Hemispheric metabolic abnormality in Rasmussen's encephalitis

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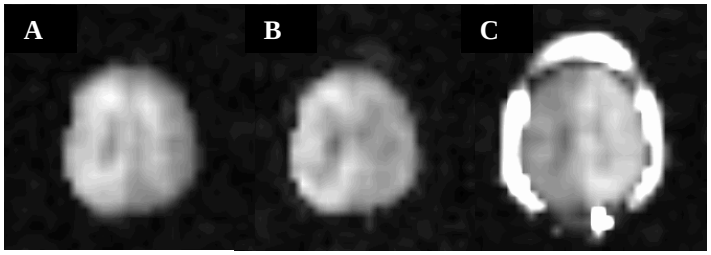

Fig. 1: 3-year-old female with left sided focal seizures for 6 months. Spectroscopic images of Cho (A), Cr (B), and NAA (C) demonstrate increased Cho and Cr and decreased NAA in the right hemisphere.

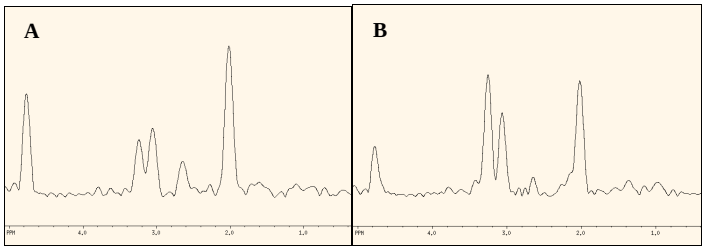

Fig. 2: MR spectra of the same patient as Fig.1. A, Normal MR spectra from the left parietal gray matter. B, MR spectra of the right parietal gray matter with increased Cho peak and decreased NAA peak.

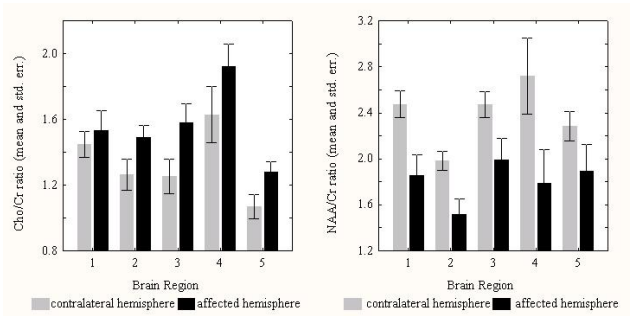

Fig. 3: Mean Cho/Cr and NAA/Cr ratio of brain regions (1: centrum semiovale, 2: insular cortex, 3. parietal white matter, 4. frontal white matter, 5. parietal gray matter) between the affected and contralateral hemispheres.

Introduction: Rasmussen's encephalitis is a rare chronic, progressive encephalitis with epilepsy generally confined to one hemisphere of the brain. Diagnosis is made based on EEG findings, seizure characteristics, and MRI findings. T2 abnormality often begins in the insular region as a hyperintensity in the white matter and progresses to diffuse cerebral atrophy. There have been several reports of MR spectroscopy in Rasmussen's encephalitis (1,2,3,4). The purpose of the current study was to evaluate metabolic abnormalities of this disease in multiple regions of both affected and contralateral hemispheres using multi-slice proton magnetic resonance spectroscopic imaging (MRSI).

Methods: Eleven patients with Rasmussen's encephalitis were included in the study. Age ranged from 3 to 16 years with seizure duration of 6 to 66 months. Spectroscopic imaging was performed by using a multisection spin-echo sequence (TR/TE/NEX=2300/280/1, 15-mm section thickness, FOV 24 cm, 32 x 32 phase-encoding steps, rendering a nominal voxel size of 0.8 cm^3) with outer volume saturation bands. Details of the sequence were described previously (5). MRSI was performed at four levels in a plane parallel to the anterior commissure-posterior commissure line identified from the mid-sagittal localizer. Spectroscopic images of choline (Cho), creatine (Cr), and *N*-acetyl aspartate (NAA) were reconstructed using MR spectroscopy software written at our institution. NAA/Cho, Cho/Cr, and NAA/Cr ratios were calculated for the insula, frontal and parietal white matter, centrum semiovale and parietal gray matter. Axial T2-weighted (TR/TE/NEX=4000/100/1) images were reviewed and the pattern of imaging abnormalities was categorized as either diffuse or localized. A mixed general linear model (Statistica 6.1) was used to determine the effect of hemisphere side (affected or contralateral) and brain regions (two-way repeated measures: 2 hemisphere side X 5 brain regions), and MR imaging pattern (between subjects factor); covariates were age and duration of seizure. The level of significance was set at $p<0.05$.

Results: Typical MRSI demonstrated decreased NAA, increased Cho and normal to increased Cr diffusely involving the affected hemisphere (Fig.1,2). There was a significant difference in NAA/Cho ($p=0.004$), NAA/Cr ($p=0.019$), and Cho/Cr ($p=0.037$) ratios between the affected hemisphere and contralateral hemisphere. There was no significant difference among brain regions for all three ratios. However, trends of regional differences appear to hold even in the affected hemisphere (Fig.3). Duration of seizure, age, and MR imaging pattern did not produce a significant effect.

Discussion: Highly significant differences in NAA/Cho, NAA/Cr and Cho/Cr ratios between affected and contralateral hemispheres were found. These changes are consistent with the pathological changes of microglial proliferation and neuronal loss. No significant regional differences were found in the affected hemisphere, suggesting that Rasmussen's encephalitis is indeed a hemispheric disease, not a localized pathology, even though MRI abnormalities as well as seizure activity may be localized. There was no significant interaction between seizure duration and metabolite ratios, suggesting that metabolic changes occur early in the disease process. In summary, proton MRSI consistently shows metabolic abnormalities in Rasmussens encephalitis, which may be helpful in establishing the diagnosis and defining the extent of the disease process.

Reference: **1.** Matthews PM. et al. Neurology 1990;40:985-989. **2.** Cendes F. et al. Brain. 1995;118(Pt3):753-8. **3.** Sundgren PC et al. Neuroradiology 1999;41:935-940. **4.** Sener RN. J. Neuroradiol. 2000;27:179-184. **5.** Duyn JH. et al. Radiology1993;188:277 –282.