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Neuroimaging Highlight

MRI Findings in Transient Headache and Neurologic Deficits with Cerebrospinal Lymphocytosis Syndrome

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A healthy 18-year-old man presented with 1-day history of left arm incoordination and 5-day history of throbbing headache associated with photophobia and vomiting. There was a strong family history of migraine. Neurological examination findings were limited to impaired proprioception and cortical sensory loss in the left arm. CSF analysis showed 100 nucleated cells (98% lymphocytes) with mildly elevated protein at 0.5 g/L. He received initial empirical antimicrobial coverage for meningitis. MRI was remarkable for five punctate discrete cortical lesions with restricted diffusion in the right frontal, parietal, and posterior temporal regions suggestive of acute/early subacute infarctions with no abnormal enhancement (Figure 1). EEG showed slowing over the right hemisphere without epileptiform changes. Digital subtraction angiography showed no evidence of vasculitis. The CSF infectious workup was negative, as were ANA, hepatitis, and HIV serologies. Focal neurological signs resolved over the next 2 days, though headache persisted for a week. He was discharged with a diagnosis of headache and neurological deficits associated with cerebrospinal fluid lymphocytosis (HaNDL). He presented a week later with a milder exacerbation of migraine-like headache and right-sided subjective weakness and was successfully treated with ibuprofen and acetaminophen.

HaNDL was first described in 1981.[1] The ICHD-3 diagnostic criteria require episodes of migraine-like headache accompanied by focal deficits, CSF lymphocytic pleocytosis, self-resolution within 3 months, and no alternative explanation.[2] A case series of 50 patients reported onset between age 14 and 39 years with male predominance (68%).[3] Roughly, a third of these patients had a personal history of migraine.[3] There was a mean of three episodes per patient (range 1–12).[3] Some patients had contralateral hemispheric involvement in subsequent episodes similar to our patient.[3]

The pathophysiology of HaNDL remains uncertain but may involve spreading depression, neurogenic sterile inflammation, and transient cerebral vasomotor changes. This is supported by reports of focally reduced uptake in SPECT imaging,[3,4] abnormalities on transcranial Doppler[5], and focally increased mean transit time on CT perfusion.[6] MRI has mostly been reported as unrevealing in HaNDL.[3] There are single-case reports of subtle diffusion restriction in the symptomatic hemisphere[7] and gray matter swelling with minimal sulcal enhancement.[8] Our patient had several discrete lesions, widely separated over the symptomatic

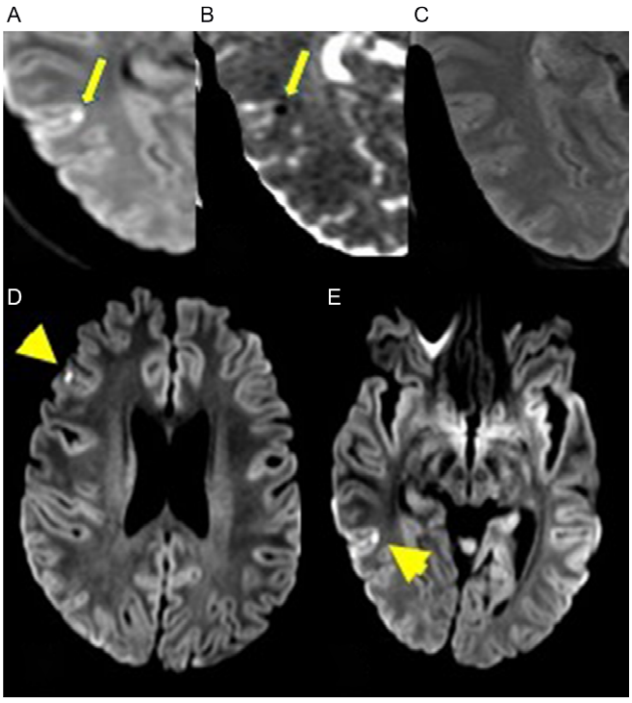

Figure 1: Brain MRI. Axial DWI (A) shows a punctate cortical hyperintense lesion with corresponding hypointensity on ADC (B) and subtle hyperintensity in FLAIR (C) compatible with ischemic lesion. Axial DWI images (D and E) show other punctate hyperintense lesions involving the cortex of the symptomatic hemisphere.

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hemispheric cortex. These were likely due to focal distal microvascular changes. Cerebral angiography ruled out arterial pathology in large or medium diameter branches. The occurrence of so many ipsilateral lesions would not be consistent with an embolic shower. In keeping with the minimal overall lesion burden, our patient had a very favorable evolution, as is the rule in HaNDL.

Though HaNDL has distinct features of CSF lymphocytosis and male preponderance, it likely belongs to the spectrum of migraine. A much broader range of at times striking MRI abnormalities has however been described in sporadic or familial hemiplegic migraine. These include chronic asymptomatic discrete subcortical hemispheric T2 lesions mimicking demyelination, venous dilation with sulcal enhancement, and posterior circulation-predominant infarctions. It is likely that the use of DWI and SWI in modern MRI protocols will lead to a wider detection of MRI abnormalities in both HaNDL and migraine.

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CT contributed to the imaging interpretation and editing of the manuscript.

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