Clinical Considerations in the Diagnosis and Treatment of Secondary Headaches (Part II)

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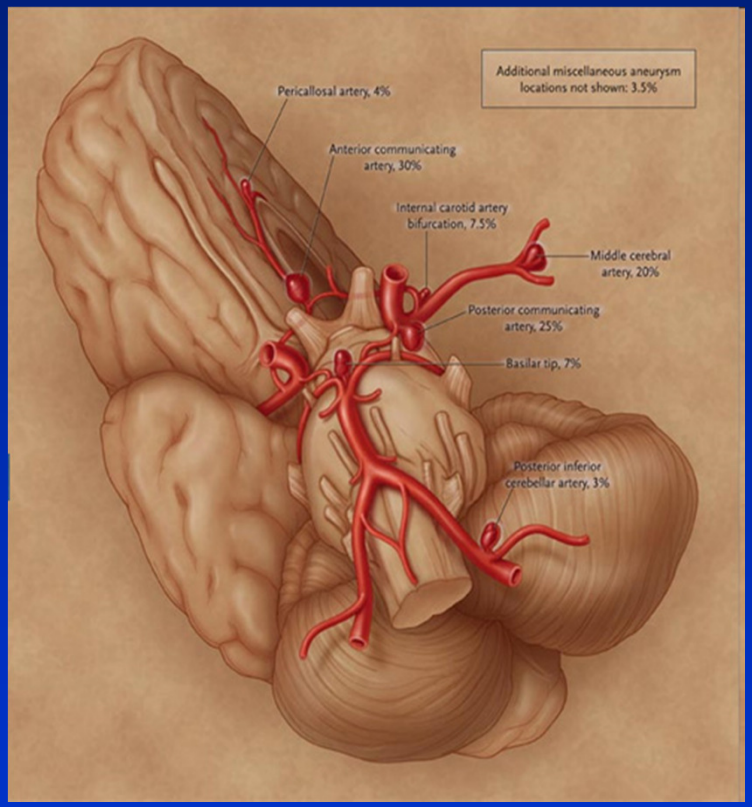
Headache Attributed to Cranial Vascular Disorder (Cont'd)

- **Unruptured vascular malformation (MRIMRA/angiography evidence) :**
  - **Saccular aneurysm: headache resolves < 72 hr)**
  - **Arteriovenous malformation (AVM): same as above**
  - **Cavenous angioma (grossly dilated blood vessels with a single layer of endothelium and an absence of neuronal tissue within the lesions. Recurrent headaches, focal neurological deficits, hemorrhagic stroke, and seizure**
  - **Sturge Weber syndrome: encephalotrigeminal or leptomeningeal angiomatosis, congenital and skin disorder. It is often associated with port-wine stains of the face, seizure, mental retardation, and ipsilateral leptomeningeal angioma, AVM.**

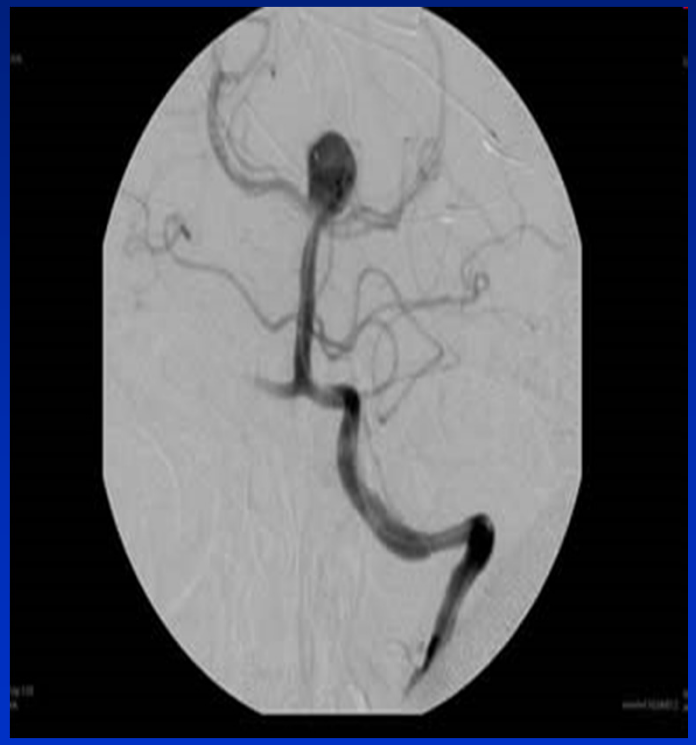
Saccular Aneurysm (Intracranial Berry Aneurysm)



