

# A Trigeminal involvement as unusual feature in a patient with Susac Syndrome



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**CASE REPORT:** a 56 years old woman was admitted to our Hospital because of the recent onset of left facial hypoesthesia, memory disturbances and personality changes. Her medical history included migraine, dizziness and bilateral sensorineural hearing loss. She had received antiplatelet therapy for an Antiphospholipid Syndrome diagnosed on the basis of mild autoantibodies titers and aspecific vascular lesions on brain Magnetic Resonance Imaging (MRI). During hospitalisation she referred photopsias and visual scotoma, so she performed a fluorescein angiography that showed branch retinal arteries occlusion in her left eye (Fig1). She repeated brain MRI that showed central callosal "holes" (Fig 5), multiple hyperintense foci in deep white matter ( Fig 4), external capsules, brainstem and a left trigeminal nerve root enhancement (Fig 3). Routine laboratory tests, cerebrospinal fluid, immunological and cancer screening were normal. Mild elevation of antiphospholipid antibodies and Factor VIII was found, indicating an endothelium damage. Brain MRI, fluorescein angiography and audiometry (Fig. 2) were highly suggestive for a Susac Syndrome (SS). The patient was therefore treated with Methylprednisolone 1g/die followed by Prednisone 25 mg/die tapered in 15 days with resolution of hypoesthesia. Visual disturbances and cognitive symptoms mildly improved whereas hypoacusia remained.

