Thunderclap headache

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definition:

- A headache that is very severe and has abrupt onset, reaching maximum intensity in less than 1 minute.
- not defined solely by its high-intensity pain, but also by the rapidity with which it reaches maximum intensity.
- Its explosive and unexpected nature.

Epidemiology:

- incidence ~43 per 100,000 adults per year.

Differential Diagnosis:

- Most Common Causes of Thunderclap Headache:

-Reversible cerebral vasoconstriction syndrome
-Subarachnoid hemorrhage

- Less Common Causes of Thunderclap Headache:

-Cerebral infection
-Cerebral venous sinus thrombosis
-Cervical artery dissection
-Complicated sinusitis
-Hypertensive crisis
-Intracerebral hemorrhage
-Ischemic stroke
-Spontaneous intracranial hypotension
-Subdural hematoma

▷Uncommon Causes of Thunderclap Headache:

-Aqueductal stenosis
-Brain tumor
-Giant cell arteritis
-Pituitary apoplexy
-Pheochromocytoma
-Retroclival hematoma
-Third ventricle colloid cyst

▷ Possible Causes of Thunderclap Headache:

-Primary or idiopathic thunderclap headache
-Unruptured intracranial aneurysm

CLINICAL PRESENTATION

- differentiated from other severe headaches, such as migraine or cluster, by the rapidity with which they reach their maximum intensity.
- cannot be differentiated from other headache types based solely upon the intensity of the headache.

Key points in PHx

- altered level of consciousness
- visual symptoms
- papilledema
- meningismus
- Fever
- tinnitus, auditory muffling
- Horner syndrome
- Hypertension
- orthostatic worsening of the headache
- seizures
- focal neurologic deficits such as focal weakness and sensory disturbance.

- altered level of consciousness, seizures, or focal neurologic symptoms and signs :
  - SAH
  - other intracranial hemorrhages
  - hypertensive crisis
  - cervical artery dissection
  - ischemic stroke
  - RCVS
  - PRES
  - cerebral venous sinus thrombosis

- Recurrent thunderclap headaches over several days to weeks:

-RCVS

-SAH (sentinel headache)

- thunderclap headache followed by orthostatic headaches and auditory muffling :

-spontaneous intracranial hypotension

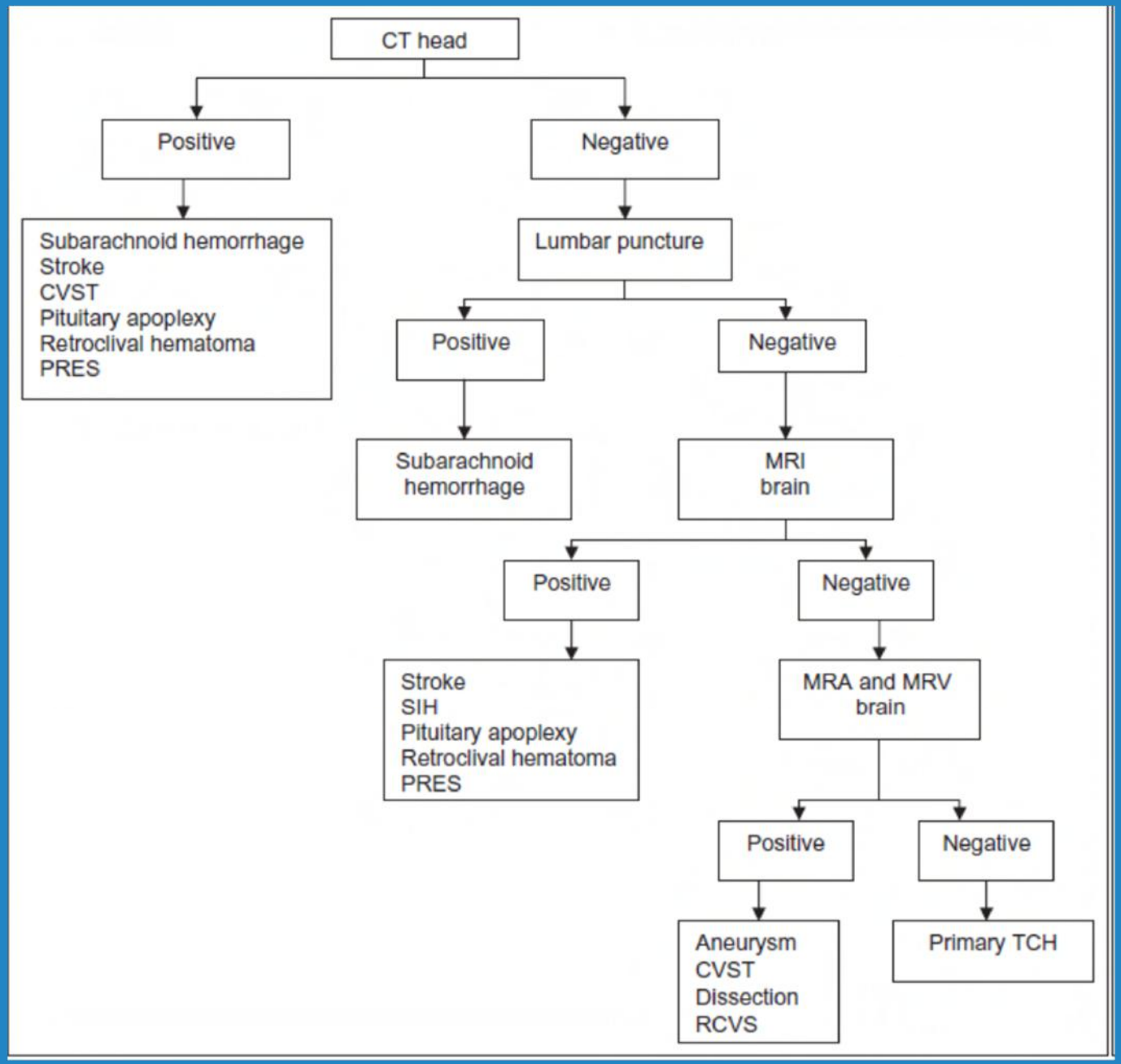
General Evaluation of the Patient With Thunderclap Headache

- Brain CT: sensitivity for detecting an aneurysmal SAH when the CT is performed within 6 hours of symptom onset is between 92% and 100%.
- 85% to 95% on day 2
- 75% on day 3
- 50% after 5 days

Lumbar puncture

- LP should be performed as soon as possible after the brain CT.
- the sensitivity of CSF analyses for detection of an SAH is higher if the LP is performed at least 6 hours, preferably 12 hours, after SAH onset.
- However, LP should not be delayed, because of the risk of a second aneurysmal rupture within 24 hours.