- a sac-like outpouching in a cerebral blood vessel, berry-shaped, hence the name.
- likely to rupture, causing a stroke.
- medical emergencies, and should be treated as soon as possible.
- incidence is 1 in 10000 people per year, mortality rate of 70-90%.
- Circle of Willis.
- Hint: Young adult with headache, double vision or loss of vision

http://emedicine.medscape.com/article/252142-overview
http://en.wikipedia.org/wiki/Intracranial_berry_aneurysm

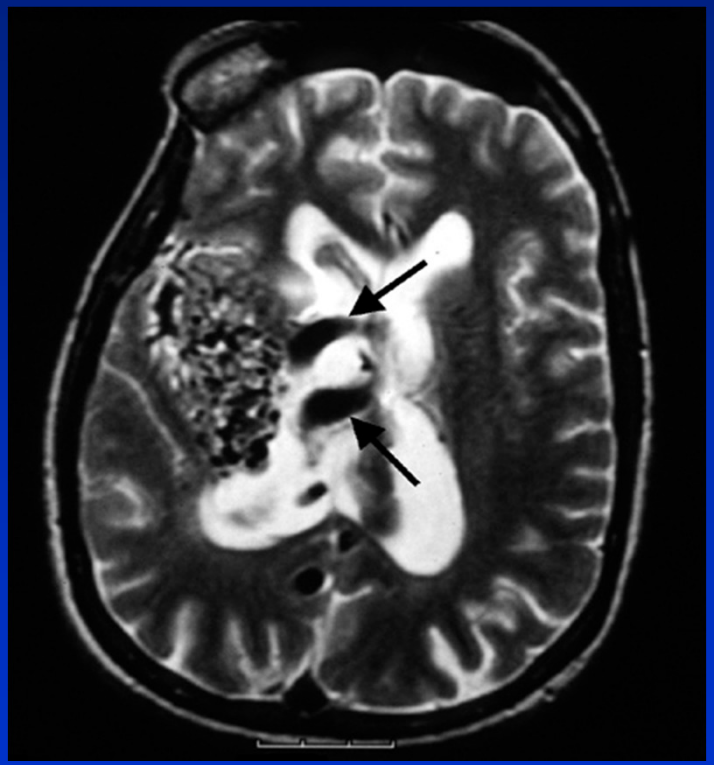
Saccular Aneurysm (Intracranial Berry Aneurysm)



Basilartipaneurysm

- **Hint: Young adult with headache, double vision or loss of vision**

Arteriovenous malformation (AVM)

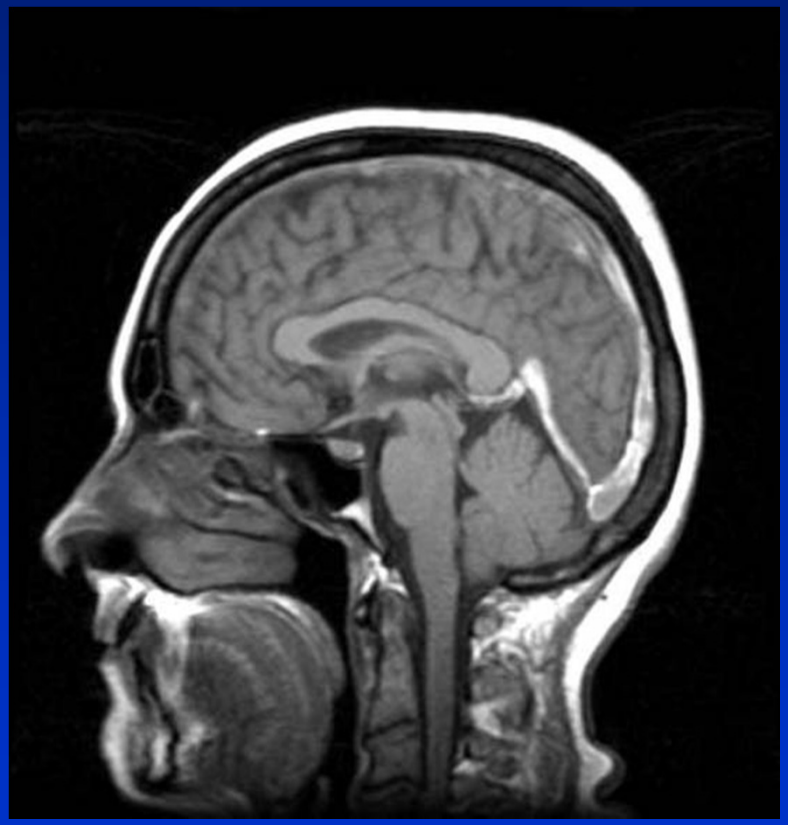


Axial T2-weighted MRI

- an abnormal connection between veins and arteries, usually congenital
- Hint: headache and epilepsy

