The headache and cranial neuralgias

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WWW

- Handouts:
- www.neuroM.pl
- International Headache Society
- http://ihs-classification.org/en/
  - Look for the IHS Classification III

IHS Classification ICHD-II

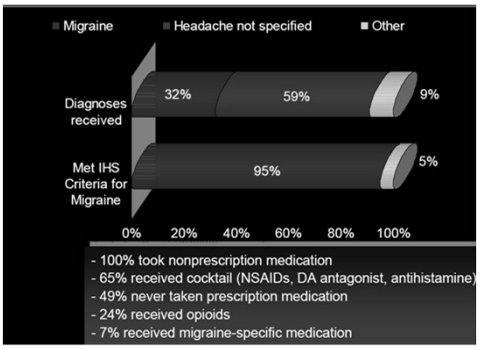
Headache in emergency department

Headache in ED

- Headache is the 5th most common complaint seen in the US EDs
- Majority primary headaches
- 10% are “repeat customers”

4

Headache in ED



Blumenthal et al. *Headache*. 2003;43(10):1026-1031.

5

Primary headaches in ED

- MIGRAINE
- Cluster headache and related syndromes (including paroxysmal hemicranias, SUNCT)
- Hypnic headaches
- Benign exertional/sex headache
- Cough headache

6

Secondary headaches in ED

TUNDERCLAP HEADACHE (TCH)

- **Traumatic**
  - simple closed head injury
  - complex with subdural /extradural/ subarachnoid/intracerebral haemorrhage
- **Vascular disease**
  - SUBARACHNOID HAEMORRHAGE (aneurysmal, perimesencephalic, other vascular anomalies)
  - unruptured aneurysms
  - acute cerebral ischaemia (TIA or stroke)
  - non-traumatic subdural/extradural/intracerebral haemorrhage
  - dissection of carotid/vertebrobasilar arteries
  - cerebral venous thrombosis
  - vasculitis (including giant cell arteritis)
- **CNS infection**
  - **MENINGO-ENCEPHALITIS (BACTERIAL, VIRAL, FUNGAL)**
  - cerebral abscess
- **Non-vascular intracranial disease**
  - intermittent hydrocephalus (e.g. colloid cyst)
  - idiopathic intracranial hypertension
  - intracranial hypotension (spontaneous or post LP)
  - intracranial tumour
  - pituitary apoplexy
  - Arnold Chiari malformations
  - optic neuritis
- **Metabolic or toxic disturbances**
  - phaeochromocytoma
  - thyroid disease
  - drug induced
  - withdrawal syndromes
  - hypercarbia
- **Hypertensive encephalopathy**
- **Cervical spine disease**
- **Dental, ENT or ophthalmic disease** (e.g. sinusitis, acute glaucoma)
- **Secondary to general medical conditions** (e.g. ischaemic heart disease, infection other than cranial, epilepsy)

7

History is the most important point in evaluation of a patient with a headache

- Acute and subacute
  - Headache appeared hours or days (acute) before
  - Several days or weeks (subacute)
  - No difference if the headache is severe or not
  - Usually symptomatic/secondary
- Chronic
  - History of months to years
  - Usually primary headaches

8

S2NOOP4 (Dodick 2003)

S	Systemic symptoms	Fever, weight loss	Infections,
S	Secondary risk factors	HIV, cancer, immune suppression	Infections, metastatic cancer, carcinomatous meningitis
N	Neurologic symptoms/signs	Altered consciousness, focal deficits	Encephalitis, mass lesion, stroke
O	Onset	Split-second, thunderclap	Subarachnoid hemorrhage, and others
O	Older	New onset after 50	Giant cell arteritis
P4	Prior history	First, newly progressive, or different from usual	Any secondary cause can lead to change in pattern
	Positional	Dramatic change upright vs. recumbent	Intracranial hypotension, orthostatic dysautonomia
	Papilledema	Visual obscurations	Increased intracranial pressure
	Precipitants		

9

History

- When did your actual headache started?
- Have you ever had this type of headache before?
- How did the headache begin?
- How has the pain changed since its onset?

10

History

- Age of onset
  - New onset in the 6th or 7th decade → secondary!
- Prior history of headaches
  - stereotypic, reccurent → benign
  - Changes in pattern → secondary!
  - **„Once a benign headache not always a benign headache"**
- First ever headache → investigate!

11

Systemic symptoms

- Fever
- Weight loss
- HIV
- Immune suppression
- Cancer

12

Temporal profile of headache

- Acute single headache
  - SAH,
  - meningitis,
  - sinusitis,
  - glaucoma,
  - posttraumatic,
  - medication side-effect

13

Subacute headache

- subdural hematoma
- neoplasm
- brain abscess
- pseudotumour cerebri (idiopatic intracranial hypertension)
- giant cell arteritis

14

Headache triggers

- Cough → posterior fossa lesions
- Coital headache → may be SAH
- Sleep induced
  - benign: hypnic, migraine, cluster
  - secondary: ↑ intracranial pressure (tumor), sleep disorder (sleep apnea, snoring?)
- Positional induced:
  - low CSF pressure
  - Chiari malformation

15

Family history

- Suggests benign headache
- ask for history of
  - aneurysm
  - connective tissue disorders
  - coagulopathies

16

Past medical history

- neoplasms
- connective tissue disorder
- past strokes
- known aneurysm
- seizure disorder
- recent lumbar puncture

- Above suggest secondary headache

17

General medical examination

- Vital signs
  - Blood pressure
  - Increased body temperature
  - Heart rate
- Head:
  - Temporal arteries: palpate for tenderness and presence of pulses
  - Inflamed tympanic membrane
- Cervical exam:
  - Enlarged cervical lymph nodes
  - Range of motion; palpation over greater occipital notches, palpating for pain trigger points
- Cardiac status
- Skin: rash

18

Neurological examination

- Meningismus (neck stiffness; Kernig's, Brudzinski')
  - May be non-specific
  - May be absent (SAH!!)
- Mental status:
  - level of consciousness, presence of delusions or hallucinations
- Neurological abnormality
  - Pupillary asymmetry & reactivity
  - Diplopia, disconjugate gaze, visual field defect
  - Limb weakness or sensory loss (hemiparesis), asymmetric reflexes, Babinski sign
  - Limb ataxia, gait and station, ataxia, Romberg sign
- Fundoscopy: papilledema, retinal hemorrhage
- Neurovascular examination – auscultation for cervical, cranial and orbital bruits

19

Case 1

History CC

- An ambulance brings a 39-year-old woman complaining of a severe occipital headache and vomiting from her workplace to the emergency department (ED).

21

HPI

- She describes the headache as "the worst ever headache of my life" and states that it started suddenly, after a stressful situation at work.
- The intensity of the headache did not diminish after taking 200 mg of ibuprofen
- denies any head trauma, intense physical exertion, fever, changes in vision, photophobia, or any other associated symptoms (other than vomiting).
- 2 days before presentation, she had a headache of similar intensity, but it only lasted a few seconds before resolving spontaneously.

22

PMH

- Her medical history is remarkable for 2 normal pregnancies with uncomplicated deliveries.
- She denies any prior surgeries
- Meds:
  - oral contraceptives, which she has taken for about 15 years.

