

# Multifocal Cerebral and Bilateral Middle Cerebellar Peduncle Infarctions in CADASIL

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A premorbidly high functioning 46-year old male with NOTCH3 confirmed cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) presented with a four-day history of abrupt cognitive decline with associated hypersomnolence, emotional lability and a mild gait ataxia. A magnetic resonance imaging scan (MRI) on admission showed multi-focal diffusion weighted imaging lesions that were associated with reduced apparent diffusion coefficient in white matter of both cerebral hemispheres and middle cerebellar peduncles, characteristic of acute infarcts (Figure 1). Acute multi focal ischemic stroke has been observed to occur in CADASIL<sup>1,2</sup> but, to our knowledge, such a rapidly diffuse case of ischemic encephalopathy involving middle cerebellar peduncles has not been previously reported in CADASIL. Typical neuroradiological findings of CADASIL on MRI are multifocal and bilateral FLAIR/T2 hyperintensities in the periventricular and deep white matter. The lesions are mainly in the temporal pole, frontal and parietal lobes, external capsule, pons and basal ganglia.<sup>3</sup> In our case, neuroimaging with MRI confirmed acute DWI lesions in deep white matter of the cerebrum. This case is a rare presentation of ischemic encephalopathy in a case of genetically confirmed CADASIL. This case strikingly demonstrates the acute simultaneous multifocal location of diffusion weighted lesions involving the white matter tracts of the cerebrum and, uncharacteristically, the cerebello-pontine connections, which has not been previously reported. The differential diagnosis for the imaging pattern might

include proximal embolic shower, hypoglycaemia and other rare metabolic conditions.<sup>4</sup> However, the location of the stroke, specifically involving the white matter, negative diagnostic cardiac tests, and a previous acute presentation of subcortical stroke in a man known to have the NOTCH 3 mutation for CADASIL supported our conclusions that this unusual presentation was related to the underlying genetic mutation.

Common clinical features of CADASIL include migraine with aura, transient ischemic attacks (TIAs) and fixed focal neurological deficits caused by lacunar infarctions, and cognitive decline of subcortical type.<sup>3</sup> Though many patients found to have CADASIL first present with ischemic stroke, to our knowledge, six cases of CADASIL presenting acutely with multiple simultaneous infarctions have been reported to date. Clinical features of these reported cases had a large heterogeneity, with their symptoms dependent on the location of their subcortical strokes.<sup>1,5</sup> In our case, the patient's bilateral middle cerebellar peduncle infarcts would explain his gait ataxia. Acute stroke typically presents with focal neurological deficits, but abrupt changes in executive function can be a rare and under recognized presentation. The acute bilateral subcortical lesions seen in our case likely contribute to the profound slowing of processing speed and executive dysfunction, in addition to perhaps accounting for the patient's hypersomnolence. Acute cognitive decline with CADASIL is extremely rare; generally the cognitive decline is gradual and progressive.<sup>3,6</sup> Deterioration presenting with a rapidly progressive diffuse ischemic form is a striking feature of this case.

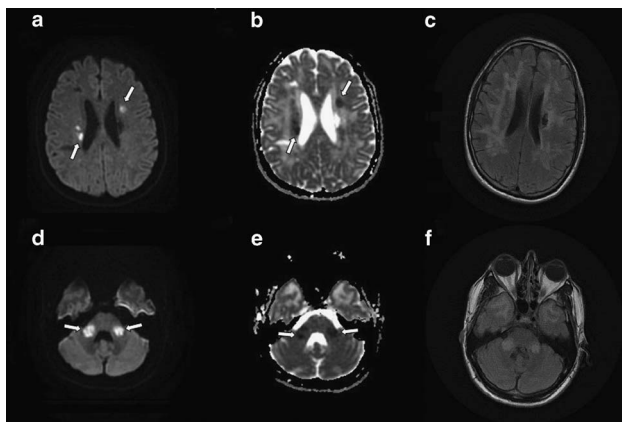
## STATEMENT OF AUTHORSHIP

Kelvin Au - Manuscript draft, data acquisition. Ramana Appireddy - Manuscript revision and analysis and interpretation of MRI findings.

Dr. Philip Barber - Study supervision and manuscript revision, data interpretation of MRI findings.

## DISCLOSURES

Kelvin Au, Ramana Appireddy, and Phil Barber do not have anything to disclose.



**Figure 1:** Multiple diffusion hyperintensities in periventricular white matter with corresponding low ADC values (a,b) and T2 FLAIR images (c). Bilateral middle cerebellar peduncle restricted diffusion lesions (d,e) with T2 FLAIR images (f).

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