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Atypical Spontaneous Intracranial Hypotension With a Head-Shaking Headache

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Spontaneous intracranial hypotension (SIH) is typically characterized by orthostatic headache; however, various atypical manifestations of SIH have been reported recently. We report here the case of a 46-year-old man with headache secondary to SIH, which was nonorthostatic, triggered only when the patient shook his head. We suggest that SIH should be suspected in patients with headache induced by head-shaking, even without orthostatic features, especially when the headache is accompanied by other symptoms commonly associated with SIH.

Key words: head-shaking headache, spontaneous intracranial hypotension

Abbreviations: CSF cerebrospinal fluid, MRI magnetic resonance imaging, SDH subdural hematoma, SIH spontaneous intracranial hypotension

Spontaneous intracranial hypotension (SIH) is typically characterized by orthostatic headaches which occur or worsen less than 15 minutes after assuming an upright position, and disappear or improve less than 30 minutes after resuming a recumbent position. However, various atypical clinical features have been reported recently, including the absence of headache and nonorthostatic headaches.¹⁻³ These diverse presentations of SIH lead to a high frequency of initial misdiagnoses by physicians. We describe here a unique case of SIH with nonorthostatic headache occurring only when the patient shook his head.

CASE REPORT

A 46-year-old previously healthy man was admitted to our hospital for evaluation of the sudden

development of headache refractory to common analgesics. The headache was throbbing in nature with moderate intensity (visual analog scale of 5) and was localized to the occipital area. Interestingly, the headache occurred only when the patient shook his head in the horizontal and vertical planes and totally disappeared within a few seconds after the head-shaking stopped. Other activities of daily living were normal, but he could not do any things that involved head movement and could not even wash his face properly because of the associated headache. The headaches were velocity-dependent, but not position-dependent. The headaches were not affected by standing or lying position. Despite repeated administration of common analgesics, this head-shaking headache occurred every day when the patient tried to move his head. On the fifth day after symptom onset, his pain was accompanied by left-sided tinnitus. He had no history of trauma, lumbar puncture, or surgery