http://emedicine.medscape.com/article/252426-overview

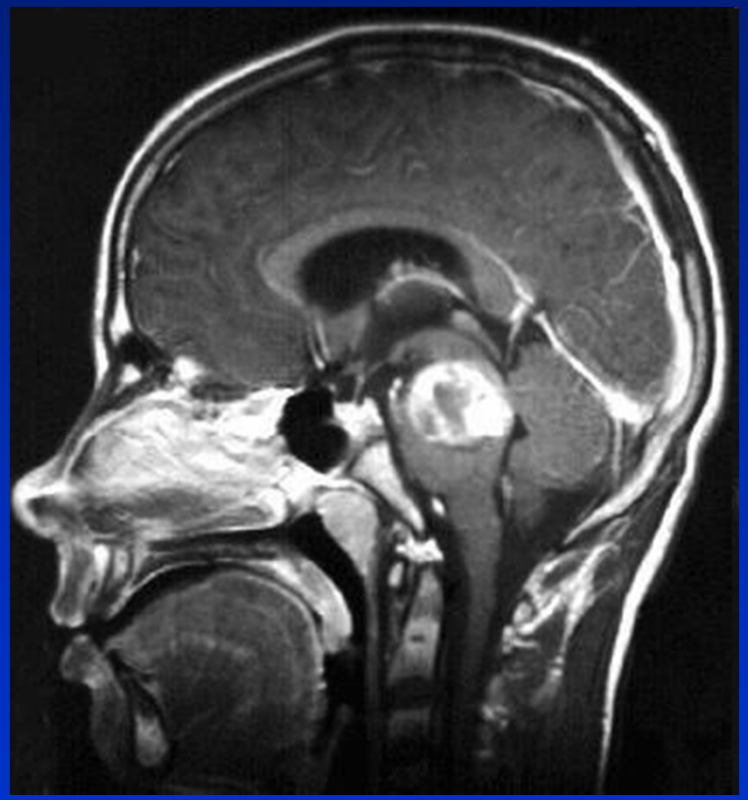
Cerebral Venous Sinus Thrombosis



- thrombosis (a blood clot) of the dural venous sinuses, which drain blood from the brain.
- Headache, abnormal vision, stroke-like symptoms (paralysis), seizure, altered mental status, increased ICP
- Hint: history or risk factors for blood clotting (e.g. pregnancy, use of OCP, abnormality of coagulation (presence of factor V leiden, deficiency of protein C and protein S, or antithrombin.

Bousser MG et al. In: Wolff's Headache And Other Head Pain. 2001.

Cavenous angioma

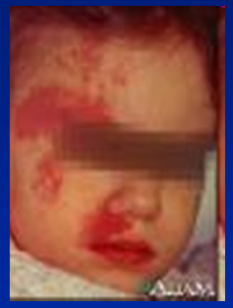


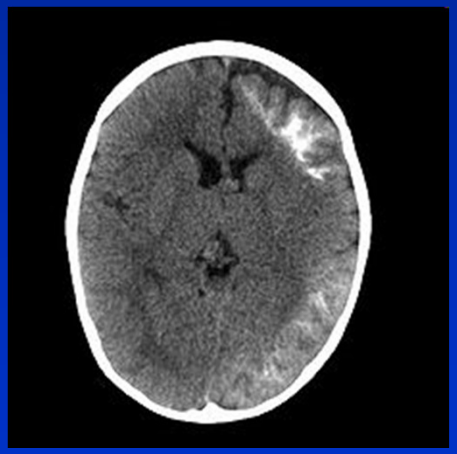
Axial T2-weighted MRI

- grossly dilated blood vessels with a single layer of endothelium and an absence of neuronal tissue within the lesions.
- Focal neurological deficits, hemorrhagic stroke, and seizure
- Hint: recurrent headache and focal neurological signs