- nondiagnostic brain CT and LP →contrast-enhanced brain MRI and noninvasive vascular imaging of the head and neck (eg, MRA, CTA).
- Venous sinus imaging via MRV or CTV →clinical suspicion for an underlying cerebral venous sinus.


CT head
Positive
Negative
Subarachnoid hemorrhage
Stroke
CVST
Pituitary apoplexy
Retroclival hematoma
PRES
Lumbar puncture
Positive
Negative
Subarachnoid hemorrhage
MRI brain
Positive
Negative
Stroke
SIH
Pituitary apoplexy
Retroclival hematoma
PRES
MRA and MRV brain
Positive
Negative
Aneurysm
CVST
Dissection
RCVS
Primary TCH

Subarachnoid Hemorrhage

- most common cause of secondary TCH.
- 11 –25 % of patients with TCH have a SAH as the etiology .
- Approximately 70 % of patients with SAH present with a headache of whom 50 % of have a TCH.
- The location of pain varies; however, the most common site is the occipital region with neck pain.

- No specific clinical features can differentiate TCH related to SAH from other etiologies, however, the presence of any combination of age >40, neck pain or stiffness, loss of consciousness or headache onset during exertion may be a useful clinical predictor of non-traumatic SAH .
- In the study by Mark et al these clinical features had a sensitivity of 97 % and specificity of approximately 23 % for the diagnosis of SAH.

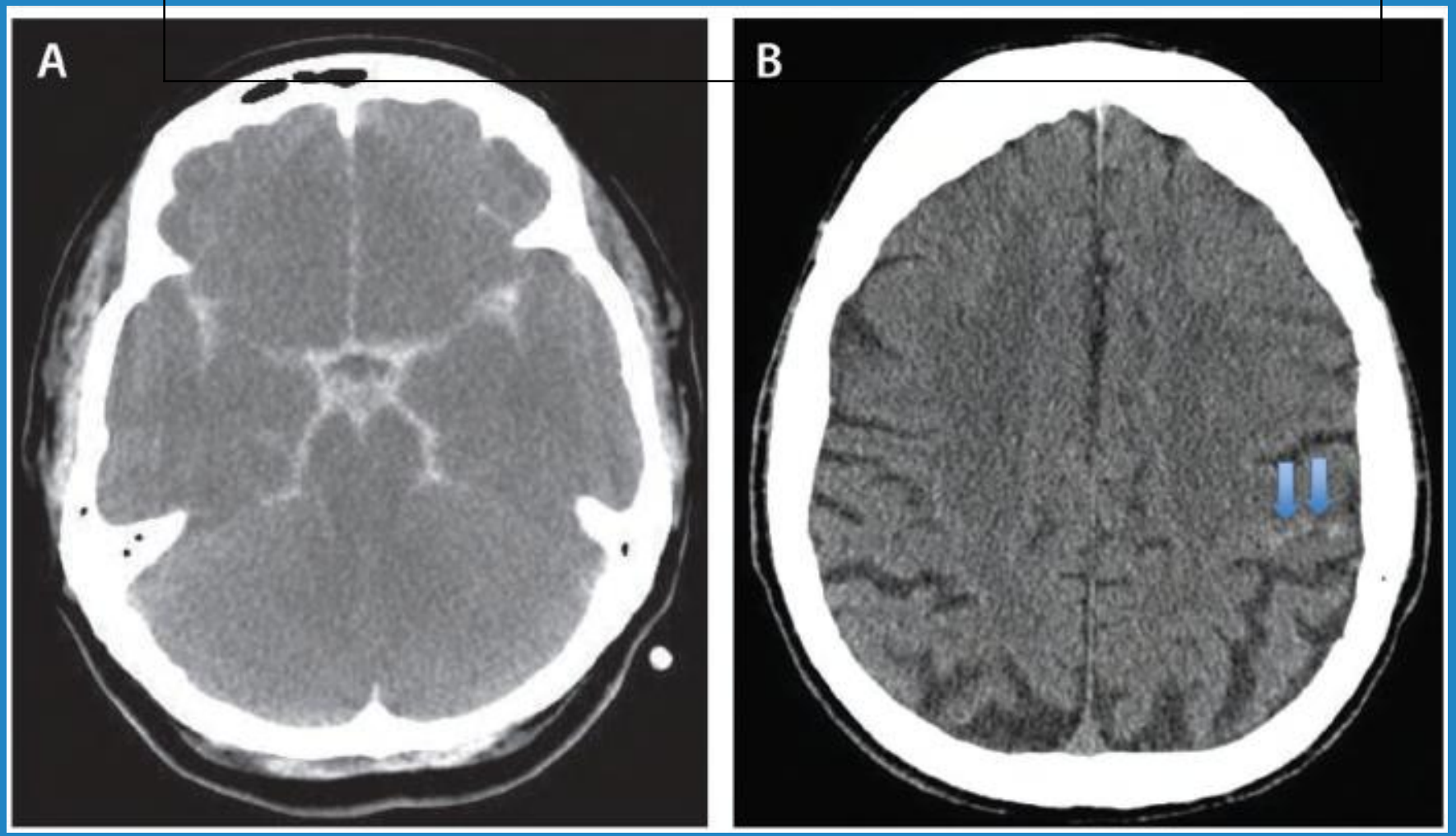
-The following features are associated with increased odds of SAH in a patient with TCH:

- impaired consciousness
- Neck stiffness
- nausea, vomiting
- exertion or valsalva immediately preceding onset of TCH
- elevated BP
- occipital headache
- History of smoking

- Whether or not these features are present, all patient with TCH need to be evaluated for SAH.
- Approximately 12 to 51 percent of patients with SAH are initially misdiagnosed.

