Hemiatrophy and seizures: a case of adult-onset Rasmussen encephalitis

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A 21-year-old man with no medical history presented in 2007 with a generalised tonic-clonic seizure. Over the previous 2 months he had experienced intermittent episodes of left-sided paraesthesia. Examination was normal. MR scan of the brain showed right-sided temporal, parietal and occipital lobe abnormalities. Differential diagnoses included infection, inflammation and tumour. Vasculitic and thrombophilia screens were negative, serum ACE level normal. Cerebrospinal fluid (CSF) was unremarkable, with negative results for culture, cytology and oligoclonal bands. Echocardiogram and CT scan of the thorax, abdomen and pelvis were also normal. He started carbamazepine for focal seizures and follow-up imaging was arranged.

Serial MRI found progressive right-sided hemiatrophy with variable areas of patchy right-sided signal change on gadolinium enhancement (figure 1A–C). He developed a left-sided homonymous hemianopia and left-sided spasticity with persistent sensory abnormalities in his upper limb. His partial-onset seizures continued despite high-dose carbamazepine and levetiracetam. In 2010, a course of methylprednisolone brought about an improvement in seizure frequency and disability but required a high oral dose to maintain benefit. Azathioprine was tried, but was poorly tolerated. A trial of intravenous immunoglobulin was ineffective. We considered biopsy on several occasions but this has not been performed.

Rasmussen encephalitis (RE) is a rare condition most often characterised by unihemispheric, progressive inflammatory disease causing intractable focal epilepsy, cognitive decline and hemiparesis. The aetiology remains unknown. It is most common in children with an average age of onset of 6–8 years.[1] However, there is a rarer adult-onset variant, accounting for ~10% of cases, which is characteristically less severe and slower to progress.[2]

The diagnosis of RE in our patient is based on a European consensus statement published in 2005.[3] This framework allows RE to be diagnosed without histological confirmation, but based on clinical, EEG and MRI changes. Our patient meets these criteria although the presence of gadolinium enhancement on his scans means we cannot fully exclude unihemispheric vasculitis.[4]

The treatment aims in RE are to prevent neurological decline and to control seizures—which remain drug-resistant in 80% of children.[5] Immunotherapy is often used although success is limited. Neurosurgical disconnection of the affected hemisphere from the normal one can improve seizure control and halt cognitive decline;

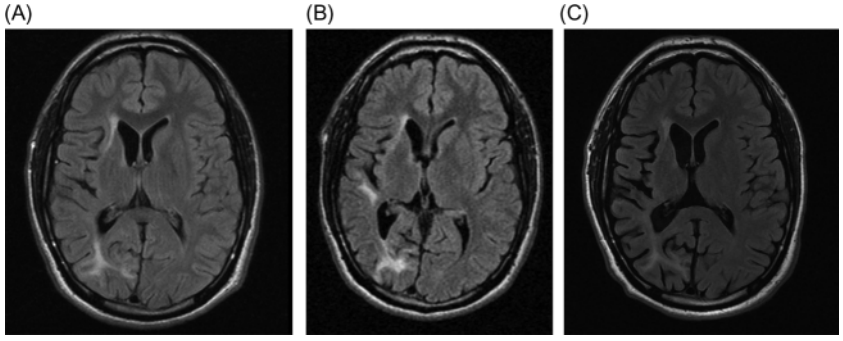

Figure 1 Axial fluid attenuated inversion recovery (FLAIR) MRI of brain from 2007 (A), 2009 (B) and 2012 (C) demonstrating variable right-sided hyper-intense signal change and progressive right hemiatrophy.

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appropriateness and timing of the procedure depend on the deficits a patient already has. Adult-onset RE is more likely to respond to immunological treatment[2] and the milder clinical course results in less residual functional deficit.[3]

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