23

SH,FH

- She works as a dressmaker
- denies illicit drug use, tobacco use, or alcohol consumption.
- She does not recall any significant medical problems in her family history.

24

Physical examination

- Obese and appears her stated age.
- In mild distress due to pain.
- Vital signs
  - T 97.8°F (36.6°C). ; HR 78 bpm;, BP 160/80 mm Hg (which she states is high for her), RR 20 breaths/min.

25

Head

- Her head is normal on inspection, without any areas of tenderness.
- The ears, nose, and throat are clear. Her pupils are both small at 2 mm, but they are reactive to light.
- Otherwise, the eyes appear normal, with normal extraocular movements and no photophobia or nystagmus.
- The fundi and optic discs appear normal.

26

Neck

- No masses are detected on examination of her neck, but significant nuchal rigidity is noted.

27

Chest & abdomen

- The chest examination is normal, with lungs clear to auscultation bilaterally and normal respiratory effort. The heart sounds are also normal.
- Her abdomen is soft and nontender.

28

Neurological exam

- she is fully alert, oriented, and mildly anxious.
- Her cranial nerves - intact.
- Motor strength is symmetrical, with brisk and symmetric deep tendon reflexes without clonus.
- Cerebellar function and sensory systems are normal.

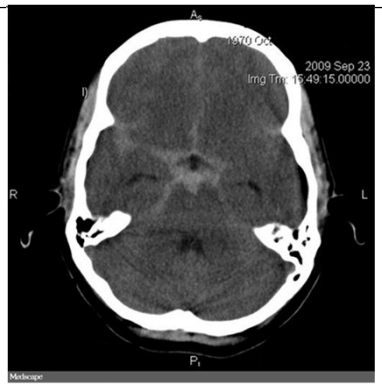
29

Labs

- Laboratory analyses, including a complete blood count, metabolic panel, and urine analysis, are normal.

30

CT



- A noncontrast cerebral computed tomography (CT) scan is performed (Figure 1).

31

Subarachnoid haemorrhage

How likely is SAH in patient presenting with acute onset headache

- Study of sudden onset headache (=reaching max intensity in 10 s.) (Landtblom et al, 2002)
  - 137 patients giving annual incidence 43 per 100,000
  - **11,3% of that SAH** (annual incidence of SAH 5/100,000)
  - Other: cerebral infarction (five), intracerebral haematoma (three), aseptic meningitis (four), cerebral oedema (one) and sinus thrombosis (one)
  - Nausea, neck stiffness, occipital location and impaired consciousness were significantly more frequent in SAH
- Other study of sudden onset headache (max intensity 60 s., lasting at least 1 hr) (Linn et al, 1994):
  - 148 patients
  - 25% (37) had SAH (annual incidence of SAH 4/100,000)

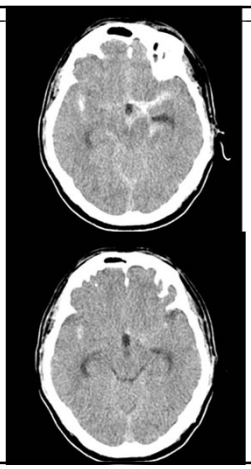
33

- Between 1 in 10 and 1 in 4 patients presenting with an **sudden** onset headache may have SAH
- other serious neurological condition may be present

34

Brain CT in SAH

- If scan done within 48 hrs, modern scanner, experienced neuroradiologist **➔ CT sensitivity 95-98% (not 100%!)**
- After 48 hrs the CT sensitivity falls
- CT returns to normal within 1-2 weeks of initial SAH



35

Imaging in SAH

- CT positive in 98% in first 24 hrs
- MRI FLAIR
  - Equally sensitive as CT
  - More sensitive then CT 3-40 days after ictus
- MRA sensitivity for aneurysm: 70-100%
- CTA sensitivity for aneurysm: 85-98%

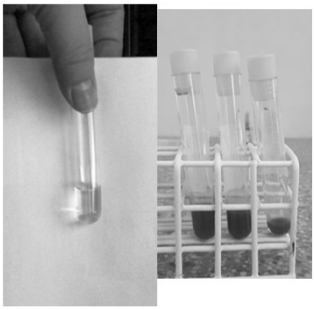
36

Lumbar puncture

- After neurologic exam and neuroimaging tests fail to uncover the cause the CFS exam is mandatory in
  - The first, worst or unusually severe pain
  - A severe rapid onset, reccurent headache
  - A subacute/progressive headache
  - An atypical chronic intractable headache
  - A headache associated with fever

37

LP & CSF examination in suspected SAH (normal CT scan)



- Delay the LP for 6 to 12 h after symptom onset
- LP should be done by an experienced operator
- Measure opening pressure
- CSF bilirubin is the key result suggestive of SAH (usually accompanied by oxyhemoglobin)
- Early on (within the first 48 hs) the red cell count is also important
  - No SAH with less then a 1000 red cell per mm^2 (or rarely)

38

Subarachnoid haemorrhage

- Sudden onset of an unusually severe generalized new headache ("the worst headache I had in my life", „like a blow to my head")
  - Develops within moments but may develop in minutes,
  - SAH is exceedingly rare cause of headache taking 10+ minutes to evolve
- Lasting at least one hour
- Though present in 85-100% cases the headache may not be severe
- SAH should be considered in anyone with a sudden severe headache, even if it resolves within one hour, particularly if there is any impairment of consciousness

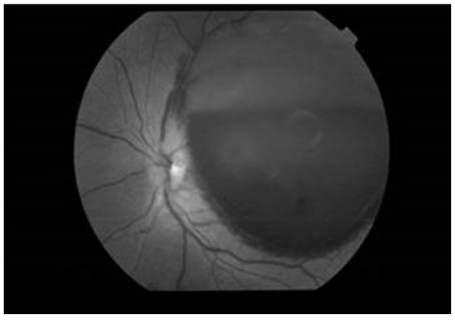
39

Non-headache features of SAH

- Sentinel headaches (10-43% Pts
- Nausea, vomiting
- Fever
- Dizziness
- Visual disturbances (intraocular haemorrhage)
- Meningeal signs
- Neck or back pain
- Papilloedema
- Subhyaloid haemorrhage
- Loss of consciousness (33%)
  - Transient
  - Coma
- Delirium (16%)
- Epileptic seizures (6-9%)
- Focal neurological signs
  - Paralysis or pareses; visual field defects
  - Cranial nerve palsies III, VI
- Sudden death
- GI bleeding
- Pulmonary edema
- Syndrome of secretion of inappropriate antidiuretic hormone
- ECG changes

40

Subhyaloid haemorrhage



41

Vascular events other than SAH

- Ischemic stroke
  - Ipsilateral to stroke
  - Non-throbbing
- Transient ischemic attack
  - About 30% cases
  - may be difficult to distinguish from migraine
  - Neurological symptoms present in second
  - do not develop in succession
  - Very rarely prominent symptom of TIA
- Intracerebral hemorrhagic stroke

42

Thunderclap headache

- Sudden onset, worst in life,
- peak intensity without latency,
  - < 60 seconds
- Duration of at least 5 minutes
- 30%-80% symptomatic
- Must out acute neurologic event

