Managing Patients With Nontraumatic, Severe, Rapid-Onset Headache

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Editor's Note: *The Expert Clinical Management series consists of shorter, practical review articles focused on the optimal approach to a specific sign, symptom, disease, procedure, technology, or other emergency department challenge. These articles—typically solicited from recognized experts in the subject area—will summarize the best available evidence relating to the topic while including practical recommendations where the evidence is incomplete or conflicting.*

INTRODUCTION

Severe rapid-onset headache, sometimes called thunderclap headache, is a relatively common emergency department (ED) chief complaint. Although these headaches have been defined as "peaking within one minute,"[1,2] some have questioned this rigid time criterion.[3] In one study of subarachnoid hemorrhage diagnosis, the interobserver agreement in the ED for "thunderclap" onset was only moderate (κ=0.49), and 6 (of 132) subarachnoid hemorrhage patients reported time to peak intensity of 1 hour.[4] For these reasons, I do not strictly use "sudden onset" in practice, but these patients generally recall exactly what they were doing at headache onset.

All headache patients with new neurologic deficits require sufficient evaluation to explain those findings. This article focuses on the diagnosis of neurologically intact patients with nontraumatic, rapid-onset, severe and unusual headaches.

DIFFERENTIAL DIAGNOSIS

The most serious cause of severe rapid-onset headache is subarachnoid hemorrhage, accounting for approximately 7% to 8% of such patients.[5,6] The converse is that nearly all awake patients with subarachnoid hemorrhage will complain of such a headache. Most subarachnoid hemorrhage patients have cerebral aneurysms, although other causes exist (Table 1). History of neck pain (positive likelihood ratio [LR+] 4.1) or stiffness on examination (LR+ 6.6) suggests subarachnoid hemorrhage.[5] Forty percent of patents with subarachnoid hemorrhage appear well and have isolated severe headache.[6]

Less commonly considered in the ED, the second most common potentially serious cause is reversible cerebral vasoconstriction syndrome, accounting for approximately 8% to 9% of cases in 2 ED-based series (personal communication, W. Y. Kim, 2017).[7] Patients with cerebral vasoconstriction were treated by nearly 5 physicians before receiving a correct diagnosis.[8] Vasoconstriction typically presents with multiple thunderclap headaches (mean of 4) during days to weeks. This presentation is probably pathognomonic.[9,10] Although the onset of these headaches is identical to those in subarachnoid hemorrhage, their duration is usually much shorter, typically several hours.[9,10] Triggers (including exposure to marijuana, vasoactive and immunosuppressive drugs, autoimmune diseases, postpartum state, conditions that abruptly raise sympathetic output, sexual activity, and exposure to cold or heat, especially showering and bathing) are common.[8-10] Diagnostic criteria exist (Figure 1).

By definition, the vasoconstriction is "reversible," and most patients have benign outcomes. However, some patients have seizures (1% to 17%) or ischemic or hemorrhagic stroke with persistent deficit (3% to 20%), making this entity an important condition for emergency physicians to recognize.[9-11] Hemorrhagic events occur early and ischemic events occur later.[9] Death is rare.[9]

Uncommon but serious causes include cervical arterial dissection, cerebral venous sinus thrombosis, and pituitary apoplexy (Table 1).[1]

A study of 970 patients with arterial dissections showed that 5% presented with thunderclap headache (3.6% carotid and 9.2% vertebral).[12] Nearly half of the 668 patients with carotid dissections had Horner's syndrome.[12] Examine these patients for mild ptosis and

Table 1. Differential diagnosis of thunderclap headache and clinical clues to the diagnosis in neurologically intact patients.

