

Diffusion tensor imaging permits detection of disjunct MD and FA changes in the basal ganglia in patients with Susac's syndrome

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Introduction: Susac's syndrome is a rare disease that is characterized by retinal artery occlusion (BRAO), sensorineural hearing loss, and encephalopathy with associated cognitive and psychiatric symptoms. Conventional structural MRI (T1w, T2w, FLAIR) findings typically indicate callosal lesions which do not correlate with the type and severity of neuropsychological deficits. We recently demonstrated that microstructural damage as detected by diffusion tensor imaging (DTI) of otherwise normal appearing white matter (NAWM) correlated more effectively with neuropsychological deficits relative to macroscopic lesions (1). Macroscopic lesions of the basal ganglia may also be detected during acute stages of the syndrome but ordinarily disappear during chronic stages. The primary objective of the present study was to investigate whether DTI could identify persisting grey matter damage of the basal ganglia during chronic stages of Susac's syndrome through analyses of fractional anisotropy (FA) and mean diffusibility (MD).

Methods: Nine patients with clinical evidence of Susac's syndrome (age: 31 y +/- 11 y; w 5, m 4) and 83 age-matched healthy control subjects (age: 35 y +/- 13 y; w 42, m 41) participated in this study. DTI was performed by using echo planar imaging at 3T (20 diffusion weighted images: $b=1000$ s/mm²; 3 b0 images). All individual DTI data were automatically pre-processed and iteratively normalized on the basis of FA and b0 contrasts by the „Münster Neuroimaging Evaluation System“ (Eval 3.0) using the „SPM normalization toolbox for voxel-based statistics on fractional anisotropy images“ (2). Voxel-based statistics (VBS) was performed using SPM8 (www.fil.ion.ucl.ac.uk/spm) to test for regionally significant group-wise differences in FA and MD.

Results: Widespread FA reductions were preferentially observed in frontopolar white matter (WM), genu of the corpus callosum, and temporal WM in patients relative to controls (fig 1). Prefrontal WM damage occurred in every patient, independently of the location of macroscopic lesions. Mean FA of total WM was significantly reduced in each patient. FA *increases* were found only in grey matter (GM) of the basal ganglia and brain stem. Increased MD was observed in widespread cortical and subcortical regions (fig 1). However, with respect to the basal ganglia, increased MD was restricted to the pallidum whilst increased FA was restricted to the putamen (fig. 2). There was therefore a circumscribed disassociation between MD and FA abnormalities in the basal ganglia in patients relative to controls.

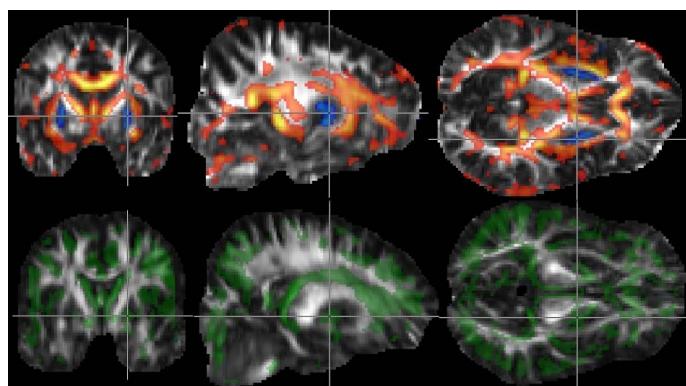


Figure 1

Upper panel: Results of the FA-VBS group analysis: Significant ($p<0.001$, uncor.) FA reductions are displayed in red-yellow and increased FA in blue-light blue (color ranges represent t-values from 3.2 to 7.0 and -3.2 to -7.0). FA was increased in the putamen of the basal ganglia (see intersection of planes) but not in the globus pallidus (compare fig. 2). The t-maps are overlaid to an FA image. **Lower panel:** Results of the MD-VBS group analysis: Significant ($p<0.05$, uncor.) MD increases are displayed in green, no MD decreases were found. MD was increased in WM, cortical and subcortical GM, including the globus pallidus (see intersection of planes) of the basal ganglia but not in the putamen (color ranges represent t-values from 1.66 to 5.0 and -1.66 to -5.0).

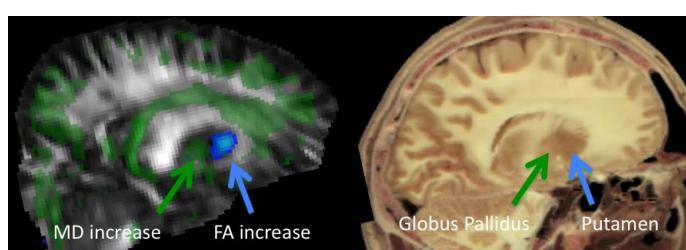


Figure 2

Disjunct MD and FA changes in the basal ganglia: MD increase in the globus pallidum (green) and FA increase in the putamen (blue).

Discussion:

Our results provide evidence indicating that DTI is sensitive in detecting damage to WM that is of normal appearance in Susac's syndrome using conventional MRI methods. GM alterations in Susac's syndrome are also detectable by circumscribed FA and MD increases in the basal ganglia that are also not observed using conventional MRI. The alterations in basal ganglia MD and FA are differentially localized to the pallidum and putamen, respectively.

References:

1. Kleffner I, Deppe M, Mohammadi S, et al. Diffusion tensor imaging demonstrates fiber impairment in Susac syndrome. Neurology 2008;70:1867-1869.
2. Mohammadi S, Glauche V, Deppe M. SPM normalization toolbox for diffusion weighted images. ISMRM 2010 Abstract # 5504