Serial proton MRS and DWI of Balo's concentric sclerosis in a child

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Introduction:

Balo's concentric sclerosis is a demyelinating disorder and considered as a rare variant of MS [1]. The condition is neuropathologically uniquely characterized by consecutive, circular layers of demyelinated and myelinated white matter. On T2-weighted MRI this pattern pathognomonically appears as concentric rings or whorled areas with alternating hyperintense and hypointense bands [2]. Serial MRS has so far only been performed in adult patients over a period of 23 months not considering absolute concentrations [3].

Here, we report a combined serial MRS and DWI study over 3 years 9 months of a brain lesion of a child diagnosed with Balo's concentric sclerosis. The study was carried out to better understand the underlying biochemical changes and their correlations with the evolution of clinical symptoms. A further scope of the study was to evaluate the combination of both methods for these clinical questions.

Clinical findings:

The 13 years 6 months old girl was admitted to the hospital with a sudden onset of right homonym hemianopsia, dysarthria, akalulia, sensoric aphasia and alexia. The initial MRI revealed left-hemispheric widespread edema with multiple, whorled, cocardic appearing, contrast-enhancing signal alterations which were most pronounced in peritrigonal white matter. In addition, a small right frontal edematous zone was appreciable. In CSF pleozytosis and oligoclonal IgG were detectable. Despite several therapeutic trials (dexamethason, aciclovir, foscavir, rocephin i.v., predison p.o.) the clinical condition first worsened and subsequent MRI exams showed lesion progression. During the following three months all symptoms subsided completely or partially with the exception of hemianopsia which persists up to this day. Five months later an acute relapse occurred with abulia and hypokinesia and subsequent remittance. On MRI a new contrast-enhancing, concentric lesion in the right parietal region extending precentrally could be distinguished. The left-hemispheric lesions appeared diminished in size. Meanwhile the girl received an immunmodulatory therapy (ß-interferone). The so far last relapse occurred recently, 3 years 9 months after the first onset of symptoms with an acute left hemipareses and another contrast-enhancing, concentric signal alteration within the right basal ganglia region. The diagnosis of Balo's concentric sclerosis was reached considering the relapsing and partially remitting course, the MRI appearance, and the CSF findings.

Methods:

Localized proton MRS of cerebral WM (STEAM, TR/TE/TM = 2000/20/10 ms, 64 accumulations, volume-of-interest [VOI] = 4.85 and 4.1 ml) was performed 1, 2, and 4 months as well as 3 years 5 months and 3 years 9 months after onset of symptoms. Three examinations were performed at 2 T (Siemens Vision) and two at 2.9 T (Siemens Trio). The VOI was placed within the left parieto-occipital (LPO) lesion (**Fig. MRI**). Absolute concentrations of N-acetylaspartate and N-acetylaspartylglutamate (tNAA), creatine and phosphocreatine (tCr), choline-containing compounds (Cho), inositol (Ins) and lactate (Lac) were determined by LCModel [4]. Concomitant DWI (4 months and 3 years 9 months after disease onset) were obtained with single-shot STEAM at 2x2x4 mm^3 resolution (24 gradient directions, b=0 and 1000 s/mm^2) at 2 and 2.9 T, respectively. Maps of the relative anisotropy (RA) and apparent diffusion coefficient (ADC) were calculated.

Results and Discussion:

The **Table** shows metabolite concentrations as well as RA and ADC values in the LPO lesion. Initially, MRS revealed very low tNAA and elevated Cho and Lac concentrations but normal tCr and Ins (**Fig. MRS)**. During the course of the disease tNAA significantly but not fully recovered, Cho and Lac decreased to normal values and Ins raised well above normal (**Fig. MRS**). DWI resulted in reduced RA and increased ADC values which remained comparatively unchanged.

The initial biochemical alterations detected by MRS indicate an acute inflammatory process (confirmed by contrast-enhanced MRI) compromising myelin as well as neuroaxonal structures. As the inflammation seemed to subside, the partial recovery was accompanied by gliotic changes. However, the focal symptom of hemianopsia persisted unchanged. On the other hand, DWI showed no pronounced changes during the course of the disease. The persistence of low RA values suggests that the main myelin and/or axonal structures remain severely impaired, although it cannot be fully excluded that current methods fail to depict minor recovery processes. Further studies including more follow-up exams are warranted to discern these disease mechanisms.

Table: RA and ADC of the third and fifths examination and absolute concentrations of brain metabolites in mmol/l of all five examinations given as time after onset of symptoms and age matched controls (mean ± 2SD).

	1 months	2 months	4 months	3 years 5 months	3 years 9 months	control
tNAA	1.68	2.83	3.77	5.01	5.2	7.9 ± 0.6
tCr	3.59	4.07	5.43	4.92	4.97	4.8 ± 0.4
Cho	2.33	3.0	2.54	2.18	1.73	1.6 ± 0.2
Ins	2.55	6.84	8.2	7.99	6.44	3.9 ± 1.2
Lac	8.0	4.06	4.12	1.48	1.4	<1
RA			0.5		0.6	
ADC			1.5		1.6	

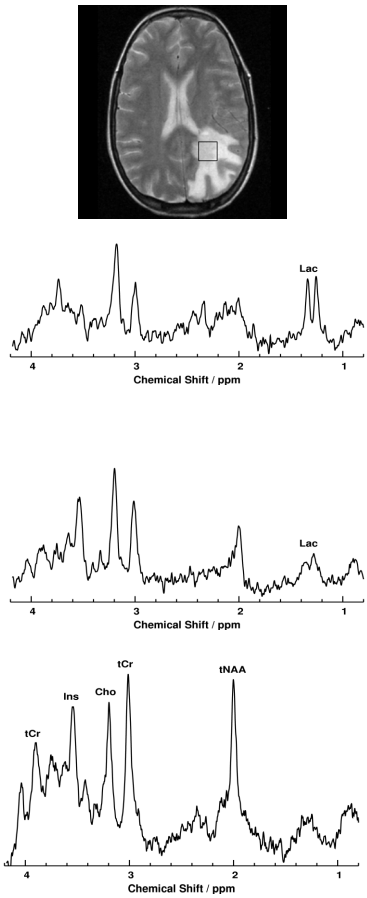

Figure: Patient with BCS: axial T2W MRI (VOI) and serial proton MRS 1 and 4 months and 3 years 9 months after onset of symptoms.

References: 1. Kastrup, O. et al., *J Neurol*, 249, 811, 2002; 2. Karaaslan, E. et. al., *Am J Neuroradiol*, 22, 1362, 2001; 3. Chen, C.-J. et. al., *J Comput Assist Tomogr*, 25, 713, 2001; 4. Provencher, S.W., *MRM*, 30, 672, 1993.