

CASE REPORT



Finger trembling improvement after surgery in Hirayama disease: a case report

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INTRODUCTION: Finger trembling is a characteristic physical finding in Hirayama disease. Although conservative treatment is recommended to stop disease progression, surgery is optional in some cases. However, the postoperative recovery of finger trembling is scarcely reported.

CASE PRESENTATION: A 26-year-old Japanese female patient whose chief complaint was left finger trembling with active finger extension presented at our hospital. Hand weakness without muscle atrophy of the left arm was observed. MRI showed left-side oriented intramedullary signal change with concomitant cord atrophy at C4-5 and C5-6. The CT myelogram (CTM) on flexion showed anterior cord compression and anterior shift of posterior dura matter from C4 to C6. And CTM on extension showed the resolution of both findings. Electrophysiological studies showed active and chronic neuronal damage and preserved motor neuron pool of hand muscle. Since she had exhibited a gradual aggravation of symptoms over a period of 5 years, she underwent anterior cervical discectomy and fusion after careful assessment of both conservative and surgical treatment. Finger trembling recovered soon after surgery.

DISCUSSION: Finger trembling is an unfamiliar physical finding in terms of postoperative recovery prediction. Anterior horn cell impairment is postulated as a cause of finger trembling. Postural restoration of spinal cord shape and cerebrospinal fluid around the cord with preserved neural function could facilitate functional recovery.

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INTRODUCTION

Juvenile muscular atrophy of the unilateral upper extremity (Hirayama disease) is a rare disease entity, first reported in 1959 [1]. A unilateral upper arm weakness and atrophy develop during adolescence and lead to spontaneous arrest, several years after onset. Due to its natural history, conservative treatment using a cervical collar is adopted in the early stages of the disease to prevent it from progressing any further [2]. On the contrary, surgical treatment has also been reported as a treatment option and researchers have described functional recovery such as grip power improvement after surgery [3–5].

Finger trembling [6] or polyminimyoclonus [7] aggravated by active finger extension is a well-known physical feature of Hirayama disease. However, the treatment response of finger trembling has not been reported. We herein describe a case of improved finger trembling after surgery.

Illustrative case

A 26-year-old, right-handed Japanese female was referred to our hospital. She complained of a gradual aggravation of left finger trembling over a period of 5 years without specific diagnosis. The patient worked as an office worker and tended to have neck flexion posture due to deskwork at a computer. She experienced finger trembling, which intermittently became severe in recent

years. Her physical findings demonstrated a severe weakness (MMT 2 out of 5) in the left-hand intrinsic muscles such as the first dorsal interosseous (FDI) muscle and second palmar interosseous muscle. Other muscles displaying weakness were the extensor digitorum communis (MMT 4/5) and abductor pollicis brevis (MMT 4/5). We observed fine finger trembling during finger extension on the middle and ring fingers (Video 1). The patient was unable to keep active left finger adduction on the left side (positive finger escape sign) [8]. The results of a 10-s grip and release test on the right and left side were 25 and 10, respectively. The grip powers were 28 and 18 kg on the right and left sides. However, the atrophy of the forearm muscle, so-called “oblique amyotrophy [9],” was not evident. The circumferences of the right and left forearms were 22.5 and 21.5 cm, respectively. In addition, hand intrinsic muscle atrophy was not apparent. She also complained of left-hand ulnar side numbness which had gradually developed over the past 6 months. The stretch reflexes were normal and no pathological reflexes such as Babinski reflex were observed.

