Anterior temporal lobe involvement: Useful magnetic resonance imaging sign to diagnose Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy

Prasanna Venkatesan Eswaradass, Balakrishnan Ramasamy, Ramadoss Kalidoss, Gnanagurusamy Gnanashanmugham

Department of Neurology, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India

We report a 32-year-old male who was admitted with speech disturbance. The patient presented to us with confusion, headache, and urinary incontinence. On examination, patient had no deficits except for aphasia. We proceeded with investigations. Magnetic resonance imaging (MRI) brain revealed extensive white matter ischemia and acute infarcts in both centrum semiovale regions. Fluid-attenuated inversion recovery (FLAIR) images showed anterior temporal lobe and external capsule involvement bilaterally [Figures 1 and 2]. Cerebrospinal fluid analysis done to rule out encephalitis was normal. After the investigations, we reviewed the history and found that the patient had recurrent episodes of migraine for several years, and his mother and maternal uncle had repeated episodes of the stroke leading to dementia. We suspected the inherited cause for stroke. We tested the blood for NOTCH3 gene mutation. It was sequenced from exons 2, 3, 4, 5, and 6 and patient was found to be heterozygous for p.C144s mutation confirming the diagnosis of Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL).

CADASIL is an acronym for Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy. CADASIL is a hereditary disease of small vessels predominantly affecting middle-aged individuals. It has been reported in nearly 500 families, but the actual prevalence is unknown. [1] The most frequent and earliest abnormalities are a hyperintense signal on T2-weighted/FLAIR appearing as punctiform lesions, in periventricular areas and in the centrum semiovale, which later become diffuse and symmetrical. Lacunar infarcts of basal ganglia, thalamus, pons and cerebral microbleseds are also seen frequently. [2] Involvement of the anterior part of the temporal lobes and the external capsule is highly suggestive of CADASIL. According to Markus et al., anterior temporal lobe involvement has

Address for correspondence:
Dr. Prasanna Venkatesan Eswaradass,
Department of Neurology, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.
E-mail: eprasanna2k1@gmail.com

For reprints contact: reprints@medknow.com

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

How to cite this article: Eswaradass PV, Ramasamy B, Kalidoss R, Gnanashanmugham G. Anterior temporal lobe involvement: Useful magnetic resonance imaging sign to diagnose Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy. J Neurosci Rural Pract 2015;6:622-3.
Eswaradass, et al.: Anterior temporal lobe involvement in CADASIL

The gold standard for diagnosis of CADASIL is genetic analysis, but it is expensive and may be false negative if only a cluster of most probably affected exons is examined. MRI involvement of the anterior temporal lobes is a useful sign to suspect CADASIL in patients with an appropriate history and clinical findings. However, this could be false positive, and CADASIL is a diagnosis of exclusion in the absence of genetic testing.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**References**