Teaching Video NeuroImages: Susac syndrome's acute onset callosal disconnection

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Figure 1 T1-weighted sagittal brain MRI

Punched-out holes through the genu (A), corpus (A, B), and splenium (B) of the corpus callosum (yellow arrows). Remarkably, the calloso-septal interface is spared. Although some authors consider this finding pathognomonic of Susac syndrome, Marchiafava-Bignami disease could be a differential diagnosis.

A 59-year-old right-handed hypertensive diabetic and previously alcoholic man presented acute confusional state followed by apathy, dysexecutive syndrome, clumsy left hand, and apraxic gait. A year later, his wife noticed impaired hearing. Neurologic examination revealed marked callosal apraxia (nondominant limb ideomotor apraxia, disconnection variant) and left stereoagnosia due to callosal disconnection (video). Left hand agraphia was present without aphasia. MRI showed punched out holes through corpus callosum, sparing the calloso-septal interface (figure 1). Audiometry revealed left neurosensorial loss. Fluorescein retinography demonstrated hyperfluorescence of arterial vessel wall (figure 2), confirming the hypothesis of Susac syndrome.¹

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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Figure 2 Retinal fluorescein angiography

Hyperfluorescence of arterial vessel wall (yellow circle). This examination should be performed whenever there is suspicion of Susac syndrome because patients may not have visual symptoms. Follow-up may be necessary: at disease onset, only 13% of patients have the Susac triad¹ (encephalopathy,² neurosensorial loss, and retinal endotheliopathy).

Appendix Authors

Name	Location	Contribution
Lucas Oliveira Mourão, MD	University of São Paulo	Primary clinical care of the patient, caption and video edition, literature review, drafting and revision of the manuscript
Jacy Bezerra Parmera, MD	University of São Paulo	Primary clinical care of the patient and revision of the manuscript
Eduardo de Novaes Costa Bergamaschi, MD	University of São Paulo	Primary clinical care of the patient and revision of the manuscript
Douglas Mendes Nunes, MD	University of São Paulo	Neuroimaging revision
Ricardo Nitrini, MD, PhD	University of São Paulo	Data drafting and revision of the manuscript
Sonia M.D. Brucki, MD, PhD	University of São Paulo	Data drafting and revision of the manuscript

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