43

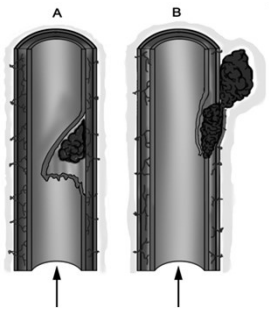
Thunderclup headache differential diagnosis

- Intracranial hemorrhage (SAH or intracerebral h.)
- Sentinel bleed
- Arterial dissection: carotid or vertebral)
- Cerebral venous thrombosis
- Acute stroke
- Cerebral vasculitis
- Spontaneous intracranial hypotension/hypovolemia (CSF leak)
- Pituitary apoplexy
- RCVS or Reversible cerebral vasoconstriction syndrome
- Malignant hypertensive crisis
- PRES or Posterior reversible leukoencephalopathy s.
- Colloid cyst of the third ventricle
- Sinusitis (especially sphenoid)
- Meningitis
- Myocardial infarction
- By exclusion: Primary thunderclap headache (idiopathic)

44

Dissection of the carotid or vertebral artery

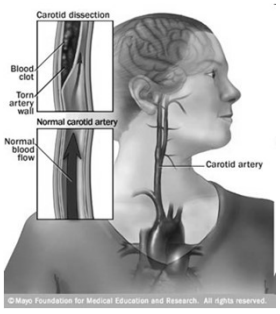
- Dissections mostly spontaneous
  - Secondary to major trauma (whiplash injury) or to minor trauma (coughing, sneezing, normal flexion of neck)



45

Carotid artery dissection

- ipsilateral headache anterior part of the head in the orbit or periorbital (84% patients),
- focal ischemic signs (61%)
- Horner's syndrome (53%)
- neck pain (23%)
- light-headedness (21%)
- bruits (objective or subjective) 45%



46

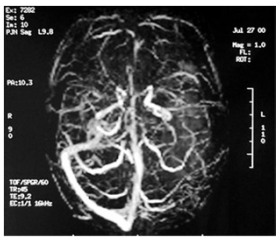
Vertebral artery dissection

- Symptoms:
  - headache – in the base of the head
  - +/- neck pain
  - Focal ischemic symptoms (lateral medullary syndrome)
  - some patients only posterior circulation stroke or TIA
  - some only headache or neck pain
  - 20% present as a thunderclap headache

47

Cerebral venous thrombosis

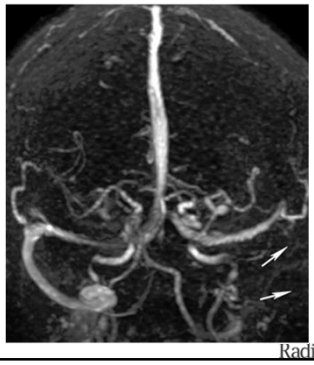
- Presentations:
  - Headache
  - Raised intracranial pressure
  - Other neurological symptoms
    - Mental status changes,
    - Seizures
    - Stroke
  - Cavernous sinus thrombosis: painful third or sixth n. palsy
  - Mostly progressive headache
  - 2%-10% TCH



Axial view of MR venogram demonstrating lack of flow in transverse sinus.
(Medscape)

48

Acute thrombus in a 35-year-old woman with a severe headache for 5 days.



Leach J L et al. Radiographics 2006;26:S19-S41

RadioGraphics

Axial unenhanced CT

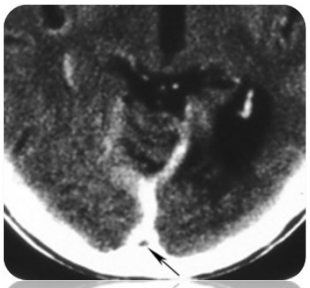
Axial unenhanced CT image shows areas of abnormal hyperattenuation consistent with thrombi in both transverse sinuses (arrows).

Leach J L et al. Radiographics 2006;26:S19-S41

RadioGraphics

Contrast-enhanced CT image in a patient with superior sagittal sinus thrombosis

- Contrast-enhanced CT image in a patient with superior sagittal sinus thrombosis shows a central filling defect in the superior sagittal sinus (arrow), surrounded by intensely enhanced dura mater.



Leach J L et al. Radiographics 2006;26:S19-S41

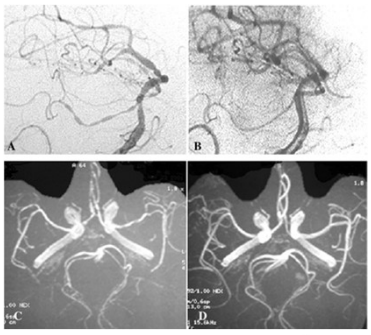
RadioGraphics

Reversible cerebral vasoconstriction syndrome (Call-Fleming syndrome)

- Segmental cerebral vasoconstriction
- Resolves in few weeks
- Diagnosis: angiography (conventional, MRA, CT)
- Transcranial Doppler for monitoring
- Nimodipine (Ca blocker) help relieve symptoms & may prevent stroke
- Glucocorticoid Tx

52

Vascular imaging in RCVS. (A) Catheter angiography at disease onset: multiple narrowing and dilatations, (B) Control catheter angiography at 3 months, (C) MRA at disease onset and (D) Control MRA.

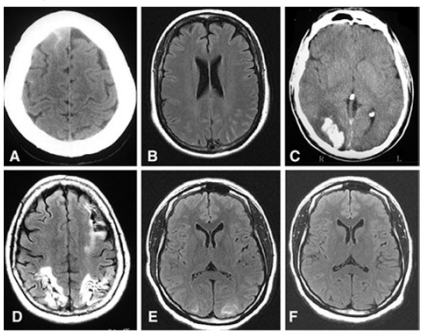


Ducros A et al. Brain 2007;130:3091-3101

BRAIN A JOURNAL OF NEUROLOGY

Brain imaging in RCVS.

(A) CT scan showing a small cSAH, (B) MRI (FLAIR sequence) showing a small cSAH, (C) CT scan showing an occipital intracerebral haemorrhage, (D) MRI showing sequelae of bilateral occipital infarcts and left frontal-parietal infarct, (E) MRI (FLAIR) showing hypersignals consistent with a RPLS and (F) Control MRI in the same patient after 28 days showing resolution of the RPLS.