- SAH from aneurysmal rupture must be differentiated from a cortical SAH that can be seen with RCVS.
- Aneurysmal SAH is typically seen within the sylvian fissures and basal cisterns, while SAH from RCVS is seen in the hemispheric convexities

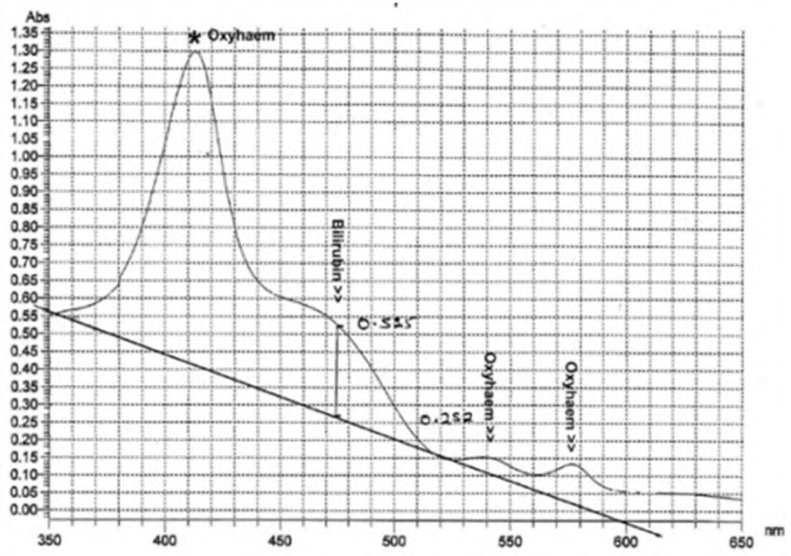
Axial noncontrast CTs showing aneurysmal subarachnoid hemorrhage versus cortical subarachnoid hemorrhage (seen in up to one-third of patients with RCVS

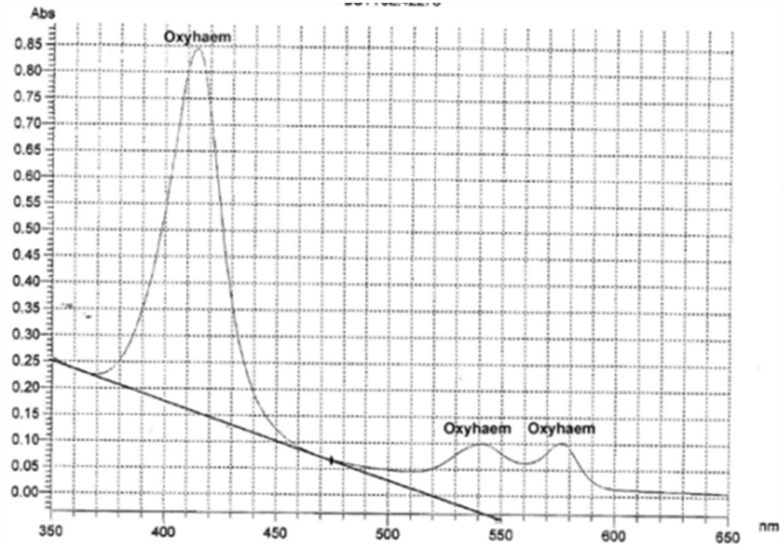


- Approximately 2% to 15% of patients with thunderclap headache with normal brain CT scans who are ultimately diagnosed with aneurysmal SAH have evidence for SAH on CSF analysis.

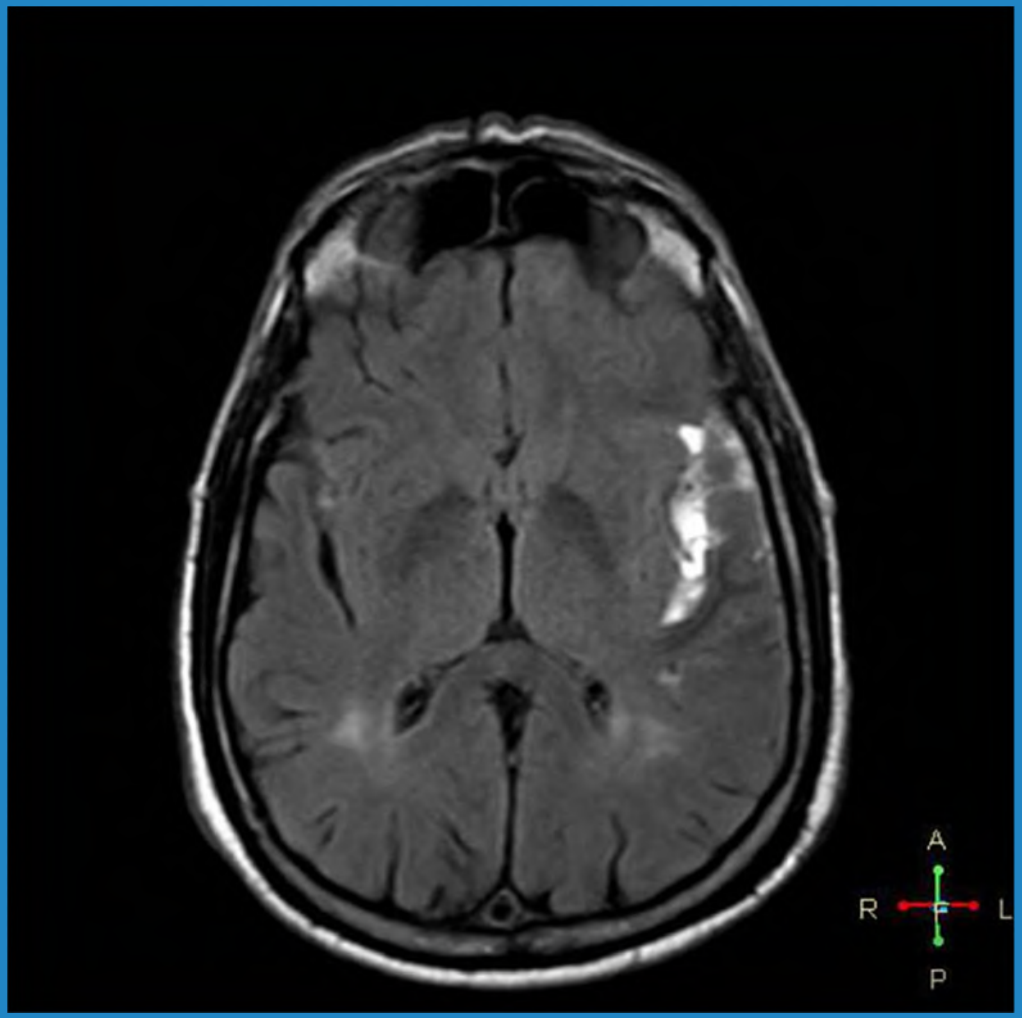
- The CSF diagnosis of SAH is made via measurement of red blood cells in tube 1 and tube 4, visual inspection of the CSF for xanthochromia and spectrophotometry (if available)

