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

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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 stereoaagnosis 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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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.

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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 triad (encephalopathy, neurosensorial loss, and retinal endotheliopathy).

References
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