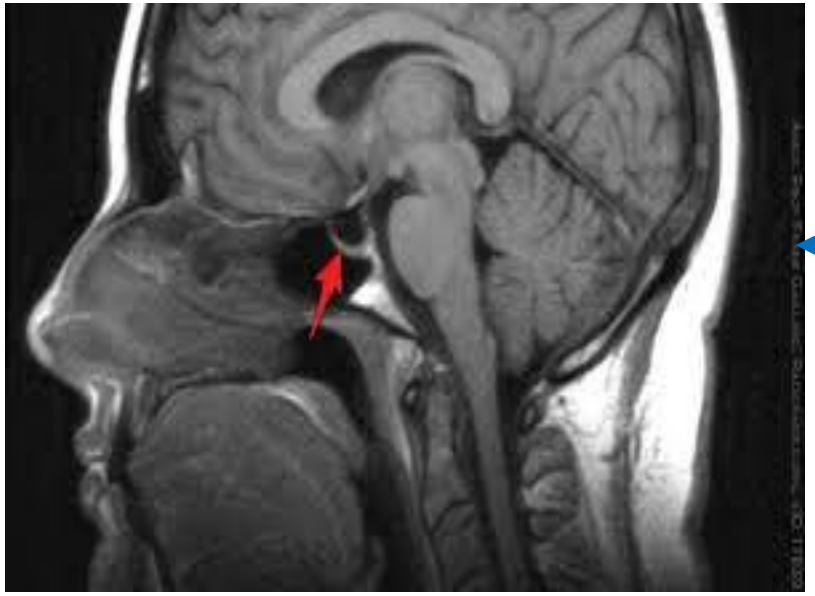
MRI



IIH

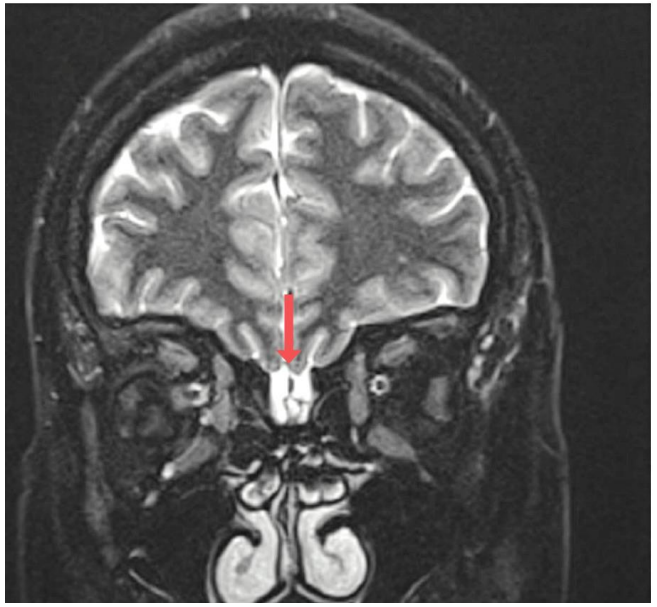


Axial T2 MR scans through orbits of the patient suffering from IIH show posterior globe flattening with mild bulging of the optic nerve disc head due to papilledema.



Sagittal T1 midline brain MRI scan shows partially empty sella in a female patient with suspected IIH.

Coronal STIR scan shows trans-ethmoid meningocele.



Headache attributed to increased cerebrospinal fluid (CSF) pressure

2. Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal cause

- acute hepatic failure,
- renal failure,
- hypercarbia,
- acute hypertensive crisis,
- hepato-cerebral syndrome
- Cerebral venous sinus thrombosis
- right heart failure
- substances (including thyroid hormone as replacement in children, all-trans retinoic acid, retinoids, tetracyclines and chlordecone)
- vitamin A toxicity
- corticosteroid withdrawal

Headache attributed to increased cerebrospinal fluid (CSF) pressure

3. Headache attributed to intracranial hypertension secondary to chromosomal disorder

- Chromosomal disorders associated with intracranial hypertension include Turner syndrome and Down syndrome.

