

**Unusual presentations of Lyme neuroborreliosis**

When laboratory tests for diagnosis of Lyme borreliosis (LB) became available, a plethora of case reports on different neurological symptoms and syndromes that were linked to infection with *Borrelia burgdorferi* (Bb) were published. In these, the diagnosis of LB was primarily based on the demonstration of Bb antibodies in the patient's sera. Clear evidence for a causal relationship between syndromes observed and Bb infection was lacking in most of the cases. Furthermore, their overrepresentation in the literature had obscured the fact that just a few syndromes of the early disseminated stage of NB such as Bannwarth's syndrome in adults and facial palsy or meningitis in children make up the vast majority of cases with NB. Also, some of the rare manifestations are clinically well characterised such as chronic neuropathy in patients with acrodermatitis chronica atrophicans, or are well known and can be diagnosed easily when the CSF is examined - as it is the case in chronic meningitis or progressing encephalomyelitis. Nevertheless, beside these rare but well known manifestations, other rare but unusual presentations exist. They may mimic well described clinical syndromes - such as normal pressure hydrocephalus, benign intracranial hypertension, ataxia and opsoclonus, extrapyramidal syndromes, motor neuron disease, psychiatric syndromes - or present as stroke due to cerebral vasculitis. Careful diagnostic evaluation and the exclusion of other possible causes are essential before such unusual and extremely rare manifestations should be attributed to LB. In cases with central nervous system involvement demonstration of infection with Bb - either directly (culture, PCR) or indirectly (intrathecal Bb specific antibody synthesis) - and of active disease (cerebrospinal fluid pleocytosis) is mandatory before a definite diagnosis of NB can be established. Lack of therapeutic response to an adequate antibiotic treatment should raise doubts on the diagnosis of NB and should lead to a diagnostic re-evaluation.