A Case of Cortical Multiple Sclerosis

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Dear Editor,

Multiple Sclerosis (MS) is a chronic progressive demyelinating disease of the central nervous system. Cognitive dysfunction (CD) is usually observed at advanced stages of the disease but CD may be rarely observed at the initial stages of MS. Very unusually, MS presents primarily as CD; when this presentation occurs, it is referred as "cortical" or "cerebral" MS in the relevant literature (1). A case of a patient diagnosed with cortical MS, beginning with complaints of forgetfulness, and clinical benefit from treatment with donepezil was reported in this article.

The male patient with right hand dominance was aged 47 years and graduated from primary school. He was admitted with complaints of forgetfulness and difficulty in walking. We learned from his story that he had gradually increasing complaints of forgetfulness for 5 years; he could not remember words that he wanted to say since the last year, and he was unable to perform daily tasks and had impaired self-care. Also, he had weakness in the legs during the last 14 months. Psychomotor retardation, disorientation, hypophonia, faltering speech, bilateral temporal pallor at fundus, left central facial paralysis, reduction in vibration sensation in all four extremities, loss of position sense in the lower extremities, +4/5 monoparesis at right lower extremity, and ataxia at tandem gait were observed on conducting neurological examination.

Minimental state examination was scored as 17/30, digit span forward score of 4, digit span backward score of 3, verbal fluency of 7/min (perseveration, 2/min), phonemic fluency of (character K) 5/min (perseveration, 0/min), abstraction of 1/3, clock drawing score of 7/10, verbal learning test score at the 5th

min of 1/3, recall memory score of 1/3 on performing neurocognitive assessment. It was reported as temporolimbic memory, attention, and executive function defects.

Progressive dysfunction of cognitive abilities, affected pyramidal system, and affected deep sensory system were observed in this patient. Therefore, neurodegenerative processes avitaminosis, chronic infections, chronic metabolic and systemic diseases, mitochondrial diseases, and paraneoplastic processes were investigated. Abnormality was not detected in these tests. Cerebrospinal fluid (CSF) biochemistry, cytology, and IgG index was observed in the normal range. Oligoclonal bands were not detected in CSF. With bilateral PR stimulation, late latency was observed at visual evoked potentials and deformed response on the left side. EEG examinations that were performed in a 2-month period were within the normal range. Cerebral and cervical MR angiography examinations were within normal limits. On T2 sequence brain magnetic resonance imaging (MRI), multiple hyperintense lesions in both cerebral hemispheres and pericallosal areas were observed. On T2 sequence cervical MRI, mild, limited, multiple hyperintense lesions from level C2 to the conus medullaris were detected. As a result of suspected demyelinating disease, intravenous methylprednisolone therapy was administered for 7 days at a dose of 1 g/day. There was no change in cognitive findings, partial recovery was observed in pyramidal signs and walking. Donepezil was started at 10 mg/day for cognitive symptoms.

The patient was diagnosed as primary progressive MS during the follow-ups. Minimental state examination score was 21/30, digit span forward score was 5, digit span backward score was 3, verbal fluency was 11/min (perseveration, 0/min),

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phonemic fluency was (character K) 5/min (perseveration, 0/min), abstraction was 3/3, clock drawing score was 7/10, verbal learning test score at 5th min was 2/3, and recall memory score was 2/3 at control neurocognitive assessment performed after 6 months.

Similar to other cortical MS patients in the literature, this case started with complaints of forgetfulness and showed a progressive nature. Besides the impairment of memory, attention and executive function were observed at neuropsychological assessment (2). We observed in this case, similar to that observed in the literature, increased latency of VEP without visual complaints (3). Negative oligoclonal bands have been reported at 16% in the literature in cortical MS patients, and the patient in this case also had negative oligoclonal bands. Unfortunately, there have been limited studies about treatments of CD in MS. There are conflicting results in the studies of donepezil's effect on cognitive function in MS, but predominant effects were reported as positive (4,5,6,7,8). On the other hand, rivastigmine, memantine, and ginkgo biloba did not show any beneficial effect on CD in MS (9).

This patient had benefited from treatment with donepezil for CD in MS. This case and other cases reported in the literature were evaluated; finally, we concluded that donepezil may positively effect CD in MS, and there is a need for detailed, clearly defined and planned long-term studies.

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