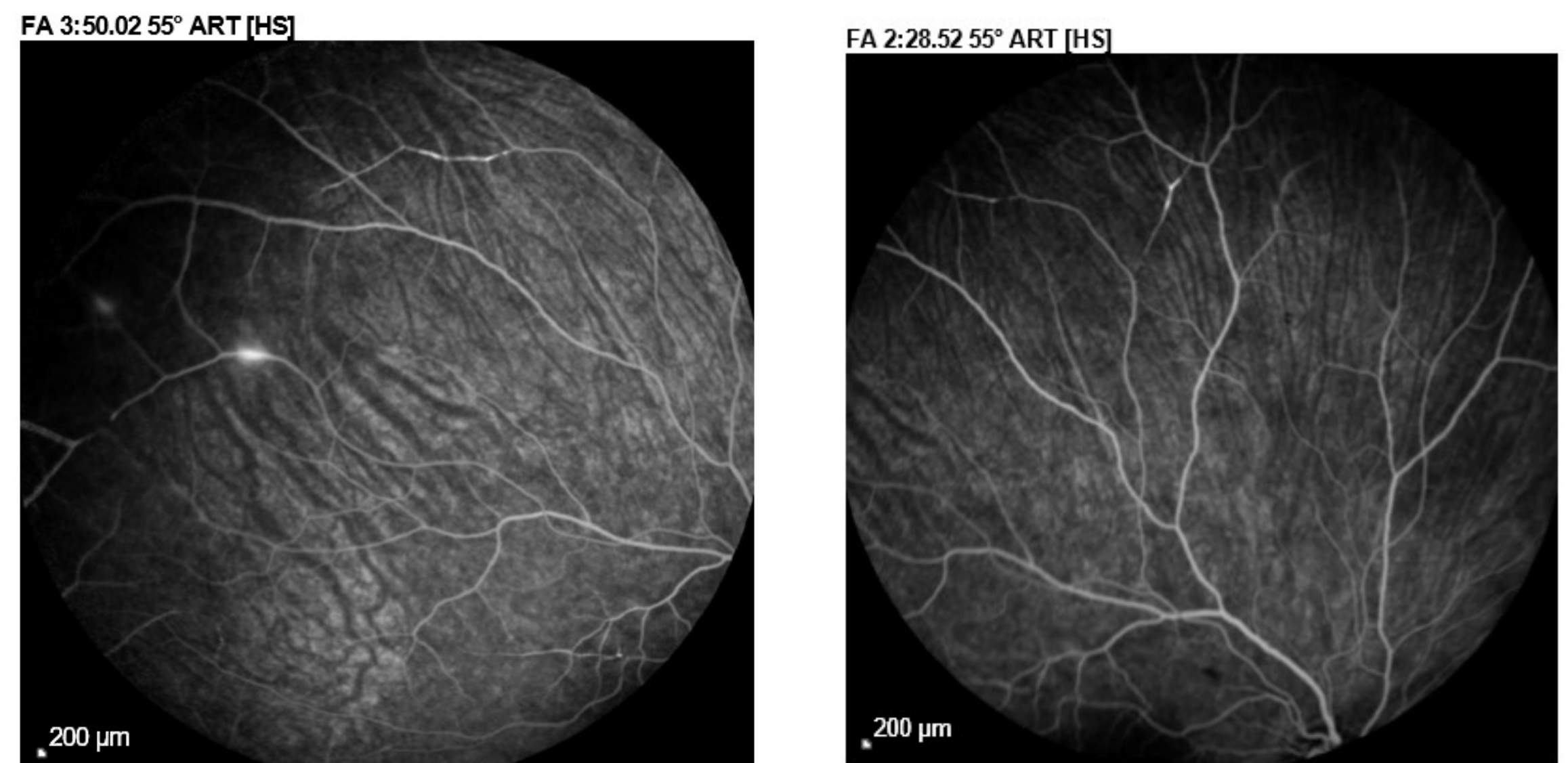


Fig 1 Fluorescein angiography that showed branch retinal arteries occlusion in left eye

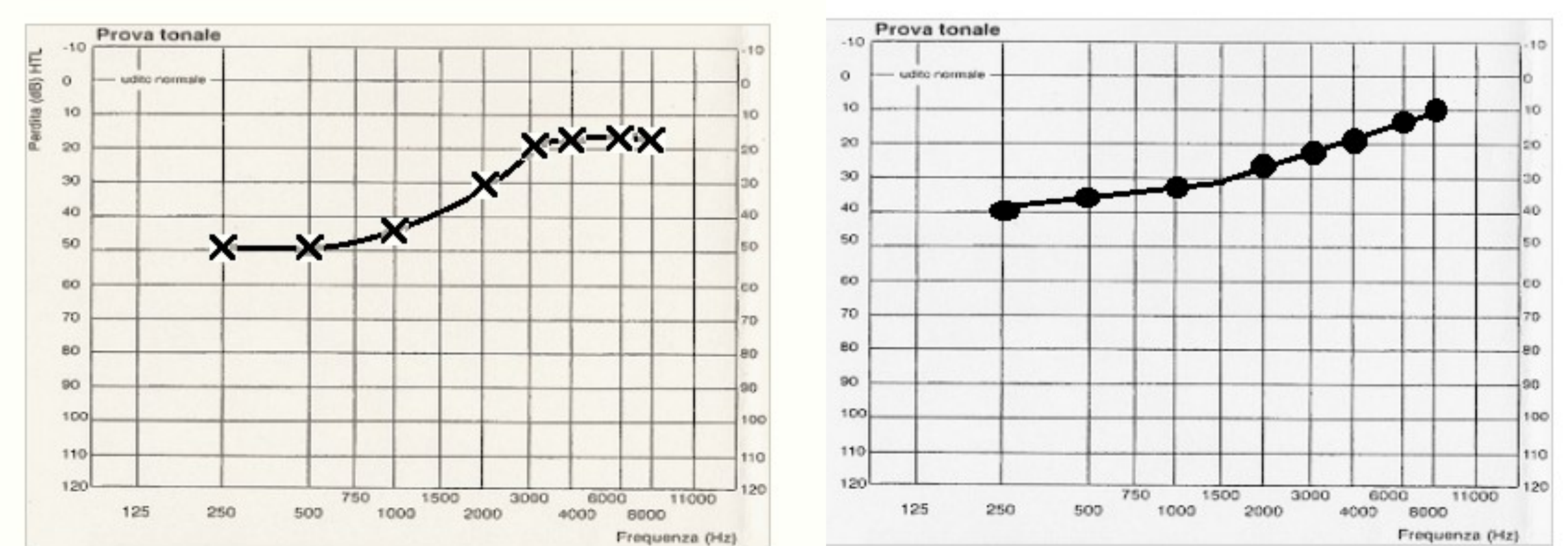


Fig 2 Audiometry : sensorineural hearing loss for mid-low-in left ear (A) and in right ear (B)

