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Letter to the Editor

Steroid therapy ameliorated cataplexy in three children with recent-onset of narcolepsy

To the Editor:

Autoimmune mechanism is implicated in the pathology of narcolepsy [1]. We reported favorable effects of steroid therapy on three recent onset children who suffered from narcolepsy with cataplexy. Three children showed excessive daytime sleepiness and cataplexy, and were diagnosed as narcolepsy by undetectable hypocretin 1 levels in the cerebrospinal fluids (CSF) and the presence of sleep onset rapid eye movement (REM) period. They aged from 5 to 13 years, and patient 1 was a girl, while patients 2 and 3 were boys. All three patients carried HLA-DRB1*1501/DQB1*0602 haplotypes. The titer of Trib2-specific antibody was not elevated in any of the patients. Polysomnography and multiple sleep latency test (MSLT) studied in two patients also supported a diagnosis of narcolepsy. The steroid therapy was initiated at 1 month in patients 1 and 2, and 20 months in patient 3, respectively, after the onset. Prednisolone (1mg/kg/day) was given continuously for 2 or 3 weeks, and tapered gradually. The parents of the patients gave the written informed consent to the unauthorized use of prednisolone in children with narcolepsy; the ethics committee approved this trial. After the steroid therapy, sleepiness and cataplexy were ameliorated, though the CSF hypocretin-1 levels were unchanged. The Japanese version of the Epworth sleepiness scale decreased after the steroid therapy in all. Patient 1 recovered independent walking, which was disturbed by cataplexy. In patient 2, the frequency of cataplexy decreased. Patient 3 recovered fluency of speech, which was disturbed by cataplexy in the tongue. Improvement was found in atypical cataplexy, such as continuous hypotonia without emotional triggers and repetitive tongue protrusion [2]. No side effects were observed except for a temporary increase of intraocular pressure. Patients 1 and 2 were followed for 4 years, while patient 3 was followed for 1 year. It is noteworthy that the CSF hypocretin-1 levels remained low in two patients even after the amelioration of cataplexy and drowsiness probably caused by the steroid therapy, although the exact mechanism was not clarified. We speculated the possibility that the steroid may improve abnormalities in the immune system and/or the hypothalamic–pituitary–adrenal axis, which is involved in maintaining alertness and modulating sleep [3]. It is also possible that the steroid therapy may modulate the carnitine metabolism, which was reported to be disturbed in patients with narcolepsy [4], resulting in the amelioration of cataplexy and excessive daytime sleepiness [5]. The steroid therapy may be one effective treatment for recent onset children with narcolepsy.

Conflicts of interest

The author declares no conflict of interest. The author has indicated no financial support.