Global Hemispheric CT Hypoperfusion May Differentiate Headache With Associated Neurological Deficits and Lymphocytosis From Acute Stroke

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Headache with associated neurological deficits and lymphocytosis (HaNDL) is characterized by temporary recurrent neurological deficits, moderate–severe headache, cerebrospinal fluid lymphocytosis, elevated protein, and increased opening pressure. Although CT and MRI should be normal, single photon emission CT may indicate focal hypoperfusion and electroencephalogram may reveal slowing or even epileptiform activity resolving once the patient is symptom-free (3 months). Catheter angiography also yields normal results but may trigger an acute neurological episode.

Because HaNDL is a benign, self-limited syndrome, it is important to differentiate it from cerebrovascular disease to avoid unnecessary interventions such as catheter angiography and thrombolysis. We present a case initially thought to be acute stroke in which the patient was considered for thrombolysis. CT perfusion changes atypical for stroke made stroke diagnosis questionable and thrombolysis was withheld.

Case Report
A 31-year-old man with hypertension, dyslipidemia, and sleep apnea (treated with continuous positive airway pressure) but no history of prior migraine, was brought into our emergency department by ambulance with suspected acute ischemic stroke. He had become aphasic at work so his coworkers called 911. On examination, he was globally aphasic without focal weakness or obvious sensory changes aberrant for a right-sided temporal lobe. He had no further neurological deficits, moderate–severe headache, cerebrospinal fluid lymphocytosis, elevated protein, and increased opening pressure.

Routine blood work, toxicology, and MRI, including diffusion-weighted imaging, MR angiography, and fluid-attenuated inversion recovery (Figure B), were normal. A lumbar puncture revealed elevated opening pressure of 270 mm H20, increased protein (834 mg/L), and pleocytosis (447×10⁹/mL) with 96% lymphocytes. Glucose and Gram’s stain/culture were normal as was a complete viral workup, including herpes simplex, West Nile, cytomegalovirus, syphilis, and HIV. An electroencephalogram revealed intermittent delta wave slowing and transient sharp waves in the left temporal lobe.

The patient was discharged on carbamazepine as a precaution for possible seizures. He had no further neurological episodes or headaches and returned to work. Repeat clinical examination, CT perfusion (see Figure C), electroencephalogram, and formal detailed neuropsychological testing all performed 4 to 7 months later were within normal limits. The carbamazepine was tapered and he remains symptom-free. A final diagnosis of HaNDL was given because his presentation met the diagnostic criteria in the absence of a more likely diagnosis.

Discussion
It is important for neurologists to be aware of HaNDL syndrome. The presentation of sudden-onset neurological signs and symptoms, particularly headache and associated neurological deficits, should be evaluated using the appropriate diagnostic tests to rule out cerebrovascular disease. Once those become normal, further evaluation is indicated to rule out other potential etiologies such as HaNDL.
deficits illustrates the clinical similarities between HaNDL and acute stroke. A plain CT may be unremarkable in both conditions. However, CT perfusion in acute stroke should reveal significant reductions in cerebral blood flow and possibly cerebral blood volume with a prolongation of mean transit time in an arterial territory. Ischemia of 3 vascular territories is unusual, but theoretically may be produced where an occluded proximal internal carotid artery supplies a fetal-type posterior communicating artery in the absence of an anterior communicating artery. Such an arrangement was excluded by CT angiography as was any vascular occlusion. Furthermore, hemiparesis would be expected with this distribution of ischemia. We speculate that our patient did not experience hemiparesis with such a large affected distribution because the degree of perfusion impairment (i.e., oligemia rather than ischemia) may not have reached a unique threshold sufficient to impair motor function. Furthermore, cortical spreading depression may affect a more focal area of the brain reflected by the temporal lobe changes seen on electroencephalogram despite a larger distribution of oligemia that was observed on perfusion imaging. This may explain why language, and not motor function, was impaired in this case.

Our findings corroborate those of Fuentes and colleagues who demonstrated globally reduced left hemispheric uptake on single photon emission CT during acute aphasia in a HaNDL case. Other reports describing more focal single photon emission CT changes after the acute event may have captured resolving perfusion deficits. Transcranial Doppler performed postevent in HaNDL has revealed decreased flow velocities in the ipsilateral middle cerebral artery. Taken together, the changes in blood flow and associated electroencephalogram activity further supports the hypothesis that a HaNDL neurological attack may represent a cortical spreading wave of depression with associated oligemia, presumed to be due to a viral or postviral etiology. Interestingly, perfusion is similarly decreased in migraine aura, although only within the occipital lobe.

To our knowledge, this is the first report of global hemispheric CT perfusion changes during an acute neurological attack of HaNDL. Given the benign, self-limited nature of this syndrome, such a finding in the absence of vascular occlusion may differentiate it from acute stroke avoiding unnecessary interventions. Compared with single photon emission CT, CT perfusion is a more accessible, available, and rapid form of blood flow assessment with less radiation exposure. We chose CT perfusion for the investigation of this case because it is routinely used at our center for the evaluation of suspected acute stroke due to its rapid acquisition and processing characteristics. However, it is likely that MR perfusion would have yielded similar hemispheric hypoperfusion findings. We conclude that CT perfusion may help differentiate stroke from the more benign HaNDL syndrome in the acute setting.

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Disclosures
None.

References

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