Neurology Resident Teaching Series: Thunderclap Headaches

**Objective**

To familiarize Neurology clerkship medical students with the differential for severe, sudden onset headaches.

To organize etiologies of thunderclap headaches by pathologic mechanism.

To identify the etiologies that can cause permanent neurologic deficit.

To identify the etiologies that can lead to sudden or rapid progression to death.

To determine the appropriate diagnostic testing.

**-itis, -oma, -emia**

Thunderclap headaches are sudden onset headaches and are usually severe in intensity. The headache may be the only presenting symptom, thus requiring a good history and physical examination to identify subtle neighboring signs and advanced diagnostic testing to confirm the diagnosis.

Thunderclap headaches arise from inflammation, mass effect, or vascular occlusion or rupture. This is an oversimplification of their mechanisms, but it helps to break down an otherwise considerably lengthy list of potential diagnoses into a few groupings. A few, more benign etiologies do not readily fit into these categories (migraine, primary thunderclap headache, and spontaneous intracranial hypotension.)

Several of these etiologies can lead to permanent neurologic damage through ischemia, hemorrhage, or mass effect. Those that cause ischemia include CNS vasculitis, cervical artery dissection, cerebral venous sinus thrombosis, and occasionally reversible cerebral vasoconstriction syndrome (RCVS). Antithrombotic therapy is important for cervical artery dissection and cerebral venous sinus thrombosis (CVST), while vasospasm prophylaxis may be useful in CNS vasculitis, RCVS, and subarachnoid hemorrhage (SAH). Antihypertensive therapy is the mainstay of treatment for hypertensive encephalopathy and subarachnoid hemorrhage. Both SAH and colloid cyst of the third ventricle may require decompressive surgical intervention.

Of these etiologies, the two etiologies that can lead to sudden or rapid progression to death are SAH and third ventricle colloid cyst (by acute hydrocephalus through a ball-valve obstruction of CSF flow).

**Thunderclap Headaches**

1. **CNS vasculitis** refers to conditions which cause inflammation and vasospasm of intracerebral blood vessels. This includes both primary angiitis of the CNS as well as secondary causes of vasospasm (systemic vasculitis, infection, malignancy, connective tissue
diseases, etc.). These are relatively uncommon conditions that may be identified initially through vessel imaging (which may show “beading” or “pruning” of several intracerebral branches). They may require body imaging, systemic tissue biopsy, and brain biopsy to confirm the diagnosis.

2. **Colloid cyst of the third ventricle** is a condition that presents as a sudden onset headache at any age. The headache occurs when the cyst obstructs CSF flow by acting as a ball-valve. This headache may be temporarily relieved by lying down, but it is a dangerous neurosurgical condition that may rapidly progress to death if not addressed quickly.

3. **Pituitary apoplexy** results from sudden onset hemorrhage into the pituitary gland, usually in the setting of a pituitary adenoma. In addition to the headache, individuals may develop diplopia from third nerve palsies, visual field or acuity deficits from pressure on the optic pathways, and hypotension from adrenal crisis. Although not necessarily likely to cause death by a neurologic cause (as in SAH or third ventricle colloid cyst), the hypotension can be very dangerous.

4. **Cervical artery dissection** can affect either carotid or vertebral arteries. These typically occur in the setting of neck trauma or manipulation (as with chiropractic, deep neck massage, or extreme traction injury with exercise). Vertebral artery dissections are more likely to be accompanied by neck pain, although this is also relatively common in carotid artery dissection. This is a relatively common cause of embolic ischemic stroke in young adults who have no other vascular risk factors, so this headache may be accompanied by focal neurologic deficits.

5. **Cerebral venous thrombosis** may occur in small cortical veins or in the larger sinuses, causing venous hypertension, edema, and ischemic and hemorrhagic damage to adjacent brain parenchyma. These typically occur in the same settings as systemic deep venous thrombosis, such as dehydration, hypercoagulable states (including pregnancy and malignancy), tobacco use, and estrogen-containing contraceptive use. It is important to note that individuals with focal neurologic deficits that do not fit an arterial vascular distribution may have deficits referable to a venous vascular distribution (including bilateral thalamic edema causing a comatose state that may resolve with treatment).

6. **Hypertensive encephalopathy** is also known as posterior reversible leukoencephalopathy syndrome (PRLS) and posterior reversible encephalopathy syndrome (PRES). While many individuals describe their headaches as “hypertensive” headaches, they do not meet the criteria for this condition until neurologic deficits accumulate. Classically, this is described as a condition causing vasogenic edema and damage to bilateral parietal-occipital regions with associated visual, sensory processing, and cognitive deficits. Closely controlled (and not excessively rapid) lowering of the blood pressure is the treatment.

7. **Subarachnoid hemorrhage** is the most well-known and feared thunderclap headache. Some individuals, if they survive the hemorrhage, describe the headache onset as “being struck by a brick wall.” Typically, the headache, which is always present, progresses to rapid diminishment of consciousness and then coma. SAH can cause CSF flow obstruction and acute obstructive hydrocephalus, leading to death if not addressed with decompressive neurosurgical intervention. Seizures may also occur. Vasospasm and associated ischemia is a late consequence. SAH is usually caused by trauma, but individuals who present with
thunderclap headache in the absence of trauma or neurologic deficits may have a sentinel hemorrhage of an unruptured (but leaky) intracranial aneurysm that proceeds rupture by days to weeks. Noncontrast head CT is the initial first test that is highly sensitive for the detection of intracranial hemorrhage early in the onset of headache, but often a lumbar puncture is performed to exclude the diagnosis of SAH.

8. **Reversible cerebral vasoconstriction syndrome** is a group of conditions (including Call-Fleming syndrome) which result in segmental, reversible vasospasm of intracranial arteries. These often occur after pregnancy, when taking sympathomimetic medications, or with a history of migraine. While these conditions are reversible without intervention, they may be complicated by ischemia, hemorrhage, or seizure.

9. **Migraine** may present as a sudden onset, severe headache. In the case of migraine with aura, this may be accompanied by transient focal neurologic symptoms, but these often involve a gradual spread over minutes rather than over seconds (seizure) or having a maximal distribution at onset (stroke). The history and normal (or resolving) neurologic examination are the most important parts of the evaluation.

10. **Primary thunderclap headache** is a group of benign conditions associated with sudden onset, severe headache that may occur in the setting of exertion (including sexual intercourse or exercise), Valsalva maneuvers, or even spontaneously without an identified trigger. There are no neurologic deficits associated with this condition.

11. **Spontaneous intracranial hypotension** may result from a small leak in the meninges (from a dural tear or malformation), resulting in a low pressure in the CSF which causes traction on the meninges and pain. This may occur with (even minor) trauma. This headache is postural and worsens when standing or sitting up. This condition usually does not cause permanent neurologic damage (although rarely it is possible with severe reduction in CSF pressure with hemorrhage, compressive damage, etc.), but it can transform into a very difficult to treat chronic daily headache.

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**References**


