Advanced Hirayama Disease with Successful Improvement of Activities of Daily Living by Operative Reconstruction

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Abstract

Juvenile muscular atrophy of the distal upper extremity (Hirayama disease) is a benign and non-progressive motor neuron disease. Application of a cervical collar is believed to prevent progression of symptoms in the early stages, but there is no effective therapy for the advanced disease. We found that tendon transfer improved the activities of daily living (ADL) of a patient with advanced Hirayama disease. An operative reconstruction can be valuable, even in patients with Hirayama disease who have developed impaired ADL due to extensive intrinsic hand muscle atrophy.

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Key words: Hirayama disease, advanced stage, tendon transfer, ADL

Introduction

Juvenile muscular atrophy of the distal upper extremity (Hirayama disease) is a sporadic juvenile-onset disease that presents with the gradual onset of unilateral weakness and atrophy in the hand and forearm muscles. Generally, this disorder is considered a benign, non-progressive motor neuron disease. Application of a cervical collar is believed to minimize neck flexion and prevent progression of muscle weakness and atrophy in the early stages, but there is no effective therapy for the advanced disease. We found that operative reconstruction by tendon transfer improved the activities of daily living (ADL) of a patient with advanced Hirayama disease who showed marked intrinsic muscle atrophy of the left hand.

Case Report

The patient was a right-handed 28-year-old. When he was eighteen years old, he developed slowly progressive muscle weakness of the left hand with focal atrophy of the ulnar side-forearm and intrinsic hand muscles for 6 months after the onset. Subsequently, he experienced cold paresthesia of his left forearm and hand during winter, and he underwent a left sympathetic nerve block under intrathoracic endoscopy, which produced a partial clinical improvement. In March 1999, he visited our clinic because of impairment of his routine work due to left finger clumsiness. On admission, he had significant atrophy of the left ulnar side-forearm muscles excluding the brachioradialis muscle. Atrophy of the intrinsic hand muscles including the interossei, thenar and hypothenar muscles was also marked. This characteristic distribution of the muscle atrophy was compatible with “oblique atrophy” (Fig. 1). There was neither fasciculation nor muscle cramp of the affected muscles. He showed bilateral fine postural hand tremor. Manual muscle testing disclosed 4-/5 strength in the left opponens pollicis and intrinsic hand muscles. His grip power was 20.8 kg (right) and 14.4 kg (left), respectively. Cold paresthesia of the left hand was also apparently observed. The rest of the neurological examination was entirely normal. Motor and sensory nerve conduction velocities were normal, however, the amplitude of compound muscle action potentials obtained from the left thenar and hypothenar muscles was significantly decreased. F wave analysis obtained by bilateral median nerve stimulation disclosed normal latencies and velocities, however, single high amplitude potentials were occasionally observed in the right abductor digiti minimi muscle. There was no increase of F wave persistency while the neck was flexed for 10 minutes. Motor evoked potentials of the bilateral abductor digiti minimi muscles induced by transcranial magnetic stimulation showed no

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amplitude change before or after neck flexion. An electromyogram demonstrated left-dominant neurogenic changes from bilateral forearm and hand muscles, which ranged from myotome C7 to Th1.

The cervical MR images in the neutral position revealed an atrophic change of the lower cervical cord. There was neither anterior shift of the dorsal cervical dura nor enlargement of the extradural space. Cerebrospinal fluid examination revealed a mild increase in protein and IgG concentration.

Based on the above findings, the patient was diagnosed as having advanced Hirayama disease. According to the electrophysiological findings, there was no indication for the neck collar treatment. From the aspects of the ADL of this patient, his main problem was due to impaired opposition of

the thumb (pinching) during fine intricate hand movements. Orthopaedic evaluation was carried out to assess the possibility of operative reconstruction. The patient had no contracture of the finger and wrist joints nor of the first web space. The main lesion of the left hand was sufficiently covered with soft tissues, and there was an available functioning muscle and tendon to restore opposition of the thumb. Finally, the patient was judged to be suitable for operative tendon transfer using the palmaris longus.

The operative procedure is shown in Fig. 2. The palm was incised as shown in the illustration, and a strip of palmar fascia in continuity with the palmaris longus was removed. A subcutaneous tunnel was created from the volar surface of the distal forearm to the tendon of the abductor pollicis brevis. The palmar fascia was then pulled into the short incision in the area of the thumb MP joint and sutured into position. After the operation, continuous rehabilitation therapy was provided. Evaluation of the left finger movements 1 year post surgery indicated improvements both in grip power of the left hand (from 14.4 to 20.2 kg) and in pinching power (from 0.3 to 0.8kg). The improvements of his ADL became particularly obvious in the left finger movements, e.g., pinching, turning over pages (Fig. 3). The patient was able to return to work as a clerk.

