Thunderclap Headache

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45YO female with recurrent severe headache

- Abrupt pulsatile headache
- Bifrontal → entire crania
- Very severe headache
- x 1-2/day, for a week
- Duration: about 3 h
- NSAIDs: not effective → getting worse.

Cephalalgia 2008

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45YO female with recurrent severe headache

- nausea/vomiting/photophobia/phonophobia: deny
- Conjunctival injection/nasal obstruction/lacrimation: deny
- Sweating/palpitation/anxiety/pallor: deny
- Trauma/operation/smoking/alcohol drinking: deny

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Evaluation

- BP 114/79, PR 79/min
- N/Ex: normal
- Brain CT, MRI: normal
- CSF tapping: normal
- Conventional angiography: normal
- Diagnosis ?

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International Classification of Headache Disorder 3rd edition (beta version)

Part one: the primary headaches

1. Migraine
2. Tension-type headache
3. Trigeminal autonomic cephalalgias
4. Other primary headache disorders

Part two: 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

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International Classification of Headache Disorder 3rd edition (beta version) continued

Part two: the secondary headaches

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

Part three: painful cranial neuropathies, other facial pains and other headache

13. Painful cranial neuropathies and other facial pains
14. Other headache disorders

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ICHD-3 beta: Primary headache disorders presenting as thunderclap headache

- High-intensity headahce of abrupt onset, mimicking that of ruptured cerebral aneurysm, in the absence of any intracranial pathology

4.1 Primary cough headache
4.2 Primary exercise headache
4.3 Primary headache associated with sexual activity
4.4 Primary thunderclap headache

A Severe head pain fulfilling criteria B and C
B Abrupt onset, reaching maximu intensity in $<$ 1 minute.
C Lasting for $\geq$ 5 minutes
D Not better accounted for by another ICHD-3 diagnosis.

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Diagnosis

- Intermittent SBP elevation to 190mm Hg associated with headache $\rightarrow$ improvement of headache with SBP reduction
- Vanillymandelic acid and metanephrine in 24 h urine: elevated
- Abdominal CT: retroperitoneal solid mass $\rightarrow$ Pheochromocytoma

10.3.1 Headache attributed to phaeochromocytoma

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Diagnosis

- Intermittent SBP elevation to 190mm Hg associated with headache → improvement of headache with SBP reduction
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10.3.1 Headache attributed to phaeochromocytoma

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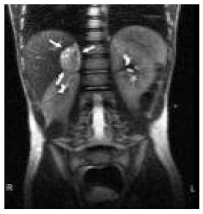
Pheochromocytoma

- Pheochromocytoma should be considered in patients with headache and
  - Hypertension, paroxysmal or persistent
  - Autonomic disturbances
  - Panic attacks
  - Adrenal incidentaloma
  - Predisposing familial disorders (MEN II, neurofibromatosis type 1, ..)
- 2-8/1 milion person/yr
- 10% rule: extraadrenal, malignant, bilateral



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36YO female with recurrent severe headache

- Cesarean delivery of twins 19 days ago
- Headache developed 9 days ago
  - Intermittent, throbbing, bifrontal area
  - VAS 8/10, BP 150/72
  - Improved spontaneously and recurred in the evening
  - BP 190/80
  - Brain CT/MRI and laboratory test: normal
  - Analgesics and BP 168/70 → discharge

N Engl J Med 2009;360:1126-37

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36YO female with recurrent severe headache

- Headache recurred 4 days ago
- Severe headache with photophobia and nausea 2 days ago
  - VAS 10/10, BP 204/96
  - HCT 34.7%, protein 30mg/dl on UA
  - GTCS in the ER
  - Brain CT: normal
  - CSF: WBC 0, RBC 0, protein 64mg/dl, glucose 54mg/dl
  - Afterwards, headache persisted (VAS 4) and SBP ranged from 140 to 160.
- Transient right hemiparesis, aphasia, and confusion for 5 min on the day of admission
  - 136/91, PR 94/min, RR 24/min, BT 36.5

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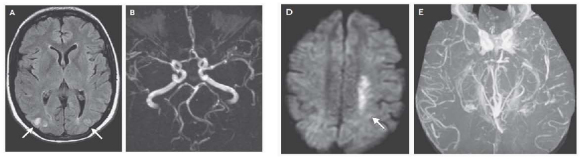
36YO female with recurrent severe headache

- HD 1
  - Rt. Side ataxic hemiparesis, intermittent
  - EEG: normal
- HD 2
  - Weakness in right arm and both legs. Level of consciousness decreased.
  - Intermittent slowing on EEG monitoring



