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Short communication

Acute vestibular syndrome in a patient with cerebral autosomal dominant leukoencephalopathy with subcortical infarcts and leukoencephalopathy (CADASIL)

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Abstract

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is a hereditary form of small vessel disease in which the pons may show lacunar infarcts and leukoaraiosis. Acute pure vestibular syndrome may be due to caudal pontine lesions and is probably underestimated. We describe a case of CADASIL with acute vestibular syndrome mimicking peripheral vestibulopathy, and evidence of focal infarction in the ponto-medullary junction at gadolinium-enhanced MRI including diffusion-weighted imaging, involving the area of the right vestibular nucleus and root entry zone of the ipsilateral vestibular nerve bundle. In CADASIL, both focal brainstem lesions and leukoaraiosis may parallel supratentorial white matter changes and may be related to poor outcome. Their actual extent should be evaluated in longitudinal studies that might predict clinical outcome and progression of disability.

Keywords: CADASIL; Acute peripheral vestibular syndrome; Lacunar infarcts; Pons

1. Introduction

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is a monogenic form of small vessels disease [1], characterized clinically by recurrent stroke, progressive cognitive impairment and MRI evidence of lacunar infarcts and diffuse subcortical white matter (WM) changes also known as leukoaraiosis [2].

Although the brainstem is often affected, the real extent and prognostic relevance of its involvement has never been investigated in detail in the disease.

Large brainstem infarctions are uncommon, however both lacunar infarctions and leukoaraiosis resulting from damage to perforating arteries arising from branches of the vertebral arteries, is frequently seen, and well documented by MRI [3] in subjects with CADASIL.

In this respect, besides micro-lacunar infarcts, the central pons is particularly susceptible to chronic hypoperfusion related to small vessel disease, as confluent areas of myelin rarefaction are also reported in this region in patients with supratentorial ischemic stroke and leukoaraiosis [4].

Here, we report a patient with genetically confirmed CADASIL and clinical history of recurrent strokes, who complained of acute vestibular syndrome (AVS), mimicking peripheral vestibulopathy, associated with MRI evidence of focal infarction at the ponto-medullary junction, involving the area of the right vestibular nucleus and/or the root entry zone of the ipsilateral vestibular nerve bundle. Additional oculomotor dysfunctions included, horizontal gaze-evoked nystagmus and saccadic pursuit. Further neurological abnormalities which had been occurred earlier and

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