4 Headache attributed to intracranial hypertension secondary to hydrocephalus

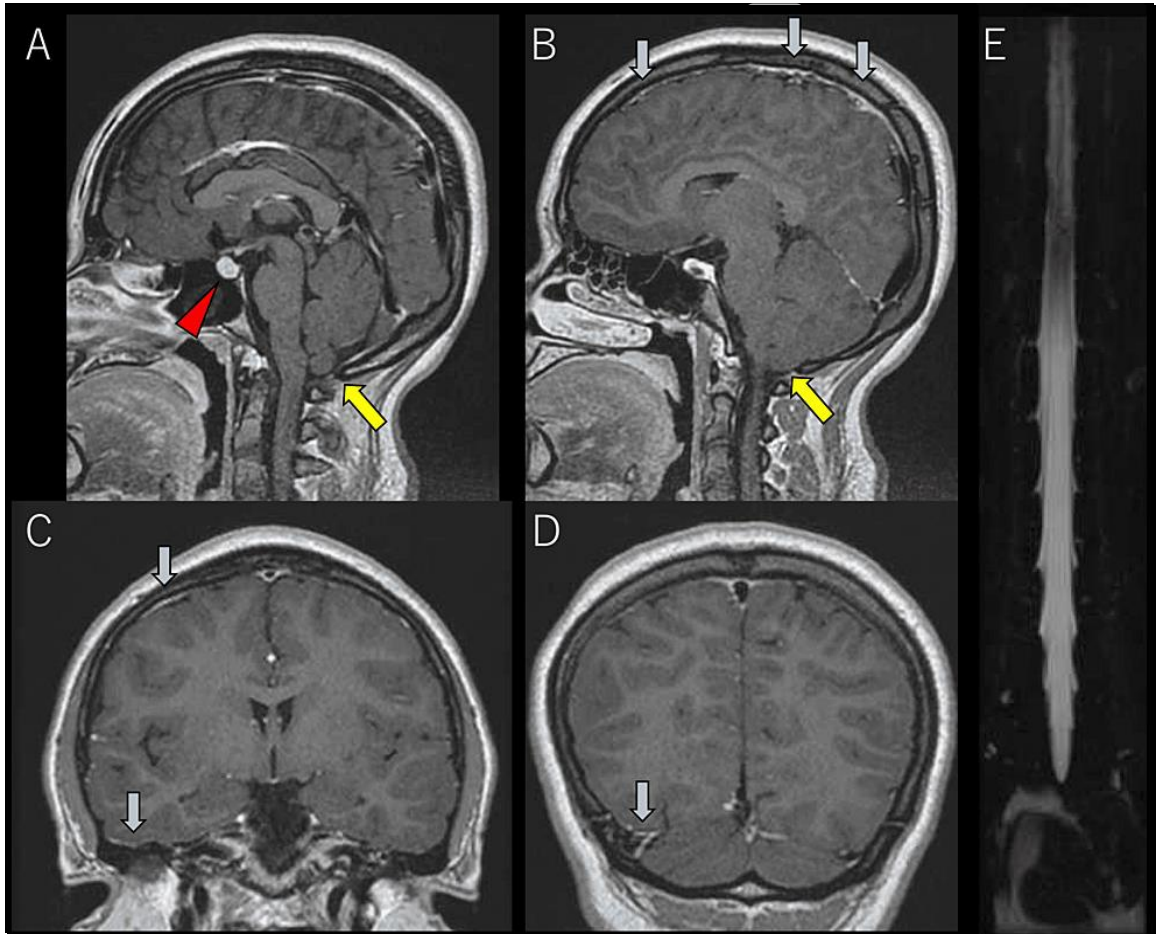
- caused by intracranial hypertension secondary to hydrocephalus

Headache attributed to low cerebrospinal fluid (CSF) pressure

- neck pain
- tinnitus
- changes in hearing
- Photophophia
- nausea
- significantly worsens soon after sitting upright or standing and/or
- improves after lying horizontally is likely to be caused by low CSF pressure
- remission after normalization of CSF pressure or successful sealing of the CSF leak.

The gadolinium contrast-enhanced T1-weighted magnetic resonance (MR) imaging showed partially enhanced dura matter (gray arrows in A, B, C, and D)

pituitary enlargement (red arrowhead in A)



low-lying tonsils (yellow arrows in A and B),

Intracranial Hypotension

Headache attributed to low cerebrospinal fluid (CSF) pressure

1. Post-dural puncture headache

- Headache occurring within five days of a lumbar puncture, caused by cerebrospinal fluid (CSF) leakage through the dural puncture.
- Usually accompanied by neck stiffness and/or subjective hearing symptoms.
- Remission spontaneously within two weeks, or after sealing of the leak with autologous epidural lumbar patch.

