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Isolated Bilateral Superior Cerebellar Peduncular Lesion Presenting Square-Wave Jerks and Ataxia

Jooheon Kong[a]
Sun-Uk Lee[a]
Sungwook Yu[a]
Ji-Soo Kim[b,c]

[a]Department of Neurology, Korea University Medical Center, Seoul, Korea
[b]Department of Neurology, Seoul National University College of Medicine, Seoul, Korea
[c]Dizziness Center, Seoul National University Bundang Hospital, Seongnam, Korea

Dear Editor,

The superior cerebellar peduncle (SCP), also known as the brachium conjunctivum, is the main conduit for the cerebellar efferent fibers.[1] Lesions that involve the SCP can induce various neurotologic abnormalities, including central positional nystagmus,[2] saccadic contrapulsion,[3] and ocular tilt reaction.[4] We present a patient with frequent square-wave jerks (SWJs) as well as limb and truncal ataxia due to a discrete lesion involving both SCP in isolation.

A 75-year-old man with chronic kidney disease, hypertension, and diabetes mellitus presented with disequilibrium and dysarthria for 3 days. The patient had a pontine infarction presenting with dysarthria 16 years previously, but without neurologic sequelae. He experienced a brief loss of consciousness for less than 1 min while walking 3 days before the presentation, since when he had been dysarthric and unable to walk on his own.

Examination revealed SWJs during visual fixation in a dimly lit room. Bedside head impulse tests were normal. The patient showed exophoria of 6 prism diopters during primary gaze, and convergence was impaired. The patient had profound truncal ataxia and was unable to stand or walk even with a support. He also showed limb ataxia in all extremities, which was more profound on the left side. The patient showed no motor weakness, sensory changes, or Horner syndrome.

Video-oculography documented frequent horizontal SWJs at an abnormally high rate of 111.6/min without visual fixation (Fig. 1A). Right-beating nystagmus was elicited at a peak velocity of 7.1°/s after head shaking, but there was no nystagmus during positional maneuvers. Horizontal and vertical saccades were hypometric but with a normal velocity (Fig. 1B). Smooth pursuit was impaired in both horizontal (pursuit gain of 0.33 to the right and 0.27 to the left for a moving target with a peak velocity of 10°/s) and vertical directions (0.32 upward and 0.35 downward). Brain MRIs revealed discrete lesions that involved the bilateral SCPs in isolation (Fig. 1C). Holter monitoring and echocardiography did not yield any evidence of cardioembolism. Cerebrospinal fluid analyses and serologic tests did not reveal any infectious, autoimmune, or demyelinating diseases. Under the suspicion of infarction, the patient received 100 g of aspirin and 40 mg of atorvastatin. The patient became able to walk with a cane 2 weeks later. Follow-up video-oculography performed 6 months later indicated partial improvement of the SWJs (Fig. 1A). The patient did not develop tremor, rigidity, bradykinesia, or oculopalatal tremor initially or during the follow-up.

Our patient presented frequent SWJs, hypometric saccades, and profound limb and truncal ataxia due to lesions that involved the bilateral SCPs.

The SCP also contains efferent fibers from the cerebellar nuclei to the red nucleus, thalamus, and pontine reticular nuclei, which are crucially important in equilibrium and locomotion.[1] Similar to our findings, mechanical severance of bilateral SCPs resulted in profound truncal and limb dysmetria in monkeys, probably due to disconnection between the cerebellar nuclei and mesencephalic locomotor region.[5]

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Correspondence
Sun-Uk Lee, MD, PhD
Department of Neurology, Korea University Medical Center, 73 Goryeodae-ro, Seongbuk-gu, Seoul 02841, Korea
Tel +82-2-2199-3808
E-mail sulee716@gmail.com