- In contrast to a traumatic LP, red blood cell counts in tube 4 should be similar to or higher than tube 1 in patients with SAH.
- Spectrophotometry is useful, when available, as it has a sensitivity of 98% for detecting SAH when performed between 12 hours and 2 weeks after symptom onset.





- FLAIR and gradient echo/susceptibility-weighted images are very sensitive for detecting SAH.
- Brain MRI is considered to be equally as sensitive as brain CT for detecting SAH within the acute phase and more sensitive than CT after the acute phase.
- Thus, brain MRI is an important tool for the evaluation of patients suspected to have SAH who present for evaluation several days after symptom onset.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- Although catheter angiography is still considered the gold standard, MRA or CTA may be considered for the initial angiographic evaluation.
- MRA and CTA on modern scanners have high sensitivity for detecting aneurysms, especially those larger than 3 mm.

- Both the vasoconstriction of RCVS and the vasospasm of aneurysmal SAH can be delayed findings, being maximal several days to 1 week after aneurysmal SAH and up to 3 weeks following onset of RCVS.
- The vasospasm of aneurysmal SAH is confined to the blood vessels in the area of the SAH, while the vasoconstriction of RCVS involves multiple arteries, often including vessels in both hemispheres and in both the anterior and posterior circulation.

Unruptured Intracranial Aneurysm—“Sentinel Headache”

- “warning headache” that occurs days to weeks prior to a SAH .
- typically present with a TCH without meningismus or altered level of consciousness or focal neurological symptoms and signs.

- Ten to 43 percent of patients with aneurysmal SAH report a history of a sentinel or warning headache.
- sentinel headaches are likely caused by small aneurysmal leaks of blood into the subarachnoid space or physical changes within the aneurysm wall.

- Sentinel headache is typically diagnosed retrospectively where the patient had TCH days to weeks prior to the intracranial aneurysmal rupture with SAH.
- In a prospective review, 75%of patients with a sentinel headache, had the TCH within 2 weeks of the SAH with a peak incidence at 24 hours.

- The decision of whether to treat the unruptured aneurysm should be based on current guidelines for the management of unruptured intracranial aneurysms.
- For patients who do not undergo aneurysm treatment, close follow-up is indicated.

Reversible Cerebral Vasoconstriction Syndrome

- RCVS is more common in women (3:1) than men and typically present between the ages of 20 and 50 years.

- In early RCVS, vasoconstriction may not be seen and may lead to the erroneous diagnosis of primary thunderclap headache.

- Thunderclap headache(s) with or without focal neurologic deficits or seizures
- Monophasic course without new symptoms more than 1 month after initial onset of symptoms
- Multifocal, multivessel, segmental vasoconstriction of cerebral arteries
- Absence of aneurysmal subarachnoid hemorrhage
- Normal or near-normal CSF

  Protein less than 100 mg/dL

  White blood cells less than 15 per mm^3

  Glucose normal
- Complete or substantial normalization of cerebral arteries within 12 weeks of symptom onset

trigger

- migraine
- postpartum period
- exposed to different pharmacologic agents including ergotamine, triptans ,SSRI , pseudoephedrine, cocaine, amphetamine, ecstasy, cannabis and bromocriptine
- other agents (IVIG, cyclophosphamide, tacrolimus)
- catecholamine-secreting tumor (pheochromocytoma)
- precipitants include vascular surgery and trauma.

