Excessive daytime sleepiness in Behçet’s disease with diencephalic lesions and hypocretin dysfunction

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A 31-year-old man with Behçet's disease suffered from acute diplopia, hypersomnia (>12 h sleep/day) and sleepiness (Epworth sleepiness scale: 14 points). Cranial MRI revealed diencephalic lesions with left-sided subthalamic gadolinium enhancement (arrow; fig. 1A–C). Wake-promoting hypothalamic hypocretin-1 in the cerebrospinal fluid (CSF) was decreased (215 pg/ml; normal: >320 pg/ml) [1]. Following treatment with prednisone for 1 month and with azathioprine, diplopia, sleepiness and hypersomnia disappeared within 2 months. Hyperintense lesions vanished on cranial MRI (fig. 1D). CSF studies revealed a normal hypocretin-1 level (400 pg/ml). This is the first reported patient with Behçet's disease with a low and treatment-responsive CSF hypocretin-1 level.

Reference

Key Words
Excessive daytime sleepiness • Hypersomnia • Behçet’s disease • Hypocretin

Fig. 1. Cranial MRI images revealed diencephalic lesions with left-sided subthalamic gadolinium enhancement (arrow) (A–C). After treatment, hyperintense lesions vanished on cranial MRI (D).