

Multiphasic presentation of Rasmussen's encephalitis

Andreja Avberšek¹, Anna Miserocchi², Andrew W. McEvoy²,
Ayesha V. Patel³, Eleonora Aronica⁴, Ingmar Blümcke⁵,
Thomas S. Jacques⁶, James Acheson⁷, Maria Thom⁸,
Sanjay M. Sisodiya¹

¹ Department of Clinical and Experimental Epilepsy, Institute of Neurology, UCL Institute of Neurology, Queen Square, London

² Department of Neurosurgery, National Hospital for Neurology and Neurosurgery, Queen Square, London

³ Aston University, Birmingham, United Kingdom

⁴ Department of (Neuro)Pathology, Academic Medical Center; Swammerdam Institute for Life Sciences, Center for Neuroscience, University of Amsterdam; SEIN - Stichting Epilepsie Instellingen Nederland, Heemstede, The Netherlands

⁵ Department of Neuropathology, University Hospital Erlangen, Erlangen, Germany

⁶ Developmental Biology and Cancer Programme, UCL Institute of Child Health and Department of Histopathology, Great Ormond Street Hospital for Children NHS Foundation Trust, London

⁷ Department of Neuro-Ophthalmology, National Hospital for Neurology and Neurosurgery, Queen Square, London

⁸ Department of Neuropathology, UCL Institute of Neurology, Queen Square, London, United Kingdom

Received November 23, 2014; Accepted May 19, 2015

Typical vs. atypical clinical course of RE

Typical clinical course of RE

1. Early (prodromal) stage: infrequent seizures arising from one hemisphere, mild hemiparesis.
2. Acute stage: frequent seizures (epilepsia partialis continua can be seen in 50% of the cases) and progression to hemiparesis, hemianopia and cognitive decline.
3. Chronic stage: the neurological deficits are fixed.

Atypical clinical course

- Onset in adolescence or adulthood.
- Less aggressive or fluctuating course and MRI changes.
- Unusual clinical features, such as: occipital lobe involvement, bilateral involvement, or absence of epilepsia partialis continua.

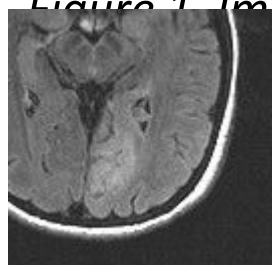
Characteristic MRI changes of RE

1. Initial MRI changes:
swelling and hyperintense signal in affected areas of cortex and/or subcortical white matter and deep white matter on T2-weighted and fluid attenuation inversion recovery (FLAIR) images.
2. Resolution of swelling and later atrophy, with persistence of the hyperintense signal.
3. MRI changes in the late stage:
progressive atrophy and normal signal.

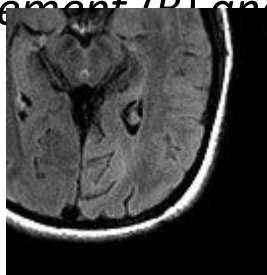
Atypical MRI changes of RE

1. Improvement of hyperintense signal abnormalities without atrophy
2. Improvement and re-occurrence of hyperintense signal abnormalities (illustrated in Figure 1)
3. Sustained increased signal

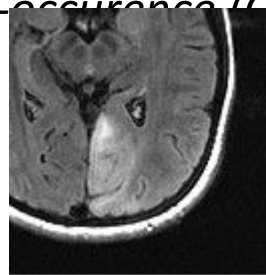
Figure 1. Improvement (B) and re-occurrence (C) of hyperintensities.



A



B



C