European Headache School
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Differential diagnosis of secondary headaches

Cases

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Case 1

M. D. ♂ 51 years

- Medical history: hypertension
- Social history: former smoker, denies alcohol use
- Family history: father had hypertension and diabetes mellitus, deceased. No family history of headaches

PRESENTING COMPLIANT

- 8 months history of right-sided headache, localized at the occipital region and behind the ear
- The pain is mostly dull, middle grade intensity
- It occurs only by night, a few hours after falling asleep, and lasts several hours.
- 4 months later he noticed hearing disturbances on the right and also developed ipsilateral nasal congestion

PRESENTING COMPLIANT

- Ears/nose/throat specialist (ENT): Otitis chr. adhesiva lat. dex, Pansinusits
- In the next three months headaches gradually increased in frequency and intensity
- He takes analgesics every night

NEUROLGIST

- Hypnic headache
- Cervicogenic headache
- Headache attributed to disorder of ears/ rhinosinusitis
- Further investigation is needed!

LATER…

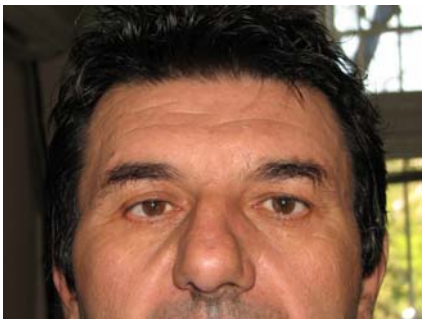
- Brain CT: normal

Two months later he developed double vision

Neurological examination:

- right-sided Horner's syndrome
- weakness of the right m. rectus lateralis
- conductive hearing loss on the right
- pyramidal, extrapyramidal and cerebellar signs normal, no sensory loss

M.D.



Differential diagnosis

- Headache attributed to disorder of ears/ rhinosinusitis
- Nasopharyngeal tumor
- Tolosa Hunt syndrome

DIAGNOSTIC TESTING

- Blood tests: normal, except ESR 40 mm/h
- Ophalmologist: right abducent nerve palsy
- Chest X-Ray: normal
- Radiography of cervical spine: spondylosis
- Carotid Duplex Ultrasound Examination and Transcranial Doppler: normal

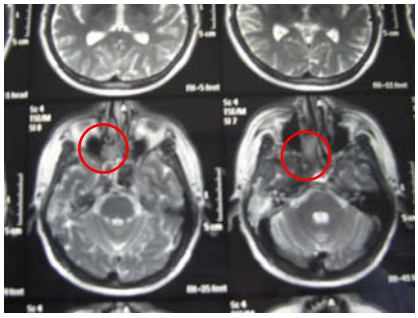
DIAGNOSTIC TESTING

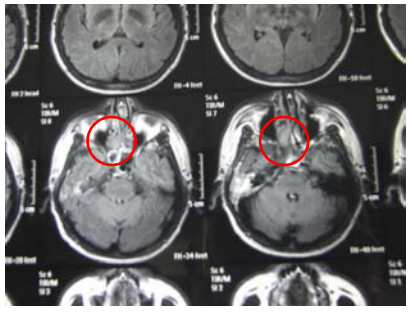
- Auditory evoked potentials: conductive hearing loss, changes not consistent with acoustic neurinoma
- Lyme antibody: negative
- HIV test: negative
- Lumbar puncture:
- CSF analysis: proteins 0,69 g/l, glucose 3,9 mmol/l, lymphocytes 2, erythrocyte 32
- Isoelecrtic focusing: normal

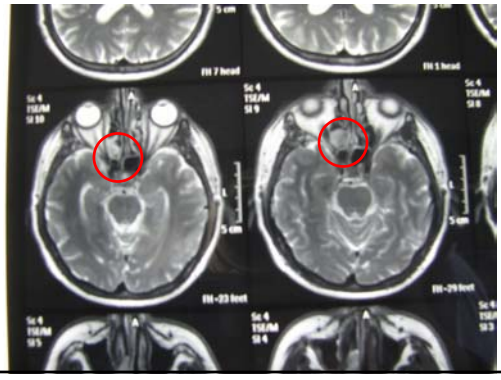
DIAGNOSTIC TESTING

- Brain MRI:

- Tumorous mass in the nasopharynx with palatum infiltration, infratemporal propagation, propagation to the sphenoid sinus, infiltration of the foramen magnum and epidural propagation at the craniocervical junction. Infiltration of the right sinus cavernosus.
- With gadolinium administration, tumorous mass significantly enhances
- Mastoiditis lat. dex, pansinusitis

