HYPERSONMIA IN WILSON´S DISEASE

S. Kim

Department of Neurology, Seoul National University Bundang Hospital, Seong-Nam, Republic of Korea

Background & significance: Wilson’s disease (WD) shows a wide heterogeneity in symptoms. In this case report we present hypersomnia as an unusual symptom in an unusual case of WD.

Case: A 48-year-old woman was admitted because of cognitive impairment for 2 years. Her complaints as fatigue, decreased level of concentration, and depression started at 3 years ago. There was no family history of psychiatric or other neurologic illness. No abnormality was found at neurologic examination. A moderate elevation in liver function tests was found, but the viral hepatitis markers were negative. The diagnosis of WD was based on decreased ceruloplasmin level, increased urinary excretion of copper, and decreased level of serum copper. Kayser-Fleischer ring was not detectable. Brain MRI disclosed increase T1 signal lesions in both globus pallidus. Electroencephalography showed diffuse slow waves of bilateral hemisphere without epileptiform discharge. Liver biopsy showed the presence of nuclear glycogen a cytoplasmic brownish pigment in hepatocytes and a minimal degree of periportal fibrosis. At 8-10 months after the initiation of the D-penicillamine therapy her complaints gradually resolved. In one year since resolution, she became drowsier and more hypersomnolent. She spent more than 2/3 of the day sleeping. After the administration of Methylphenidate 20mg, she became almost alert and gradually improved negativism.

Conclusions or comments: Hypersomnia was the main symptom in this patient. Although neurologic presentation of WD without Kayser-Fleischer rings have been already reported, lack of Kayser-Fleischer rings and the unusual clinical symptom demonstrate that this patient is a highly exceptional WD-patient.