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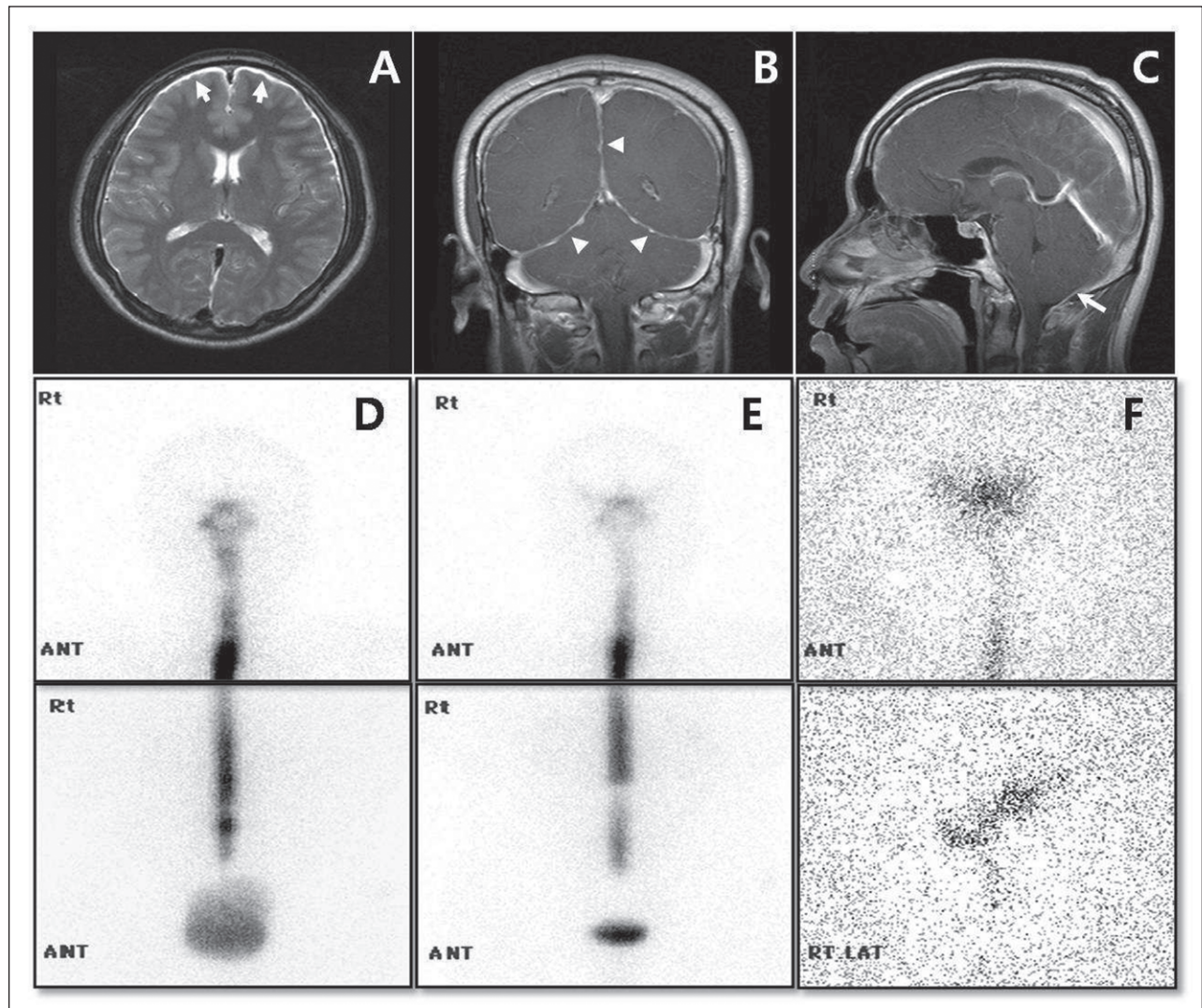


Figure.—Brain magnetic resonance imaging with gadolinium enhancement shows bilateral subdural fluid collections (A, arrows), diffuse pachymeningeal enhancement (B, arrowheads), and sagging of the brain (C, long arrow). Radionuclide cisternography 2 hours (D), 4 hours (E), and 24 hours (F) after injection of the tracer demonstrates early bladder uptake (D, E) and slow ascent of radionuclide, as evidenced by no apparent radionuclide activity over the cerebral convexities (F). These findings are suggestive of intracranial hypotension.

of any kind. Routine neurological examination was normal without meningeal inflammation signs.

Magnetic resonance imaging (MRI) of the brain with gadolinium enhancement showed diffuse, smooth pachymeningeal enhancement and small bilateral subdural fluid collections along the cerebral convexities and tentorium cerebella (Figure A-C). Cerebrospinal fluid (CSF) analysis revealed the presence of a clear fluid, with an opening pressure of 3 cmH₂O, a white blood cell count of 2 cells/ μ L, a slightly elevated CSF protein level of 65 mg/dL

(normal range < 45 mg/dL), and a normal CSF/serum glucose ratio, suggesting decreased intracranial CSF pressure. Further evaluation with radionuclide cisternography to confirm the diagnosis demonstrated rapid accumulation of radiopharmaceutical in the bladder at 1 hour and 4 hours, with faint renal uptake and without accumulation in the cerebral convexities by 24 hours, suggesting CSF leakage (Figure D-F).

The diagnosis of SIH induced by specific action, ie, head-shaking, was made. Concerning the treatment, despite 4 days of hydration and supine bed rest,

the patient's head-shaking headache remained. After autologous epidural blood patching was performed using 15 mL of blood injected at the L3-L4 level, however, the headache completely subsided within a day. He was discharged after 5 days of hospitalization. One month after the blood patch, he no longer complained of headache and returned to work full time.