Flexion and extension radiographs showed hypermobility of the subaxial cervical spine; the C2-7 Cobb angle was a 57-degree kyphotic angle on flexion and a 58-degree lordotic angle on extension. T2-weighted 1.5T-MRI demonstrated focal intramedullary high signal changes with concomitant cord atrophy at C4-5 and C5-6; both located only on the left side (Fig. 1). The CT

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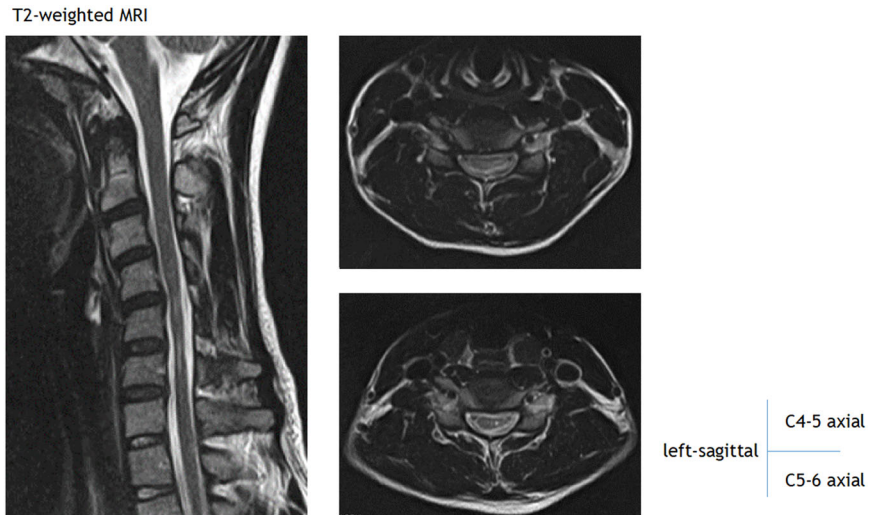


Fig. 1 The preoperative MRI. The left panel shows the intramedullary signal changes at C4-5 and C5-6. The right upper and lower panels respectively show intramedullary high intensity changes with concomitant cord atrophy located on the left side.

