Neuroimaging Highlight

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Serial MR Imaging of Adult-Onset Rasmussen's Encephalitis

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A 52-year-old woman was referred for a progressive neurological condition which started a year before with continuous irregular twitching of the right facial and arm muscles as well as the tongue and palate, followed by progressive dysphagia, right hemiparesis, ataxia, dysphasia and dysarthria. Though magnetic resonance imaging (MRI) at clinical presentation and a year after were both normal (Figure A1), Rasmussen's encephalitis (RE) was strongly suspected and treatment with corticosteroids and immunoglobulins were begun. A third MRI, two years after onset, revealed mild T2 hyperintense subcortical white matter changes over the left perisylvian region (Figure A) which confirmed our initial suspicion of RE as she now met clinical, electrophysiological and morphological criterias for RE. While waiting for a cerebral biopsy after unsatisfactory response from antiepileptic drugs, corticosteroids and immunoglobulins, her condition markedly worsened less than three years after onset with the sudden occurrence of status epilepticus requiring intubation and continuous infusions of midazolam and propofol. Electroencephalography (EEG) monitoring disclosed left temporal periodic lateralized epileptiform discharges, several left temporal non-convulsive seizures, and independent left facial jerks with no obvious EEG correlate except for muscle artifacts. While epilepsia partialis continua continued unabated, left temporal complex partial seizures were eventually controlled within a week using a combination of phenytoin, levetiracetam, clonazepam and immunoglobulins. Magnetic resonance imaging in this acute setting revealed mild subcortical frontal operculum enhancement (Figure A3), mild progression of the T2/FLAIR frontal opercular hypersignal (Figure A4), left hemispheric gyriform increased signal on T2-weighted images and restricted diffusion on DWI/ADC images (Figure B), and crossed cerebellar diaschisis (Figure C). Stereotactic needle biopsy (which unfortunately could not be performed in the highly functional perisylvian area where maximal MR abnormalities were) showed inflammation with increased microglial cells (but only a few microglial nodules), T lymphocytes, mild perivascular inflammation, neuronal loss and gliosis. B cells were extremely rare. No inclusion bodies suggestive of a viral infection were found. No mitotic activity was found. Repeat MRIs showed reversal of status-induced signal changes one month after status resolution and appearance of mild left perisylvian atrophy at three months (Figure D).

Rasmussen's encephalitis is a chronic inflammatory disease of unknown origin affecting one hemisphere. It is clinically characterized by intractable focal onset seizures (usually epilepsia partialis continua) and deterioration of functions associated with the affected hemisphere[1]. Although the majority of cases occur in children, an adult variant has been described[2,3]. An auto-immune origin is suspected. Recent work from our group has shown that adult humanized mice (NOD/SCID/IL2rg^{null}) engrafted with CD34+ stem cells isolated from the peripheral blood of RE patients developed seizures and brain gliosis[4]. Serial MRI of RE patients reveal a characteristic longitudinal sequence of changes: normal volume and signal (stage 0), swelling and hyperintense T2/FLAIR signal (stage 2), atrophy and hyperintense signal (stage 3), and progressive atrophy and normal signal (stage 4)[5]. Our patient also presented transient unilateral hemispheric gyriform T2/FLAIR hyperintensity and restricted diffusion during complex partial status.

These periictal signal changes have been reported previously in status due to other etiologies and are thought to be related to vasogenic and cytotoxic oedema[6]. Finally, crossed cerebellar diaschisis was also observed in our patient. Diaschisis is classically defined as a sudden inhibition of function in an intact area of the brain due to a lesion in a remote area that is however anatomically connected through fiber tracts[7]. In status, it may be related to excessive neuronal transmission from prolonged excitatory synaptic activity via the cortico-pontine-cerebellar pathways[8,9]. With the natural progression of the disease, permanent crossed cerebellar degeneration may occur[10]. Treatment options to alleviate the seizure disorder and cease the progressive neurological deficit include corticosteroids, immunoglobulins, plasma exchanges, tacrolimus and functional hemispherectomy/hemispherotomy[1].