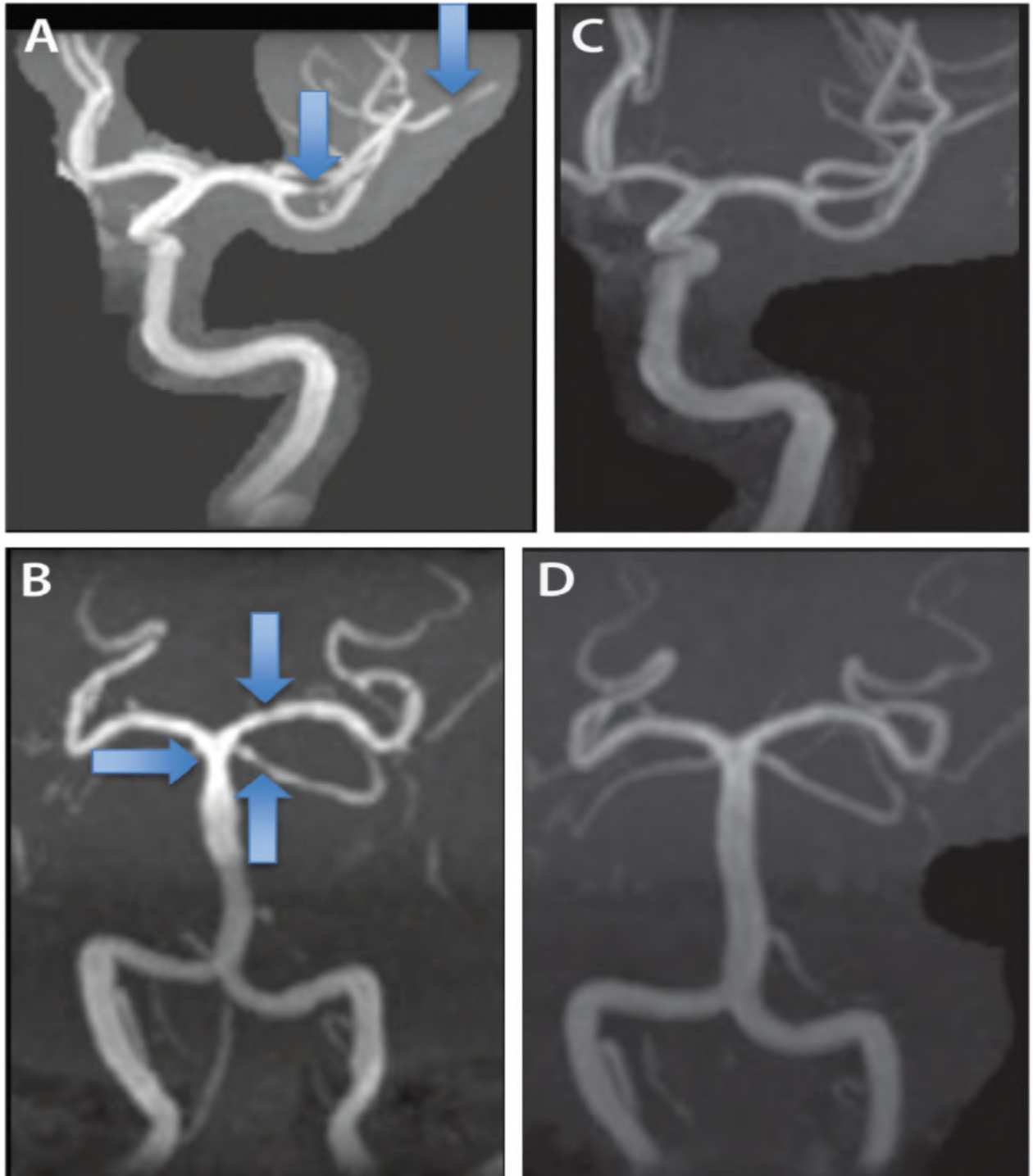
- RCVS is a common cause of TCH.
- severe and often recurrent headaches over a period of 1– 2 weeks and diffuse segmental cerebral arterial vasoconstriction.
- Multiple TCH recurring over a mean period of 1 week has been reported in up to 94 % of patients
- A pattern of recurrent thunderclap headaches (between 2 and 10 thunderclap headaches) over approximately 1 to 2 weeks is very suggestive of RCVS.

other symptoms

- a continuous mild to moderate headache
- nausea, vomiting
- photophobia, phonophobia
- cognitive dysfunction
- alterations in consciousness
- Seizures
- transient focal neurologic deficits
- permanent neurologic deficits from ischemic or hemorrhagic stroke.

- normal Brain CT and MRI in 30% to 70% of patients.
- cortical SAH in 22% to 34%
- intracerebral hemorrhage in 6% to 20%
- ischemic stroke in 4% to 39%
- cerebral edema such as that seen in PRES in 9% to 38%.

- Angiography reveals multifocal vasoconstrictions of multiple intracranial arteries in a ‘‘string of beads’’ appearance .
- Vasoconstrictions are maximal at about 2 to 3 weeks after symptom onset.



Numerous areas of vasoconstriction (arrows) are apparent within the anterior (A) and posterior (B) circulation on magnetic resonance angiography. Normalization of the intracranial arteries is seen 10 weeks later (C, D).

- vasoconstriction might start distally and move more proximally during the first several weeks after symptom onset→ normal vascular imaging performed early after the onset of symptoms→ repeat vascular imaging after several weeks

- DDx: primary CNS vasculitis

Cerebral Venous Sinus Thrombosis

- Headache occurs in over 80 % of cases and is subacute (2 days to 1 month); however, 2 %–14 % of patients have reported TCH at onset.
- Although 25% of patients with cerebral venous sinus thrombosis present with headache alone, the majority of patients present with additional features such as abnormal neurologic examinations, papilledema, altered mental status, seizures, and focal neurologic deficits.

Cervical Artery Dissection

- spontaneous internal carotid or vertebral artery dissections, headache was present at clinical presentation in about 70% of patients.
- Thunderclap headaches are present in 9.2% of patients with vertebral artery dissections and 3.6% of patients with internal carotid artery dissections.

- Headache or neck pain may precede other neurologic symptoms and/or signs by a median time of 4 days for a carotid dissection or 14.5 hours for a vertebral artery dissection .
- Thunderclap headache and neck pain were more common in vertebral artery dissections.

