

Patient with vertigo, dizziness and depression

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SUBMISSION: 14/6/2017 | ACCEPTANCE: 19/3/2018

PART A

A 54-year-old woman presented with dizziness and vertigo as an outpatient at the Ear-Nose-Throat (ENT) Clinic. The patient claimed transient numbness in both hands and exhibited mild depression. Clinical examination was unremarkable. Imaging workup included a temporal bone

and brain magnetic resonance imaging (MRI). Temporal bone imaging did not disclose any abnormality. Axial FLAIR (**Fig. 1a-c**), sagittal FLAIR (**Fig. 1d**), coronal T2-W (**Fig. 1e**) and axial susceptibility weighted imaging (SWI) (**Fig. 1f**) MRI sequences are shown.



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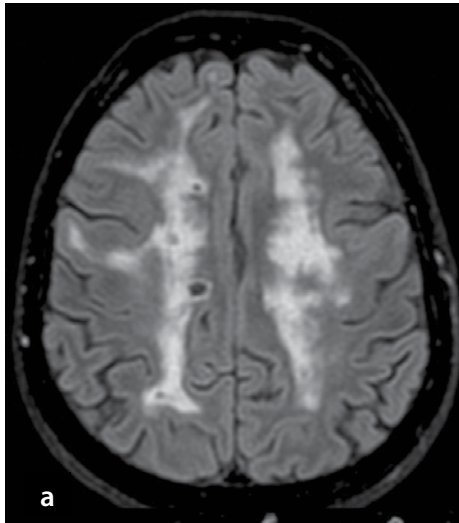
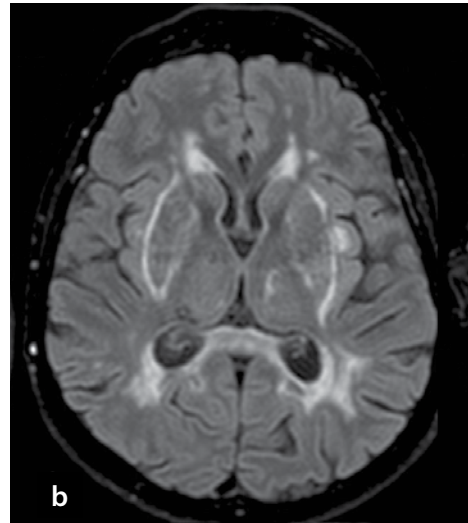
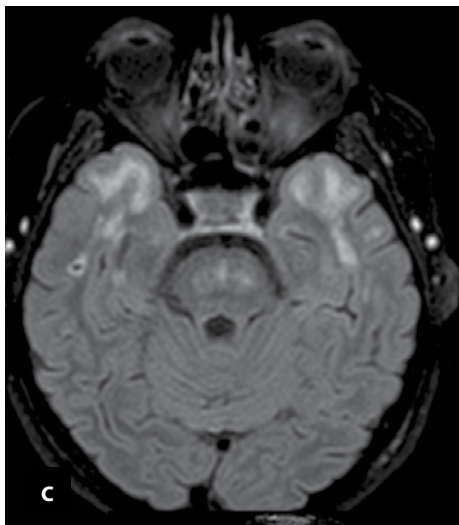