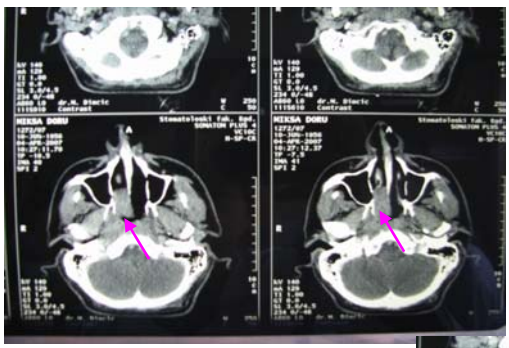




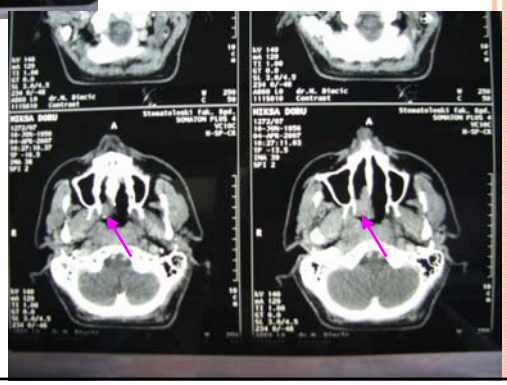


DIAGNOSTIC TESTING

- nasopharyngoscopy: tumor infiltrating posterior upper nasopharyngeal wall spreading to right lateral wall.
- Nasopharyngeal mass biopsy-pathohistology: nasopharyngeal carcinoma
- Radiotherapy was initiated which partially ameliorated most of his symptoms



Follow up CT



CASE 2

P.S., ♀ 39 years

Past history: menstrual migraine without aura

Presenting symptoms: headache and neck pain

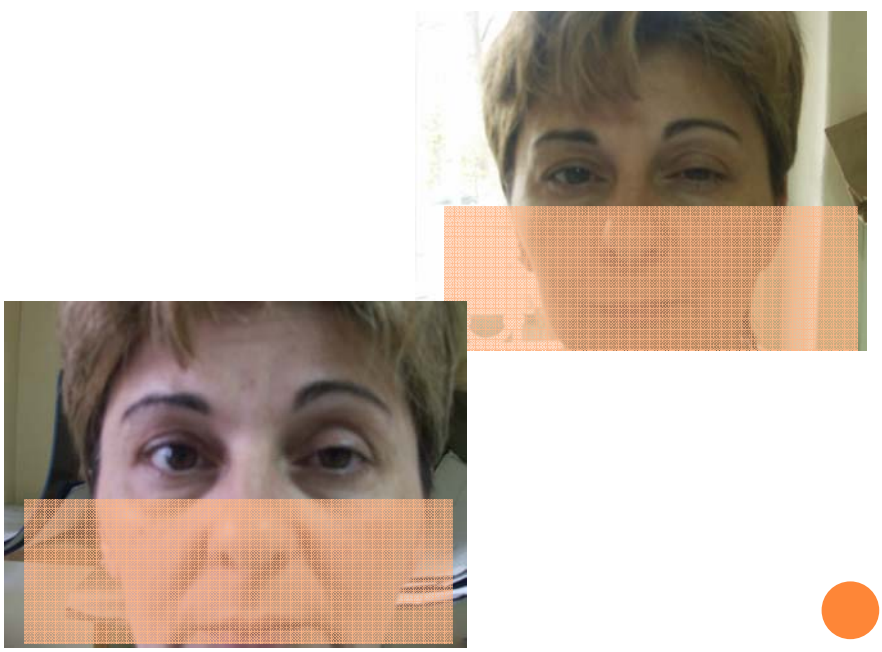
Patient presents to the emergency department 24 hours after onset of sudden headache and neck pain

CASE HISTORY

- Dull, constant headache of moderate intensity localized mostly behind left eye, with left temporal, ear and neck pain
- Worsening of pain by swallowing
- No associated photophobia, phonophobia, nausea, or vomiting

CASE HISTORY

- Physical examination: BP 150/100 mmHg
- Neurological examinaton:
  - left-sided mild ptosis and miosis (Horner's syndroma)
  - no other neurological abnormalities were found
- Brain CT: normal



Differential diagnosis

- Headache attributed to arterial hypertension
- Headache and neck pain attributed to cervical artery dissection
- Headache attributed to temporomandibular joint (TMJ) disorder
- Headache attributed to peritonsillar abscess

Diagnostic testing

- Carotid Duplex Ultrasound Examination:
  Normal diameters of CCA and ICA (left ICA smaller). No signs of atherosclerosis, normal velocity patterns in carotid and vertebral arteries. No signs of dissection!
- Transcranial Doppler: normal
- Blood tests: within a normal range

A DAY LATER…

- Persistent left-sided headache, moderate to severe, Horner's sign still present
- Mild fever
- Acute tonsillitis and otitis media lat.sin.
  ENT specialist: antibiotic treatment