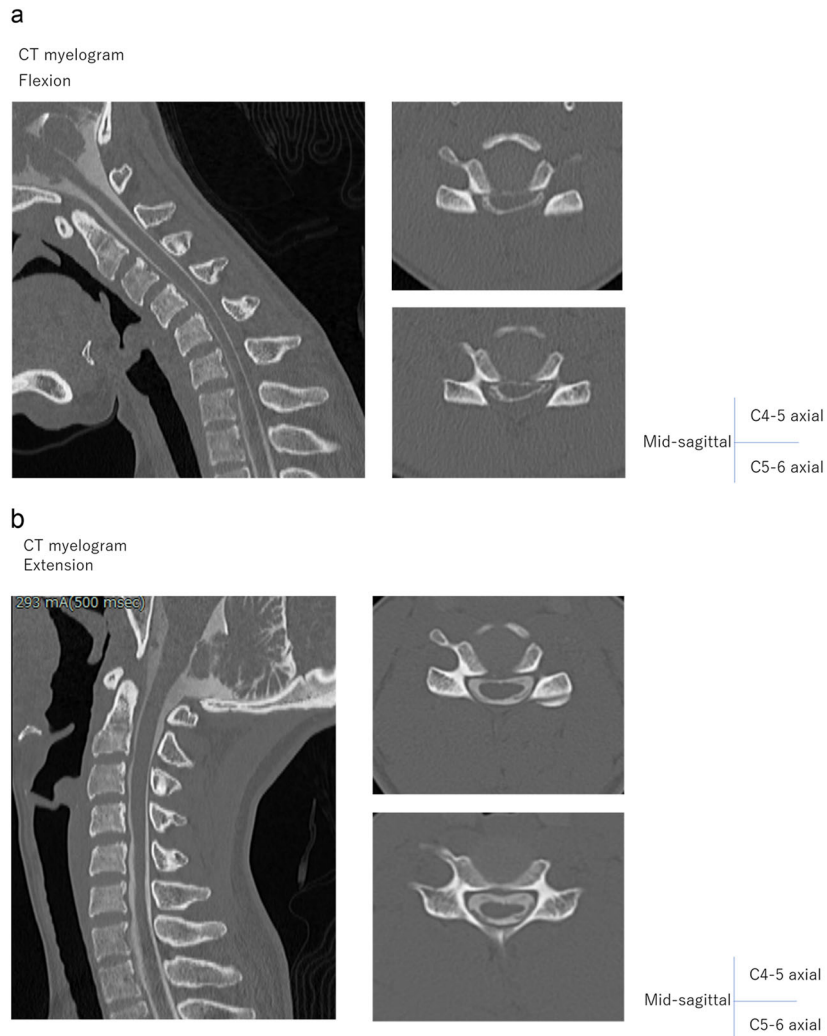


Fig. 2 CT myelogram with flexion and extension. **a** The left panel shows the spinal cord compression due to anterior structures: vertebrae and disc, and anterior shifting of the posterior dural sac with epidural space enlargement on flexion posture. The right upper and lower panel show the disappearance of anterior cerebrospinal fluid (CSF) space and a flattening of the spinal cord. **B** The left panel shows resolution of the spinal cord compression in the extended position. The right upper and lower panel also show CSF surrounding the spinal cord circumferentially at C4-5 and C5-6.

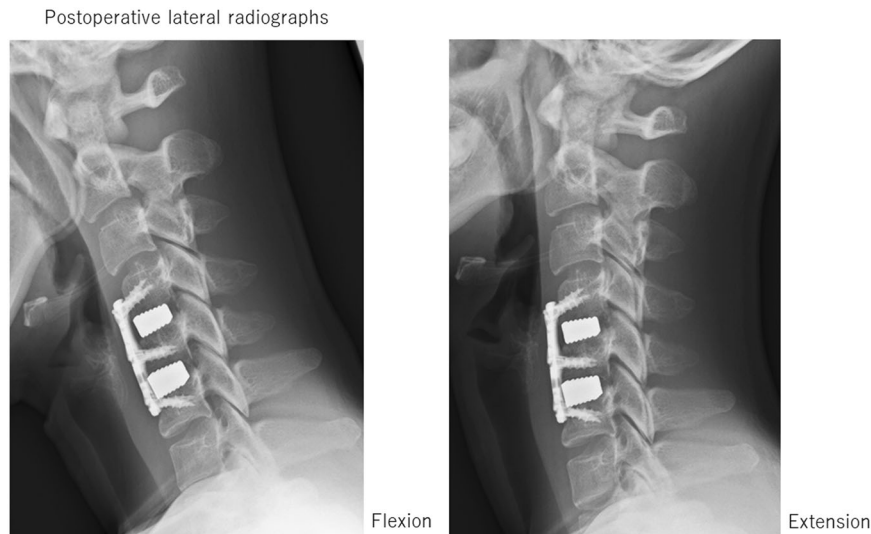


Fig. 3 The radiographs taken twelve months after surgery. The left and right panels respectively show the postoperative flexion and extension lateral radiographs. Both C4-5 and C5-6 levels were successfully fused.

myelogram (CTM) with flexion showed the spinal cord flattening due to anterior compression and the cerebrospinal fluid disappearance in front of the spinal cord from C4-5 to C5-6. The spinal cord flattening and the cerebrospinal fluid space in front of the spinal cord were both restored with extension (Fig. 2a, b). Both direct cord compression by anterior structures such as the vertebral bodies and intervertebral discs and indirect cord compression by the anterior shift of posterior dura matter making the dural canal tight were considered as the lesions responsible for the patient's condition. An electrophysiological study supported the left-sided anterior horn cell (AHC) damage; needle EMG showed acute and chronic denervation findings in the left FDI muscle. Fortunately, the amplitude of compound muscle action potentials (CMAPs) had been maintained; amplitude on the affected side of the abductor digiti minimi and abductor pollicis brevis were equivalent to those on the normal side. Thus, this patient was diagnosed with Hirayama disease. Ulnar and median nerve sensory nerve action potentials also showed no side-to-side difference, which suggested left-sided spinal cord damage as the cause of numbness.