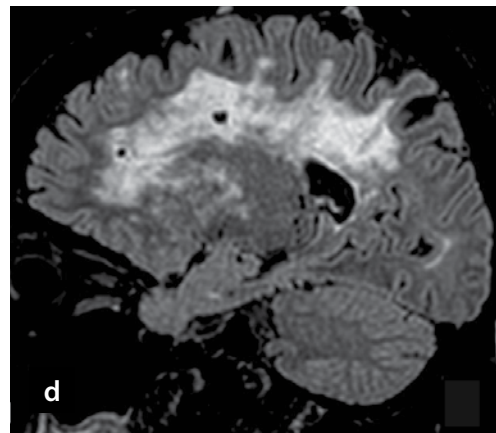
Fig. 1. Brain MRI. a. Axial FLAIR



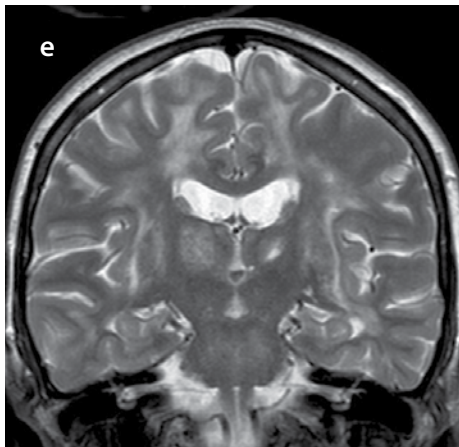
b. Axial FLAIR



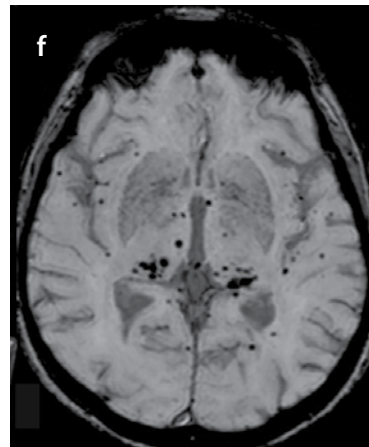
c. Axial FLAIR



d. Sagittal FLAIR,



e. Coronal T2-W



f. Axial susceptibility weighted imaging (SWI).

PART B

PART B

Diagnosis: Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)

CADASIL is a hereditary disease and belongs to the wide spectrum of Cerebral Small Vessel Diseases (CSVD). CSVD refers to a group of pathological processes with various aetiologies, affecting small arteries, arterioles, venules and capillaries of the brain. Most common predisposing factors are cerebral amyloid angiopathy, hypertension and aging [1]. Major clinical consequences of CSVDs are stroke and vascular dementia [2]. Genetic factors have a well-established impact in CSVD aetiology. Of various hereditary aetiopathologic mechanisms, CADASIL and Fabry's disease are among the most prominent and could be used as models for CSVDs' pathogenesis understanding [1]. Specifically, CADASIL is an inherited non-arteriosclerotic and amyloid-negative small-vessel disease with autosomal-dominant transmission related to a mutation of the NOTCH3 gene on chromosome 19 [3]. CADASIL is the most frequent monogenic SVD in European adults, with a prevalence of 1.3 to 4/100 000 [4]. In this condition, there is deposition predominantly of granular osmiophilic material in the walls of small arterioles of the brain [4]. These pathological changes in the small vessels may result in both ischaemic and haemorrhagic events. Imaging findings include white matter hyperintensities, that are characteristically distributed at the anterior temporal lobe, the external capsule and the superior frontal gyrus [3, 5]. The condition should be differentiated from hypertensive leukoencephalopathy, normal aging brain, amyloid angiopathy, where the characteristic distribution of lesions is lacking.

Patients with CADASIL disease may present with recurrent subcortical ischaemic attacks, migraine with aura, cognitive decline or mood disorders. Our patient was initially investigated for vertigo, thus imaging focused on the cerebellopontine angle and the inner auditory canal. Vertigo and dizziness might have a vascular background. Clinical examination revealed depression, a mood disorder that is usually related with CADASIL. Imaging pattern of the white matter involvement illustrated the extensive leukoencephalopathy associated with lacunar infarcts, indicating a severe CSVD, while the anatomic distribution was characteristic for CADASIL (Fig. 1a-d). As with other CSVDs, similar lesions are also common in the brain stem (Fig. 1e). As microbleeds are not unusual in these conditions, our patient manifested many small spots of micro-haemorrhage, revealed in the SWI sequence (Fig. 1f).

No specific treatment has yet been proposed for CADASIL and there are no data to support any of the evidence-based treatments for stroke (aspirin, thrombolysis, admission to a stroke unit) [1]. Although thrombolysis may have implications because of the haemorrhagic risk in CADASIL, it cannot be considered as a contraindication for treatment [5]. In the chronic phase, the use of aspirin for secondary prevention for strokes in older patients is common, but there is no evidence for or against its use. Whether this strategy is appropriate in CADASIL is undetermined and will require further investigation, given the possible increased haemorrhagic risk [6]. **R**

Conflict of interest

The authors declared no conflicts of interest.



