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A neuropathological study on the limbic system in dentatorubral-pallidolysian atrophy

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Dentatorubral-pallidolysian atrophy (DRPLA) is an autosomal-dominant neurodegenerative disorder characterized by progressive myoclonic epilepsy (PME) in early-onset form and dementia with cerebellar ataxia in adult-onset form. Patients with DRPLA have an expanded CAG triplet repeat on the short arm of chromosome 12. We previously performed the neuropathological analysis of the brainstem in 9 autopsy cases of early-onset DRPLA with PME and 5 autopsy cases of adult-onset DRPLA without PME, and failed to find the PME-specific lesions. To investigate epileptogenesis in DRPLA, we examined the expressions of glutamate transporters and calcium-binding proteins (parvalbumin and calbindin D28K), which are speculated to reflect parameters of glutamate neurotoxicity and gamma-aminobutyric acid (GABA)ergic inhibitory interneurons, respectively, in the hippocampus, amygdaloid complex and temporal cortex in the same DRPLA autopsy cases. About half of the cases of both early-onset and adult-onset DRPLA showed a reduction in the expression of glial excitatory amino acid transporter 1 (EAAT1), while that of another glial glutamate transporter EAAT2 was comparatively preserved in most of the cases. The expression of neuronal glutamate transporter EAAT3 was identified in the remaining neurons in most of the cases. The number of interneurons immunoreactive for either parvalbumin or calbindin D28K in cases of early-onset DRPLA with PME seemed to be inferior to that in cases of adult-onset DRPLA without PME. These data suggest that occurrence of PME in DRPLA may be related to the alterations in the GABAAergic interneurons but not disturbed glutamate transport in the limbic system.

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A case of parietal lobe epilepsy with ictal smile

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We report a 7-year-old boy with parietal lobe epilepsy (PLE) with ictal smile. He was born after a normal pregnancy, with no family history of epilepsy. At the age of 6, he started to have drop seizures, which developed into sensory fits. Interictal electroencephalography (EEG) showed frequent spikes on the C3, C4, P3, Cz (international 10-20). No epileptogenic lesions were detected by magnetic resonance imaging, and the routine neurological examinations revealed no other abnormalities. Although he was treated with antiepileptic drugs, he often fell down in seizures after feeling abnormal sensations in his right shoulder. Ictal video EEG at the age of 7 years showed the following: (1) he became motionless and complained of fear and pain in the right hand (2) he had clonic seizures of the right upper limb and fell down to the left side (3) he laughed to himself though he did not feel funny (4) he had clonic seizures of the right upper eyelid. Phenytoin improved his symptoms remarkably. The clinical features of PLE include somatosensory symptoms as subjective manifestations, and motor phenomena as objective ones. In addition, most symptoms of PLE reflect seizure spreading outside the parietal lobe. As a rule, ictal smile has been associated with three pathologic situations: (1) hypothalamic hamartomas, (2) infantile spasms, (3) complex partial seizures of suspected temporal origin. We determined that his ictal smile resulted from temporal lobe seizures spreading from the parietal lobe. Therefore PLE may include ictal smile.