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Acute Confusional Migraine associated with probable genetic vascular leukoencephalopathy presenting as a CADASIL phenotype: a case report

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Introduction
Acute confusional migraine is a rare cause of transient impairment of consciousness, characterized by defects in sensorium, impaired awareness, disattention, agitation, and amnesia. Although often reported in childhood, confusional migraine has rarely been associated with cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL). Diagnosis is made by excluding other more common and threatening causes of confusional states (e.g., epilepsy, stroke, neoplasm, intoxication, and encephalitis).

Objective
To present a case of acute confusional migraine in a patient with vascular leukoencephalopathy of probable genetic origin, evaluated in the neurology service of an academic hospital linked to the Universidade de Pernambuco.

Case report
A 43-year-old man presented with a history of probable migraine without aura since childhood, occurring with a frequency of four attacks per month. He had experienced confusion during more severe headache episodes for the past two years. During these episodes, he became disoriented, agitated, aggressive, and exhibited some inappropriate social behavior, which resolve with pain relief. The patient did not remember these episodes. One year before admission, following a mild Sars-CoV-2 infection, his wife observed progressive forgetfulness, affecting various aspects of his life, such as forgetting relatives, friends, objects, and work schedule, accompanied by psychomotor slowness, which significantly impacted his daily functioning. They sought psychiatry evaluation, leading to a diagnosis of depression and he was treated with sertraline.
Neurological examination revealed apathy, fluent and disoriented speech, a score of 12/30 points in the mini- mental status examination, global bradykinesia and mild rigidity with cogwheel phenomenon, and gegenhalten in his wrists, mild dysmetria, and a wide-based parkinsonian gait. Brain magnetic resonance imaging showed bilateral hyperintensities on T2-weighted/FLAIR sequences in the periventricular white matter, corona radiata, temporal POLES, and external capsule, with preservation of the subcortical and U- shaped fibers, without evidence of diffusion restriction or contrast enhancement. There were also focal hyperintensities in the external capsules and thalamus, consistent with prior ischemic insults, although brain and neck vascular studies and echocardiogram were unremarkable. Cerebrospinal fluid analysis revealed an elevated protein level (87mg/dL) and normal cell count. The electroencephalogram demonstrated diffuse slowing of basal activity, without epileptic activity. Inflammatory, autoimmunity, and infectious investigations were negative. The set of findings were consistent with vascular leukoencephalopathy of probable genetic origin (CADASIL), presentig as episodes of confusional migraine and evolving into subcortical dementia signs. Genetic testing or skin biopsy were unavailable. The patient was prescribed aspirin and clopidogrel, along with low doses of quetiapine, and amitriptyline, in addition to language and physical rehabilitation. He responded well to treatment and was referred to the cognition and neurovascular outpatient clinic.

Conclusion
We present an unusual case of an adult man with acute confusional migraine and vascular leukoencephalopathy exhibiting a CADASIL phenotype.

Keywords: Migraine without Aura; Acute Confusional Migraine; Subcortical Leukoencephalopathy; CADASIL.