Ducros A et al. Brain 2007;130:3091-3101

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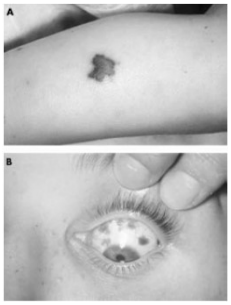
Meningitis & encepahlitis

- May present with true sudden onset headache
- Usually subacute
- Bacterial
  - Fever >38° C
  - septic shock
  - purpuric rash



55

Meningitis classic S&S

- malaise,
- headache,
- fever,
- photophobia,
- meningeal signs,
- nausea and vomiting,
- obtundation
- may be only incompletely present or absent

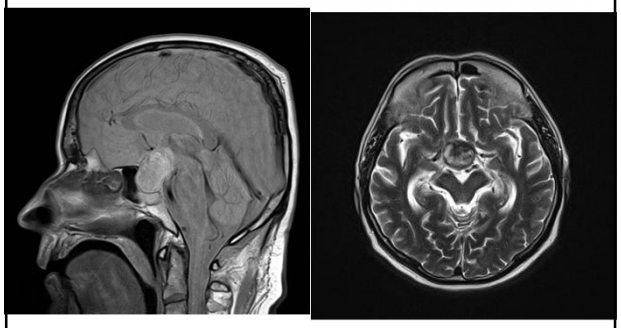
56

Pituitary apoplexy

- Combination of
  - Acute headache
  - Nausea
  - Decreased visual acuity
  - Ophtalmoplegia
  - Reduction of visual fields
- Mild symptoms → adrenal crisis → sudden death

57

Pituitary tumor and pituitary apoplexy



58

Post-dural puncture headache

- Occurs in approximately 20–25% of patients who undergo lumbar puncture,
- regardless
  - of a traumatic tap
  - of the amount of CSF removed.
- Due to low CSF pressure

http://www.flickr.com/photos/43437033@N03/4163169487/

59

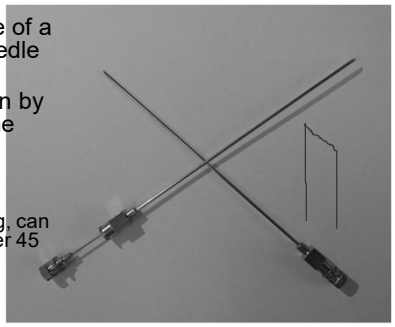
Post-dural puncture headache features

- Marked increase in pain in the upright position
- Occipital
- 24-48 hrs after the lumbar puncture
- Lasts 1-2 days (may last up to two weeks)

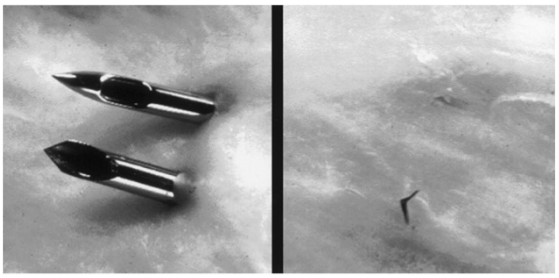
60

Risk of post-LP headache

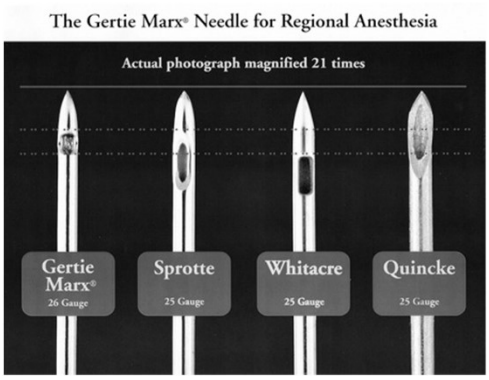
- Reduced by use of a small gauge needle (22 G)
- No risk reduction by lying flat after the procedure
- Treatment:
  - caffeine sodium benzoate iv 0,5g, can be repeated after 45 minutes
  - IV fluids



61



62



63

Hypertension and headache

- Unlikely the etiology of headache even at moderately elevated blood pressure levels
  - 180 mm Hg systolic, 110 mm Hg diastolic
- Ambulatory monitoring of moderate elevations of blood pressure, along with headache diaries, fails to establish a cause-and-effect relationship between blood pressure and headache.
- hypertensive encephalopathy ➔ distinct syndrome

64

Headache in hypertensive encephalopathy

- Diffuse pain, pulsating,
- Signs of encephalopathy:
  - Confusion
  - Reduced level of consciousness
  - Visual disturbances (other then aura, incl blindness)
  - seizures

65

(Post)coital headache

- Sudden, severe headache which occurs at the moment of orgasm in men or women.
- ! first attack → distinguish it from SAH
- a history of previous attacks under similar circumstances→ RVCS!
- not associated with loss of consciousness or vomiting.
- tends to disappear spontaneously
- beta-adrenergic blockers can be a very effective preventive treatment.

66

Giant cell arteritis (GCA) or temporal arteritis

Giant cell arteritis (GCA) or temporal arteritis

- Subacute granulomatous inflammation of the external carotid arterial system (superficial temporal artery)
- Women twice as frequently affected as men
- Uncommon before age 50
- Average age at onset 72 yrs

68

Temporal arteritis

69

Typical GCA symptoms

- Headache
- Jaw claudication
- Fever
- Malaise
- Weight loss
- Anemia
- Polmyalgia rheumatica
- Diplopia
- Visual loss

70

Headache in GCA

- Unilateral or bilateral
- Severe
- Boring in quality
- Scalp tenderness
- Temporal arteries are non-pulsating & tender on examination

71

GCA diagnosis

- Biopsy of temporal artery
  - Should not delay treatment
- ESR usually around 100 mm/h (ESR increased in 97% of patients)
- Mean value 87mm/h ± 32 SD
- CRP more sensitive
- Normochromic normocytic anemia
- Thrombocytosis
- AP ↑ 15% patients
- Alfa-2 globulins in 72% patients

72

GCA treatment

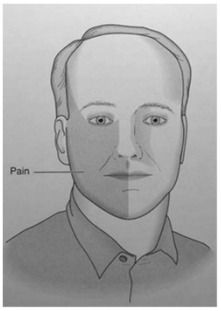
- Prednisone 1-1.5 mg/kg/day or loading dose of methylprenisolone 1-2 /day for 3 days followed by prednisone 1-1.5 mg/kg/day
- Prednisone slowly tapered when ESR or CRP is within normal limits, typically after 2 weeks after initiation of the therapy
- ESR/CRP monitoring required
- Therapy needed for 1-2 years (43% at steroids after 5 years)
- Complications of prolonged therapy

73

Trigeminal neuralgia

Trigeminal neuralgia

- Unilateral disorder characterized by brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more trigeminal nerve divisions
- Usually in persons of age 50 or more



75

Trigeminal neuralgia diagnostic criteria

- Paroxysmal attacks of pain lasting seconds to 2 minutes in the distribution of one trigeminal nerve
  - usually 2nd or 3rd,
  - only 5% of 1st div. of the 5 CN)
- Pain has one of the following characteristics:
  - Intense, sharp, superficial or stabbing
  - Precipitated from trigger areas or by trigger factor
- Attacks are stereotyped in the individual patient
- No clinically evident neurological deficit
- Not attributed to another disorder

76

Trigeminal neuralgia trigger areas and trigger factors

- Trigger areas:
  - the cheek,
  - nose,
  - mouth
- Trigger factors:
  - Cold wind on the face,
  - Washing
  - Shaving
  - Teeth-cleaning
  - Talking
  - Eating
  - Drinking

77

Trigeminal neuralgia – additional features

- The pain never crosses the midline
- May be bilateral
- Between paroxysms the patient is usually asymptomatic but a dull background pain may persist
- The pain often evokes a spasm of the muscles on the affected side (*tic douloureaux*)

