Surgical Treatment of a Patient with Prolonged Exacerbation of Hirayama Disease

Sho Dohzono[1], Hiromitsu Toyoda[2], Akiko Tamura[3], Kazunori Hayashi[2], Hidetomi Terai[2] and Hiroaki Nakamura[2]

1) Department of Orthopaedic Surgery, Yodogawa Christian Hospital, Osaka, Japan
2) Department of Orthopaedic Surgery, Osaka City University Graduate School of Medicine, Osaka, Japan
3) Department of Neurology, Sumitomo Hospital, Osaka, Japan

Juvenile muscular atrophy of the distal upper extremity, known as Hirayama disease, typically progresses before stabilization, with the clinical course plateauing within 5 or 6 years[1-3]. We present a case of surgical treatment for Hirayama disease >30 years after the onset of symptoms.

A 48-year-old woman presented with progressive weakness of the right distal upper hand and forearm. Her symptoms began at the age of 15 years with coldness in the right hand. She had difficulty in extending her right hand fingers and experienced a clumsy hand during a cold weather 3 years later. Exacerbation of muscle weakness proceeded, and she experienced slow progressive weakness of the proximal lower ipsilateral limb, for which she visited our institution. Neurological examination revealed marked atrophy of the right distal upper extremity and claw hand deformity (Fig. 1). The deltoid, biceps, and brachioradialis were normal. The deep tendon reflexes were sluggish in the affected upper limb, but they were brisk in the bilateral lower extremities. Moderate to severe sensory disturbance was observed on the ulnar side of the affected forearm and hand, but her sensory modalities were normal in the other extremities.

Electromyography demonstrated severe chronic neurogenic changes characterized by high-amplitude, long-duration motor unit potentials in the affected muscles. Conduction velocity and amplitude of the sensory nerve in the bilateral median, ulnar, tibial, and sural nerves were normal. Sensory evoked potentials (SEPs) were examined using previously described methods[4]. Her median and tibial nerve SEPs indicated that her central sensory conduction times were normal (Fig. 2). Cervical magnetic resonance imaging (MRI) showed asymmetric anteroposterior flattening with high-intensity areas on T2-weighted images at the C4-5, C5-6, and C6-7 levels. Cervical MRI in the flexion position revealed a forward shift of the spinal cord without displacement of the posterior dural wall, and the spinal cord was compressed at the C4-5 and C5-6 levels (Fig. 3).

Anterior discectomy and fusion at C4-5 and C5-6 were performed. Eight degree lordotic polyetheretherketone cages were placed in each interbody space to achieve cervical lordosis. Cervical MRI in the flexion position showed no evidence of spinal cord compression by the vertebral bodies 6 months postoperatively (Fig. 4). Two years postoperatively, her right leg had regained full strength, but atrophy of the right upper extremity and claw hand deformity remained.

The pathological mechanism of Hirayama disease is considered to be repetitive compression of the anterior cord due to forward displacement of the posterior dural sac on neck flexion[2,5]. Hirayama disease progresses before becoming stable, with the clinical course plateauing within 5 or 6 years in majority patients[1-3]. However, we are not aware of cases with a highly unusual progressive course of >30 years that was observed in our patient.

The present study did not show displacement in the posterior wall of the dura mater in the later stage of the disease, although it is unknown whether a tight dural canal presented in the early stage of the disease. A previous study has reported that neurological symptoms were severe in patients with a tight dural canal on flexion compared with those in patients without a tight dural canal[6]. These findings may help in explaining how Hirayama disease without a tight

Figure 1. The right hand showed severe atrophy of the intrinsic muscles and claw deformity.
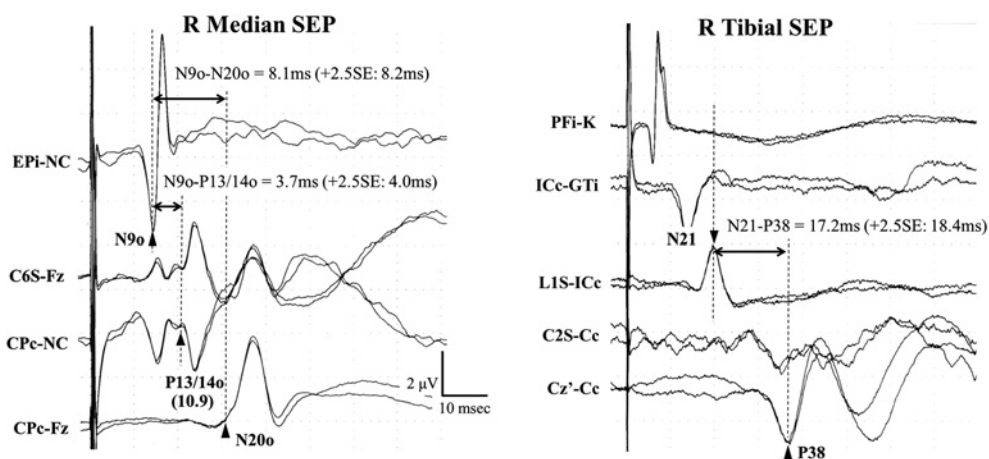
Figure 2. Somatosensory evoked potential of the right median nerve and right tibial nerve. Central sensory conduction times (CSCT), such as conduction times of N9o-P13/14o, N9o-N20o, and N21-P38 intervals, were normal. The normal limit value of CSCT was defined as between −2.5 and +2.5 SE.