Headache attributed to low cerebrospinal fluid (CSF) pressure

2. Cerebrospinal fluid (CSF) fistula headache

- after a procedure or trauma causing a persistent cerebrospinal fluid (CSF) leakage resulting in low intracranial pressure

3. Headache attributed to spontaneous intracranial hypotension

- Spontaneous cerebrospinal fluid (CSF) leak has been associated with heritable connective tissue disorders.
- Patients with CSF leaks should be screened for connective tissue and vascular abnormalities.

B. Headache attributed to non-infectious inflammatory intracranial disease

Usually with lymphocytic pleocytosis in the cerebrospinal fluid

1 Headache attributed to neurosarcoidosis

- aseptic meningitis, cranial nerve lesions, intracranial space-occupying lesion(s) on brain MRI,
- Periventricular inflammatory focal lesions and/or homogeneously enhancing mass lesions on brain or spinal MRI that are confirmed on biopsy as non-caseating granulomas.

2 Headache attributed to aseptic (non-infectious) meningitis

- Symptoms and/or clinical signs of meningeal inflammation including neck stiffness (meningismus) and/or photophobia
- CSF in patients with aseptic meningitis shows lymphocytic pleocytosis, mildly elevated protein and normal glucose in the absence of infectious organisms.
- Aseptic meningitis may occur after exposure to certain drugs, including ibuprofen or other NSAIDS,
- immunoglobulins, penicillin or trimethoprim, intrathecal injections

B. Headache attributed to non-infectious inflammatory intracranial disease

3 Headache attributed to other non-infectious inflammatory intracranial disease

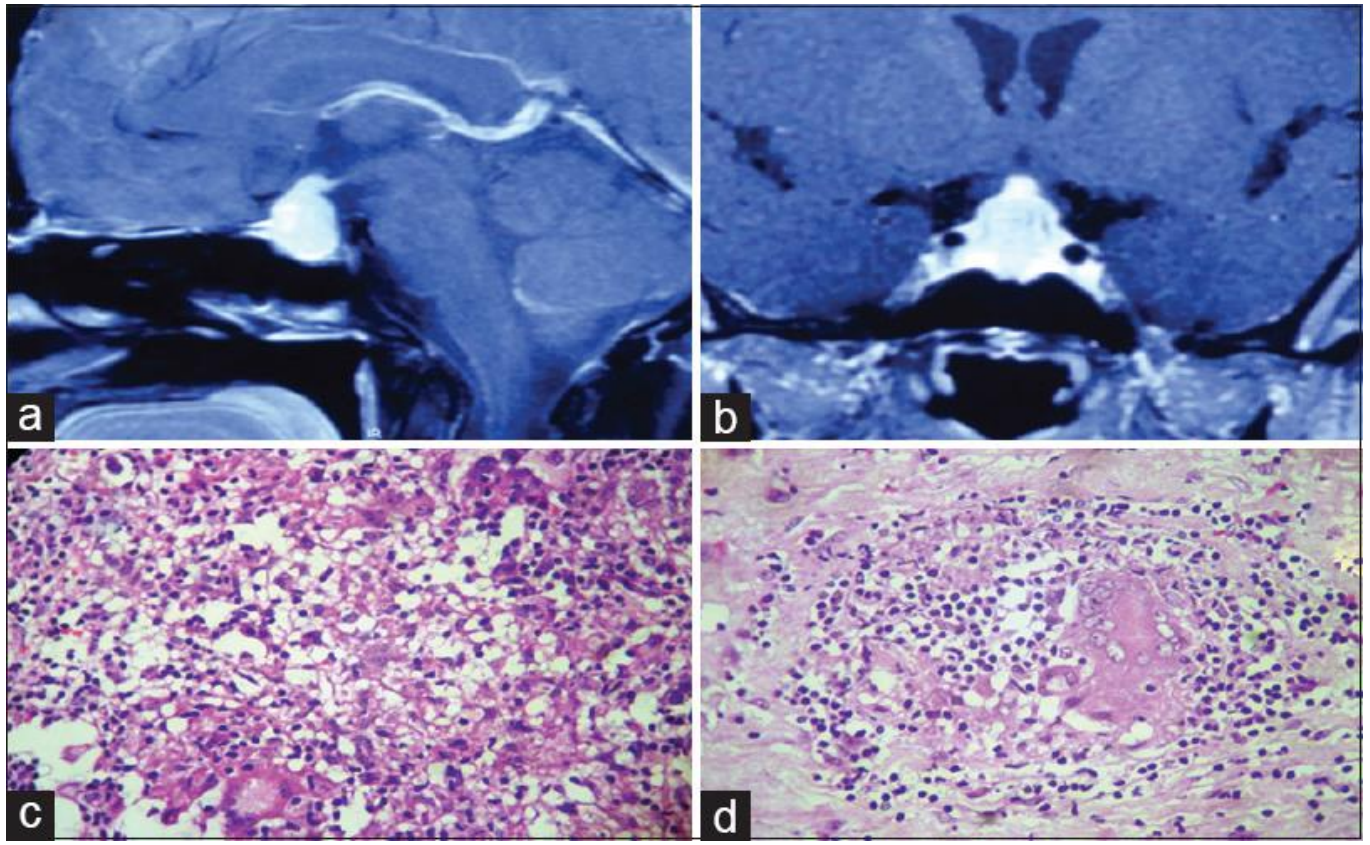
- a presenting or prominent symptom of any of a variety of autoimmune disorders

