A NEW IMAGING TECHNIQUE TO MONITOR DISEASE ACTIVITY IN SUSAC SYNDROME?

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Background: Susac syndrome is a rare auto-immune disorder. Often (though not exclusively) it affects young women. Pathophysiologically, it is a non-vasculitic microangiopathy, with involvement of the small vessels of the brain, retina and inner ear, leading to headache, encephalopathy, visual disturbances and sensorineural hearing loss as key clinical symptoms¹. It can cause significant morbidity if not treated adequately. Aggressive immunosuppressive treatment is therefore justified, but disease course and duration are highly variable. Treatment and disease activity monitoring is through clinical and imaging follow-up.

Methods: High resolution black-blood contrast-enhanced T1 images² were acquired in a patient with Susac syndrome upon diagnosis and during treatment follow-up.

Results: In our patient, there were several sulcal

enhancements visible at the time of diagnosis (*Fig 1a*).

Upon follow-up MRI, these black blood sequences were repeated, and each time, sulcal enhancement became less visible. It disappeared completely 6 months after treatment initiation (*Fig 1b*). The patient was clinically stable at this time. One year after diagnosis, our patient had a relapse. At that time, fluo-angiography and black blood imaging demonstrated disease activity (*Fig 1c*), although on DWI no new lesions were detected.

Conclusion: High resolution black-blood contrast-enhanced T1 images might be useful as an imaging marker of disease activity and appears more sensitive than DWI images in Susac syndrome. Drawback is the repeated administration of contrast. Further studies are needed to confirm these results.

References:

1. Dörr J, Krautwald S, Wildemann B et al. Characteristics of Susac Syndrome: a review. Nat Rev Neurol 2013. 9(6): 307-316.
2. Obusez EC, Hui F, Hajj-ali RA et al. High-Resolution MRI Vessel Wall Imaging: Spatial and Temporal Patterns of Reversible Cerebral Vasoconstriction Syndrome and Central Nervous System Vasculitis. AJNR 2014. 35(8):1527-32.