Invited Comments

In this issue of Neurology India, Deb et al. report on a cohort of four patients with Rasmussen encephalitis (RE) from their center, admitted over a period of 3 years and all finally undergoing hemispherotomy. Their report provides data on the neuropathology of brain tissue obtained on epilepsy surgery and also some clinical data. This is the first internationally accessible neuropathological RE series from India. Its results confirm earlier observations on RE mainly made in North America and Europe which can be summarized as follows: RE is a rare disease. It is estimated that major referral centers encounter one to two cases every year. The disease mainly affects children with a peak of incidence at the age of 6. It is a strictly unihemispheric brain disease characterized by a progressive cerebral hemiatrophy (visible on serial magnetic resonance imaging [MRI]), frequent one-sided simple partial motor (and other) seizures, often in the form of Epilepsia partialis continua, and progressive loss of unilateral neurological function. There is a maximum of disease activity during the first year after onset of the acute disease stage as evident from the high density of inflammatory cells observed in brain specimens, the rapid destruction of inflammatory cells and the loss of neurological and neuropsychological function during this period. Finally, the patients enter a residual stage with lower (but usually still high) seizure frequency, a stable, often severe neurological deficit and low-grade cerebral inflammation.

Neuropathological studies have identified T cells and microglial cells as the main components of brain inflammation, and neurons as the targets of the inflammatory attack. By functional hemispherectomy or hemispherotomy, high-seizure freedom rates (80–85%) and low-complication rates have been achieved, however at the price of irreversible neurological deficits associated with the disconnected hemisphere.

These features are fairly constantly found in all patients.
described so far. This has led to a recent proposal of formal diagnostic criteria and the suggestion of a therapeutic pathway for RE patients. From a practical point of view, the most urgent question regarding RE is now the following: What is the optimal treatment for patients in the early disease stage, i.e., with still rather well-preserved neurological deficits? Those patients are usually considered not to be eligible for hemispherectomy. They are, on the other hand, at high risk of developing severe neurological deficits. Immunosuppressive and immunomodulatory treatment regimens (steroids, intravenous immunoglobulins, plasma exchange, tacrolimus) are emerging as promising options for these patients. Early immunotreatment seems to be able to stop or slow down the progressive tissue and function loss. However, the optimal treatment still needs to be determined. Beyond this, the most challenging question is that regarding the cause of the inflammatory process. Is it indeed – as usually assumed – autoimmune in nature? Or does an infection with a so far unidentified agent trigger the immune process? Does the epilepsy as such contribute to or even initiate the chronic brain destruction? The answer to these questions may finally bring about an explanation for the most conspicuous and puzzling phenomenon of this disorder: the fact that it affects and destroys only one brain hemisphere.

To pursue these goals, future researchers will rely on a solid foundation of clinical and neuropathological data regarding this condition. Deb et al. should be credited with having broadened this foundation.

C. G. Bien

Department of Epileptology, University of Bonn, Bonn, Germany.
E-mail: christian.bien@ukb.uni-bonn.de

Reference