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Longitudinally extensive transverse myelopathy in a patient with CADASIL

Mielopatia transversa longitudinalmente extensa em um paciente com CADASIL

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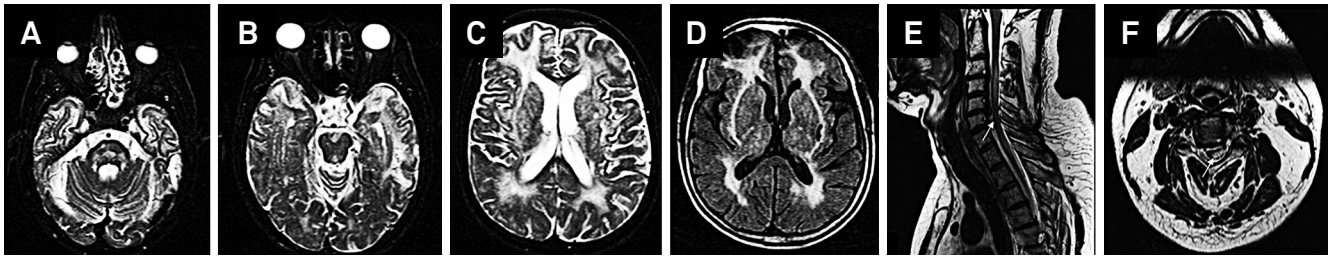
A 52-year-old Brazilian man presented with a 3-year-history of progressive cognitive decline, seizures and tetraparesis with sphincter disturbances. Familial history was positive for stroke-like episodes. Examination disclosed spastic tetraparesis with posterior cord syndrome. Neuroimaging revealed a longitudinally extensive centromedullary hyperintensity in the spinal cord between C2-T1 levels and a diffuse leukoencephalopathy (Figure), highly suggestive of Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL).

CADASIL is a common hereditary vasculopathy in adults characterized by chronic migraine, subcortical infarcts and cognitive and behavioral disturbances[1,2]. Spinal cord involvement is extremely rare, making this a differential diagnosis of demyelinating diseases[2,3].



Axial T2-weighted brain MR images (A-C) and FLAIR image (D) disclosing diffuse hyperintense signal changes in the white matter, involving the external capsule, anterior temporal lobes and pons. (E-F) T2-weighted spine MRI disclosing longitudinally extensive signal change in the cervical spinal cord level (white arrow).

Figure. Spinal cord involvement in CADASIL.

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