Lesion Morphology at 7 Tesla MRI differentiates Susac Syndrome from Multiple Sclerosis

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Introduction: First described in the '70s, Susac syndrome consists of the clinical triad of visual disturbances due to branch retinal artery occlusions, sensorineural hearing loss and encephalopathy.[1] It is considered to be an autoimmune mediated microangiopathy, which consequently leads to vascular occlusions and microinfarctions. Although an orphan disease, Susac syndrome has yet to be considered as differential diagnosis in numerous conditions. In particular, the differentiation from multiple sclerosis (MS) can be challenging, since its clinical presentation and paraclinical findings including routine MRI findings partially overlap.[2] Since ultrahigh field MRI at 7T depicts white matter lesions with great anatomical details,[3,4,5] we here studied a potential benefit of 7T MRI for i) the differentiation between Susac syndrome and MS and ii) the clarification of pathogenesis of Susac syndrome.

Methods: Four patients suffering from Susac syndrome (table), eight sex and age matched patients with relapsing-remitting MS (median EDSS 1.5) and 12 matching healthy controls (HC) were investigated at 7 Tesla (T) MRI.
The protocol included T1-weighted MPRAGE (TE 2.98 ms; TR 2300 ms; TI 900 ms, acquisition time 9:14 min, spatial resolution 1.0 x 1.0 x 1.0 mm3), T2*-weighted FLASH (TE 25.0 ms; TR 1820 ms; acquisition time 12:11 min, spatial resolution 0.5 x 0.5 x 2 mm), and TIRM sequences (TE 90 ms; TR 16000 ms; TI 2925.5 ms, acquisition time 12:50 min, spatial resolution 1.0 x 1.0 x 3.0 mm^3).
Callosal lesions showing signal intensity values not higher than 15% of mean cerebrospinal fluid (CSF) signal intensity were considered CSF-isointense. In addition, volume of corpus callosum was calculated.

Results: In total, we detected 190 (Mean±SD, range: 48±37, 6-95) white matter lesions in 4 patients with Susac syndrome and 259 (Mean±SD, range: 32±29, 5-76) plaques in 8 MS patients using T2* FLASH. Healthy controls did not show any brain pathology. 7T MRI distinguished Susac syndrome from MS by revealing differences in the morphology of white matter lesions (figure 1) and callosal damage (figure 2), additional to atrophy of the corpus callosum. At 7T T2* FLASH, MS plaques were nearly exclusively centered around a small central vein (242 lesions; 93%) and often showed a characteristic hypointense rim (110 lesions; 42%). Contrarily, white matter lesions in Susac syndrome exhibited significantly less frequently a perivascular setting (101 lesions; 53%, p=0.006), and very rarely a hypointense rim (11 lesions; 6%, p=0.01).

Conclusion: At 7T MRI, lesions in MS patients and patients with Susac syndrome differed with respect to morphology and structural organization. Thus, lesion morphology at 7T i) may serve as a marker to distinguish Susac syndrome from MS and ii) reflects a different pathophysiological mechanism underlying Susac syndrome, e.g. microinfarction rather than demyelination. In order to confirm our preliminary results further studies including more patients with Susac syndrome are needed.

References: 1 Susac JO et al. Neurology 1979. 2 Rennebohm RM et al. Curr Treat Options Neurol 2008. 3 Kollia K et al. AJNR Am J Neuroradiol 2009. 4 Tallantyre EC et al. Neurology 2008. 5 Pitt D et al. Arch Neurol 2010.

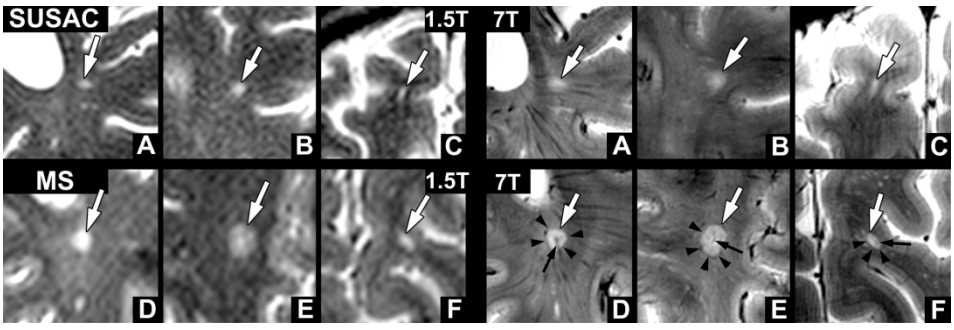

Figure 1. *Structure of T2* white matter lesions differentiates Susac syndrome from MS: Comparison between 1.5T and 7T.* At conventional MRI, white matter lesions (white arrow) in both Susac syndrome (A-C) and MS (D-F) appear as hyperintense lesions and cannot be distinguished from each other. At 7T T2* FLASH, perivenous (black arrow) MS plaques particularly express a characteristic hypointense rim (black arrow head). In contrast to their clearly centered position within MS plaques, small veins (black arrow) rarely identified in Susac syndrome lesions were most commonly located in the lesion periphery (A).

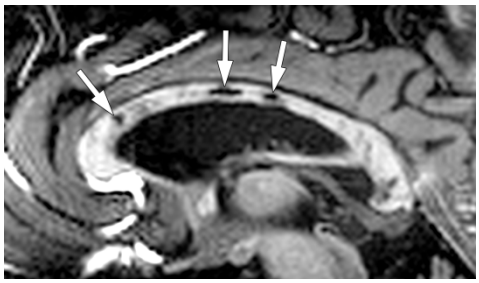

Figure 2. *7T T1 weighted MPRAGE reveals callosal damage.* All Susac patients showed CSF-isointense lesions (arrows) within the central part of corpus callosum that were not commonly seen in MS