ACUTE FIRST EVER HEADACHE: THE VASCULAR CAUSES

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DISCLOSURE : NONE
Secondary headaches?

- A new headache occurs for the first time in close temporal relation to another disorder the new headache is coded as a secondary headache (ICHD-3 beta).
Part II: The secondary headaches

5. Headache attributed to trauma or injury to the head and/or neck
6. Headache attributed to cranial or cervical vascular disorder
7. Headache attributed to non-vascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache attributed to infection
10. Headache attributed to disorder of homoeostasis
11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
12. Headache attributed to psychiatric disorder
When to WORRY?

RED FLAGS:

- Systemic features
- Neurological deficits
- Over 50 years old
- Thunderclap Onset

4Ps:
- Precipitation with Valsalva
- Positional
- Pattern change
- Pregnancy
Thunderclap Onset

- Severe head pain
- Abrupt onset
- Reaching maximum intensity in <1 minute
- Lasting for 5 minutes or more
- ICHD-3 Beta
### Thunderclap Onset

#### Incidence:

43 per 100,000 adults per year

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<th>NON-VASCULAR</th>
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How long did it take for “the worst headache of your life” to reach maximal intensity?

Duration?

Single or recurrent?

The location and type of pain is nonspecific.
Subarachnoid Hemorrhage

- SAH is the most common cause of secondary TCH.

- 11 %–25 % of patients with TCH.

- Serious condition (mortality is 40–50%)
Clinical predictor of non-traumatic SAH

Combination of:

- Age >40,
- Neck pain or stiffness,
- Loss of consciousness or
- Headache onset during exertion.

Sensitivity of 97 % - Specificity 23%
Pathophysiology of SAH

Combination of:
- Extravasation of blood from ruptured aneurysm
- Rapid rise in ICP “Brain tamponade” due to:
  - Rise in intracranial volume
  - Vasoparalysis
  - CSF obstruction due to blood clot
- Sharp and sudden reduction in CPP responsible for syncope (CPP = MAP-ICP)
- Marked acute cerebral vasoconstriction
SAH Diagnosis

Non-contrast-enhanced CT scan:

- Sensitivity is close to 99% in the first six hours after onset.
- Dropping to 50% at seven days.
SAH Diagnosis

**Lumbar Puncture:**
- Collected between 12 hours and two weeks after the onset.
- Analysed spectro-photometrically.
- Xanthochromia.
Misdiagnosis

Initial misdiagnosis:
- One quarter to one half of patients.
- The most common specific misdiagnosis being **migraine**.

Due to failure of:
- Appropriate neuroimaging.
- Perform lumbar puncture.
- Interpretation.
Sentinel Headache

- Typically present with a TCH without meningismus or altered level of consciousness.
- Due to sudden enlargement of the arterial malformation
Sentinel Headache

Incidence:

- 10%–43% in patients with aneurysmal SAH
- 50% of patients with aneurysmal SAH reported SH within the four weeks prior to diagnosis.
- Re-bleeding prior to aneurysm repair was ten times in those patients with SH.
Management of SAH:

- Securing the aneurysm (coiling vs clipping) as early as we can.
- Management of complications:
  - Vasospasm
  - Hydrocephalus
  - Electrolytes disturbance
  - Cardiovascular complications
  - Pain (TCH)
Cervical Artery Dissection

Headache Incidence:

60-95% CAD (frontal or temporal regions)

70% VAD (Occipital, parieto-occipital)

Predisposing factors:

- Minor Cervical Trauma
- Recent infection
- Hyperhomocysteinaemia
- Migraine
- Low concentrations of $\alpha_1$ antitrypsin
- Oral contraceptives
- Seasonal variations (highest rates during autumn or winter)
Predisposing factors

Genetic risk factors

- CAD may occur in patients with monogenic disorders:
  - Ehlers-Danlos syndrome
  - Marfan’s syndrome
  - Osteogenesis imperfecta
Pathophysiology

The intramural haematoma can expand towards the intima or the adventitia, resulting in:

- Luminal stenosis or occlusion
- Aneurysmal dilation of the artery
Pathophysiology

Cerebral or retinal ischemia may result from:

- Emboli from intramural thrombi at the site of the intimal tear.
- Hemodynamic infarcts related to hypoperfusion and diameter reduction of affected artery.
Clinical presentation:

Time course of symptoms and signs:

- Patients with CAD typically present with local symptoms and signs and subsequently develop an ischaemic event.