Figure 3. Preoperative T2-weighted cervical MRI.
Mid-sagittal in neutral position (A); axial at the C4-C5 (B), C5-6 (C), and C6-7 levels (D); and mid-sagittal in flexion (E) and extension position (F). Segmental kyphotic angles in flexion were calculated by drawing lines through the center of each vertebra perpendicular to the long axis of each vertebra.
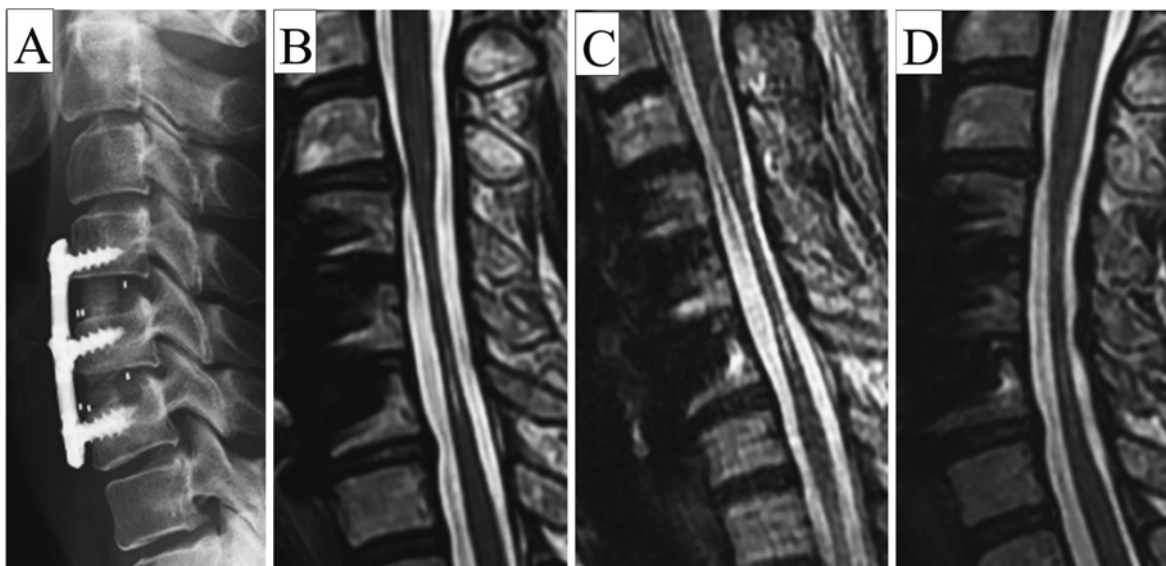
Figure 4. Postoperative X-ray on lateral view (A) and mid-sagittal T2-weighted MRI in neutral (B), flexion (C), and extension positions (D).

dural canal can progress very slowly. Therefore, neurological deterioration in our patient for >30 years might be due to excessive local kyphosis in flexion without a tight dural canal.

In conclusion, the present case suggests that hyperflexion without a tight dural canal is a significant factor in determining the long-term prognosis in patients with juvenile muscular atrophy of the unilateral distal upper extremity.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Author Contributions: S. Dohzono wrote and prepared the manuscript, and all authors participated in preparing the study design. All authors have read, reviewed, and approved the article.

Informed Consent: The patient was informed that data concerning the case would be submitted for publication, and she provided consent.

References

1. Tashiro K, Kikuchi S, Itoyama Y, et al. Nationwide survey of juvenile muscular atrophy of distal upper extremity (Hirayama disease) in Japan. Amyotroph Lateral Scler. 2006;7(1):38-45.
2. Hirayama K. Juvenile muscular atrophy of distal upper extremity (Hirayama disease). Intern Med. 2000;39(4):283-90.
3. Zhou B, Chen L, Fan D, et al. Clinical features of Hirayama disease in mainland China. Amyotroph Lateral Scler. 2010;11(1-2): 133-9.
4. Miura T, Sonoo M, Shimizu T. Establishment of standard values for the latency, interval and amplitude parameters of tibial nerve somatosensory evoked potentials (SEPs). Clin Neurophysiol. 2003; 114(7):1367-78.
5. Iwasaki Y, Tashiro K, Kikuchi S, et al. Cervical flexion myelopathy: a "tight dural canal mechanism." Case report. J Neurosurg. 1987;66(6):935-7.
6. Kitagawa M, Tashiro K, Kikuchi S, et al. [Correlation between clinical features and neuroradiological findings in juvenile muscular atrophy of unilateral upper extremity (Hirayama disease)--with and without "tight dural canal in flexion".] Rinsho shinkeigaku = Clinical neurology. 1992;32(5):479-82. Japanese.