78

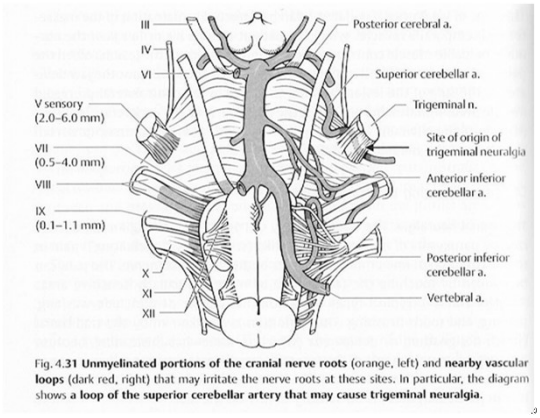


Fig. 4.31 **Unmyelinated portions of the cranial nerve roots** (orange, left) and **nearby vascular loops** (dark red, right) that may irritate the nerve roots at these sites. In particular, the diagram shows **a loop of the superior cerebellar artery that may cause trigeminal neuralgia.**

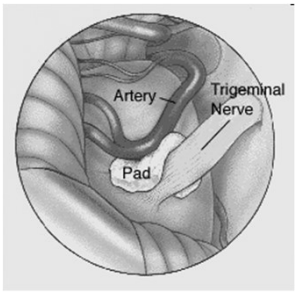
Trigeminal neuralgia treatment

- Gabapentin – 900 -3600 mg/d orally 3 doses
- Carbamazepine, 400–1200 mg/d orally in three divided doses occurs within 24 hours in such a high percentage of cases that some believe it to be diagnostic.
- Intravenous administration of phenytoin, 250 mg, will abort an acute attack,
- Phenytoin, 200–400 mg/d orally, may be effective in combination with carbamazepine if a second drug is necessary.
- Lamotrigine 100-400 mg/d
- Baclofen 10 mg three times daily–20 mg four times daily has been used in refractory cases.
- Posterior fossa microvascular decompressive surgery has been used in drug-resistant cases.

80

Trigeminal neuralgia

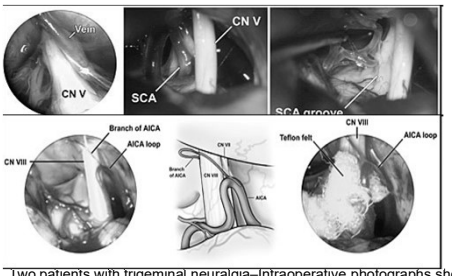
- The true cause of TN is uknown
- Many patient may have compression of the trigeminal nerve root by tortuous or aberrant vessels leading to demyelination



http://www.aans.org/education/journal/neurosurgical/november98/5-5-p1.asp

81

Microvascular decompression



- Two patients with trigeminal neuralgia–Intraoperative photographs show vessels compressing the trigeminal nerve. Note the groove on the nerve after microvascular decompression.
- 55 yr. old male presented with positional vertigo who underwent microvascular decompression of CN VIII with Teflon felt interpostion between the nerve and the contact vessel.

82

Treatment of headache in the ED

Treatment options

- Primary or secondary HA
- Secondary
  - directed treatment depending on cause
  - analgesics
- Primary
  - treatment depends on type of the headache
- Treatment for headache
  - IV treatment
  - **IV fluids** !
  - Options:
    - Magnesium
    - Neuroleptics
    - NSAIDS
    - Sodium valproate
    - Corticosteroids

84

Magnesium

- 250 mg – 1,0 g IV of magnesium sulfate over 30 min
- dissolve in saline if burning sensation
- monitor for hypotension
- 30% of patients will respond
- May be effective for secondary headaches – posttraumatic, intracerebral bleed, post infectious and non-vasoactive

85

Neuroleptics (1)

- Metoclopramid (Reglan)
  - 10 – 20 mg IV, can be repeated in 4 hrs if necessary
- Promethazine (Phenergan 50 mg/ml 1 ml)
  - 12,5-25 mg IV q 4 hrs

86

Neuroleptics (2)

- **Attention**:
  - Need ECG to check QTc
  - Pretreat with fluids IV (500-1000ml of normal saline– risk of hypotension
  - pretreat with diphenhydramine (25 mg IV) - risk of akathysia/dystonia
- **Prochlorperazine** (Compazine 5 mg/ml 10 ml)
  - 10 mg IV q4-6 hrs pretreat with diphenhydramine
- **Chlorpromazine** (25 mg/ml 1 ml)
  - 25 mg IV (1 ml) diluted in 4 ml of saline give 1 ml (5 mg) every 15 min until pain relief or max 25 mg
- Droperidol most effective highest risk – do not use

87

Valproate, Sodium

- Minimal adverse events,
- few contraindications (pregancy, lacation, liver disease)
- Preps:
  - Depacone 500 mg 5 ml vial
  - Poland: valproic acid: Depakine 400 mg 4 ml
- Dosing: 250 – 1000 mg IV slow infusion

88

NSAIDS

- Migraine
  - Acetylosalicylic acid (ASA)
    - IV 1000 mg ASA with or without metoclopramide
  - dihydroergotamine 2 mg (nasal spray or suppositories) is recommended for severe migraine attacks
  - metamizol superior to placebo but can cause severe arterial hypotension and allergic reactions

89

Steroids

- For the treatment of a status migrainosus,
- prednisone;
  - 50–100 mg IV
  - dexamethasone 10 mg IV
- recommended by expert consensus (EFNS G.)

90

Imaging in ED

- All patients with the thunderclap headache
- Initial headache or worst in life
- Headache that changes in quality, frequency, duration or severity
- Abnormal neurological examination
- Headache assoc. with seizures or EEG abnormalities
- Headache assoc. with fever or nuchal rigidity
- Planned LP (to exclude space occup. lesion)
- Headache in HIV positive patient
- New cough or exertional headache

91

Other headaches in the ED

92

Pseudotumor cerebri

Pseudotumor cerebri (PTC, idiopatic intracranial hypertension)

- a diffuse increase in intracranial pressure causing headache, papilloedema, and diminished visual acuity.
- most cases are idiopathic.
- In the idiopathic variety, women are affected much more commonly than men, with a peak incidence in the third decade.

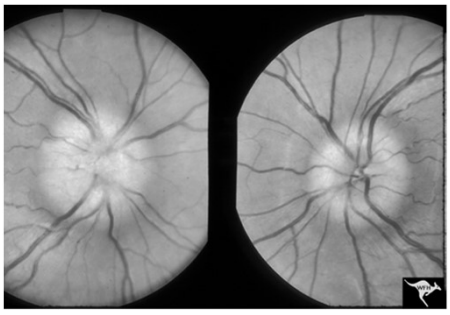
94

PTC S&S:

- Diffuse headache - almost always as a presenting symptom,
- diplopia (abducens nerve palsy) and blurred vision occur in 60% of cases.
- moderate to severe papilloedema is seen in almost 90%.
- Visual loss from increased intracranial pressure can occur

95

Moderate to severe papilledema is seen in almost 90% case of PTC.