- The delay between onset of local symptoms and ischaemic manifestations can vary from a few minutes to several weeks, and is usually less than a month.
Clinical presentation

Local symptoms and signs:
- Horner syndrome
- Neck pain
- Headache (severe intensity and throbbing)
- Cranial nerve palsy (Hypoglossal, Glossopharyngeal and Vagus)
- Tinnitus
- Cervical-root injury (vertebral-artery dissections)

Patients who have only local signs or symptoms, without cerebral or retinal ischemia are about 33% of all cases.
Radiological diagnosis Cervical dissection
Narrowed lumen from dissection

Hematoma in carotid wall

Normal internal carotid artery
Duplex images
Treatment

- Thrombolysis
- Antithrombotic therapy
- Duration of antithrombotic therapy
- Endovascular therapy
Treatment

Safety and Functional Outcome of Thrombolysis in Dissection-Related Ischemic Stroke
A Meta-Analysis of Individual Patient Data

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Background and Purpose—The safety and efficacy of thrombolysis in cervical artery dissection (CAD) are controversial. The aim of this meta-analysis was to pool all individual patient data and provide a valid estimate of safety and outcome of thrombolysis in CAD.

Methods—We performed a systematic literature search on intravenous and intra-arterial thrombolysis in CAD. We calculated the rates of pooled symptomatic intracranial hemorrhage and mortality and indirectly compared them with matched controls from the Safe Implementation of Thrombolysis in Stroke–International Stroke Thrombolysis Register. We applied multivariate regression models to identify predictors of excellent (modified Rankin Scale=0 to 1) and favorable (modified Rankin Scale=0 to 2) outcome.

Results—We obtained individual patient data of 180 patients from 14 retrospective series and 22 case reports. Patients were predominantly female (68%), with a mean±SD age of 46±11 years. Most patients presented with severe stroke (median National Institutes of Health Stroke Scale score=16). Treatment was intravenous thrombolysis in 67% and intra-arterial thrombolysis in 33%. Median follow-up was 3 months. The pooled symptomatic intracranial hemorrhage rate was 3.1% (95% CI, 1.3 to 7.2). Overall mortality was 8.1% (95% CI, 4.9 to 13.2), and 41.0% (95% CI, 31.4 to 51.4) had an excellent outcome. Stroke severity was a strong predictor of outcome. Overlapping confidence intervals of end points indicated no relevant differences with matched controls from the Safe Implementation of Thrombolysis in Stroke–International Stroke Thrombolysis Register.

Conclusions—Safety and outcome of thrombolysis in patients with CAD-related stroke appear similar to those for stroke from all causes. Based on our findings, thrombolysis should not be withheld in patients with CAD. (Stroke. 2011;42:2515-2520.)
Treatment

Antiplatelets Versus Anticoagulation in Cervical Artery Dissection

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Background and Purpose—The widespread preference of anticoagulants over antiplatelets in patients with cervical artery dissection (CAD) is empirical rather than evidence-based.

Summary of Review—This article summarizes pathophysiological considerations, clinical experiences, and the findings of a systematic metaanalysis about antithrombotic agents in CAD patients. As a result, there are several putative arguments in favor as well as against immediate anticoagulation in CAD patients.

