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Is Corticospinal Tract Degeneration Caused by Sjögren Syndrome?

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Dear Editor,

A 36-year-old woman with a history of dry eyes and recurrent oral ulcers presented with an 8-month history of progressive motor weakness of both legs. She was documented as experiencing mild weakness (4+/5) of both legs with no sensory loss 2 months after onset. Cervical MRI revealed a T2 hyperintense lesion from C2 to C7 without cord swelling or contrast enhancement (Fig. 1A). The rest of the spinal cord and the brain MRI were normal. A cerebrospinal fluid (CSF) examination was unrevealing. She was treated with intravenous and oral corticosteroids based on the presumptive diagnosis of transverse myelitis (TM).

Her motor weakness continued to worsen. On admission examination, she showed spastic paraplegia with 3/5 strength with no sensory or bladder/bowel signs or symptoms. Her deep tendon reflexes were all brisk, and the Babinski sign was positive bilaterally. Follow-up cervical MRI revealed focal areas of atrophy along the bilateral lateral columns, with sparing of the posterior columns (Fig. 1B). Workup to exclude metabolic or infectious causes of myelitis were unremarkable. Electromyography findings excluded amyotrophic lateral sclerosis, and no pathologic variants related to hereditary spastic paraplegia were found on whole-exome sequencing. Serologic studies revealed positive anti-Ro/SSA (102.3 U/mL), IgG β2-glycoprotein 1 antibody, and anticardiolipin antibody, and negative for the AQP4 antibody. The ocular staining scores were 4/3 in both eyes, and salivary scintigraphy showed delayed excretion in the parotid and submandibular glands. A salivary gland biopsy confirmed chronic lymphocytic sialadenitis, consistent with primary Sjögren syndrome (pSS). The patient was ultimately diagnosed with progressive cervical myelopathy due to pSS.

Her paraplegia gradually worsened (1/5) despite continued immunotherapy including corticosteroids and cyclophosphamide. Cervical MRI performed 1 year later revealed prominent atrophy in the regions of the lateral and anterior corticospinal tracts (CSTs) bilaterally (Fig. 1C). Her symptoms were confined to pure motor paraplegia during the 5-year follow-up after the treatment.

This case fulfilled the diagnostic criteria for definite pSS,[1] with selective atrophy of CSTs following chronic progressive cervical myelopathy, which has not been reported previously. In previous reports of myelitis due to pSS, patients presented with various courses including chronic progressive myelitis, acute or subacute TM, and multiple sclerosis-like diseases.[2] There is one previous report of pSS presenting as selective atrophy of dorsal columns within the cervical cord.[3] The mechanism underlying spinal cord atrophy in the present case might have been Wallerian degeneration or delayed axonal loss following demyelination from chronic progressive cervical myelitis.[3,4] Notably, the MRI and CSF examinations revealed no evidence of inflammation, and her disease course appeared to be unresponsive to corticosteroids, which suggests noninflammatory mechanisms as work.

There are numerous accounts of neurodegeneration in pSS and other systemic rheumatologic diseases without evidence of inflammation,[5] suggesting that pSS could alone have

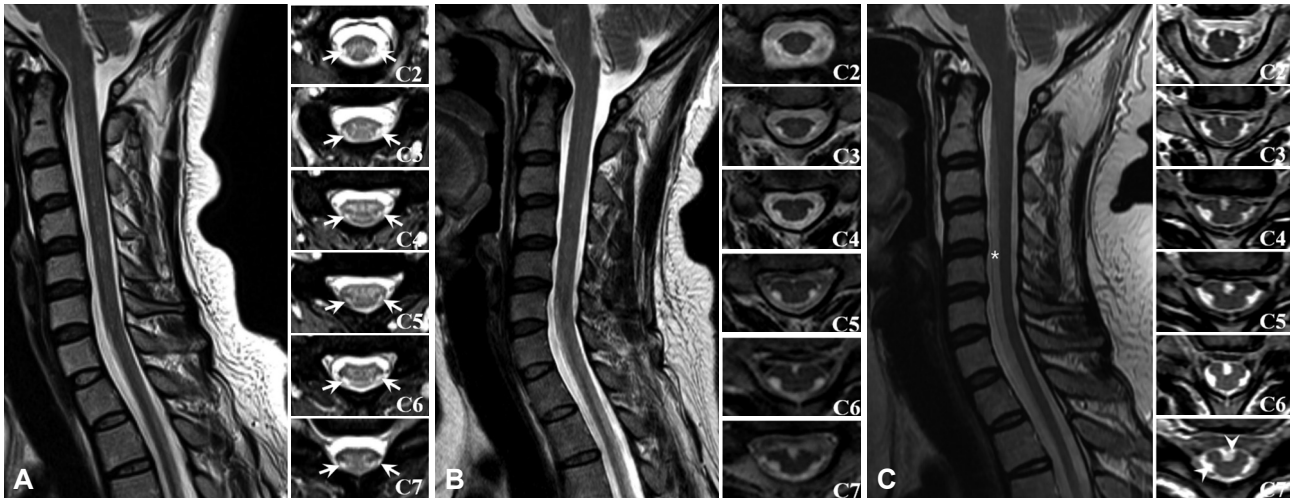

Fig. 1. Cervical MRI scans of the patient. A: At 2 months after symptom onset, a T2-weighted sagittal image shows no significant abnormality, but T2-weighted axial images show suspicious focal hyperintensities (arrows) in lateral motor tracts bilaterally in the cervical spinal cord. B: MRI images obtained 8 months after the onset show spinal cord atrophy primarily of the lateral motor tracts, with preservation of the posterior columns. C: At 20 months after the onset, a T2-weighted sagittal image shows a linear hyperintensity (asterisk) extending to the upper thoracic levels of the vertebral column (from C2 to T1), and T2-weighted axial images show more-prominent atrophy in both the lateral and ventral columns (arrow heads), which correspond to the area of lateral and anterior corticospinal tracts.

been responsible for the selective noninflammatory CST degeneration in this case.[5] Additional possibilities include vascular insufficiency or primary lateral sclerosis (PLS). pSS-associated vascular myelopathy is more compatible with an etiology of pure motor paraparesis following selective degeneration of CSTs. CSTs in the spinal cord have greater spinal cord blood flow and metabolic activity, which make them more vulnerable to ischemic injury.[6] This case also technically fulfills the diagnostic criteria for PLS proposed by Pringle et al.[7] However, this case seems less likely to be PLS considering the young age at onset, rapid worsening of paraplegia and CSTs atrophy, lack of progression to other body regions, and the presence of another disease (pSS) that was more like to underlie the myelopathy.[8]

The study protocol was approved by the Institutional Review Board at Chonnam National University Hospital, and the subject consented to the publication of her case.

Conflicts of Interest

The authors have no financial conflicts of interest.

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