patients with multiple sclerosis has not been evaluated, and there are no longitudinal studies comparing different techniques (ie, echocardiography, radionuclide ventriculography and magnetic resonance imaging). Therefore, prospective studies for monitoring cardiac function in patients with multiple sclerosis under mitoxantrone are urgently needed. Appropriate techniques should assess systolic and diastolic cardiac functions. Any decisions concerning the discontinuation of mitoxantrone owing to presumed cardiotoxicity should be based on a reliable and accurate method of assessment, as patients often have no therapeutic alternative.

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F Paul
Institute of Neuroimmunology, Charité – Universitaetsmedizin Berlin, Berlin, Germany

J Dörr
Department of Neurology, Helios-Klinikum, Berlin, Germany

J Würfel
Institute of Neuroimmunology, Charité – Universitaetsmedizin Berlin, Berlin, Germany

H-P Vogel
Department of Neurology, Helios-Klinikum, Berlin, Berlin, Germany

F Zipp
Institute of Neuroimmunology, Charité – Universitaetsmedizin Berlin, Berlin, Germany

Correspondence to: Professor F Zipp, Institute of Neuroimmunology, Clinical and Experimental Neuroimmunology, Neuroscience Research Center, Charité – Universitaetsmedizin Berlin, 10098 Berlin, Germany; frauke.zipp@charite.de

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Rasmussen encephalitis with ipsilateral brain stem involvement in an adult patient

Rasmussen encephalitis is a rare unihemispheric inflammatory disease of the brain that leads to intractable seizures, cognitive decline and progressive neurological deficits associated with the affected hemisphere. It predominantly affects children, with the onset in adults having a milder course. Immunotherapy has been suggested to improve the outcome of Rasmussen encephalitis.[1 2]

Case report

In November 2000, a left-handed 37-year-old woman experienced a head trauma with brief loss of consciousness. Shortly after, she had mild clumsiness of her right leg, which went on for the next 2 years. No magnetic resonance imaging (MRI) study was performed at that time. Her family also noticed a change in her character (all of which was retrospectively interpreted as the ''prodromal stage'' of Rasmussen encephalitis[2]).

In February 2002, the patient started having epilepsia partialis continua (EPC) of her right hand (later on interpreted as an onset of the ''acute stage'' of Rasmussen encephalitis[2]). Apart from EPC and impaired motor function of her right leg and hand (due to the EPC), the neurological examination and electroencephalogram were normal at this time. Cerebrospinal fluid contained 10 cells/μl, had a normal protein level and showed oligoclonal bands. Microbiological studies showed no sign of an infectious agent. MRI of the brain showed a mild left temporal atrophy. A steroid pulse treatment was given. However, the motor deficit progressed, accentuated in the right hand and leg, with central sensory deficit.

In December 2002, Jacksonian motor seizures of the patient's right hemibody started evolving from the EPC. Brain MRI showed left-sided supratentorial atrophy (most pronounced around the Sylvian fissure) and increased fluid-attenuated inversion recovery or T2 signal of the white matter. The brain stem, however, was neither atrophic nor did it show an increased signal (fig 1A, D, G).

In April 2003, the patient presented to our department (Department of Epileptology, University of Bonn, Bonn, Germany). MRI scans showed progression of hemiatrophy of the left hemisphere and involvement of the left mesencephalon (fig 1B, E, H). Biopsy specimens of the brain biopsy, obtained from the left superior frontal gyrus, showed perivascular and parenchymatous CD3+ CD8+ T lymphocytes (partly in close apposition to neurones), microglial activation and astrogliosis. The patient received a total of 1.2 g intravenous immunoglobulins (IvIg) per kilogram body weight.

