Susac syndrome

Fernando Freua¹, Leandro Tavares Lucato², Flávio Villela³, Getúlio Daré Rabello⁴,⁵

Susac syndrome is a rare autoimmune endotheliopathy caused by a microangiopathic occlusive arteriolar disease¹ characterized by the triad of encephalopathy, visual loss and sensorineural hearing loss².

A 41-year-old man presented with migraine-like headache, transient visual loss and tinnitus. Brain MRI showed small scattered FLAIR-hyperintense lesions involving the corpus callosum, centrum semiovale and periventricular white matter bilaterally (Figures 1 and 2). Some of them presented restricted diffusion (Figure 3). CSF analysis revealed 6.0 cells/mm³, a lymphomononuclear pattern, and elevated protein levels with an increase in gamma fraction (18.35%). Angiofluorescein study showed retinian vasculitis (Figure 4). He was treated with immunoglobulin and methylprednisolone pulse⁵, presenting good results.

¹Departamento de Neurogenética, Faculdade de Medicina, Universidade de São Paulo, Sao Paulo SP, Brazil; ²Departamento de Neuroradiologia, Faculdade de Medicina, Universidade de São Paulo, Sao Paulo SP, Brazil; ³Setor de Córnea e Doenças Externas, Departamento de Oftalmologia, Faculdade de Medicina, Universidade de São Paulo, São Paulo SP, Brazil; ⁴Departamento de Neurologia, Faculdade de Medicina, Universidade de São Paulo, Sao Paulo SP, Brazil; ⁵Departamento de Neurologia, Hospital Samaritano, Sao Paulo SP, Brazil.

Correspondence: Fernando Freua; Rua Mato Grosso, 306 Cj 1601; 01239-040 São Paulo SP, Brasil; E-mail: fernando.freua@gmail.com

Conflict of interest: There is no conflict of interest to declare.

Received 27 May 2014; Received in final form 12 June 2014; Accepted 02 July 2014.
Figure 4. Fluorescein angiograms showing hemorrhages, staining of the vessel walls and leakage.

References