

Spontaneous Low Cerebrospinal Fluid Pressure Syndrome Can Mimic Primary Cough Headache

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Key words: spontaneous low cerebrospinal fluid pressure syndrome, spontaneous intracranial hypotension headaches, cough headache

(*Headache* 2005;45:374-377)

When evaluating cough headache, if you obtain an MRI scan of the brain without contrast only, you may miss the diagnosis.

CLINICAL HISTORY

This is a 66-year-old white male with occasional mild headaches in the past. He presented with a 4-month history of headaches occurring one to four times per day, brought on by having a bowel movement, stooping, or getting up from a sitting position. He did not know if the headache was triggered by coughing because he had not coughed at all. The headache was a bifrontal and bitemporal sharp, aching pain with a 7/10 intensity and occasionally a 9/10 intensity with an average duration of 1 minute and a range of 30 seconds to 1 hour. About 20 of the headaches had lasted more than 1 minute, with most in a range of 1 to 2 minutes. He had tried ibuprofen and acetaminophen with questionable help. For the prior 5 days, he had increased his dose of aspirin from 81 mg per day to 325 mg per day. The headache was then different with a constant bifrontotemporal pressure with an intensity of 1/10 but he had not had the brief headaches with activity exacerbation. He had a CT scan of the sinuses on June 14, 2004 with essentially negative findings.

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There was a past medical history of insulin-dependent diabetes with sensory neuropathy and hypertension. Neurological examination was normal except for diminished pinprick distally of both lower extremities and absent deep tendon reflexes diffusely.

Question.—What is the diagnosis? What tests would you recommend?

EXPERT OPINION

The initial history was suggestive of cough headache, which occurs with straining. It would seem a misnomer to use the term cough headache here, given that he did not mention cough as a trigger, but the IHS criteria for primary cough headache allow precipitation by coughing *or* straining and thus it is the best nomenclature (Table 1).¹

Primary cough headache can be triggered by other sudden Valsalva maneuvers, but is typically not triggered by sustained physical exercise.² This is in contradiction to the typical circumstance in primary exertional headache, and thus the IHS has a separate criteria for primary exertional headache.

Headache brought on by coughing has long been known to be a symptom of intracranial disease. In 1932, Tinel described four patients with headache brought on by coughing, nose blowing, breath holding, and bending the head forward.³ Before the reports of Symonds and Rooke, cough and exertional headaches were always considered ominous symptoms, and there was no clear recognition that benign or primary types

Table 1.—International Headache Society 2nd Edition 2004 Diagnostic Criteria for Primary Cough Headache¹

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- A. Headache fulfilling criteria B and C.
 - B. Sudden onset, lasting from 1 second to 30 minutes.
 - C. Brought on by and occurring only in association with coughing, straining, and/or Valsalva maneuver.
 - D. Not attributed to another disorder.*
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*Cough headache is symptomatic in about 40% of cases and the large majority of these represent Chiari type I malformations. Other reported causes of symptomatic cough headache include carotid or vertebrobasilar disease and cerebral aneurysms. Diagnostic neuroimaging plays an important role in differentiating secondary cough headache from primary cough headache.

of these headaches existed. In 1968, Rooke noted that “in every patient with this complaint, an intracranial lesion of potentially serious nature, such as brain tumor, aneurysm, or vascular anomaly, has been suspected; and even when no such lesion could be identified, an uneasy uncertainty usually has remained.”⁴ The landmark paper entitled “Cough headache” by Sir Charles Symonds in 1956 brought attention to this disorder.⁵ Symonds clearly described cases of both secondary and primary cough headache. He presented patients with headache provoked by coughing, and noted that in these same patients, sneezing, straining at stool, laughing, or stooping could also provoke the headache. He did not describe headache precipitated by physical exercise. In 1968, Rooke reviewed 93 patients with primary exertional headache.⁴ He did not separate cough headache from headaches caused by running. However, his data underscored Symonds’ concept that cough headache could be benign.

Before a diagnosis of primary cough headache can be made, an intracranial lesion and specifically a Chiari I malformation must be ruled out. Pascual and colleagues found clinical differences between patients with primary and secondary cough headache.² Symptomatic or secondary cough headache started earlier in life (age 39 ± 14 years), had a longer attack duration (seconds to days), was initially associated with posterior fossa symptoms or signs in 14 of 17 patients, and did not respond to indomethacin. Of the three patients who presented with isolated cough headache, posterior fossa symptoms or signs appeared

Table 2.—Some Secondary Causes of Cough Headache (After 7)