Diagnosis	Clinical Clues to Diagnosis	Best Diagnostic Test
Subarachnoid hemorrhage	Neck pain by history or meningismus on physical examination, transient loss of consciousness Acute hypertension	CT followed by LP (depending on timing of the CT) CTA to show the offending vascular lesion*
Unruptured cerebral aneurysm	Third nerve palsy, usually with pupillary dilatation	CTA
Reversible cerebral vasoconstriction syndrome	Multiple thunderclap headaches during days or weeks is pathognomic There is usually a "trigger" to the headache (see text) Acute hypertension	CTA or MRA (may be falsely negative during the first week)
Hemorrhagic stroke	Focal neurologic deficit or altered mental status is usually present	CT
Cerebral venous sinus thrombosis	Any hypercoagulable risk factor, including pregnancy, postpartum state, and oral contraceptive use Papilledema	CTV or MRV LP will often show elevated opening pressure
Cervical artery dissection	Headache or neck pain, recent (even minor) head or neck trauma, subtle physical examination findings (see text) of posterior ischemia Horner's syndrome (carotid dissection)	CTA or MRA (need both head and neck)
Posterior reversible encephalopathy syndrome/hypertensive encephalopathy	Acute hypertension (compared with patient's baseline), pregnancy or postpartum state (blood pressure elevation in these patients may be modest) Seizure and visual symptoms	MRI
Spontaneous intracranial hypotension	Positional headache (worse on standing up, resolves with lying down) Recent spine surgery or vaginal delivery Tinnitus, sound distortion, and dizziness	LP with opening pressure MRI with gadolinium
Pituitary apoplexy	Prominent visual symptoms: ptosis, various degrees of ophthalmoplegia, and decreased vision or field cut(s)	MRI with dedicated cuts of the sella turcica
Colloid cyst of third ventricle	Associated vomiting, dizziness, and visual symptoms; transient loss of consciousness Headache is often paroxysmal and positional	CT and MRI
Sphenoid sinusitis	Preceding URI symptoms, fever	CT
Meningitis/encephalitis	Fever, neck pain, or stiffness	LP
Isolated acute-onset headache would be a rare presentation	Altered mental status (encephalitis)	

LP, Lumbar puncture; *CTA*, CT angiography; *CTV*, computed tomographic venography; *MRV*, magnetic resonance venography; *MRA*, magnetic resonance angiography.
This table does not list various rare conditions that have been reported to cause severe rapid-onset headache as case reports. These include myocardial infarction, aortic dissection, temporal arteritis, retroclival hematoma, and rare acute ischemic stroke (although these patients should by definition have some measurable neurologic deficit). Some of the listed conditions such as meningitis rarely present with severe acute-onset headache, but are important considerations.
*Aneurysms cause approximately 80% of nontraumatic subarachnoid hemorrhages. Approximately 10%, called perimesencephalic hemorrhages, are nonaneurysmal and probably due to venous bleeding. The other 10% are caused by a wide range of vascular abnormalities, including arteriovenous malformations, blood vessel abnormalities such as moyamoya, and vasculitis or coagulapathies.[6]

anisocoria (easier to appreciate in a dark room so that the unaffected pupil dilates, accentuating the difference in size). Patients with vertebral dissections may have subtle vestibular findings, including limb ataxia, nystagmus, or abnormal gait.[13] Intracranial dissections not involving the cervical portions of the relevant artery can occur.[14]

Of patients with cerebral venous sinus thrombosis, 5% to 15% present with severe rapid-onset headache.[15,16] Some patients have papilledema. In a recent meta-analysis, the negative likelihood ratio (LR–) of a sinus thrombosis after a negative D-dimer result for patients presenting with isolated headache was 0.03 (95% confidence interval 0.0 to 0.18), suggesting that venography is unnecessary in this population.[17] For patients with risk factors, including those receiving oral contraceptives, there are currently insufficient data to use D-dimer as a rule-out test.

Pituitary apoplexy occurs from bleeding into a previously undiagnosed pituitary adenoma and can be fatal without treatment. Nearly all individuals with pituitary apoplexy have thunderclap headache, often with vomiting. Ptosis, diplopia, or decreased vision is commonly present.[18]

Table 1 summarizes clinical clues and suggested testing for these conditions. Pregnant and postpartum