On discharge, the patient had a 4/5 right hemiparesis with hypoesthesia. Further, monthly courses of 0.4 g IvIg/kg were recommended.[3] The patient's compulsory health insurance, however, refused to cover the costs for this kind of treatment. Three months later, the patient's hemiparesis had markedly progressed (arm, 2–3/5; leg, 4/5). Despite reinstitution of monthly IvIg by inpatient treatments in our department, the patient was hemiplegic by October 2003 and became seizure free at about the same time (onset of the ''residual stage''[2]). Fortunately, the patient's language abilities were preserved, obviously owing to atypical dominance (functional MRI scan disclosed bilateral, predominantly right-sided activation of frontotemporal regions during language tasks). IvIg treatment was stopped.

In April 2004, the patient was admitted because of swallowing and speech problems. On cranial nerve examination, she had a newly observed deviation of the uvula to the left side and reduced soft-palate elevation; gag and cough reflexes were normal, and speech showed signs of a flaccid dysarthria. Neither oculomotor abnormalities nor other signs of upper brain stem were affected. No cerebellar signs on the unaffected side were noted. The MRI scan showed an ongoing progression of the supratentorial left-sided hemiatrophy and an increase in signal extending subcortically to the left mesencephalon and pons, without contrast enhancement. This strictly unilateral signal increase in the left pons was newly observed (fig 1C, F, I). A high-dose long-term oral steroid treatment was started. One year later, swallowing and speech problems as well as the palatal velus paresis had resolved. MRI was unchanged. The patient is now 41 years old and remains seizure free.

Discussion

To the best of our knowledge, this is the first published biopsy-proven case of an adult-onset