DISCUSSION

Spontaneous intracranial hypotension has a wide spectrum of clinical manifestations. Headache occurring only during rapid oscillatory head movements in the horizontal and vertical axes and totally disappearing shortly after cessation of shaking, as in this case, is very unique in SIH. We made the diagnosis of SIH with headache, typically induced by head-shaking movement, based on the findings of low CSF pressure, diffuse pachymeningeal enhancement and subdural fluid accumulation on MRI, the presence of CSF leakage on radionuclide cisternography, and his excellent treatment response to epidural patching.⁴⁻⁶

Orthostatic headache has been recognized as a characteristic manifestation and definite diagnostic criterion for SIH.^{4,5,7} In classic SIH, the typical orthostatic headache is caused by compensatory vasodilation of the dural sinuses and meningeal blood vessels and by traction or distortion of various anchoring pain-sensitive structures because of a downward displacement of the brain caused by reduced hydrostatic force.^{5,7} Various other headache patterns, however, have also been suggested to be rare but possible presenting symptoms of SIH, including nonpostural headaches and the absence of headaches.¹⁻³ In the present case, our patient's headache had no orthostatic component and did not disturb his daily activities as long as he did not shake his head. Some authors postulate that atypical findings might result from a milder form of SIH or a compensatory response to typical SIH.^{3,8,9} Recently, in a case of SIH with nonpostural headache accompanied by subdural hematomas (SDH), SDH was postulated to prevent the development of orthostatic headache by correcting the abnormally low intracranial pressure or volume, in accordance with the Monro-Kellie doctrine.² What is unique in this case is that the headache occurred only with rapid oscillatory movement of the

head about either the horizontal or vertical axis. We hypothesized that this was a mild form of SIH, because clinical symptoms were not evident in specific postures or during usual activity, but were only triggered by specific actions. We suggest that the brain, suspended in a bath of CSF, tends to oscillate out of phase with the calvarium during head-shaking movements and consequently strains the meninges causing headache. These mechanical effects, including stretching of sensitive meningeal structures, can disrupt the established compensatory response and lead to a sudden rise in headache intensity. Another possible explanation for a head-shaking headache is that rapid oscillatory horizontal movement of the head forms "whirlpool" also known as a vortex, within the calvarium. The swirling motion tends to suck everything within the fluid toward its center, according to Bernoulli's principle.¹⁰ Therefore, we hypothesized that vortex formation during head-shaking pulls the brain downward and precipitates sagging of the intracranial structures, inducing headache.

Other supportive symptoms of SIH include stiff neck, nausea, tinnitus, hyperacusis, and photophobia.⁵⁻⁷ Our patient had left-sided tinnitus that persisted independent of head shaking. Vestibular and auditory symptoms may be caused by direct transmission of abnormal CSF pressure to perilymphatic fluid.⁷ It should be noted that a newly developed headache in middle age accompanied by minor neurologic symptoms such as tinnitus, which is unusual in primary headache, suggests the possibility of a secondary headache disorder.

Appropriate imaging studies are essential for the diagnosis of rare secondary headaches. Brain MRI with contrast enhancement is the initial investigation of choice in suspected SIH.¹¹ If brain MRI findings support SIH, radionuclide cisternography to identify a potential CSF leak should be considered, especially if symptoms persist despite conventional treatment. The characteristic radionuclide cisternographic signs of CSF leak were described as either direct or indirect sign at early and late images. Although direct signs of CSF leak were not found in our patient, radionuclide cisternography documented indirect sign of CSF leak (ie, 24-hour paucity of activity over cerebral convexi-

ties and early appearance of urinary bladder), that is sufficient to establish a diagnosis of CSF leakage.¹²

In summary, we reported a case of specific task-induced SIH. We suggest that SIH should be suspected in patients with only head-shaking headaches even without orthostatic features, especially when the headache is accompanied by other symptoms associated with SIH including tinnitus, as in our case. Moreover, performance of appropriate imaging studies to establish an accurate diagnosis should be considered in these situations.

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