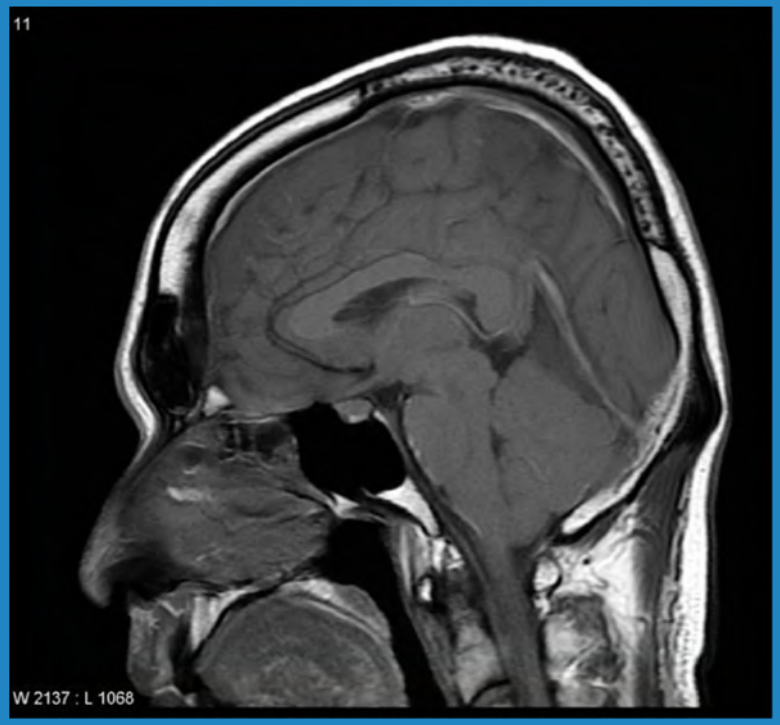
- Pain is ipsilateral (91 %) to the carotid dissection and often localized to the frontal or temporal regions, jaw, ear, and/ or orbit .
- In contrast, occipital or parieto-occipital pain is noted in approximately 50 % of patients with vertebral artery dissections.

- Symptoms related to cerebral ischemia are present in 84% to 90% of patients with vertebral artery dissection,and symptoms related to cerebral or retinal ischemia are present in 70% to 73% of patients with internal carotid artery dissection.
- Horner syndrome is present in 47.2% of patients with internal carotid dissection.

Spontaneous Intracranial Hypotension

- The hallmark feature :an orthostatic headache, a headache that is worsened when a person is upright (standing or sitting) and relieved when lying down .
- However, about 15% of patients with spontaneous intracranial hypotension initially present with a thunderclap headache.
- common symptoms of spontaneous intracranial hypotension include auditory muffling, tinnitus, nausea, vomiting, neck stiffness, dizziness, and visual changes.

- Brain MRI :diffuse, smooth, and continuous pachymeningeal enhancement ,cerebellar tonsil and optic chiasmal descent, flattening of the anterior pons and tectum, pituitary enlargement, and dilation/engorgement of the cerebral venous sinuses. Subdural fluid collections (hematomas or hygromas)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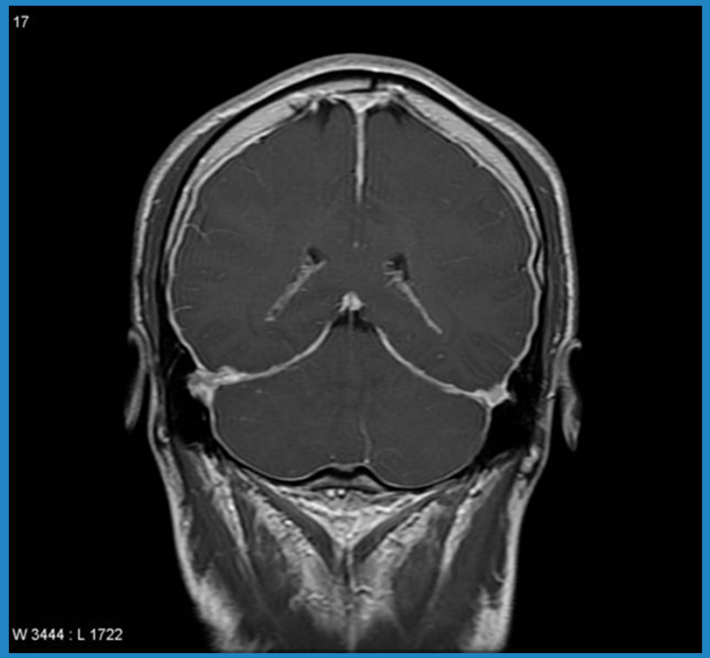
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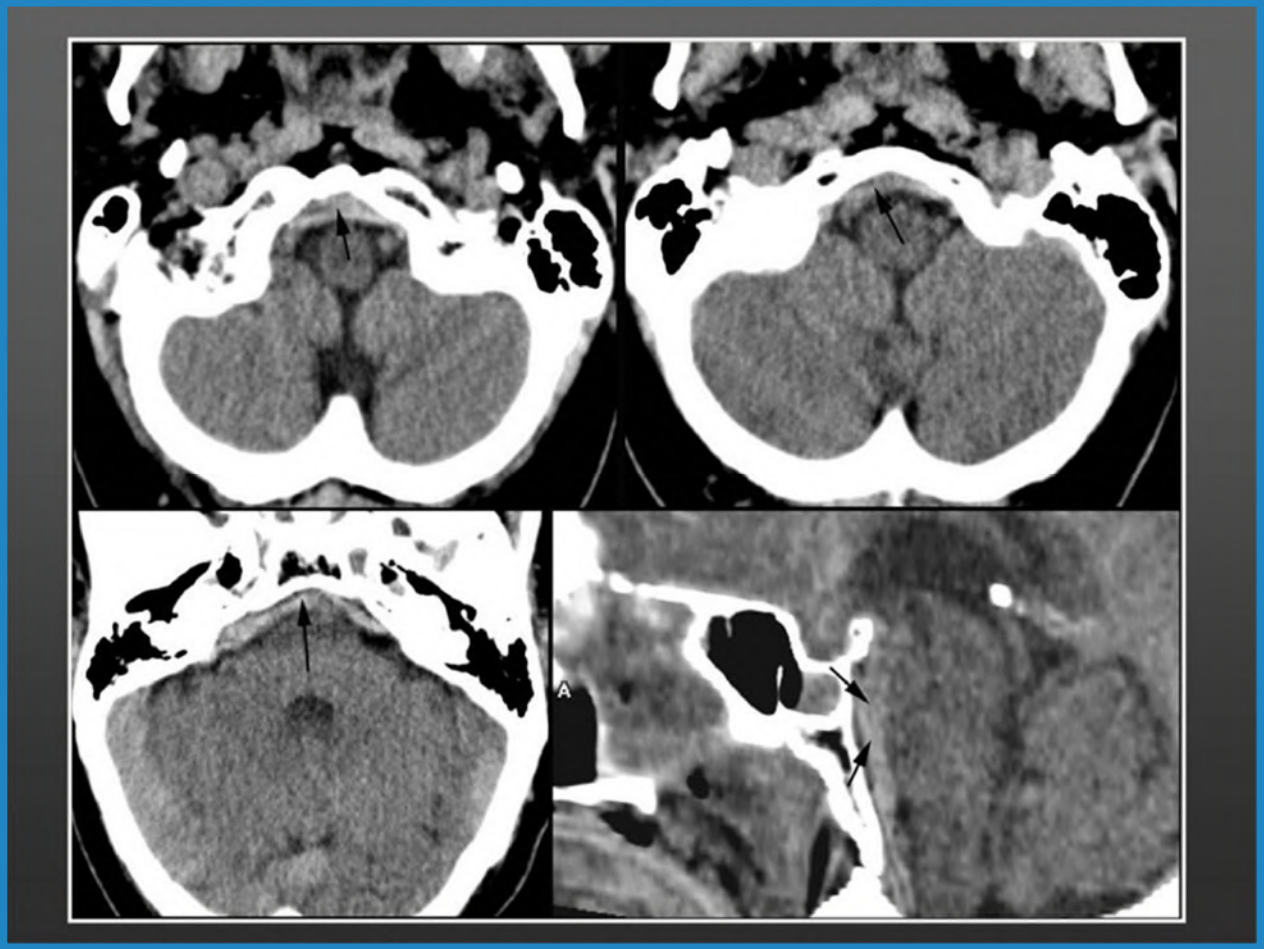
- Spine MRI might show extra-arachnoid spinal fluid collections coursing along the cervical, thoracic, or lumbar spine; venous engorgement.

