Cerebral amyloid angiopathy presenting as recurrent superficial siderosis

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Introduction: Cerebral amyloid angiopathy (CAA) is characterized by the deposition of β-amyloid in cortical and leptomeningeal vessels where intracerebral macro and microhaemorrhages are the most frequent presentations. Recently, superficial siderosis (SS) emerged as a possible manifestation of CAA. The association of CAA with Alzheimer’s disease (AD) has also been increasingly recognized.

Case Report: We report a case of a 67-year-old woman with a previous hospitalization in 2013 due to cerebral lenticulo-caudate haemorrhage. Both cerebral magnetic resonance imaging (MRI) and MR-angiography were unremarkable. In September 2016, she presented with sensory deficits, diagnosed as subarachnoid haemorrhage (SAH) and later discharged without neurological deficits. In March 2017, she returned to the emergency department with dizziness and persistent vomiting. Once again, the brain computed tomography revealed SAH. Brain and spinal cord MRI revealed deposition of hemosiderin in cerebellar and hemispheric sulci. Conventional cerebral angiography excluded aneurysmal malformation and Pittsburgh compound B positron emission tomography showed a moderate increase of β-amyloid deposition. Cerebrospinal fluid evaluation identified a decrease of β-amyloid and an increase of both Tau and phospho-Tau levels. The neuropsychological assessment emphasized a marked defect of the interfered, immediate, semantic and visual verbal memories with moderate impairment of associative verbal memory and a low verbal initiative. A diagnosis of cerebral amyloid angiopathy was proposed.

Conclusion: Our patient presented with an lenticulo-caudate haemorrhage and recurrent SS associated with cognitive impairment due to CAA. With this case, we aim to demonstrate that SS can be an important indicator of CAA and subsequent cognitive impairment due to AD.