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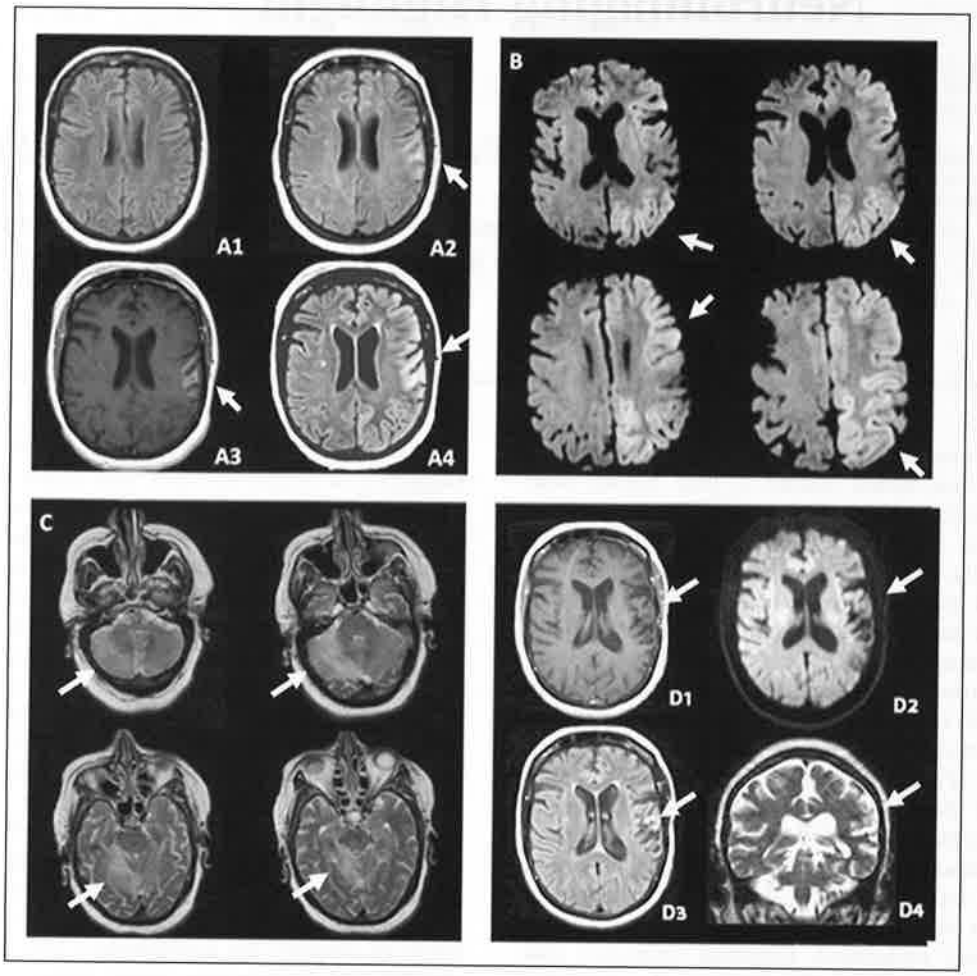

Figure: *Serial MRI findings in a 52 year-old woman with adult-onset Rasmussen's encephalitis. (A1) Axial FLAIR image at one year disclosing no obvious abnormalities; (A2) Axial FLAIR image at same level at two years showing perisylvian hyperintense signal; (A3/A4) MRI findings at three years during acute complex partial status epilepticus; (A3) Axial contrast-enhanced T1-weighted image revealing mild subcortical frontal operculum enhancement; (A4) Axial FLAIR image showing mild progression of frontal operculum hypersignal. (B) DWI axial images demonstrating restricted diffusion throughout the left cerebral cortex. (C) Axial T2-weighted images through cerebellum showing signal abnormalities in the right cerebellum hemisphere consistent with crossed cerebellar diaschisis. (D) Control images at three months after status control showing (D1) resolution of opercular enhancement on axial T1-weighted image post-gadolinium; (D2) resolution of left hemispheric restricted diffusion on axial DWI axial image as well as crossed cerebellar diaschisis (not shown); (D3/D4) and mild left perisylvian atrophy axial FLAIR image and coronal T2 image.*

References

1. Bien CG, Granata T, Antozzi C, et al. Pathogenesis, diagnosis and treatment of Rasmussen encephalitis. Brain. 2005;128:454-71.
2. Hunter GRW, Donat J, Pryse-Phillips W, Harder S, Robinson CA. Rasmussen's encephalitis in a 58-year-old female: still a variant? Can J Neurol Sci. 2006;33:302-5.
3. Hart YM, Andermann F, Fish DR, et al. Chronic encephalitis and epilepsy in adults and adolescents: a variant of Rasmussen's syndrome? Neurology. 1997;Feb;48(2):418-24.
4. Carmant L, Ouardouz M, Fontaine F, et al. Auto-immune basis of rasmussen encephalitis: development of a new animal model. Epilepsia. 2009;50 Suppl 11:477.
5. Bien CG, Urbach H, Deckert M, et al. Diagnosis and staging of Rasmussen's encephalitis by serial MRI and histopathology. Neurology. 2002;58:250-7.
6. Huang YC, Weng HH, Tsai YT, et al. Periictal magnetic resonance imaging in status epilepticus. Epilepsy Res. 2009;86:72-81.
7. Nguyen DK, Botez MI. Diaschisis and neurobehavior. Can J Neurol Sci. 1998;25:5-12.
8. Biaou O, Abderahim R, Soto Ares G, Jissendi Tchofo P. Diaschisis cortico-cérébelleux croisé post-état de mal épilpetique. J Radiol. 2008;89(4):507-9.
9. Samaniego EA, Stuckert E, Fischbein N, Wijman CA. Crossed cerebellar diaschisis in status epilepticus. Neurocrit Care. 2010;12:88-90.
10. Baheti NN, Bansal AR, Rathore C, Kesavdas C. Diaschisis. Is it always reversible? Neurology. 2009;72:e79.

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