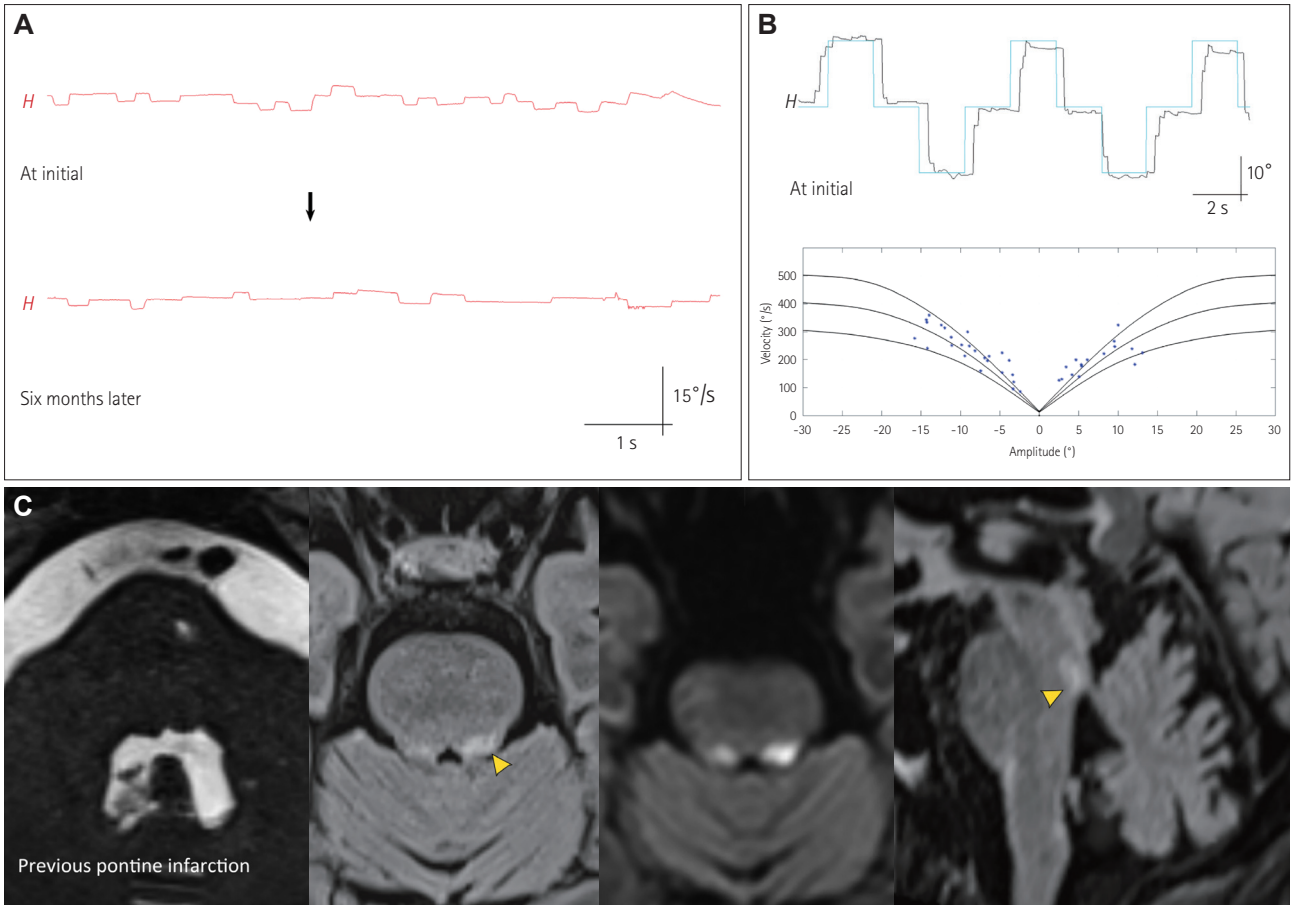

Fig. 1. Findings of the patient. A: Video-oculographic recording (SLVNG, SLMED, Seoul, South Korea; 120 Hz sampling rate) of the frequent square-wave jerks (SWJs) without visual fixation. Frequent SWJs were observed at a frequency of 111.6/min and an amplitude to 1.32° without visual fixation (83.7/min and 0.98° with fixation) at the initial presentation. The frequency of the SWJs was then decreased to 65.2/min (45.0/min with fixation) at the 6-month follow-up. B: Horizontal saccades were hypometric with normal velocities, but were frequently interrupted by SWJs. C: Fluid-attenuated inversion-recovery and diffusion-weighted images revealed discrete lesions that involved the bilateral superior cerebellar peduncles (yellow arrowheads).