96

Headaches associated with brain tumours

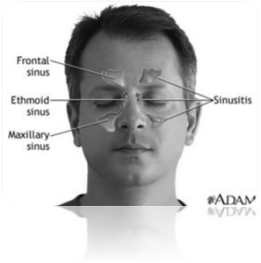
Headaches associated with brain tumours

- most often nonspecific in character,
- mild to moderate in severity,
- dull and steady in nature, and intermittent.
- bifrontal, worse ipsilaterally,
- aggravated by a change in position or by Valsalva maneuver that increase intracranial pressure, (coughing, sneezing, and straining at stool)
- classically maximal on awakening in the morning and is associated with nausea and vomiting.

98

Sinus Headache (rhinosinusitis)

- The pain of frontal, sphenoid, or ethmoid nasal sinusitis
  - felt in the middle of the forehead and above the eyes.
- maxillary sinusitis
  - radiates to the upper jaw and zygomatic region and worsens when the patient bends forward



99

- Sinusitis is treated with vasoconstrictor nose drops (eg, phenylephrine, 0.25%, instilled every 2–3 hours), antihistamines, and antibiotics.
- In refractory cases, sinus drainage may be necessary.

100

Chronic sinus pain vs migraine

- Rhinosinusitis is overdiagnosed as cause of headache
- Patients who complain of recurrent pain on the sinuses rarely have recurrent inflammation of the sinuses;
- they are much more likely to have migraine or tension headaches

101

Glossopharyngeal neuralgia

- Uncommon pain syndrome
- 100 x less prevalent then trigeminal n.
- Pain:
  - a paroxysmal pain that is identical in quality to that of trigeminal neuralgia,
  - or a continuous burning or aching discomfort

102

Glossopharyngeal neuralgia pain localization

- The pain is localized to
  - the oropharynx,
  - the tonsillar pillars,
  - the base of the tongue,
  - or the auditory meatus.

103

Trigger areas

- The trigger areas are:
  - around the tonsillar pillars,
  - symptoms are initiated by swallowing or by talking.

104

Associated symptoms

- Syncopal episodes caused by transient bradyarrhythmias.
- Men are affected more often than women
- Examination reveals no abnormal neurologic signs.

105

GPN treatment

- Carbamazepine or phenytoin therapy (as described above for trigeminal neuralgia) usually produces dramatic relief;
- microvascular decompression

106

Master and Margarita Chapter 2. Pontius Pilate

- Early in the morning on the fourteenth of the spring month of Nisan the Procurator of Judaea, Pontius Pilate, in a white cloak lined with blood-red, emerged with his shuffling cavalryman's walk into the arcade connecting the two wings of the palace of Herod the Great.
- **More than anything else in the world the Procurator hated the smell of attar of roses. The omens for the day were bad, as this scent had been haunting him since dawn. It seemed to the Procurator that the very cypresses and palms in the garden were exuding the smell of roses, that this damned stench of roses was even mingling with the smell of leather tackle and sweat from his mounted bodyguard.**
- A haze of smoke was drifting towards the arcade across the upper courtyard of the garden, coming from the wing at the rear of the palace, the quarters of the first cohort of the XII Legion ; known as the ' Lightning', it had been stationed in Jerusalem since the Procurator's arrival. The same oily perfume of roses was mixed with the acrid smoke that showed that the centuries' cooks had started to prepare breakfast.
- 'Oh gods, what are you punishing me for? . . . No, there's no doubt, **I have it again, this terrible incurable pain . . . hemicrania, when half the head aches . . . there's no cure for it, nothing helps. ... I must try not to move my head. . . . '**

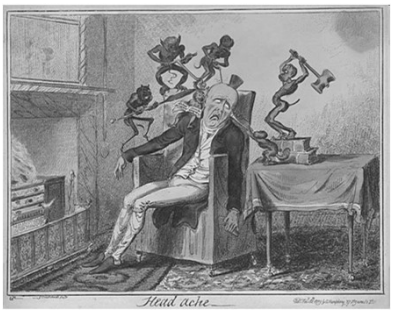
107

Diagnostic criteria of migraine (IHS classification)

- A. At least five attacks fulfilling criteria B–D
- B. Headache lasting 4–72 h (untreated or unsuccessfully treated)
- C. Headache has at least two of the following characteristics:
  - 1. Unilateral location
  - 2. Pulsating quality
  - 3. Moderate or severe pain intensity
  - 4. Aggravation by or causing avoidance of routine physical activity (e.g. walking or climbing stairs)
- D. During headache at least one of the following:
  - 1. Nausea and/or vomiting
  - 2. Photophobia and phonophobia
- E. Not attributed to another disorder

108

Migraine sufferer by George Cruikshank



109

Types

- Migraine without aura
- Migraine with aura
  - Migraine with typical aura
  - Familial hemiplegic migraine
  - Basilar migraine
  - Migraine aura without headache
  - Migraine with acute onset aura
- Ophthalmoplegic migraine
- Retinal migraine

110

Identification of migraine (self screener)

- Are you nauseated or sick to your stomach when you have a headache?
- Have the headaches limited your activities for a day or more in the last 3 months?
- Does light bother you when you have a headache?

- If 2 of 3 answers are affirmative, the positive predictive value of that patient having migraine headache is 93%.
- If all 3 answers are affirmative, the chance of that patient having migraine is 98%.

111

Childhood periodic syndrome

- May be precursor to or associated with migraine
  - Benign paroxysmal vertigo of childhood
  - Alternating hemiplegia of childhood
  - Cyclic vomiting

112

Migraine with typical aura

1. At least 2 attacks with the following
2. Aura consisting of visual, sensory and/or speech/ language symptoms, each fully reversible, but no motor, brainstem or retinal symptoms
3. At least two of the following:
   1. At least one symptom develops gradually over >5 minutes and / or two or more symptoms occur in succession over >5 min
   2. Each aura symptom lasts 5-60 min. (Duration of aura can be 180 min with successive symptoms
   3. ≥1 aura symptom is unilateral (Aphasia is considered unilateral. Dysartria may or may not be considered unilateral)
   4. the aura is accompanied, or followed within 60 minutes, by headache
- Not better accounted for by another ICHD-3 diagnosis, and transient ischemic attack has been excluded.

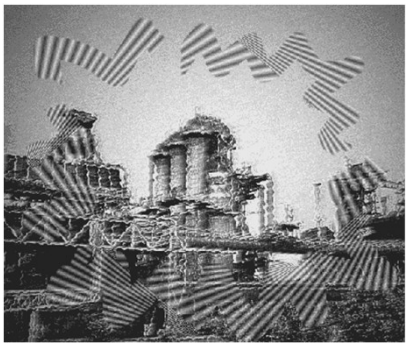
113

Aura

- Complex of neurological symptoms that occurs just before the headache
  - Reversible focal neurological deficit
- Usually develops gradually over 5-20 minutes
- Lasts for less then 60 minutes
- Afterwards the headache with migrainous features develops
- Less frequently there is no headache at all or it lacks features of migraine

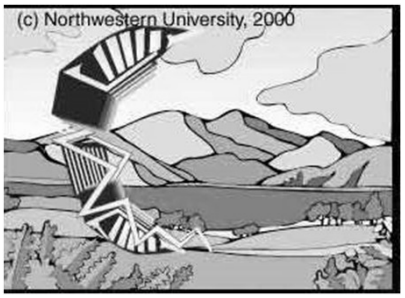
114

Visual aura - fortification spectra 1



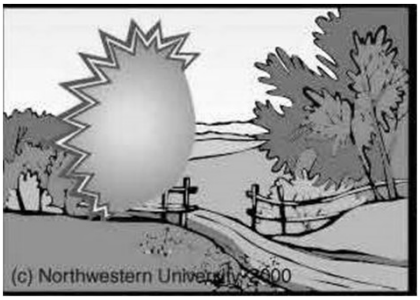
115

Fortification spectra as might be seen in migraine with aura.