http://en.wikipedia.org/wiki/File:Cavernous_hemangioma_t2.jpg

Sturge Weber Syndrome





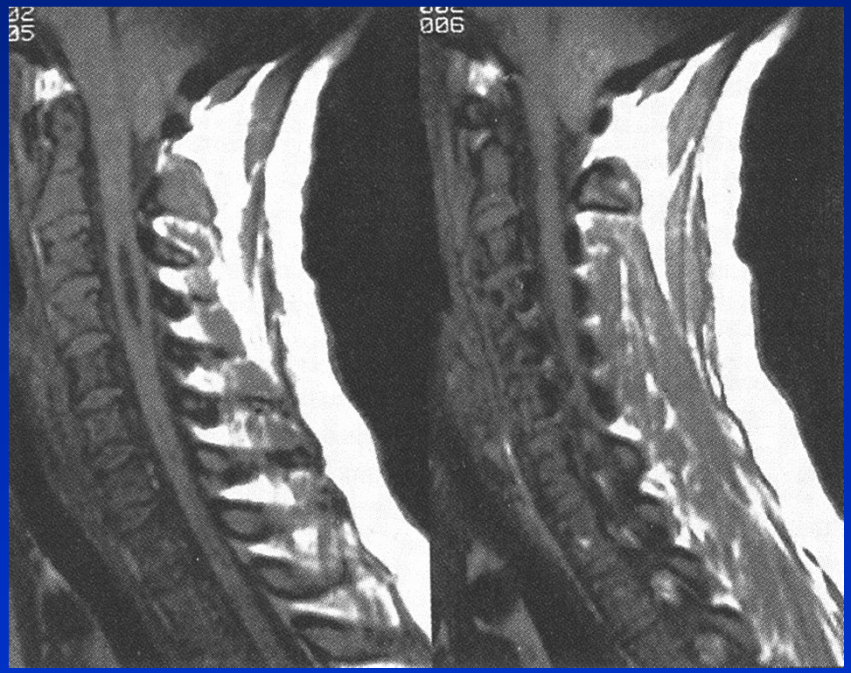
CT

- Encephalotrigeminal or leptomeningeal angiomatosis, congenital, and skin disorder.
- Associated with port-wine stains of the face, seizure, mental retardation, and ipsilateral leptomeningeal angioma, AVM
- Hint: headache with facial skin stain

http://rarediseases.about.com/od/rarediseasess/a/sturgeweber.htm

http://en.wikipedia.org/wiki/Sturge%E2%80%93Weber_syndrome

Arnold-Chiari Malformation



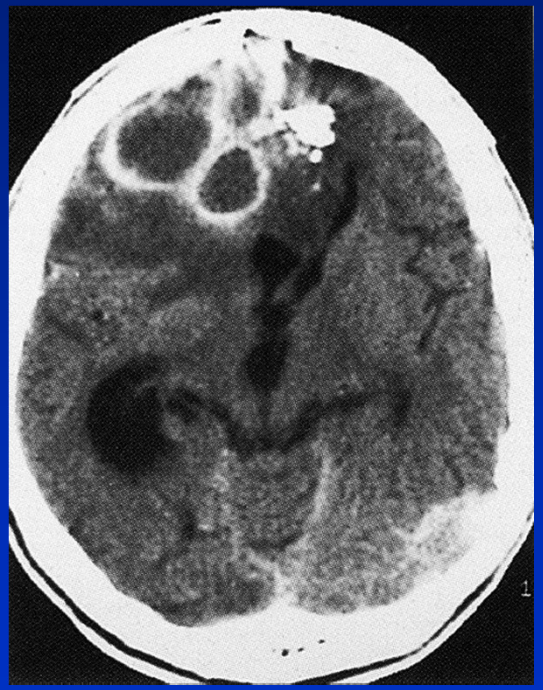
- A downward displacement of the cerebellar tonsils > 5 mm below the foramen magnum, with or without syrinx
- Sometimes causing non-communicating hydrocephalus as a result of obstruction of CSF outflow.
- Headaches, neck pain, weakness, ataxia
- Hint: persistent occipital headache, tinnitus
- Tx: decompression surgery

Silberstein SD et al. Headache in Clinical Practice. 2002.