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- Chiari I malformation
 - Spontaneous low CSF pressure/volume headache (spontaneous CSF leak or spontaneous intracranial hypotension or CSF hypovolemia or CSF volume depletion)
 - Middle cranial fossa or posterior fossa meningiomas
 - Medulloblastoma
 - Pinealoma
 - Chromophobe adenoma
 - Midbrain cyst
 - Basilar impression
 - Platybasia
 - Subdural hematoma
 - Brain tumor not otherwise specified
 - Unruptured intracranial aneurysm
 - Sinusitis with pneumocephalus on CT head
 - Carotid stenosis (questionable)
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after 1 to 5 years. Chiari I malformation was the only cause for symptomatic cough headache in this study, with syringomyelia in five cases. Indomethacin response should not be used to differentiate primary from secondary cough headache, as a patient with a Chiari I malformation-associated cough headache that responded completely to indomethacin has been reported.⁶ Table 2 lists some of the reported causes of cough headache. Given the cough headache differential diagnosis, every patient with cough headache should have an MRI of the brain. The MRI should be done with gadolinium looking for pachymeningeal enhancement given that spontaneous low CSF pressure/volume headache can present as cough headache alone with no orthostatic component.⁸ Whether a patient with an unruptured aneurysm can present with cough headache is not clear, but it seems reasonable to obtain an MRA of the intracranial circulation in most cases. I do not typically do carotid ultrasounds or MRAs of the extracranial circulation in the evaluation of cough headache, unless the patient gives a history consistent with transient ischemic attacks.

The patient’s headache changed after increasing his aspirin dose in that he had a continuous dull pain no longer triggered by straining. This was an alteration from his prior headache pattern, however, and I would still pursue the neuroimaging outlined above.

FOLLOW-UP

An MRI scan of the brain was normal except for smooth diffuse dural enhancement around both cerebral convexities and, to a milder degree, in the posterior fossa. A lumbar puncture produced an opening pressure of 11 cm of water. Cerebrospinal fluid analysis revealed 0 white blood cells, 1 red blood cell/ μ L, a glucose of 101 mg/dL (with a serum glucose of 156 mg/dL), and a protein of 109 mg/dL. The VDRL was nonreactive. An erythrocyte sedimentation rate was 34. An ANA, RA factor, Sjogren's antibodies, Lyme antibodies, and an angiotensin-converting enzyme level were negative or normal. An MRI scan of the cervical, thoracic, and lumbar spine revealed degenerative changes but no evidence of extra-arachnoid fluid collections, extradural extravasation of fluid or meningeal diverticula.

Questions.—What is the diagnosis? What treatment would you recommend?

The investigations support a diagnosis of spontaneous low CSF pressure/volume headache. The smooth pachymeningeal enhancement on MRI is consistent with this diagnosis.⁹ The *diffuse* enhancement in spontaneous low CSF pressure/volume headache is both supratentorial and infratentorial, is linear and non-nodular, and is typically uninterrupted and bilateral.⁹ One might consider inflammatory CNS diseases (sarcoidosis, Wegener's granulomatosis, idiopathic hypertrophic pachymeningitis, etc.), infectious CNS diseases, or CNS lymphoma given this MRI, but the enhancement in those situations is typically patchy or relatively focal or partly nodular or asymmetric.¹⁰⁻¹² With the exception of idiopathic hypertrophic pachymeningitis, the MRI enhancement in these other CNS diseases is often leptomeningeal as well. Patients with low CSF pressure/volume headache can have normal opening pressures (thus the inclusion of the word "volume" in the description), and the CSF protein can be elevated.⁹

Epidural blood patch (20 mL) is the treatment of choice in patients who fail an initial trial of conservative management.^{9,13} Most patients with spontaneous CSF leaks have already spent a fair amount of time recumbent prior to seeing the doctor and thus in most cases I proceed directly to blood patch. I typically do at least one lumbar blood patch even without localizing

the site or level of the leak via CT myelography when the MRI of the brain is consistent with spontaneous low CSF volume/pressure headache. If this is unsuccessful, I investigate further with CT myelography to localize the site or level of the leak, since an epidural blood patch given at the level of the leak is somewhat more effective than one given at a distant site.¹³ Many patients may need more than one epidural blood patch, and some require several.⁹ The efficacy of each blood patch is about 30%.^{9,13} Epidural fibrin glue injection is a promising treatment, but more studies are needed before this can be routinely recommended.⁹ Surgery can be tried when conservative and less invasive approaches like epidural blood patch have failed.⁹

This patient's presentation emphasizes that the phenotypic spectrum of the headache associated with spontaneous CSF leaks is still being defined. Although classically orthostatic, it can present as chronic daily headache, second half of the day headache, cough headache, thunderclap headache, or even be paradoxically postural (present when recumbent, absent when upright), among other presentations.¹⁴

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