116

Scotoma with aspects of a fortification.



117

Complications of migraine

- Chronic migraine
- Status migrainosus
- Persistent aura without infarction
- Migrainous infarction
- Migraine-triggered seizures

118

S&S 1

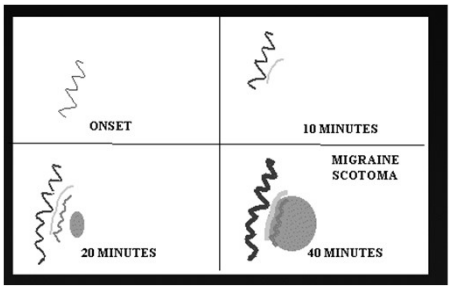
- Premonitory phase (also called *prodrome* or *warning symptoms* - not aura!)
  - Hyperactivity
  - Hypoactivity
  - Depression
  - Craving for particular foods
  - Repetitive yawing
  - Pallor
  - Blurred vision

119

Migraine triggers

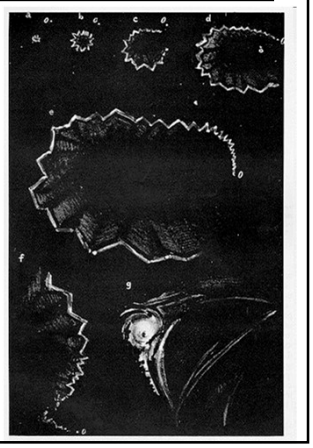
- Stress and fatigue
- Relaxation after stress ('Saturday morning migraine')
- Skipping meals
- Binge eating
- Specific foods (cheese, citrus fruits, etc.)
- Specific drinks: caffeine (too much or sudden withdrawal); red wine (too much)
- Menstruation and ovulation
- Oral contraceptives
- Early post-partum period
- Menopause
- Bright sunshine, flicker or patterns
- Strong smells
- Hypertension
- Head injury

120



121

Dr. Hubert Airy's own migraine scotomas



Other forms of the aura

- Sensory aura
  - Sensation "of pins and needles" moving slowly from the point of origin and affecting a greater or smaller part of one side of the body and face
- Speech disturbances
  - Usually dysphasia
- Motor weakness
  - ➔ hemiplegic migraine!

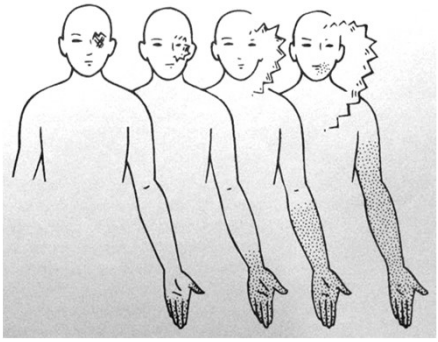
123

Brainstem aura ➔ basilar migraine

- At least two of the following
  - Dysarthria
  - Vertigo
  - Tinnitus
  - Hypoacusia
  - Diplopia
  - Ataxia
  - Decreased level of consciousness
  - Simultaneous bilateral paraesthesias

124

Sensory and visual aura



125

Current EFNS guideline on the drug treatment of migraine

- Drug treatment of migraine attacks
- Migraine prophylaxis

126

Treatment of migraine attacks

- Analgesics
- Antiemetics
- Ergot alkaloids
- Triptans (5-$HT_{1B/1D}$-agonists)

127

Acute treatment – analgesics (level A)

- Acetylsalicylic acid (ASA) 1000 mg oral or 1000 mg i.v.
- Ibuprofen 200–800 mg
- Naproxen 500–1000 mg
- Diclophenac 50–100 mg
- Paracetamol 1000 mg (oral) 1000 mg (supp.)
- ASA plus, 250 mg (oral
  - Paracetamol 200–250 mg
  - and caffeine 50 mg

128

Acute treatment – analgesics (level B)

- Metamizol 1000 mg oral or 1000 mg i.v.
- Phenazon 1000 mg (oral)
- Tolfenamic acid 200 mg (oral)

129

Antiemetics

- Metoclopramide B
- Dose (mg) and route
  - 10–20 (oral)
  - 20 (suppository)
  - 10 (intramuscular, intravenous,subcutaneous)
- Side effect: dyskinesia; contraindicated in childhood and in pregnancy; also analgesic efficacy
- Domperidon [B]
  - 20–30 (oral)
  - Side effects less severe than in metoclopramide; can be given to children

130

Acute treatment - triptans

- Sumatriptan (Imigran): 25, 50 and 100 mg (oral including rapid-release); 25 mg (suppository); 10 and 20 mg (nasal spray); 6 mg (subcutaneous)
- Zolmitriptan (Zomig): 2.5 and 5 mg (oral including disintegrating form) 2.5 and 5 mg (nasal spray)
- Rizatriptan (Maxalt RPD): 10 mg (oral including wafer form; dose 5 mg when taking propranolol
- Eletriptan (Relpax): 20 and 40 mg (oral); 80 mg allowed if 40 mg not effective
- Other:
  - Naratriptan
  - Almotriptan
  - Frovatriptan

131

Side effects for triptans

- Chest symptoms,
- Nausea,
- Distal paraesthesia,
- Fatigue.

132

Contraindications for triptans

- Untreated arterial hypertension
- Coronary heart disease
- Cerebrovascular disease
- Reynaud's disease
- History of ischemic stroke
- Pregnancy & lactation
- age under 18 (except sumatriptan nasal spray) and age above 65
- Severe liver or renal failure

133

Ergot alkaloids

- Should be restricted to patients with very long migraine attacks or with regular recurrence
- **Ergotamine tartrate** 2 mg (oral or suppositories) - the only compound with sufficient evidence of efficacy
- use must be limited to 10 days/month (can induce drug overuse headache very fast and in very low doses)

134

Prophylactic drug treatment of migraine

- High frequency of attacks (2 or more/month [OR >4/month])
- The quality of life, business duties, or school attendance are severely impaired
- Contraindication to, failure, adverse effects, or overuse of acute therapies
- Patient preference
- Presence of uncommon migraine conditions, including hemiplegic migraine, basilar migraine, migraine with prolonged aura, or migrainous infarction

135

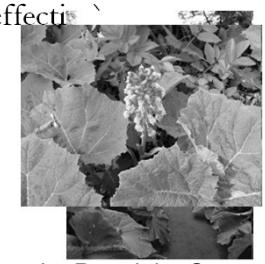
First choice drugs for the prophylactic migraine treatment

