Case report: migraine with aura in patient with cadasil

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Introduction
Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is the most frequent hereditary cause of brain ischemic small vessel disease (1). Migraine with aura (MA) is typically the presenting and most common clinical feature of CADASIL. The estimated prevalence of MA in CADASIL ranges from 20 to 40% (2).

Case Report
43-year-old woman, with previous history of hypertension, diabetes, and episodic migraine with visual aura, suffered three episodes of hemihyposthesia in 2016, 2017 and 2018 (one on her right and the other two on her left). In the last of these episodes, she reported worsening of the visual acuity of her right eye, and developed chronic migraine. Brain MRI showed extensive areas of confluent T2 hyperintensity in the white matter, as well as in the nucleocapsular and bilateral thalamic regions. In all three episodes the patient was treated with IV or oral corticosteroids and had partial improvement of the symptoms. The patient had familiar history of relatives who had suffered multiple strokes.

Discussion
The diagnosis of multiple sclerosis was firstly considered due to the evolution in clinical attacks and the response to corticotherapy. However, the patient’s family history, MRI findings and previous diagnosis of migraine with visual aura lead to the suspicion of CADASIL. This diagnosis was ultimately confirmed through genetic testing that showed C> T variation in NOTCH3 gene. Migraine was successfully treated with greater occipital nerve blocks and topiramate.

Final Comment
CADASIL stroke-like attacks remain a therapeutic challenge. It is possible that corticosteroid treatment may benefit these patients by reducing the inflammatory process that results from blood-brain barrier breakdown. More studies are needed to evaluate the efficacy and safety of corticotherapy in this population.

Keywords: CADASIL, migraine, aura, Leukoencephalopathy