KEY WORDS

Small Vessel Disease (SVD); CADASIL; Brain; MRI

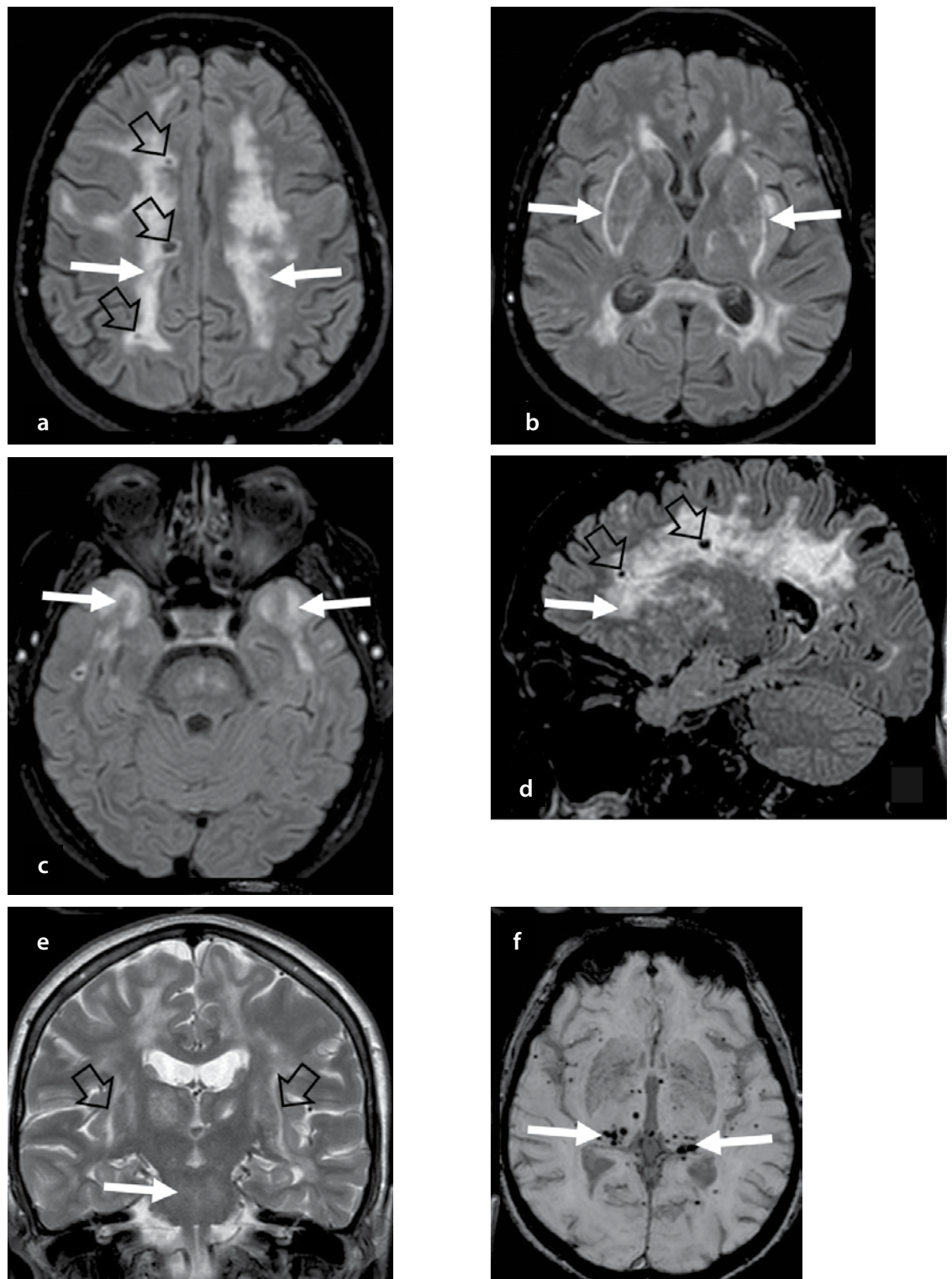


Fig. 1. **a.** Axial FLAIR image at the corona radiata level shows extensive confluent white matter lesions in both brain hemispheres (white arrows) along with several chronic subcortical infarcts, shown as areas with cerebrospinal fluid signal intensity (black open arrows). **b.** Axial FLAIR image at the basal ganglia level illustrates the striking involvement of both external capsules (white arrows). **c.** Axial FLAIR image shows increased signal intensity of the temporal lobes (white arrows). **d.** Parasagittal FLAIR image demarcates the significant extension of white matter disease (white arrow). Several lacunar infarcts are present (black open arrows). **e.** Coronal T2WI depicts brain stem lesions as high signal intensities in the central pons (white arrow) and extensive caudomeatal involvement of the external capsule (open black arrows). **f.** Axial SWI shows several punctuate spots with low signal (white arrows), representing microbleeds, not shown in other sequences.

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READY-MADE
CITATION

Mantatzis M, Argyropoulou P, Prassopoulos P. Patient with vertigo, dizziness and depression. *Hell J Radiol* 2018; 3(3): 60-64.