Headache Attributed to Giant Cell Arteritis (GCA)

- Temporal headache and temporal artery tenderness, jaw claudication
- Elevated ESR: M:age/2; F: (age+10)/2. Elevated CRP
- TA with normal ESR: 10-36%
- Treatment: prednisone

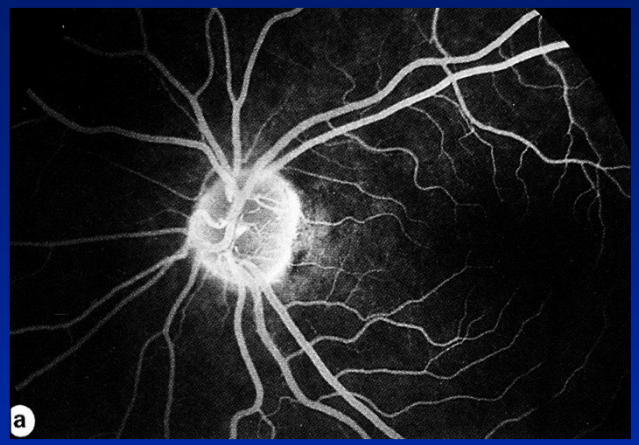
Brain Abscess



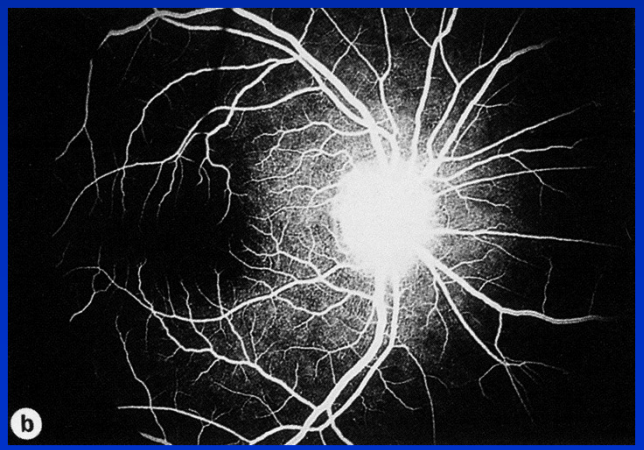
- Headache, drowsiness, confusion, seizure, hemiparesis, fever
- Hint: preceding infection or risk of infection (HIV, endocarditis, sinusitis, mastoiditis)

Silberstein SD et al. Headache in Clinical Practice. 2002.

Idiopathic intracranial hypertension (IIH)



Normal

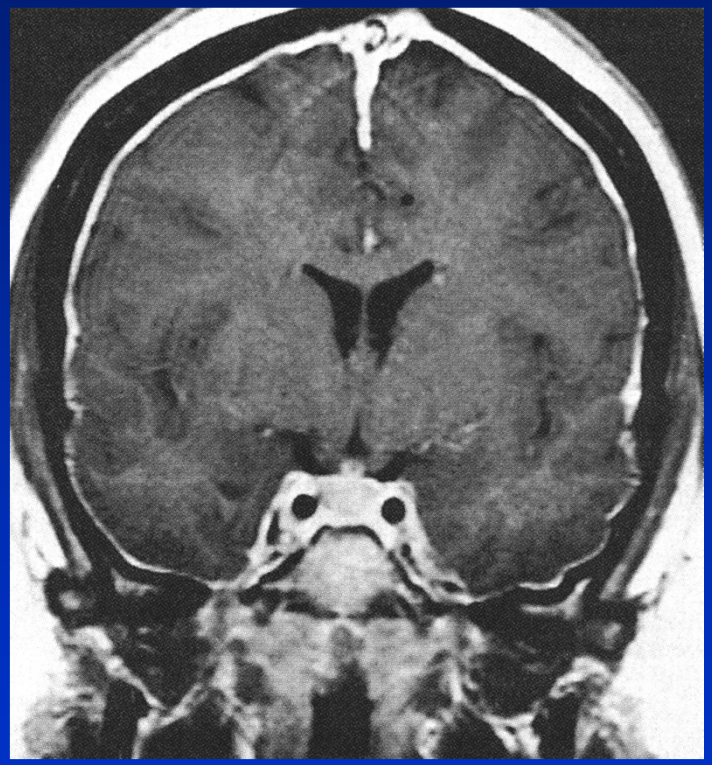


Papilledema

- Neuroimaging negative
- Opening pressure > 200 mm H2O in non-obese, > 250 mm in obese patients.
- Papilledema may be negative
- Hint: positional headache, tinnitus or hearing noise, visual disturbance
- Tx: diamox, topamax, lasix, repeated lumbar puncture (120-170 mm H20), ophthalmology consultation

Silberstein SD et al. Headache in Clinical Practice. 2002.