THEE DAYS LATER…

- transient numbness and weakness in her right arm with speech disturbances, spontaneously recovered after 10 minutes
- transient weakness repeated two hours later and lasted 5 minutes
- without impairment of consciousness
- headache persists

Differential diagnosis

- Migraine aura
- TIA
- Epileptic seizure

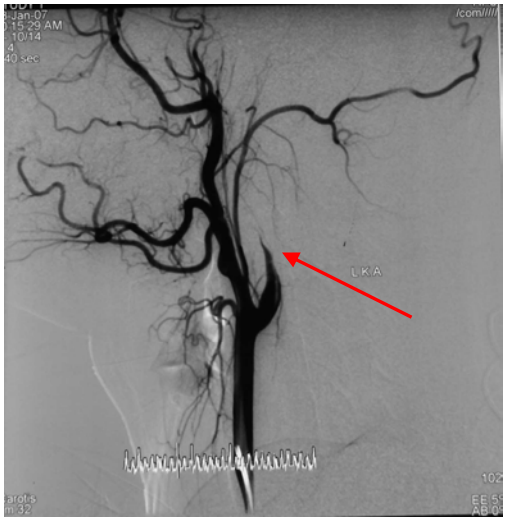
Diagnostic testing

- Electroencephalogram (EEG): normal
- ECG: normal
- Echocardiogrm: normal
- Chest X-Ray: normal
- Radiography of cervical spine: normal

TEN DAYS LATER…

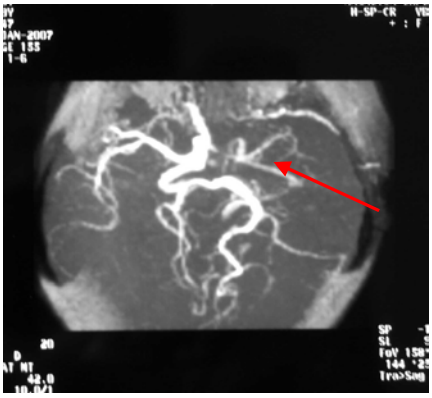
- Digital subtraction cerebral panangiogram: ***occlusion of the left ICA*** just above bifurcation.
- Repeated Carotid Duplex Ultrasound Examination confirms the absence of blood flow in left ICA, TCD shows signs of collateral circulation

DSA



SIXTEEN DAYS LATER…

- Brain MRI: normal
- MR cerebral angiography: ***extracranial subocclusion of left ICA!***



SEVERAL DAYS LATER…

- Carotid Duplex Ultrasound Examination:

- Recanalization of the left ICA with increased blood flow velocities. Medially of this flow is present hypoechogenic zone with smooth edges, 20 mm wide and several cm long.

- These findings suggested the re-establishment of blood flow after dissection!

DIAGNOSIS

Dissection of left ICA

- possible association of causal factors:
  - mild neck trauma and inflammation of the tonsil
- development of intramural thrombus
- TIA
- ICA "occlusion"
- early recanalization

CASE 3
V.B. 75 YEARS ♂

- Past medical history:
  - A year ago he suffered from recidivant herpes zoster ophtalmicus
    - Keratitis herpetica recidivans- o.d.
    - Glaucoma simplex chronicum- o.s.
  - Hypertension

PRESENT COMPLAINTS

- Presents with two-day history of constant severe pain in the right temporal area, mostly dull, sometimes throbbing
- No associated phenomena
- Painkillers were ineffective

FOLLOWING DAY….

Headache worsens

Fever, T= 37.7 °C, fatigue, mild nausea

short (10 s) left eye obscurations, repeated several times

- Emergency department
  - Neurological examination: normal

DIAGNOSTIC TESTING AT THE ED

- Brain CT
- Lumbar puncture
- Blood test: ESR

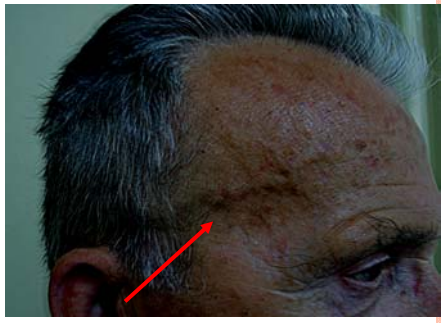
EMERGENCY DEPARTMENT

- … ESR= 82 mm/h
  CRP= 172.1 mg/l (<8)

DG: **Giant cell arteritis susp.**

PHYSICAL EXAMINATION

- Both superficial temporal arteries (STA) thick, tortuous, rigid, and painful on palpation with prominent pulsations



Clinical suspicion on giant cell arteritis
What next?