Although conservative treatment using a cervical collar to avoid neck flexion was the first-line treatment with less morbidity, it was reported to be effective in patients with a shorter disease history (under 2.5 years) and no or mild cord atrophy [2]. Since this patient had a longer disease history (over 5 years) and left-sided unilateral cord atrophy with intramedullary signal change (Fig. 1), the conservative treatment might be less effective. Considering the gradual aggravation of motor and sensory symptoms and the unfixed term for wearing the neck collar, the surgical treatment to decompress the spinal cord and to avoid further neural damage was the treatment of choice. She agreed to undergo surgery: anterior cervical discectomy with fusion (ACDF) at C4-5 and C5-6, after discussing with her family and herself the pros and cons of surgical treatment including peri-operative complications and long-term sequelae.

Soon after surgery, the finger trembling almost disappeared (Video 2). Other symptoms such as the positive finger escape sign and the stiffness of grip-and-release had also been alleviated. Postoperative radiographs and MRI respectively showed the corrected cervical spine alignment and well decompressed spinal cord with the residual intramedullary signal change and cord atrophy at C4/5 and C5/6.

Twelve months after surgery, she has maintained good hand function without any recurrent symptom. Flexion and extension

radiographs showed that solid fusion was achieved without correction loss (Fig. 3).

DISCUSSION

This case demonstrates a successful example of improvement in finger trembling after surgery. Hirayama disease typically manifests as a unilateral hand muscle weakness and atrophy and is regarded as a non-progressive disease. However, there have been reports of cases suggesting the heterogeneity of Hirayama disease and its related flexion myelopathy such as bilateral form of upper arm weakness or late onset aggravation of myelopathy [4]. Therefore, what constitutes the optimal treatment for the patient remains controversial. Conservative treatment using a cervical collar to limit neck flexion was reported to halt progression of the disease [2, 10]. Early detection of disease is important for the success of conservative treatment. On the other hand, surgical treatments such as anterior cervical decompression with fusion, posterior cervical decompression and fusion and duraplasty were also reported as a useful alternative treatment [4]. Muscle strength such as grip power improved after surgery, although hand muscle atrophy persisted [3–5]. The indications for surgical treatment are still debatable. Thus, both conservative and surgical treatment should be considered depending on each patient's condition.

In this case, muscle wasting in the hand and forearm was not apparent despite a 5-year disease history. Fluctuating finger trembling over the years was the chief complaint. MRI and CTM on flexion depict the intramedullary lesion corresponding to the hand symptoms and anterior cord compression with the anterior shift of posterior dural sac, respectively. EMG abnormalities were also consistent with the spinal cord damage. The diagnosis was rather straightforward. On the other hand, the postoperative recovery of finger trembling, an unfamiliar physical finding to spine surgeons, was of concern.

Several papers mentioned the involuntary, jerky, irregular characteristic tremor-like finger movement using different terms indicating the same finding such as finger trembling [6], polyminimyoclonus [11], and polymyoclonus [12]. Maintaining a posture of outstretched hand or action such as active finger extension triggers the abovementioned finding [13]. The hyperexcitability of AHCs was reported to account for the fasciculation [13]. CTM on flexion and extension posture delineate dynamic cord compression, exaggerated by flexion posture. The reported normal flexion angle in Japanese females in their 20 s was $28.8 \pm$

10.8 degree [14]. This patient demonstrated a large cervical flexion angle, in other words, a kyphotic angle. The large flexion angle and kyphotic posture raised the greater stretching effect on the spinal cord which lead to gray matter damage [15] or the loss of AHCs [16]. Fortunately, CMAP amplitude of involved C8 and Th1 myotome was preserved [17]. These imaging and electrophysiological findings suggested AHC damage with preserved neural function and surgically correctable conditions. ACDF could restore the cervical alignment and relieve the dynamic impairment and static cord compression.

CONCLUSION

Finger trembling is an unfamiliar physical finding in terms of postoperative recovery prediction. AHC impairment is postulated as a cause of finger trembling. The postural restoration of spinal cord shape and cerebrospinal fluid around the cord with preserved neural function could facilitate functional recovery.

DATA AVAILABILITY

The datasets generated and/or analyzed during the current study are available from the corresponding author upon reasonable request.

