Supranuclear Gaze Abnormality in Sporadic-Creutzfeldt Jacob Disease

Sporadik Creutzfeldt-Jacob Hastalığında Supranükleer Bakış Parezisi

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Dear Editor,

Sporadic Creutzfeldt-Jacob disease (sCJD) usually presents with typical clinical signs. Prodromal symptoms lasting for several weeks are followed by dementia, myoclonus, ataxia and abnormalities of vision (1). Supranuclear gaze palsies accompanying sCJD have rarely been reported. Some of these reported cases were initially diagnosed as progressive supranuclear palsy (PSP) and the diagnosis was corrected only after pathological examination had revealed spongiform encephalopathy (2,3). We aimed to report a patient who presented with supranuclear ophtalmoparesis, rapidly progressive gait ataxia and dementia as his initial manifestations of probable sCJD.

A 46-year-old man had presented with a 2 month-history of rapidly progressive gait instability, slurred speech and cognitive deterioration. He also reported diplopia and insomnia. The patient was alert but disoriented and his mini mental status examination score was 21. Voluntary vertical eye movements were almost abolished, but could be elicited by doll's eyes maneuver. In horizontal gaze examination, his smooth pursuit was replaced by micro-saccades and his eyes were unable to perform convergence. He had a severe cerebellar dysarthria, impaired rapid alternating movements and severe bilateral dysmetria of upper and lower extremities. He also had a moderate gait ataxia. Neurological examination was otherwise normal.

His routine blood chemistry was normal. Cerebrospinal fluid analysis was normal for cell counts, protein and glucose levels, oligoclonal bands, infectious agent and paraneoplastic antibody scanning. However CSF 14-3-3 protein was positive and CSF total tau level was highly

increased (1646 pg/ml). Brain MRI revealed bilateral FLAIR and T1 hyperintensities of caudate nuclei, putamina and frontal lobe involving most of the anterior cingulate gyrus and superior frontal gyrus bilaterally, with restricted diffusion (Figure 1). EEG showed decreased amplitudes but no typical periodic sharp waves. He was diagnosed as

Figure 1. Structural Magnetic Resonance Imaging (MRI) scans of reported patient with sporadic Creutzfeldt Jacob disease. Note the hyperintense signals in bilateral caudate nuclei, putamen and frontal cortex, which demonstrated restricted diffusion in DWI (a,b,c,d)

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having probable sCJD according to the “Updated clinical diagnostic criteria for sCJD” (4). Over the course of 3 weeks his neurological condition deteriorated. In addition to agitation and dysphagia his progressive ataxia rendered him bed-ridden overtime. He reportedly passed away 2 weeks after his discharge.

Vertical supranuclear ophtalmoplegia (VSO) associated with slowly progressive dementia and gait unsteadiness suggests a diagnosis of PSP (5). This entity is rarely reported in familial or sCJD. In a retrospective study, 180 cases with a clinical diagnosis of PSP underwent a standardized neuropathological assessment and two cases with a pathological diagnosis of CJD were found. Both cases had early onset of falls, VSO and Parkinsonism (3). Overall there are 6 cases who had been clinically diagnosed as to be PSP and turned out to be sCJD after post-mortem diagnosis (2,3,5,6). All of these cases had a relatively slowly progressive course, whereas our case had a typical rapidly progressive course and had VSO right after the onset of the disease. Supranuclear control of vertical eye movements is subserved by diffuse cortical projections from the frontal and supplementary eye fields reaching the rostral interstitial nuclei of the medial longitudinal fasciculus (riMLF) and the interstitial nuclei of Cajal (inC) at the level of the midbrain pretectum (7). Spongioform changes in those areas may contribute to VSO in sCJD. This case suggests that VSO can be a feature of otherwise typical sCJD.

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