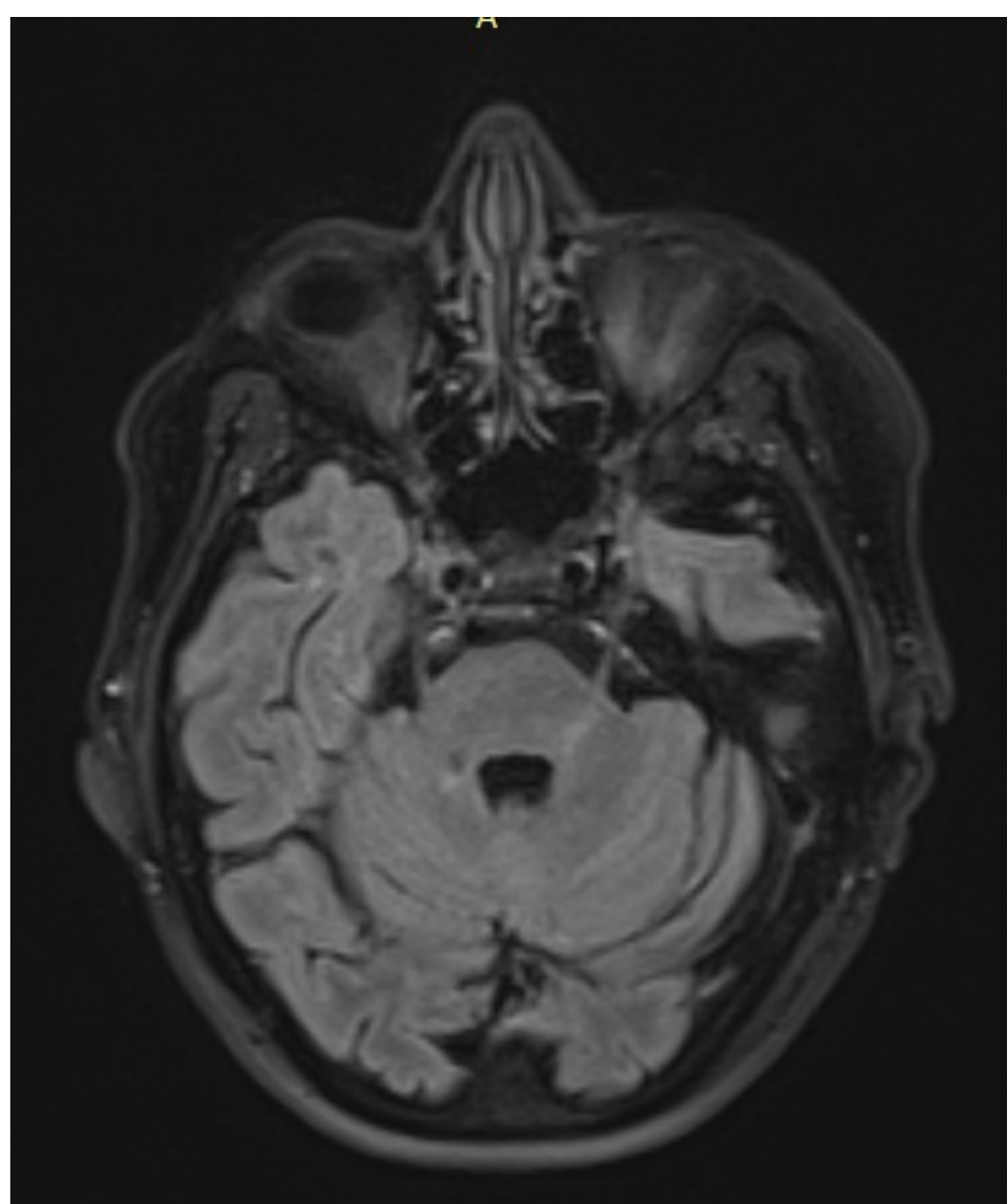


Fig 3 T2 FLAIR  
Left trigeminal nerve root hyperintense

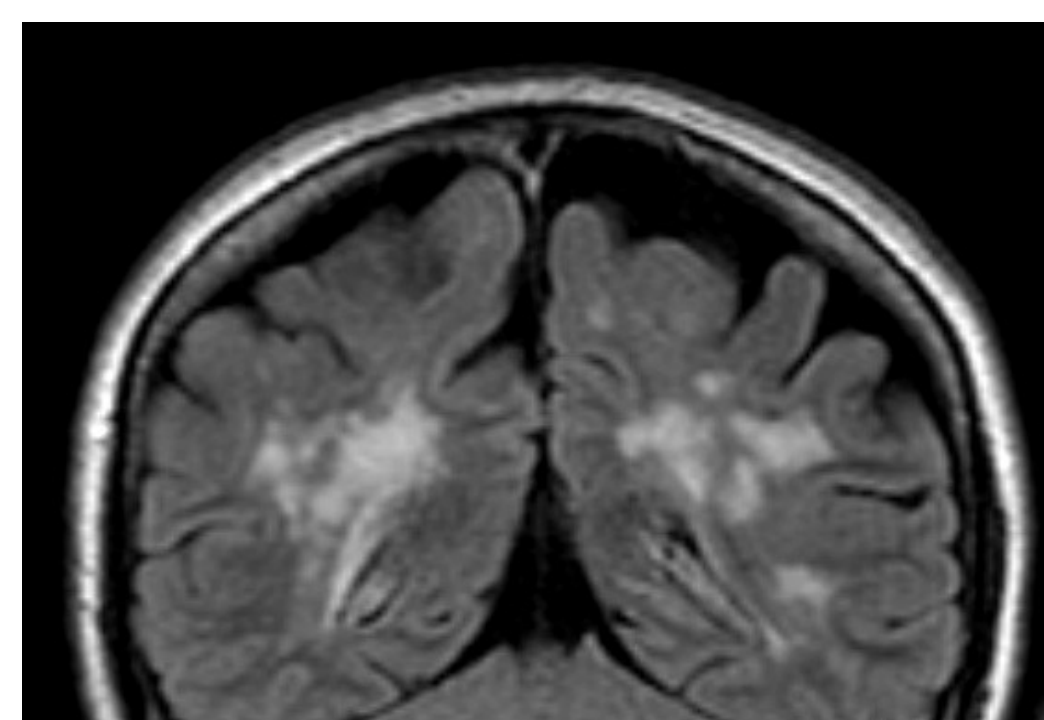


Fig 4 T2 FLAIR  
Multiple hyperintense foci in deep white matter

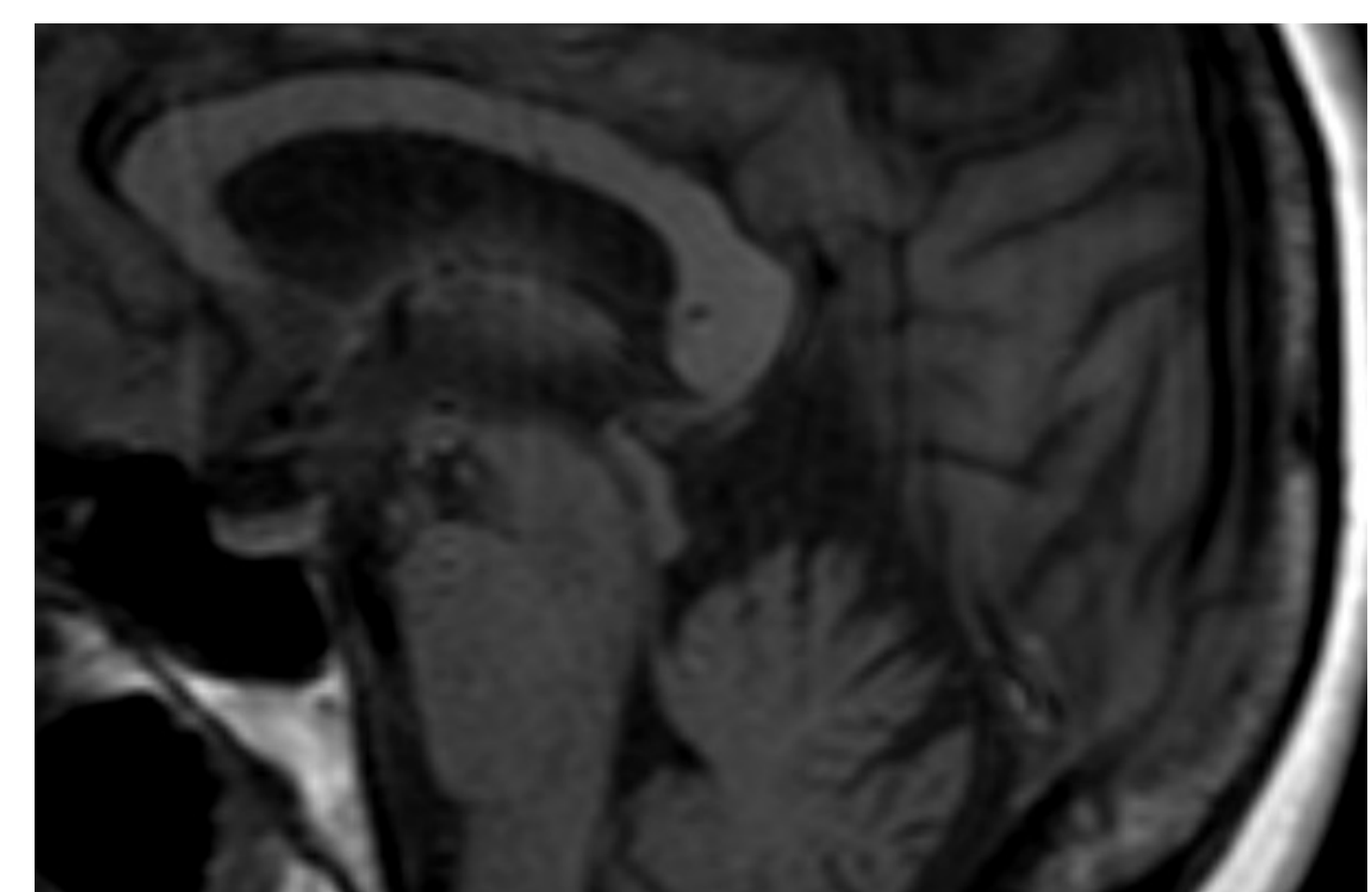


Fig 5 T1 MRI Central callosal holes

**DISCUSSION AND CONCLUSION:** Susac syndrome, with 304 reported cases worldwide, is a rare autoimmune mediated microangiopathy affecting brain, retina and ear, that can be associated with mild elevation of antiphospholipid antibodies. The blood-retinal, blood-perilymph and blood-brain barriers are similar and have unique specialized tight junctions in their endothelium; for this reason, common antigenic features could explain the limited extension of the disease at these organs with the classical triad of encephalopathy, retinopathy, and hearing loss <sup>1</sup>.

An involvement of the nerve roots of the cauda equina has recently been described as a consequence of endotheliopathy of radicolomeningeal arteries supplying the cauda <sup>2</sup>.

So it's not surprising that other nerves of central nervous system may also be involved <sup>3</sup>. We think that trigeminal involvement in our patient was due to an identical pathogenic mechanism, therefore the hemiface hypoesthesia can be included in the spectrum of Susac syndrome features.

A comprehensive knowledge of Susac Syndrome is still evolving and we don't have a complete picture of this rare condition yet; however an high clinical suspicion must be maintained in order to distinguish it from several mimicking conditions to initiate a timely treatment which can reduce debilitating sequelae.

## REFERENCES

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