4 Headache attributed to lymphocytic hypophysitis

- lymphocytic hypophysitis, associated with pituitary enlargement and, in half of cases, with hyper-prolactinaemia.
- is associated with pituitary enlargement and homogeneous contrast enhancement on brain MRI.
- It is accompanied by hyper-prolactinaemia in 50% of cases or autoantibodies against hypophyseal cytosol protein in 20% of cases

Lymphocytic Hypophysitis

Sagittal postcontrast MRI scan shows a bright contrast-enhancing lesion with infundibular thickening and dural tail with a normal sella.



(b) Coronal contrast MRI scan showing the suprasellar tongue-like projection causing compression of the chiasm

(c) Histopathology showing extensive fibrosis with lymphocytic infiltration

(d) Histopathology showing Langerhans giant cells

B. Headache attributed to non-infectious inflammatory intracranial disease

5. Syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL)

- Migraine-like headache episodes accompanied by neurological deficits including hemi-paraesthesia, hemiparesis and/or dysphasia, but positive visual symptoms only uncommonly, lasting several hours.
- cerebrospinal fluid (CSF) lymphocytic pleocytosis (>15 white cells per ml)

C. Headache attributed to intracranial neoplasia

headache has at least one of the following four characteristics:

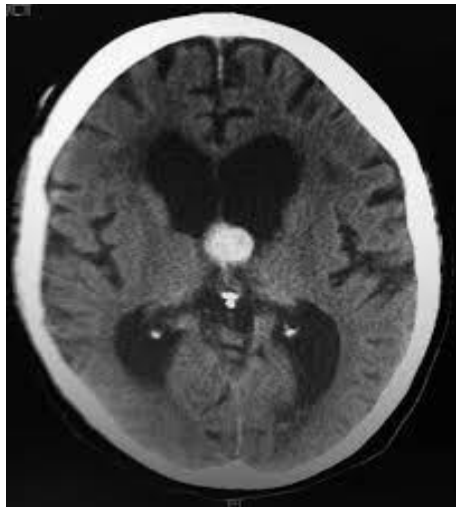
a) progressive

b) worse in the morning and/or when lying down

c) aggravated by Valsalva-like manoeuvres

d) accompanied by nausea and/or vomiting

C . Headache attributed to intracranial neoplasia



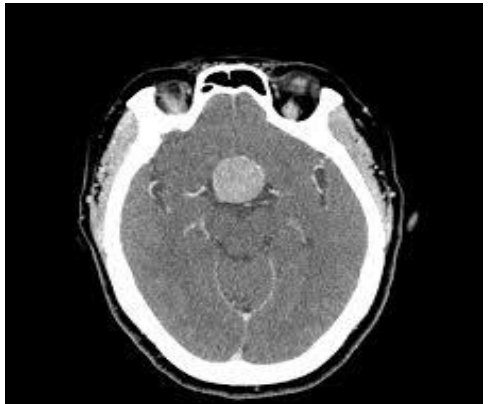
1.Headache attributed to colloid cyst of the third ventricle

- Recurrent attacks with thunderclap onset,
- often triggered by postural change or Valsalva-like manoeuvre,
- Associated with reduced level or loss of consciousness.