Low CSF Pressure Headache



Diffuse pachymeningeal enhancement

- With or without head or neck injury history
- CSF leakage detected by MRI, CT myelography or cisternography
- Opening pressure < 60 mm H2O in sitting position
- Hint: Worsened headache within15 min after sitting or standing, improved after lying, tinnitus or hearing “noise”
- Tx: epidural blood patch x 1-3

Silberstein SD et al. Headache in Clinical Practice. 2002.

Headache Attributed to epileptic seizure (IHS-2)

- **Hemicrania epileptica**
  - A. Headache lasting seconds to minutes, with features of migraine, fulfilling C and D
  - B. Having a partial epileptic seizure
  - C. Headache develops synchronously with the seizure and is ipsilateral to the ictal discharge
  - D. Headache resolves immediately after the seizure

Headache Attributed to epileptic seizure (IHS-2)

- **Post-ictal headache**
  - A. Headache with features of tension-type headache or, in a patient with migraine, of migraine headache and fulfilling C and D
  - B. Has had a partial or generalized epileptic seizure
  - C. Headache develops within 3 hours following the seizure
  - D. Headache resolves within 72 hours after the seizure

Clinical Facts of Post-Ictal Headache

- Duration: most often lasts 6-72 hours
- Migraine symptoms: In Non-migraine epileptics: 50% presents with vomiting, photophobia, phonophia; headache increased by coughing, benign and sudden headache movement; relieved by sleep.
- Headache incidence in relation to epileptic location: temporal lobe seizure (23-41%), frontal lobe (40-42%), occipital lobe (59-62%)
- Seizure type: more often associated with generalized tonic-clonic seizure (GTC)

Ito M. et al. Psychiatry and Clinical Neurosciences. 2002; 53:385-389.
Ito M et al. Acta Neural Scand. 2000;102:129-131.
Schon F & Blau JN. Journal of Neurology, Neurosgurery, and Psychiatry. 1987;50:1148-1152

Headache and Multiple Sclerosis

- Incidence of MS: > 100/100,000 in UK;
- MS population: 250,000-350,000 in USA.
- Young Caucasian adults living in temperate climates
- Female:male ratio
  - In adult: 2:1
  - In children and adolescents: 5-10:1

Pretorius PM & Quaghebeur G. Clinical Radiology. 2003;58:434-448.

Characteristics of MS

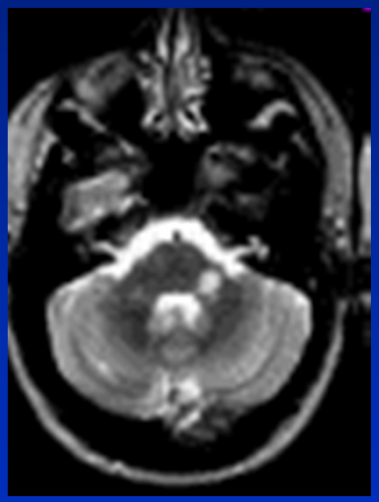
- Clinical hallmark: recurrent neurologic deficits that disseminated in space and time.
- Neurological signs:
  - Mental status: dementia, emotional labile, dysarthria
  - CN: optic atrophy, papillitis, aftterent pupillary defect, internuclear ophthalmoplegia, nystagmus
  - Motor: Spastic hemiparesis, paraparesis, or quadraparesis
  - Sensory: variable
  - Coordination: intention tremor
  - Reflexes: hyperreflexia, clonus
  - Gait: Truncal ataxia
- Hint: young female with paresthesia to the extremity and/or blurred vision, followed a few months later with loss of balance and/or muscle weakness