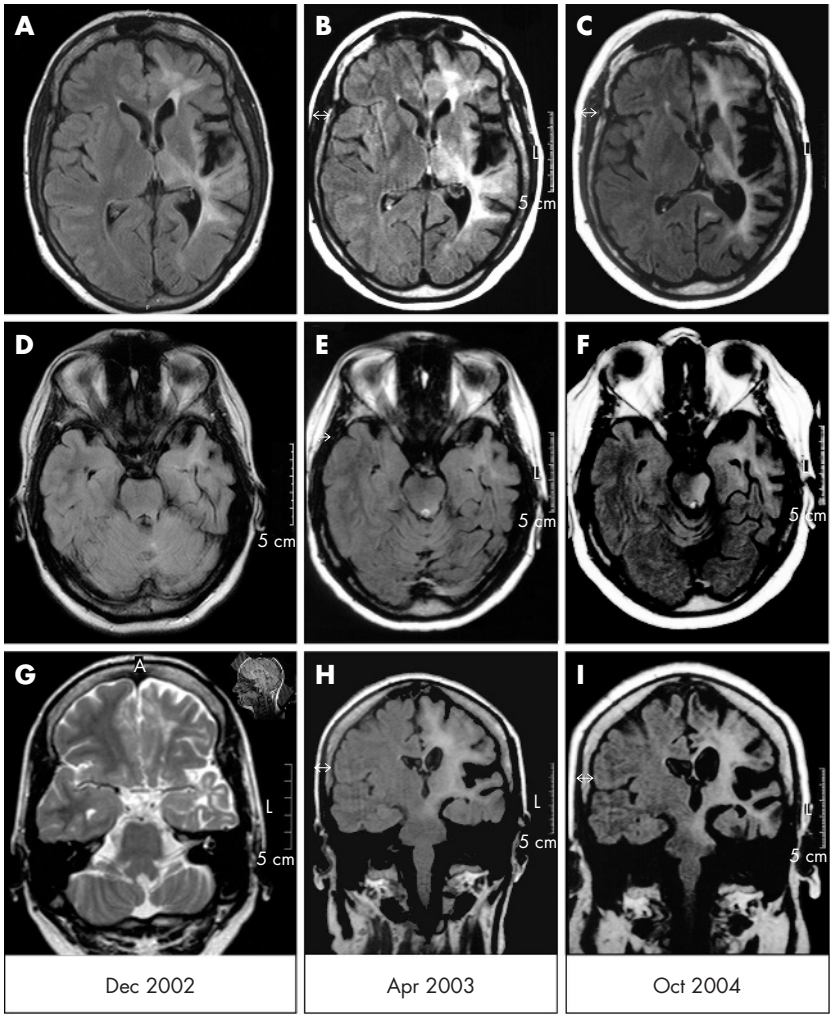

Figure 1 Serial brain fluid-attenuated inversion recovery-magnetic resonance imaging (FLAIR-MRI) scans in the patient described here. (A–C) Axial sections including the Sylvian fissure; (D–F) axial sections through the upper brain stem; (G–I) coronal sections including the pons. For the time course, see the dates on the bottom of the figure. (G) As no coronal FLAIR or T2 images from December 2002 exist, this unusually angulated T2 section showing the pons (for orientation of slices, see the small image in the upper right corner of (G)) from July 2002 is used as a substitute. A slight atrophy of the left cerebellar hemisphere is also seen.

Rasmussen encephalitis with magnetic resonance–tomographically demonstrated affection of the brain stem. The patient presented here exhibited a large increase in the fluid-attenuated inversion recovery signal, suggesting active inflammation or strong astrogliosis of the left upper brain stem in continuity with the supratentorial lesion, and clearly delineated from the right side. This is remarkable, as the characteristic and puzzling property of Rasmussen encephalitis of respecting the midline of the cerebral hemispheres is also observed here within the tight space of the brain stem. It cannot, however, be ruled out that Wallerian degeneration secondary to the supratentorial lesion contributes to this MRI presentation. The clinical improvement after resuming the immunosuppressive treatment also supports an ongoing active inflammatory process, even through the persistence of the radiological lesion. The relative mildness of the brain stem symptoms as well as the lack of topographical concordance should be noted, but does not contribute to the aetiological clarification. A possible explanation for this lower cranial nerve symptomatology without evident medullar involvement may be a supranuclear affection of the corticobulbar pathways.[4]

Infratentorial involvement in the form of cerebellar atrophy has previously been observed in childhood-onset cases, partly ipsilaterally and partly contralaterally (in the sense of a ''cerebellar diaschisis''), however, without reported clinical correlates.[5] In our patient, mild cerebellar hemiatrophy ipsilateral to the cerebral hemiatrophy without clinical correlates was observed.

To conclude, this case is an example of ongoing damage to the central nervous system, including the brain stem, even during the ''residual disease stage'', a feature indicating a late relapse of disease activity. This is particularly surprising given the fact that patients with adult-onset Rasmussen encephalitis usually experience a rather mild course and a relatively good long-term outcome.[2] This case may broaden the clinical and neuroradiological spectrum of possible courses of Rasmussen encephalitis.

C M Quesada
Department of Epileptology, University of Bonn, Bonn, Germany

H Urbach
Department of Radiology/Neuroradiology, University of Bonn, Bonn, Germany

C E Elger, C G Bien
Department of Epileptology, University of Bonn, Bonn, Germany

Correspondence to: Dr C G Bien, Department of Epileptology, University of Bonn, Sigmund-Freud-Strasse 25, Bonn 53105, Germany; christian.bien@ukb.uni-bonn.de

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Creutzfeldt–Jakob disease in a Chinese patient with a novel seven extra-repeat insertion in *PRNP*

Familial transmissible spongiform encephalopathies comprise about 14% of all cases of transmissible spongiform encephalopathy in humans. We report on a patient with a definite diagnosis of familial Creutzfeldt–Jakob disease with an insertional mutation consisting of seven extra octapeptide repeats between codons 51 and 91 in the *PRNP* gene, associated with a genotype homozygotic for methionine at codon 129 and a novel coding change of the inserted octapeptide region.

Case report

A Chinese woman developed forgetfulness at the age of 44 years, which progressed into memory deterioration 2 years later. At age 48 years, she developed gait disturbance and was admitted to hospital. On neurological examination she presented with mild dysarthria, intellectual deterioration and cognitive dysfunction. All her extremities showed hypertonia and ataxia, and no pathological reflexes were elicited. Her general health condition was good, except for the narrowed interpalpebral fissure dimension in her left eye and poor visual acuity since childhood.

A general brain magnetic resonance image showed no remarkable changes. The first electroencephalogram showed slow wave