Pituitary Apoplexy

- Sudden severe headache (87 %–97 %) , meningismus, nausea/vomiting (78 %), neuro-ophthalmological symptoms (83 %) such as visual disturbance and ophthalmoparesis and altered level of consciousness (13 %–50 %).
- Patients may rarely present with TCH alone.
- MRI identifies the pituitary hemorrhage in 88 % of cases compared with 21 % with CT head and is the imaging of choice.

Retroclival Hematoma

- Severe headache and neck trauma leading to atlantoaxial dislocation typically causes a retroclival hematoma.
- spontaneous hemorrhage has been reported.
- MRI with gadolinium and cerebral angiogram including injection of the external carotid artery has been recommended in cases of spontaneous retroclival hematoma.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Stroke

- Ischemic strokes (25 %–34 %) and more commonly intracranial hemorrhage (ICH) may present with a headache.
- TCH was found in 2%–6% of patients with ICH .
- There are cases of TCH reported in ischemic strokes ; however, the mechanism is uncertain.

Acute Hypertensive Crisis

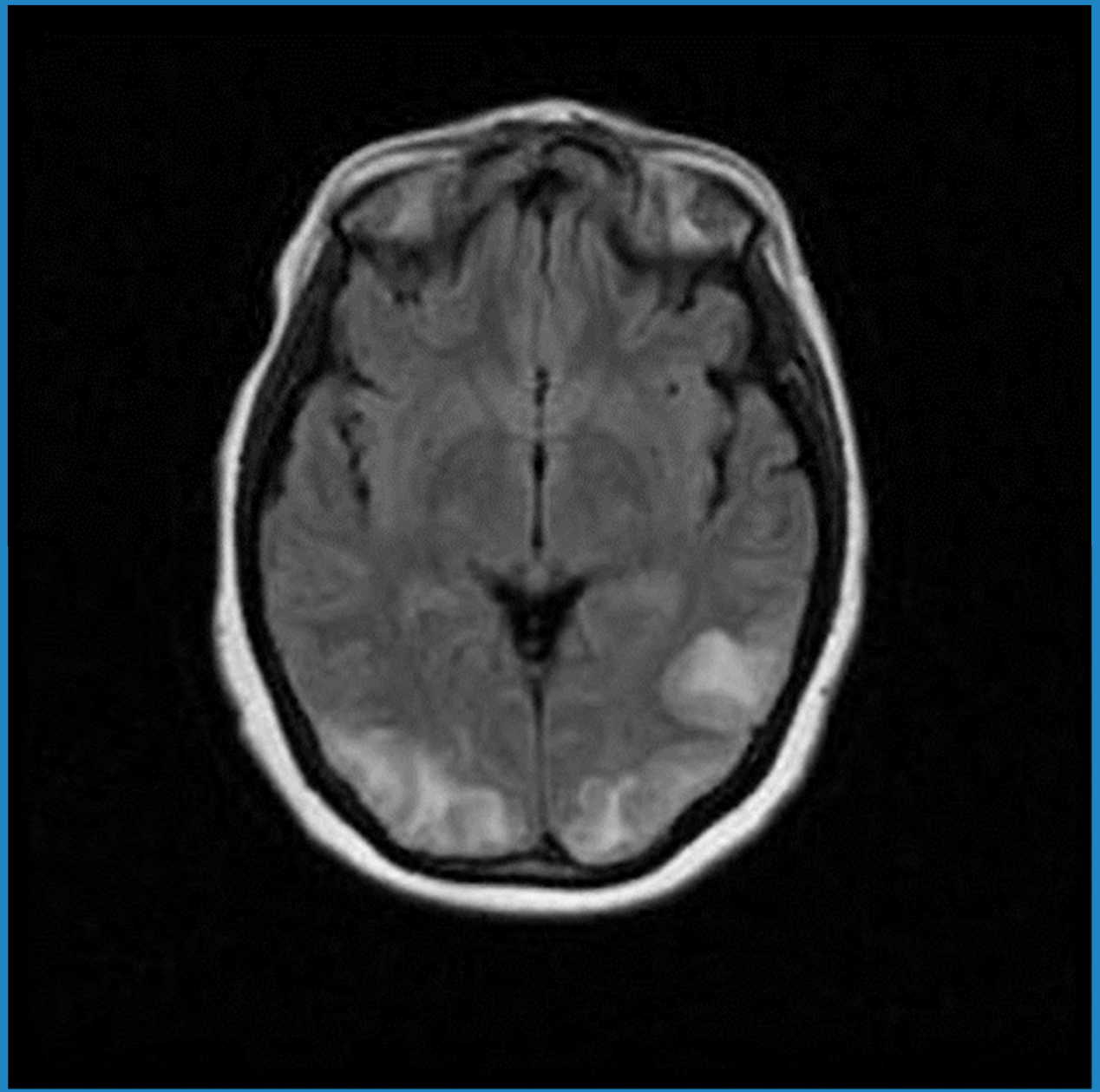
- Headache occurs in approximately 20 % of patients with acute hypertensive crisis .