Diagnosis of MS

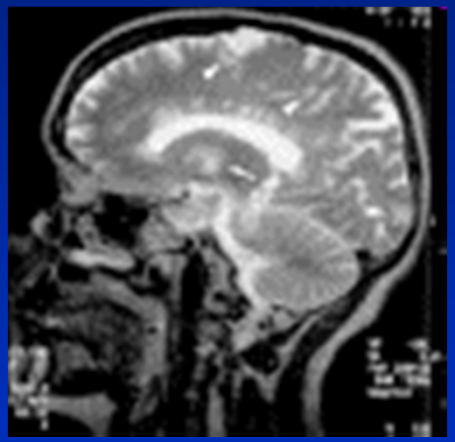
- Clinical history, symptoms, signs
- Brain MRI: subcortical white matter lesion (T2, or T1 gadolinium-enhancing lesion) in 95% cases.
- CSF: presence of oligoclonal bands (OCBs, 90% cases), IgG index > 0.7 (75% cases), myelin basic protein
- Evoked potentials (EPs)
  - Visual (VEP)
  - Somatosensory (SSEP)
  - Brainstem auditory (BAEP)

Giesser BS. Neurol. Clin. 2011;29:381-388.
Marshall RS & Mayer SA. On Call Neurology. 2nd ed. Philadelphia. 2001.
Pretorius PM & Quaghebeur G. Clinical Radiology. 2003;58:434-448.

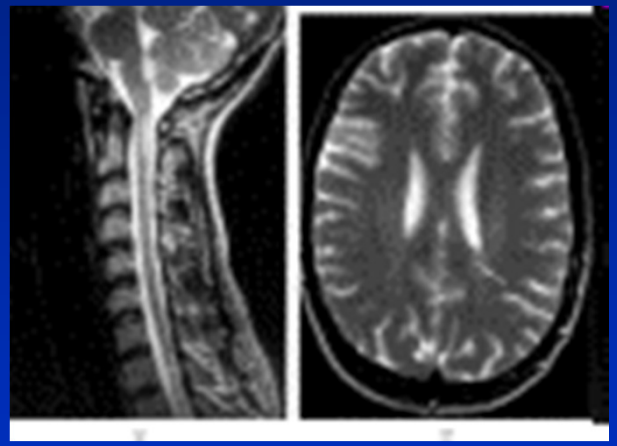
Brain MRI in MS



(Axial T2. Lesions in lateral pone and left cerebella lesion)



(Saggital T2. Lesions as "Dawson's fingers" at corpus callosum and in the thalamus.)



(Saggital T2, lesions in the posterior spinal cord and near left lateral ventricle.)

Pretorius PM & Quaghebeur G. Clinical Radiology. 2003;58:434-448.

Headache and Multiple Sclerosis

- Incidence of headache in MS patients: > 50% vs. 23% in control.
- Headache types: migraine, tension-type, cluster, occipital neuralgia
  - Most common: tension-type, migraine w/o aura
- Possible causes:
  - Demyelinating lesion in cervical spinal cord, brainstem (e.g. pontine), thalamus.
  - Side effect of medicine, e.g. interferon (disease modifying therapy)

Mantia L. Neurol. Sci. 2009;30:suppl 1:S23-6.
Gentile S et al. J. Headache Pain. 2007;8:245-7.
Vacca G et al. Neurol. Sci. 2007;28:133-5.
Gee JR et al. Headache. 2005;45:670-7.

Post-traumatic Headache

- With or without loss of consciousness after the trauma
- Acute post-traumatic headache: symptoms develops within 7 days, resolves or persists < 3 months
- Chronic: symptoms develops within 7 days, last > 3 months of head trauma
- Treatment: TCA antidepressants, venlafaxine/effexor, valproic acid, topiramate

Headache Attributed to Whiplash Injury

- History of whiplash (sudden and significant acceleration/deceleration movement of the neck)
- Acute: symptoms develops within 7 days, resolves or persists < 3 months
- Chronic: symptoms develops within 7 days, last > 3 months of the neck injury

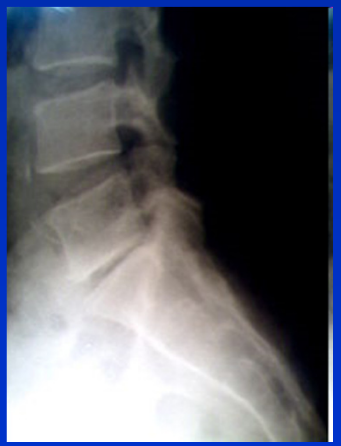
Cervicogenic Headache – IHS-2 diagnostic criteria

- A. Pain, referred from a source in the neck and perceived in one or more regions of the head and/or face, fulfilling C and D
- B. Clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck
- C. Evidence that the pain can be attributed to the neck disorder or lesion based on at least one of the following:
  - Clinical signs that implicate a source of pain in the neck
  - Abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo- or other adequate controls
- D. Pain resolves within 3 months after successful treatment of the causative disorder or lesion