- Betablockers
  - Propranolol 40–240 mg (a sustained release formulation (Inderal LA)
  - Metoprolol (Toprol XL) 50–200 mg
  - Timolol
- Calcium channel blockers
  - Flunarizine 5–10 mg
- Antiepileptic drugs
  - Valproic acid 500–1800 mg
  - Topiramate 25–100 mg

136

Drugs of second choice for migraine prophylaxis (probably effecti

- Antidepressants
  - Amitriptyline 50 –150
  - Venlafaxine 75–150
- Beta-blockers
  - Naproxen 2 x 250–500
  - Bisoprolol 5–10
- butterbur root extract formula: Petadolex® GelCaps 2 x 75 (latin: Petasites hybridus; Norwegian „pestrot", Italian: *Farfaccio maggiore)*



137

Drugs of third choice for migraine prophylaxis (possible efficacy)

- Lisinopril 20 mg
- Candesartan 16 mg
- Acetylsalicylic acid 300 mg
- Gabapentin 1200–1600 mg
- Magnesium 24 mmol / d
- Riboflavin 400 mg
- Coenzyme Q10 300 mg

138

Case **b.**

- 'It's a tight band around my head, holding it in a vice, like something's expanding inside.'
- Tension headache gives a constant feeling of pressure in the head, often described in powerful language and accompanied by an overt or unspoken fear of an underlying brain tumour.

139

Tension type headache

- Most common primary headache
- Lifetime prevalence 30-78%

140

TTH Definition

- Headache lasts from 30 minutes to 7 days
- Has at least two of the features:
  - Bilateral location
  - Pressing/tightening (non-pulsating) quality
  - Mild to moderate intensity
  - Not aggravated by routine physical activity (walking, climbing stairs)
- Both of the following
  - No nausea or vomiting (anorexia may occur)
  - No more than one of photophobia or phonophobia

141

TTH types

TTH type	Episodes per month/per year	Nausea?
Infrequent episodic	1/12	No
Frequent episodic	<15/<180	No
Chronic TTH	≥15/ ≥180	**mild** nausea

142

Symptoms

- usually bilateral with occipitonuchal, temporal, or frontal predominance or diffuse extension over the top of the cranium.
- dull and aching,
  - such as fullness, tightness, or pressure (as though the head were surrounded by a band or clamped in a vise)
  - a feeling that the head is swollen and may burst.
- waves of aching pain
  - may be interpreted as paroxysmal or throbbing
- No throbbing, nausea, photophobia, nor phonophobia
- TTH do not interfere seriously with daily activities (as migraine does)

143

Treatment

- Usually non-rewarding
- Simple analgesics, (aspirin, acetaminophen or other NSAIDs)
  - may be helpful, but only for brief periods.
- drugs that relieve anxiety or depression, especially when these symptoms are present.
- Stronger analgesic medication should be avoided.

144

Case c.

- 'Every night at 2 o'clock, like someone is squeezing my eyeball very hard.'
- The predictable diurnal timing and intense orbital pain are typical of cluster headache.

145

Cluster headache

Cluster headache eponims

- *paroxysmal nocturnal cephalalgia* (Adams),
- *migrainous neuralgia* (Harris),
- *histamine cephalalgia* (Horton),
- *red migraine,*
- *erythromelalgia of the head*,
- "*cluster headache*" (Kunkle and colleagues),

147

Cluster headache

- young adult men (range 20 to 50 years; male-to-female ratio about 5:1)
- a consistent unilateral orbital or temporal localization.
- a nightly recurrence, between 1 and 2 h after the onset of sleep, or several times during the night;
- Non-throbbing, intense

148

Cluster headache

- Cluster periods lasting 4-8 weeks 1-3 times per year
- Occurs 1-8 times per day,
- lasts 15-180 minutes (usually 1 hr)
- Associated with:
  - motor restlesness
  - ipsilateral autonomic features (concunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis, eyelid edema)
- Provoked by nitrates, alcohol

149

Cluster headache treatment

- Acute treatment
  - Inhaling high-flow oxygen (7 l/min for 15 min facial mask)
  - Triptans (sumatriptan s.c. 6 mg)
  - Steroids
- Prophylactic treatment
  - Introduce during acute treatment
  - Valproic acid 600-1800 mg/d
  - Verapamil 320-480 mg/d

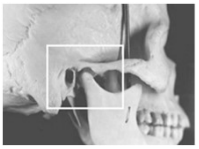
150

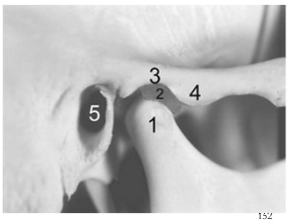
Temporo-mandibular joint disorders

151

Temporomandibular joint dysfunction (TMJ, Costen syndrome)

- A craniofacial pain consequent on dysfunction of one temporomandibular joint





152

Pain in TMJ syndrome

- Unilateral, behind or front of ear, temple, face
- Severe aching pain, intensified by chewing
- Tenderness over temporomandibular joints
- Associated
  - Malocclusion, missing molars
- Pain increased by
  - Chewing
  - Pressure over temporomandibular joints

153

TMJ syndrome - Associated diseases & Treatment

- Associated diseases
  - Loss of teeth,
  - Rheumatoid arthritis
- Treatment
  - Correction of bite
  - Surgery in some
  - Amitryptyline
  - myorelaxants

154

General tests

- CBC (+ differential)
- ESR/CRP
- Chemistry (glucose, Na, K, Ca, BUN/creatinin, bilirubin)
- TSH (?)
- Coagulation screen
- X-rays (depending on indication)
  - sinus
  - C-spine
  - chest

155

Imaging in ED

- All patients with the thunderclap headache
- Initial headache or worst in life
- Headache that changes in quality, frequency, duration or severity
- Abnormal neurological examination
- Headache assoc. with seizures or EEG abnormalities
- Headache assoc. with fever or nuchal rigidity
- Planned LP (to exclude space occup. lesion)
- Headache in HIV positive patient
- New cough or exertional headache

156

Case a.

- ‘It was like being hit on the back of the head with a baseball bat.’
- This is the typical sudden, severe history that should make you think of subarachnoid haemorrhage.

157

Case g.

- ‘Cruel, sudden, I can’t brush my teeth.’
- Brief, lancinating pains triggered by stimulation of the face or mouth suggest trigeminal neuralgia.

158

Case h.

- ‘Awful when I sit up, fine lying flat.’
- Low-pressure headache is postural and relieved by lying flat.

159

Case j.

- ‘It’s very embarrassing, doctor, it always happens just as I’m, you know. . . .’
- Repeated sudden headaches at the moment of orgasms are due to benign sex headache.

160

Case e.

- ‘All over my head, it feels so tender, I’ve never felt so bad in all my years.’
- In an elderly patient, a new headache with scalp tenderness and malaise means giant cell arteritis until proved otherwise: ESR and steroids in the meantime.

161

Case d.

- ‘Mainly in the mornings and when I bend over.’
- The headache of raised intracranial pressure.
- Morning headaches are more often due to *migraine*, *hangovers*, *anxiety* about the day ahead or *obstructive sleep apnoea*, while headaches on stooping are very common in *sinusitis*, but look hard for signs of raised intracranial pressure when you hear this story.

162