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DDx of Postpartum headache

Primary headahce
- Migraine
- TTHA
- Primary thunderclap HA

Secondary headahce
- Postdural puncture HA
- SAH/ICH
- Dissection
- Cerebral venous sinus thrombosis
- Eclampsia
- RCVS
- PRES

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RCVS

- Young adults (20 to 60)
- Severe headache with/without seizure and focal neurologic deficits
- 60%: postpartum or vasoactive drugs
- Brain MRI: normal(70%)
- Major Cx: cortical SAH(22%), infarction or ICH (7%)
- Diagnosis: string of beads on angiography → resolved within 12 weeks.
- Tx.: Nimodipine may be effective in relieving headache but not in reducing complication of infarction or ICH.

Pract Neurol 2009

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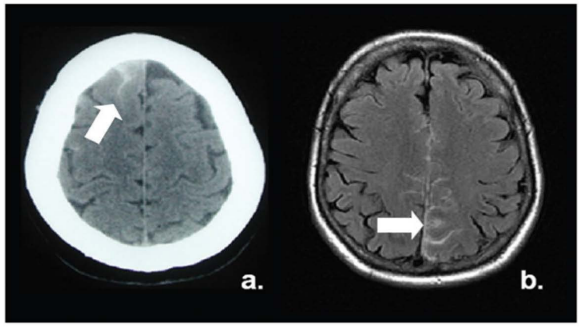
RCVS



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RCVS and related conditions

Idiopathic

No identifiable precipitating factor

Headache disorders (migraine, primary thunderclap headache, benign exertional headache, benign sexual headache, and primary cough headache)

Pregnancy and puerperium

Early puerperium, late pregnancy, preeclampsia, eclampsia, delayed postpartum eclampsia

Drugs and blood products

Phenylpropanolamine, pseudoephedrine, ergotamine tartrate, methylergonovine, bromocriptine, lisuride, selective serotonin-reuptake inhibitors, sumatriptan, isometheptene, cocaine, ecstasy, amphetamine derivatives, marijuana, lysergic acid diethylamide, tacrolimus, cyclophosphamide, erythropoietin, intravenous immune globulin, and red-cell transfusions

Miscellaneous

Hypercalcemia, porphyria, pheochromocytoma, bronchial carcinoid tumor, unruptured saccular cerebral aneurysm, head trauma, spinal subdural hematoma, post–carotid endarterectomy, postdural puncture, open neurosurgical procedures

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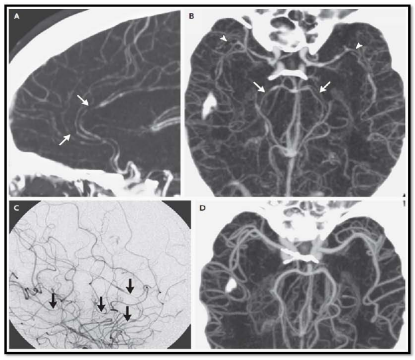
CTA and DSA



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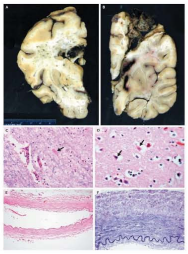
Autopsy



- Gray-White matter junctions are indistinct
- A sectionof the Rt. PCA and MCA shows normal appearance.

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ICHD-3 beta: 6.7.3 Headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)

A Any new headache fulfilling criterion C

B Reversible cerebral vasoconstriction syndrome (RCVS) has been diagnosed

C Evidence of causation demonstrated by at least one of the following:

1. headache, with or without focal deficits and/or seizures, has led to angiography (with string and beads appearance) and diagnosis of RCVS
2. headache has either or both of the following characteristics:
   a) recurrent during ≤ 1 month, and with thunderclap onset
   b) triggered by sexual activity, exertion, Valsalva manoeuvres, emotion, bathing and/or showering
3. no new significant headache occurs > 1 month after onset

D Not better accounted for by another ICHD-3 diagnosis, and aneurysmal subarachnoid haemorrhage has been excluded by appropriate investigations.