Cervicogenic Headache – IHS-2 diagnostic criteria

- Clinical features (not unique): neck pain, focal neck tenderness, history of neck trauma, mechanical exacerbation of pain, unilateralist, coexisting shoulder pain, reduced range of motion in the neck , nuchal onset, nausea, vomiting, photophobia

- Causes of cervicogenic headache:
  - tumors, fractures, infections, rheumatoid arthritis of the upper cervical spine,
    - Not formally validated
    - But accepted as valid causes in individual cases
  - cervical spondylosis, osteochondritis:
    - not accepted as valid causes
  - myofascial tender spots:
    - should be coded as tension-type headache



http://en.wikipedia.org/wiki/Spondylosis

The Fact of Cervicogenic Headache

- A survey of 30,000 persons aged 30-44 years old,
  - Prevalence of CEH: 0.17%, with a female preponderance.
  - 50% had co-occurrence of medication overuse and 42% had co-occurrence of migraine.
- A survey of 1838 persons, aged 18-65 years old,
  - Prevalence of CEH 4.1 %, with a male preponderance.

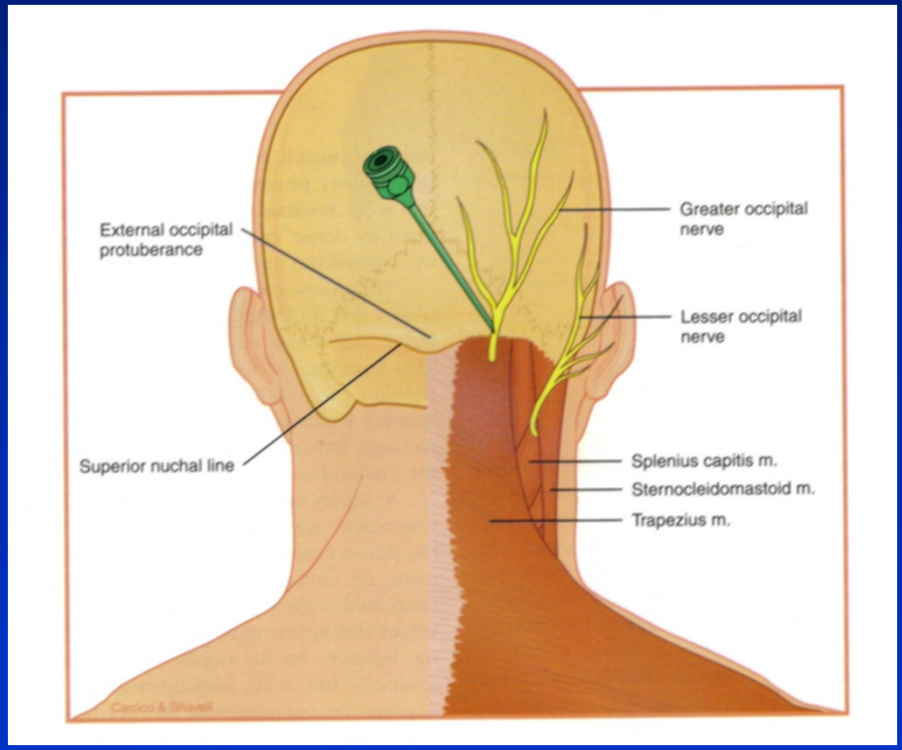
Knackstedt H et al. Cervicogenic headache in the general population: the Akershus study of chronic headache. Cephalalgia. 201;30:1468-76.
Sjaastad O & Bakketeig LS. Acta Neurol Scand. 2008;117:173-180

Cervicogenic Headache

- PE: Facet joint palpation and aggravation test
- Treatment:
  - greater occipital nerve (GON) blockage
  - neck exercise
  - C1/2, C2/3 facet joint injections, C2 (the greater occipital nerve) and C3 spinal rami (including the lesser and third occipital nerves) blockades.
    - 28/31 (90.3%) patients experienced >50% headache relief after treatment, with an average duration of 21.7 (1-90) days.
  - Botox not helpful in one DBRC study

Linde M et al. Cephalalgia. 201;31:797-807
Knackstedt H et al. Cephalalgia. 201;30:1468-76
Zhou L et al. Headache. 2010;50:657-63

Greater and Lesser Occipital Nerve Block



Waldman SD. Atlas of Pain Management Injection Techniques, 2000