Clinical commentary

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Good outcome in adult-onset Rasmussen's encephalitis syndrome: is recovery possible?

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Aetiology of Rasmussen's encephalitis (RE)

- RE is an autoimmune disease but the primary cause remains elusive
- T-cell dominated encephalitis with activated microglial cells (forming nodules)
- Possible involvement of viral agents
- *In situ* hybridization techniques and improvement using ganciclovir suggest a possible role of CMV in a number of RE cases

RE clinical features and phases

- **Prodromal phase**: variable duration with polymorphic progressive seizures resistant to AEDs, no permanent neurological deficit
- **Acute phase**: lasting several months with increasing seizures and spreading encephalitis leading to neurological deterioration and volume loss of the affected hemisphere
- **Residual phase**: long lasting with atrophy of the affected hemisphere, associated with decreasing seizures but permanent neurological deficit

RE diagnostic criteria

Part A (all 3 requested)

- **Clinical**: focal seizures and unilateral cortical deficits
- **EEG**: mono-hemispheric epileptic discharges and/or slow activity
- **MRI**: mono-hemispheric hemiatrophy with T2/FLAIR hyperintensity or ipsilateral atrophy of the head of the caudate nucleus

Part B (at least 2 of 3)

- **Clinical**: “epilepsia partialis continua” or progressive unilateral cortical deficits
- **MRI**: progressive mono-hemispheric focal cortical atrophy
- **Histopathology**: T-cell dominated encephalitis with activated microglial cells (forming nodules)

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RE pattern depends upon patient's age

- **Type 1**: developing in childhood, characterized by a more aggressive course
- **Type 2**: developing in adolescence and adulthood, characterized by a slower and less aggressive course, with a more protracted prodromal phase

almost all RE reported cases show a severe clinical course

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Clinical characteristics of this RE case

- Adult-onset RE with protracted prodromal phase; the patient fulfilled clinical, EEG, and imaging criteria for a diagnosis of RE (all three “part A” criteria)
- The clinical course reflected the prodromal, acute, and residual phases outlined by Bien *et al*, and other known conditions, including unihemispheric vasculitis, could be ruled out
- During the acute phase, anti-CMV IgG was detected in both CSF and serum samples with a high specific index for intrathecal production of anti-CMV IgG, but no CMV genome was present in CSF

Treatment and outcome of this RE case

- During the prodromal phase, the patient received a high dose of steroids and then high-dose intravenous polyvalent immunoglobulins containing a high anti-CMV titre, otherwise used to treat CMV infections in immuno-compromised subjects
- Followed over a period of more than 15 years, the patient showed a very mild clinical course. Presently, he is seizure-free and able to pursue his normal activities. He has slight right pyramidal signs and a slight loss of discriminative sensation in the right hand, but no major sensory, motor, or neuropsychological deficit.

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Conclusions

- Very mild RE cases do exist, especially in adulthood, and may have a good outcome, without the need for surgery
- To prevent the occurrence of severe brain damage, treatments (steroids, immunosuppressive agents, intravenous polyvalent Ig, and monoclonal antibodies) are more useful at the beginning of the disease
- Specific index for intrathecal production of antibodies may reveal possible involvement of specific antibodies , which may indicate the use of a particular therapeutic approach

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