Conclusions—A randomized controlled trial comparing antiplatelets with anticoagulation is needed and ethically justified. However, attributable to the large sample size which is required to gather meaningful results, such a trial represents a huge venture. This comprehensive overview may be helpful for the design and the promotion of such a trial. In addition, it could be used to encourage both participation of centers and randomization of CAD patients. Alternatively, antithrombotic treatment decisions can be customized based on clinical and paraclinical characteristics of individual CAD patients. Stroke severity with National Institutes of Health Stroke Scale score ≥15, accompanying intracranial dissection, local compression syndromes without ischemic events, or concomitant diseases with increased bleeding risk are features in which antiplatelets seem preferable. In turn, in CAD patients with (pseudo)occlusion of the dissected artery, high intensity transient signals in transcranial ultrasound studies despite (dual) antiplatelets, multiple ischemic events in the same circulation, or with free-floating thrombus immediate anticoagulation is favored. (Stroke. 2007;38: 2605-2611.)
Reversible Cerebral Vasoconstriction Syndrome (RCVS)

◆ 75% present with headache as the only symptom.

◆ Typically:

➢ TCH
➢ Recurring over one to two weeks
➢ Often triggered by sexual activity, exertion, Valsalva maneuvers and/or emotion.
Criteria for diagnosis Reversible Cerebral Vasoconstriction Syndrome

- Intense, acute headache, with or without additional neurological signs or symptoms
- Catheter, CT, or MRI angiography demonstrating segmental cerebral artery vasoconstriction
- No evidence for aneurysmal SAH
- Reversibility of angiographic abnormalities within 12 weeks of onset; if death occurs before the follow-up studies are complete, the autopsy has to rule out vasculitis, intracranial atherosclerosis and aneurysmal SAH, which can also manifest with headache and stroke
- CSF analysis normal or close to normal (protein level < 80 mg/dL, leukocytes < 10/mm3, normal glucose level)

Shih Pin et al, Therapeutic advances in Neurological disorders 2010
Reversible Cerebral Vasoconstriction Syndrome

More common

Sex: women (3:1)

Age: 20 - 50 years.

Hypertension → 50%

21% > 180/110
**Reversible Cerebral Vasoconstriction Syndrome**

**Transcranial Doppler (TCD):**

- Monitor the resolution of the vasoconstriction

- Assess for the risks of ischemic stroke and PRES.
Reversible Cerebral Vasoconstriction Syndrome

Transcranial Doppler (TCD):
Mean flow velocity (VMCA) and Lindegaard index (LI) of the middle cerebral artery:

- In RCVS, VMCA and LI are elevated.
- VMCA >120 cm/s and LI >3 indicate risk of PRES and ischemic stroke.
- Headache.
- Risk of PRES and ischemic stroke.
Reversible Cerebral Vasoconstriction Syndrome

- Self-limiting
- 1-3 months
- Disappearance of the arterial abnormalities (hence ‘reversible’) and, almost always, resolution of the headache.
Cerebral Venous Sinus Thrombosis

- 1.3 per 100,000 person
- 90% presented with headache only.
- 2-14% presented with TCH.

Pain:
- No specific characteristics:
- Mostly diffuse, progressive and severe.
- Can be: unilateral and sudden, mild, and sometimes is migraine-like.
Cerebral Venous Sinus Thrombosis

**Diagnosis:**
- MRI with MR venogram, CT venogram, and/or digital subtraction angiography (DSA).
- Visualization of venous thrombosis:
  - Attenuated vein sign (100% sensitive) or Cord sign (65% sensitive).
Dense clot sign (3)
On the left images of a patient with a hemorrhagic infarction in the temporal lobe (red arrow). Notice the dense transverse sinus due to thrombosis (blue arrows).

Empty delta sign
The empty delta sign is a finding that is seen on a contrast enhanced CT (CECT) and was first described in thrombosis of the superior sagittal sinus. The sign consists of a triangular area of enhancement with a relatively low-attenuating center, which is the thrombosed sinus. The likely explanation is enhancement of the rich dural venous collateral circulation surrounding the thrombosed sinus, producing the central region of low attenuation. In early thrombosis the empty delta sign may be absent and you will have to rely on non-visualization of the thrombosed vein on the CECT. The sign may be absent after two months due to recanalization within the thrombus.
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<th>Dense clot sign</th>
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<td>Cord sign</td>
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<td>Empty delta</td>
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<td>Loss of normal flow void on MR</td>
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<td>Venous infarction</td>
<td>Bilateral - <em>parasagittal</em></td>
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<td><em>bithalamic</em></td>
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<td>Temporal lobe infarction</td>
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<td>Cortical edema or hemorrhage</td>
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<td>Peripheral lobar hemorrhage</td>
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<td>Clinically</td>
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<td>Headache</td>
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<td>Loss of consciousness</td>
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Cerebral Venous Sinus Thrombosis