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Causes of thunderclap headache

- Subarachnoid hemorrhage
- Sentinel headache
- Cerebral venous sinus thrombosis
- Cervical artery dissection
- Spontaneous intracranial hypotension
- Pituitary apoplexy
- Retroclival hematoma
- Ischemic stroke
- Acute hypertensive crisis
- Reversible cerebral vasoconstriction syndrome
- Third ventricle colloid cyst
- Intracranial infection
- Primary headaches

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RCVS after blood transfusion

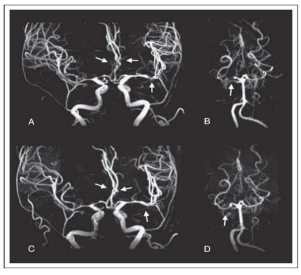

Fig 2.—(A and B) Three-dimensional (3D) time-of-flight (TOF) magnetic resonance angiography (MRA) showed multiple segmental narrowings over bilateral anterior cerebral arteries, left middle cerebral artery, and right posterior cerebral artery (arrows). (C and D) 3D TOF MRA obtained after 7 days showed significant improvement of vasoconstriction (arrows).

Headache 2014

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Thunderclap HA with orgasm: BA dissection

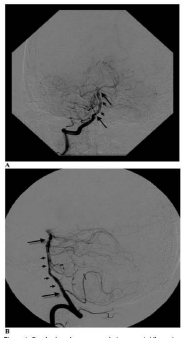

Figure 1. Cerebral angiogram revealed segmental (boundaries denoted by large arrows) irregularities (small arrows) of the basilar artery in both anterior-posterior (A) and lateral (B) views.

J of Emergency Medicine 2012

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Thunderclap HA: Cerebral Venous Sinus Thrombosis

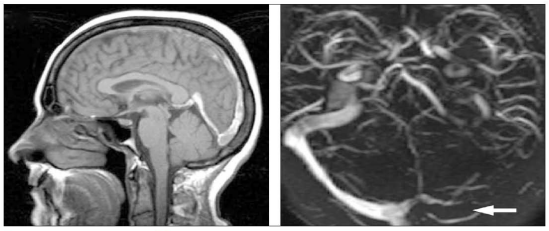

Figure 1: **Cerebral venous sinus thrombosis**
Left: T1 weighted sagittal MRI reveals hyperintense signal within the superior sagittal sinus and straight sinus. Right: Magnetic-resonance venogram reveals loss of flow signal secondary to thrombus in the central portion of left transverse sinus (arrow).

Lancet Neurol 2006

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Assessment of thunderclap headache

- Non-contrast CT: SAH
  - Sensitivity nearly 100% within 12 hrs of headache development
  - 85% on day 2, 58% after 5 days. → CSF test may be needed.
- MRI, MRA, MRV
- Conventional angiography is not usually necessary.

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Assessment of thunderclap headache

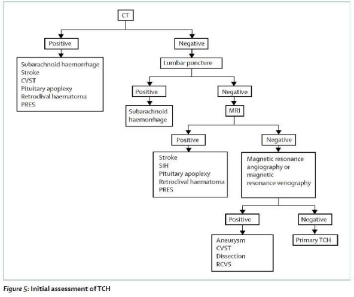

Figure 5: Initial assessment of TCH

Lancet Neurol 2006

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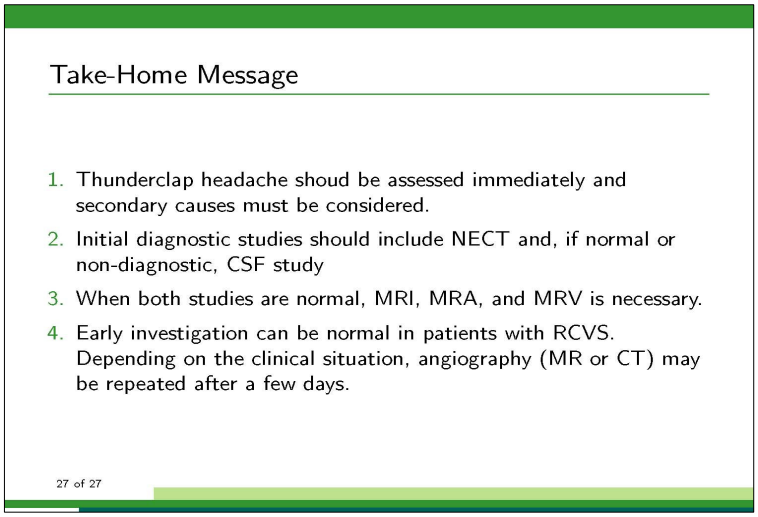
Take-Home Message
1. Thunderclap headache shoud be assessed immediately and secondary causes must be considered.
2. Initial diagnostic studies should include NECT and, if normal or non-diagnostic, CSF study
3. When both studies are normal, MRI, MRA, and MRV is necessary.
4. Early investigation can be normal in patients with RCVS. Depending on the clinical situation, angiography (MR or CT) may be repeated after a few days.
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