2 Headache attributed to carcinomatous meningitis

- headache is associated with cranial nerve palsies and/or encephalopathy

3 Headache attributed to hypothalamic or pituitary hyper- or hyposecretion



headache is associated with at least one of the following:

a) disorder of temperature regulation

b) abnormal emotional state

c) altered thirst and/or appetite

D. Headache attributed to intrathecal injection

- headache has developed within four days of the intrathecal injection
- headache has significantly improved within 14 days after the intrathecal injection
- signs of meningeal irritation

E. Headache attributed to epileptic seizure

1. Ictal epileptic headache

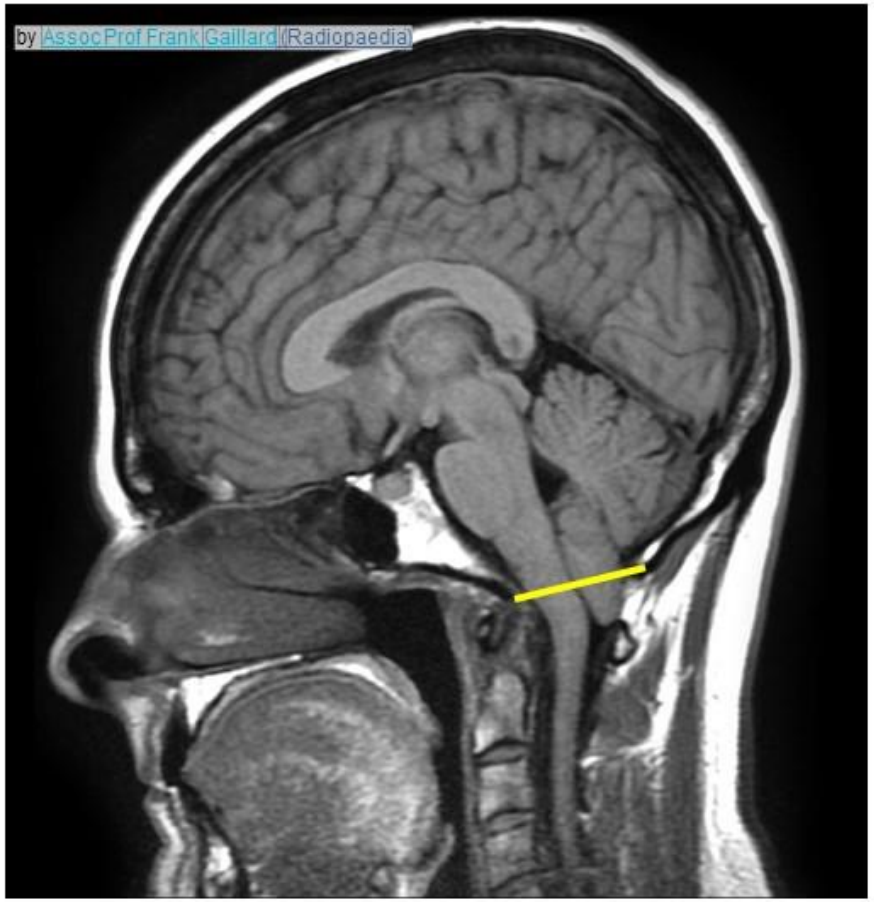
- Headache caused by and occurring during a partial epileptic seizure, ipsilateral to the epileptic discharge
- remitting immediately or soon after the seizure has terminated.

2. Post-ictal headache

- Headache has developed within three hours after the epileptic seizure has terminated
- Headache has resolved within 72 hours after the epileptic seizure has terminated

F. Headache attributed to Chiari malformation type I (CM1)

- usually occipital or suboccipital,
- short duration (less than five minutes)
- provoked by cough or other Valsalva-like manoeuvres.
- remits after the successful treatment of the Chiari malformation.
- headache is associated with other symptoms and/or clinical signs of brainstem, cerebellar, lower cranial nerve and/or cervical spinal cord Dysfunction



Chiari 1 Malformation

- 5-mm or more caudal displacement of the cerebellar tonsils inferior to the foramen magnum
- Restricts normal CSF flow across the craniocervical junction
- Commonly associated findings are cervical syringomyelia(50-75%) and, on occasion, hydrocephalus (10%)