**Treatment:**

- Should be started as early as possible.
- Heparin followed by at least six months of oral anticoagulation.
- Symptomatic treatment.
- Treatment of the underlying cause.
Pituitary Apoplexy

Neurosurgical emergency.

- TCH    87-97%
- Meningismus
- Neuro-ophthalmological symptoms 83 % such as visual disturbance and ophthalmoparesis.
- Nausea/vomiting    78 %
- Altered level of consciousness 13 %–50 %
Pituitary Apoplexy

**Diagnosis:**

The imaging of choice: **MRI** identifies the pituitary hemorrhage in 88% of cases compared with 21% with CT head.
**Pituitary Apoplexy**

Diagnosis of Apoplexy Confirmed

Maintain hemodynamic stability with IV fluids and glucocorticoids

Vision:
- Reduced acuity
- Reduced visual fields
- Altered state of consciousness

If patient becomes unstable

If NO
- Admit to hospital/ICU for close observation
- Consult experienced neurosurgeon
- Hourly neurological assessments until stable
- Assessment of endocrine status

If YES
- Admit to ICU
- Consult experienced neurosurgeon
- Consider surgical decompression within 7 days of presentation
Does Ischemic Stroke present with headache?
Let’s judge....
Case presentation (1)

- 50 years old female physician, unremarkable past history.

- She got severe TC headache, sudden convulsions and DCL GCS 12/15

- She went to large tertiary care center and patient got MRI immediately without prior CT Brain.

- MRI showed bilateral frontal infarct so aspirin was started at dose 300 mg daily…….
48 hours, she got deteriorated GCS9/15 so CT brain was done, and surprisingly showed SAH

- Video 1
Case interpretation!
Case presentation (2)

✧ 34-years old male working as a driver...
✧ Chronic heavy smoker about 2 packet / day for 20 years ……chronic cannabis addict for about 2 cigarettes / day for 10 years.

✧ Presented to ER after 1 hour with acute onset of occipital headache, left ataxic hemiparesis, left hemihypothesia including face, dysarthria and pseudobulbar.
Our patient

- Young male without risk factors apart for cannabis & smoking.

- With duplex suspected thrombotic occlusion vs. dissection of vertebral artery.

- So, dissection is highly suspected.
Case presentation (3)

- 21 yrs old female on the 3rd day postpartum. She got sever bitemporal headache
- Left hand numbness then left upper limb weakness and numbness.
- Patient had a past history of pre-eclampsia in her last pregnancy, BP 160/110
Criteria for diagnosis Reversible Cerebral Vasoconstriction Syndrome

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- Catheter, CT, or MRI angiography demonstrating segmental cerebral artery vasoconstriction
- No evidence for aneurysmal SAH
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Reversible Cerebral Vasoconstriction Syndrome

More common

Sex: women (3:1)

Age: 20 - 50 years.

Hypertension → 50%

21% > 180/110
Conclusion

TCH

- Abnormal CT Brain
- Normal CT brain + Abnormal LP
- Normal CT brain + Normal LP
Abnormal CT Brain

- Subarachnoid hemorrhage (SAH)
- Intracerebral hemorrhage (ICH)
- Acute subdural hemorrhage (SDH)
- Hydrocephalus
  - Posterior fossa pathology
  - Aqueductal stenosis
  - Third ventricle colloid cyst
Normal CT brain + Abnormal LP

- SAH

- Meningitis (2% of thunderclap headache)
Normal CT brain + Normal LP

- Reversible Cerebral Vasospasm (RCVS)
- Arterial dissection
- Cerebral Venous Sinus Thrombosis (CVST)
- Pituitary Apoplexy
- Low CSF pressure
Thank You
Thank you