- Such headaches are commonly located in the posterior regions of the head and are usually associated with other symptoms, such as dizziness, shortness of breath, chest pain, psychomotor agitation epistaxis, and focal neurologic deficits.

Reversible Posterior Leukoencephalopathy Syndrome

- Reversible posterior leukoencephalopathy syndrome (RPLS) is a clinico-radiologic syndrome characterized by acute or subacute onset of confusion, headache, seizures, and visual changes.
- It is also known as posterior reversible encephalopathy syndrome (PRES).
- Headache is severe and acute or thunderclap.
- RPLS may occur in the setting of extreme hypertension, eclampsia, thrombotic thrombocytopenic purpura, hemolytic uremic syndrome, and immunosuppressive drugs such as IVIG, cyclosporine, tacrolimus, and interferon alpha .

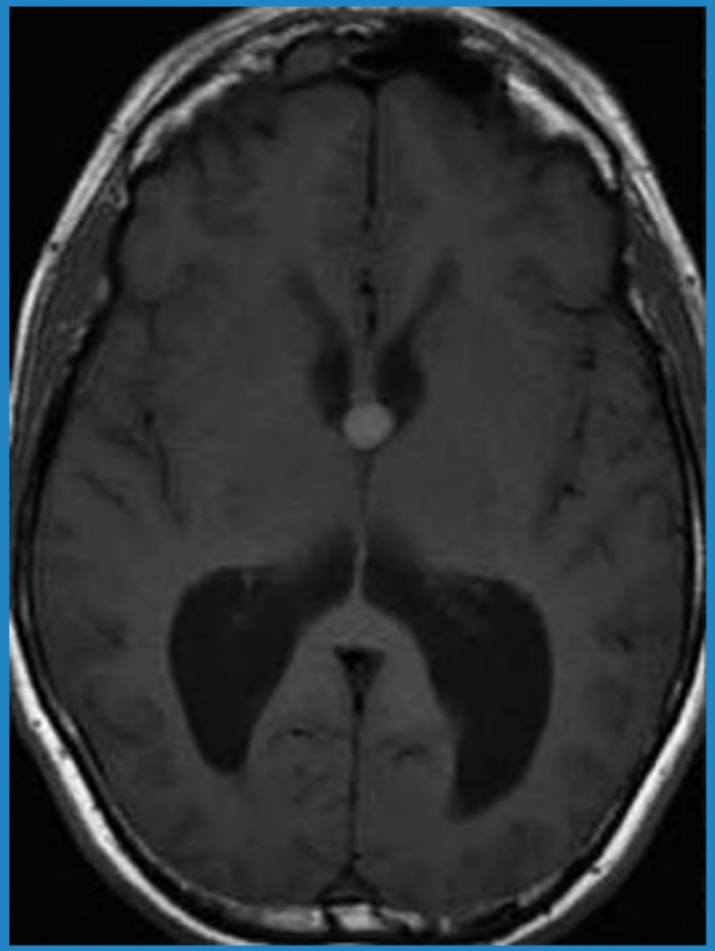
- Moderate to severe hypertension occurs in about three-fourths of patients.
- CT head and lumbar puncture is usually normal in RPLS.
- Reversible vasogenic edema predominately in the posterior white matter and cortex of the parieto-occipital region is seen on MRI brain and explains the reversibility of the neurologic symptoms.
- The frontal lobes, inferior temporal-occipital junction, cerebellum, and basal ganglia can be involved.



Third Ventricle Colloid Cyst

- Headache, often severe, sudden, paroxysmal and positional, is the most common symptom .
- TCH and the acute severe headache can be reproduced when tilting the head forward (Brun phenomenon).
- Colloid cyst should be in the differential diagnosis of recurrent brief (<30 minutes) severe headaches .

- frontal predominant severe headaches can be associated with vomiting (50 %), visual disturbance, papilledema, drop attacks, altered mental status or consciousness, gait disturbance.
- Sudden death has been reported in up to 10 % of cases .
- CT head :hyperdense mass in the third ventricle while on the MRI head, the cyst is usually hypointense on T2 weighted images and variable in intensity on T1 weighted images.



Primary Thunderclap Headache

- Primary Thunderclap Headache (TCH) is characterized as a high-intensity headache of abrupt onset, mimicking that of ruptured cerebral aneurysm, in the absence of any intracranial pathology.
- Primary TCH should be a diagnosis of exclusion

ICHD-3

-Diagnostic criteria:

A. Severe head pain fulfilling criteria B and C

B. Abrupt onset, reaching maximum intensity in <1minute

C. Lasting for ≥5 minutes

D. Not better accounted for by another ICHD-3 diagnosis.

Thanks!