- Start steroids immediately
- Confirm diagnosis: STA color-duplex sonography
- Perform STA biopsy: wait for histopathology confirmation

.....WHAT DID WE DO?

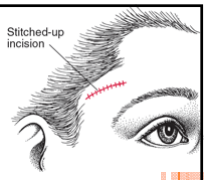
- Prednisone oral 80 mg daily initiated immediately with excellent response:
  - no headache in 1 week time
  - Blood tests: ESR= 20 mm/h
    CRP= 3.5 mg/l
    all in 2 weeks time

STA COLOR-DUPLEX SONOGRAPHY:

- Tortuous STA, with thickened, inflamed walls secondary to segmental dark halos on both sides in all STA branches, no occlusions

Findings consistent with suspected diagnosis of GCA

STA color-Duplex examination: Bilateral halo, diameter 0.6-0.9mm



BIOPSY AND HISTOPATHOLOGY

- STA biopsy (right STA, ramus frontalis, 2x 0.5 cm)
- Monocyte inflammatory infiltration of the arterial vessel walls with few giant cells

Confirms diagnosis of GCA!

FOLLOW UP IN 4 WEEKS

Follow-up STA color-Duplex examination: halo not present or < 5 mm

CASE 4

- V.B. 55 years ♂
- 4 months history of bitemporal headaches

- Past Medical History

  hypertension, chronic sinusitis, and a remote history of bleeding gastric ulcer
- Social History: He denies any tobacco, alcohol, or sedative drug use
- Family History: Multiple family members are known to have coronary artery disease and hypertension

Presenting symptoms

- Intermittent at first, aggravated with standing or sitting, resolved after lying down
- The headaches became progressively more severe and constant
- over-the-counter medications ineffective
- 2 months after headache onset he had occasional nausea, and a burning sensation in his scalp
- Headaches worsened upon coughing, sneezing
- Complained of memory difficulties

Headache red flags ?

- New-onset headache in patient older than 50 years of age
- Worsening of headache with coughing
- Worsening cognition and memory
- Bilateral character of headaches
- Accompanying nausea

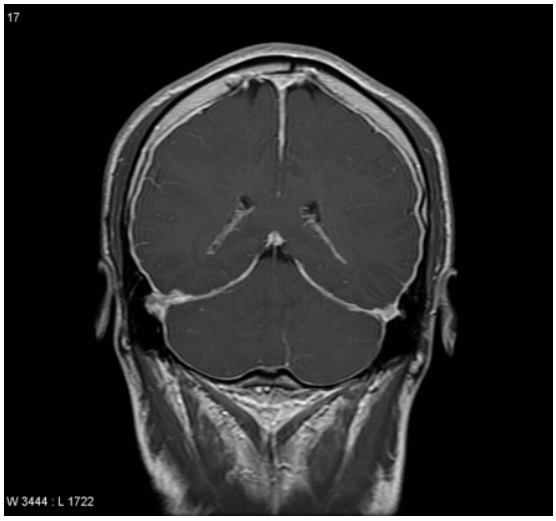
Physical examination

- afebrile, stable vital signs
- normal temporal artery pulsations bilaterally
- Normal neurological examination

- Brain MRI with gadolinium: diffuse pachymeningeal (dural) thickening with diffuse meningeal enhancement

Diagnostic testing

Brain MRI with contrast



Differential diagnosis

- Carcinomatous meningitis
- Giant cell arteritis
- Central nervous system Lyme disease
- Spontaneous intracranial hypotension
- Sarcoidosis

Diagnostic testing

- Blood tests: normal (ESR 7 mm/h)
- Chest X-Ray: normal
- HIV, hepatitis, and Lyme serologies: negative
- ANA, ANCA negative
- CSF analysis: 3 lymptocite, glucose 4,2 mmol/l, proteins 0.60 g/l
- Gram stain and bacterial culture, fungal and mycobacterial cultures were negative
- No oligoclonal bands were detected
- ACE within a normal range

Differential diagnosis

- Subdural empyema
- Spontaneous intracranial hypotension
- Neurosarcoidosis
- Cerebral venous sinus thrombosis

Diagnosis
Spontaneous intracranial hypotension

What is the most appropriate next step for this patient?

- Ventriculoperitoneal shunt to control intracranial pressure
- Cisternogram to identify area of CSF leak
- CT myelogram to identify area of CSF leak
- Strict bed rest and hydration to speed resolution of CSF leak
- Epidural blood patch to stop CSF leak

TREATMENT

- Our patient did not improve with bed rest and hydration
- We performed 2 lumbar EBPs, the second of which resulted in substantial improvement of symptoms
- Within 1-2 weeks he returned to work

BELGRADE, MAY, 2012

Neurology Clinic
Clinical Center of Serbia

UNIVERSITY OF BELGRADE
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