G. Headache attributed to other non-vascular intracranial disorder

- Non-vascular causes other than those described above

Conclusion

Key Diagnostic Features

1. a new headache occurs for the first time in close temporal relation to a non-vascular intracranial disorder → secondary headache attributed to that disorder

2. - a pre-existing headache with the characteristics of a primary headache disorder becomes chronic

   or

   - significantly worse (usually meaning a twofold or greater increase in frequency and/or severity)

   - in close temporal relation to a non-vascular intracranial disorder →

both the initial headache diagnosis and a diagnosis of headache attributed to non-vascular intracranial disorder (or one of its types or subtypes)

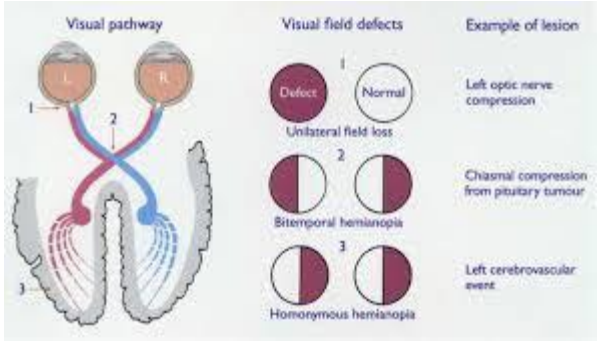
Red Flags – History taking

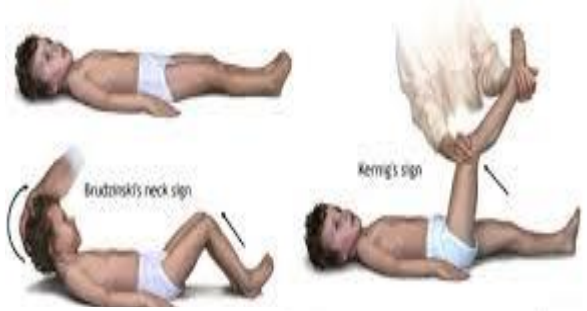
Headache

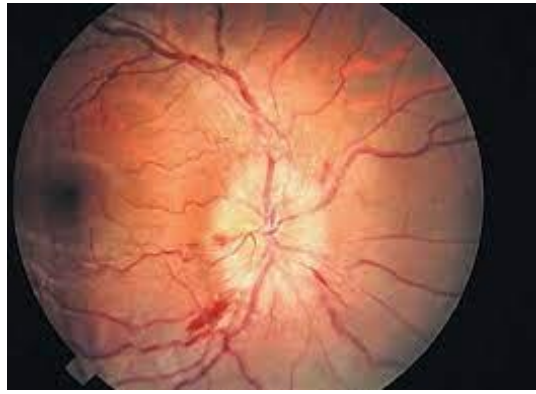
- Postural changes (diff between lying and standing)
- Valsalva maneuver
- Early morning headache
- Associated nausea and vomiting
- Double vision
- Balance problems

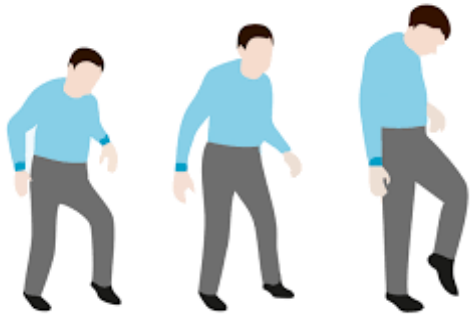
Red Flags – Physical Examination

- Reduced conscious level
- Visual impairment
- Fundus – papilloedema
- Neck stiffness
- Cranial nerve palsies
- Pyramidal tract signs
- Ataxia









Red Flags - Imaging

"Not to overlook the abnormal findings"

Choice of Imaging modality (CT/MRI/Angio/Venogram)

Choice of Imaging protocol (Pituitary/Cavernous/orbit/Brain)

Radiologist's experiences and expertise

Clinical correlation

Cost-effectiveness

Key Message

- ✓Careful history taking, physical examination , appropriate choice of investigations and looking for the red flags are the keys to have the correct diagnosis.

Ref

- Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders,3rd edition

  Cephalalgia,2018, Vol. 38(1) 1–211,


thank you