REFERENCES

- Hirayama K, Tsubaki T, Toyokura Y, Okinaka S. Juvenile muscular atrophy of unilateral upper extremity. *Neurology*. 1963;13:373–80.
- Tokumaru Y, Hirayama K. Cervical collar therapy for juvenile muscular atrophy of distal upper extremity (Hirayama disease): results from 38 cases. *Rinsho Shinkeigaku*. 2001;41:173–8.
- Kato Y, Imajo Y, Kanchiku T, Kojima T, Kataoka H, Taguchi T. Dynamic electrophysiological examination of cervical flexion myelopathy. *J Neurosurg Spine*. 2008;9:180–5.
- Fujimori T, Tamura A, Miwa T, Iwasaki M, Oda T. Severe cervical flexion myelopathy with long tract signs: a case report and a review of literature. *Spinal Cord Ser Cases*. 2017;3:17016.
- Watanabe K, Hasegawa K, Hirano T, Endo N, Yamazaki A, Homma T, et al. Anterior spinal decompression and fusion for cervical flexion myelopathy in young patients. *J Neurosurg Spine*. 2005;3:86–91.
- Tashiro K, Kikuchi S, Itoyama Y, Tokumaru Y, Sobue G, Mukai E, et al. Nationwide survey of juvenile muscular atrophy of distal upper extremity (Hirayama disease) in Japan. *Amyotroph Lateral Scler*. 2006;7:38–45.
- Meng D, Ghavami K, Chen T. Polyminimyoelonus in Hirayama disease. *BMJ Case Rep*. 2021;14:e246831. <https://doi.org/10.1136/bcr-2021246831>.
- Ono K, Ebara S, Fujii T, Yonenobu K, Fujiwara K, Yamashita K. Myelopathy hand. New clinical signs of cervical cord damage. *J Bone Jt Surg Br Vol*. 1987;69:215–9.
- Huang YL, Chen CJ. Hirayama disease. *Neuroimaging Clin N Am*. 2011;21:939–50.
- Kieser DC, Cox PJ, Kieser SCJ. Hirayama disease. *Eur Spine J*. 2018;27:1201–6.
- Misra UK, Kalita J, Mishra VN, Kesari A, Mittal B. A clinical, magnetic resonance imaging, and survival motor neuron gene deletion study of Hirayama disease. *Arch Neurol*. 2005;62:120–3.
- Ong JJ, Wang FS, Chan YC. Teaching neuroimages: a case of Hirayama disease presenting with polymyoclonus. *Neurology*. 2015;85:e156–7.
- Ganguly J, Chai JR, Jog M. Minipolymyoclonus: a critical appraisal. *J Mov Disord*. 2021;14:114–8.
- Yukawa Y, Kato F, Suda K, Yamagata M, Ueta T. Age-related changes in osseous anatomy, alignment, and range of motion of the cervical spine. Part I: radiographic data from over 1,200 asymptomatic subjects. *Eur Spine J*. 2012;21:1492–8.
- Kato Y, Kataoka H, Ichihara K, Imajo Y, Kojima T, Kawano S, et al. Biomechanical study of cervical flexion myelopathy using a three-dimensional finite element method. *J Neurosurg Spine*. 2008;8:436–41.
- Shimizu K, Nakamura M, Nishikawa Y, Hijikata S, Chiba K, Toyama Y, et al. Spinal kyphosis causes demyelination and neuronal loss in the spinal cord: a new model of kyphotic deformity using juvenile Japanese small game fowls. *Spine*. 2005;30:2388–92.
- Chiba T, Konoeda F, Higashihara M, Kamiya H, Oishi C, Hatanaka Y, et al. C8 and T1 innervation of forearm muscles. *Clin Neurophysiol*. 2015;126:837–42.

AUTHOR CONTRIBUTIONS

NT: conceptualization, writing—original draft. KH, KK, YK, NA, RT, MI: writing—review and editing.

COMPETING INTERESTS

The authors declare no competing interests.

ETHICAL APPROVAL

The patient was informed that data concerning the case would be submitted for publication, and provided her consent.

ADDITIONAL INFORMATION

Supplementary information The online version contains supplementary material available at <https://doi.org/10.1038/s41394-022-00514-w>.

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