SWJs refer to spontaneous horizontal conjugate saccades moving away from the intended fixation position followed by a return to that point with an intersaccadic interval of 200 ms.[6] They are frequently found in healthy subjects, and are known to increase with aging (mean frequency of 4.7/min in young subjects and 27/min in the elderly).[7] SWJs indicate fixation instability in the saccadic system and have been ascribed to insufficient tonic inhibition of the omnipause cell activity in pathologic conditions.[6] They have been observed in various diseases such as progressive supranuclear palsy, multiple sclerosis, and Arnold-Chiari malformation.[6] The mechanism of SWJs has been hypothesized as disconnection between the burst neurons and the superior colliculus, or the fastigial nucleus that normally chokes off drive to the saccadic burst neurons.[8] However, these burst neurons form a positive feedback loop in the brainstem, and disconnecting from these afferents often induces saccadic oscillation without intersaccadic intervals,[9] which did not occur in our patient. Alternately, the long-lead burst neuron (LLBN) projection may be disrupted by the bilateral pontomesencephalic tegmental lesion that normally turns off the omnipause neurons. In this way, sustained inhibition of the excitatory and inhibitory burst neurons is not maintained when steady fixation is required.[10] Our findings suggest that the disrupted connection between the saccadic premotor regions—such as the cerebellar fastigial nucleus, superior colliculus, or the LLBN projection in the pontomesencephalic junction—and the brainstem ocular motor network can lead to increased SWJ occurrence.

While an infectious, inflammatory, or autoimmune disorder could still have been the cause in our patient, an infarction may be inferred as the presumed etiology of the SCP lesion. The transient loss of consciousness may indicate an embolism or stenosis in the basilar artery and occlusion of the perforating arteries. The SCP is irrigated by the long circumferential arterial branches that arise from the superior cerebellar artery (SCA) or posterior cerebral artery (PCA).[1] The vascular sup-

ply to the SCP may differ among individuals depending on the arterial network formed by the SCA and PCA that further branches off the choroidal and quadrigeminal arteries.[1] The findings in our patient imply that an isolated lesion involving the bilateral SCPs can impair the modulatory role of the cerebellum in stable fixation and balance.

Ethics Statement

This study followed the tenets of the Declaration of Helsinki and was performed according to the guidelines of the Institutional Review Board of Korea University Anam Hospital (2022AN0045). This study was approved with a waiver of informed consent by the Institutional Review Board of Korea University Anam Hospital.

Availability of Data and Material

The datasets generated or analyzed during the study are available from the corresponding author on reasonable request.

ORCID iDs

Jooheon Kong	https://orcid.org/0000-0001-6589-762X
Sun-Uk Lee	https://orcid.org/0000-0001-8660-7759
Sungwook Yu	https://orcid.org/0000-0002-4224-4025
Ji-Soo Kim	https://orcid.org/0000-0002-1508-2024

Author Contributions

Conceptualization: Sun-Uk Lee. Data curation: Jooheon Kong, Sun-Uk Lee. Formal analysis: Jeeheon Kong, Sun-Uk Lee, Ji-Soo Kim. Funding acquisition: Sun-Uk Lee. Investigation: Jooehon Kong, Sungwook Yu. Supervision: Sun-Uk Lee, Sungwook Yu, Ji-Soo Kim. Visualization: Jooheon Kong. Writing—original draft: Jooheon Kong, Sun-Uk Lee. Writing—review & editing: all authors.

Conflicts of Interest

Drs. J Kong, SU Lee, and S Yu report no disclosures. Dr. JS Kim serves as an Associate Editor of *Frontiers in Neuro-Otology* and on the editorial boards of the *Journal of Clinical Neurology*, *Frontiers in Neuro-Ophthalmology*, *Journal of Neuro-Ophthalmology*, *Journal of Vestibular Research*, *Medicine*, and *Clinical and Translational Neuroscience*. Ji-Soo Kim, a contributing editor of the *Journal of Clinical Neurology*, was not involved in the editorial evaluation or decision to publish this article.

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