

Susac syndrome

Síndrome de Susac

Fernando Freua¹, Leandro Tavares Lucato², Flávio Villela³, Getúlio Daré Rabello^{4,5}

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 pulse³, presenting good results.

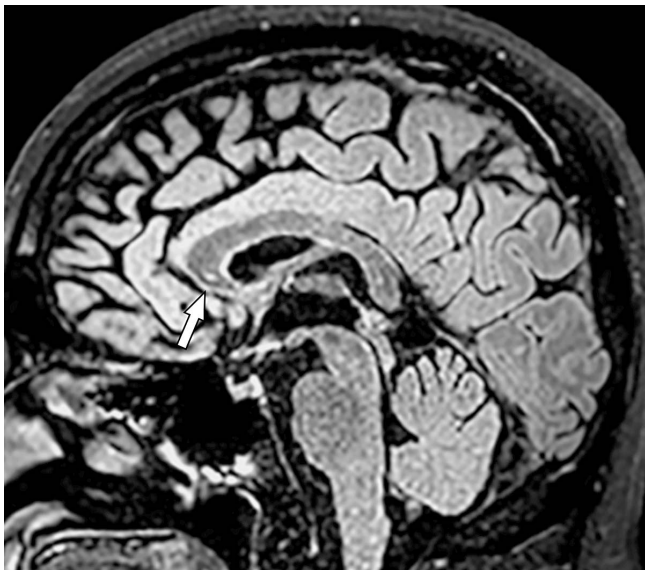


Figure 1. Sagittal FLAIR image discloses a small hyperintense lesion in the genu of the corpus callosum (arrow).

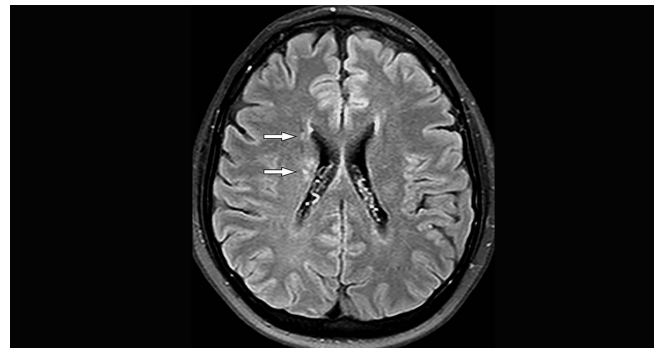


Figure 2. Axial FLAIR image shows some tiny scattered hyperintense lesions, more evident in the right cerebral periventricular white matter (arrows).

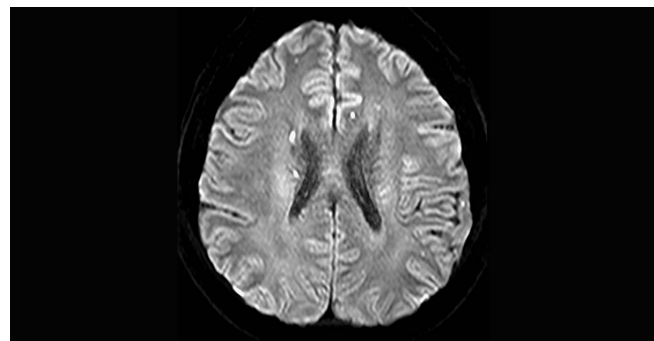


Figure 3. Axial diffusion-weighted image demonstrates restricted diffusion in some of the lesions, characterized by hyperintensity in this sequence.

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

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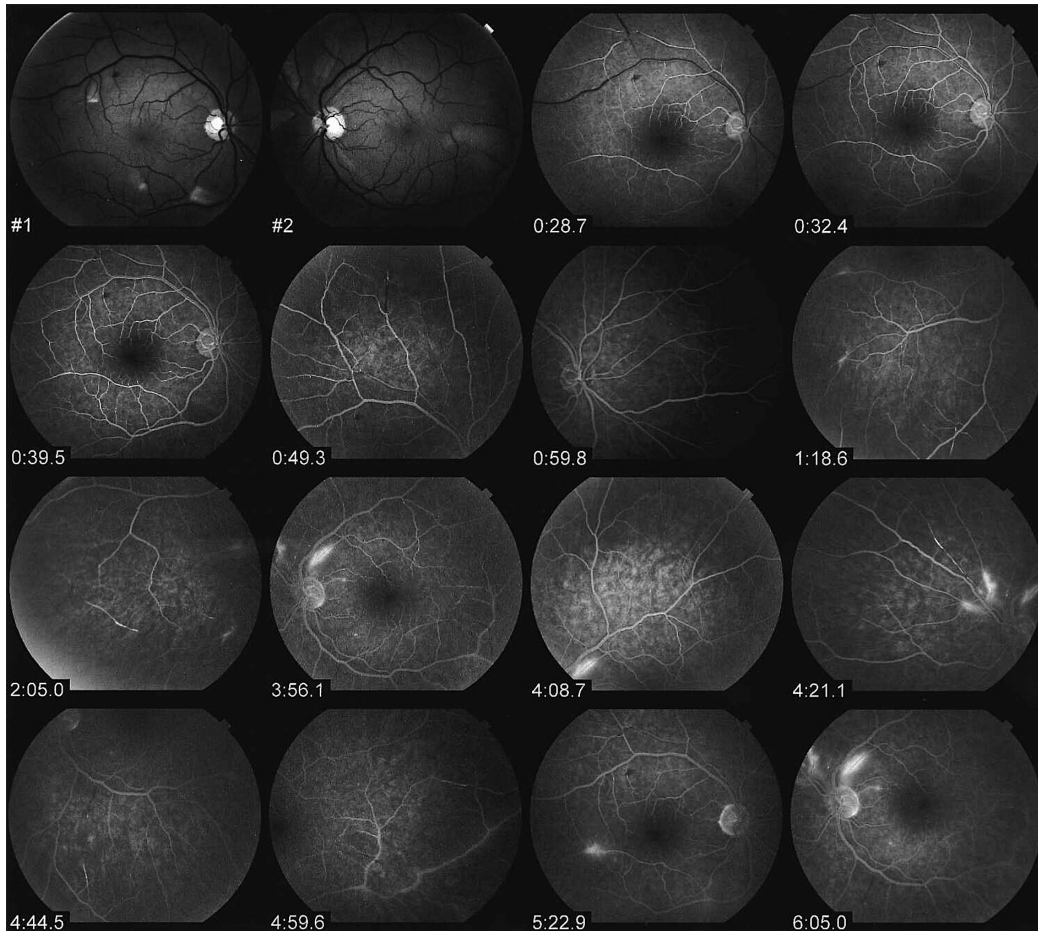


Figure 4. Fluorescein angiograms showing hemorrhages, staining of the vessel walls and leakage.

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