Discussion

Hirayama disease occurs predominantly in men in the second decade of life and presents with unilateral muscle atrophy and weakness involving the fingers, hands and medial forearm (1, 2). Most of the reported cases are from Asia and India, although there have been some reports from other countries (3). Regarding the etiology, recent radiological investigations have proved that compressive flattening of the lower cervical cord due to forward displacement of the cervi-

Figure 1. The characteristic distribution of the left ulnar side forearm and the intrinsic hand muscle atrophy was compatible with "oblique atrophy".

Figure 2. The surgical procedure for reconstruction of opposition of the left thumb (see text).

Figure 3. The improvements of the ADL became particularly obvious in the left finger movements, which were achieved by improved thumb opposition.

cal dural sac and spinal cord induced by neck flexion is responsible. These findings suggest that sustained or repeated neck flexion may cause ischemic changes in the cervical anterior horn (4). A cervical collar application to minimize neck flexion prevents progressive muscular weakness in the early stages of the disease. To determine the value of cervical collar application, electrophysiological studies, especially of F wave and/or motor evoked potentials is thought to be useful. In cases of Hirayama disease, F wave studies show minimal increase in latency, low persistency, and occasionally, rare single high amplitude potentials suggesting denervation/reinnervation of the corresponding cervical cord. While the neck is flexed, F wave persistency decreases in the progressive phase of the disease. These phenomena should be indicators to start and/or stop cervical collar therapy (5). Additionally, motor evoked potentials after transcranial magnetic stimulation show an increased latency and decreased amplitude, which is temporarily aggravated by neck flexion (6). These parameters not only suggest a pathogenetic role of neck flexion in this disease but also give a rational basis for neck collar therapy. In regard to surgery, Konno et al proposed the usefulness of cervical duraplasty with posterior spinal fusion in patients who showed no improvement after wearing a cervical collar for more than 3 months (7). This procedure should be applied only to patients whose radiological findings include compression of the middle and lower cervical cord against the vertebral body, narrowing of the ventral subarachnoid space, and a forward shift of the posterior wall of dura during neck flexion. According to the above criteria, the present case had neither an indication for neck collar application nor for cervical surgery.

Practically, the main problem for this patient was impaired opposition of the left thumb. Loss of ability to oppose the thumb to the fingers is a devastating functional loss to the hand. We proposed the tendon transfer as an operative reconstruction to improve the intricate finger movements. The requirements for tendon transfer are as follows: 1. The synergistic muscle is preserved. 2. The strength of the available muscle is more than 4/5. 3. The range of motion of transferred tendon and muscle resembles the affected muscle. 4. The operation is relatively simple and noninvasive (8). According to the above criteria, we decided the palmaris longus was available because the strength of this muscle was unchanged for several years. Finally, this patient underwent operative tendon transfer. Evaluation of the left finger movements 1 year after the operation indicated improvements of power both in grip and pinch. We conclude that tendon transfer was of significant benefit in this patient.

In summary, in the early stage of Hirayama disease, the application of cervical collar is necessary and cervical surgery should be carried out in some cases. As far as we know, operative reconstruction has not been reported in cases of advanced stage. We would like to emphasize that operative reconstruction can be valuable, even in patients with Hirayama disease who have developed impaired ADL due to extensive intrinsic hand muscle atrophy.

References

- 1) Peiris J, Seneviratne K, Wickremasinghe H, Gunatilake S, Gamage R. Non familial juvenile distal spinal muscular atrophy of upper extremity. *J Neurol Neurosurg Psychiatry* **52**: 314–319, 1989.
- 2) Sobue I, Saito N, Iida M, Ando K. Juvenile type of distal and segmental muscular atrophy of upper extremities. *Ann Neurol* **3**: 429–432, 1978.
- 3) Chan YW, Kay R, Schwartz MS. Juvenile distal spinal muscular atrophy of upper extremities in Chinese males: a single fibre electromyographic study of arms and legs. *J Neurol Neurosurg Psychiatry* **54**: 165–166, 1991.
- 4) Hirayama K, Tokumaru Y. Cervical dural sac and spinal cord in juvenile muscular atrophy of distal upper extremity. *Neurology* **54**: 1922–1926, 2000.
- 5) Kuwabara S, Nakajima M, Hattori T, Hirayama K. Electrophysiology of juvenile muscular atrophy of unilateral upper limb (Hirayama's disease). *Rinsho Shinkeigaku (Clin Neurol)* **39**: 508–512, 1999 (in Japanese, Abstract in English).
- 6) Shizukawa H, Imai T, Kobayashi N, Chiba S, Matsumoto H. Cervical flexion-induced changes of motor evoked potentials by transcranial magnetic stimulation in a patient with Hirayama's disease: juvenile muscular atrophy of unilateral upper extremity. *Rinsho Shinkeigaku (Clin Neurol)* **34**: 500–503, 1994 (in Japanese, Abstract in English).
- 7) Konno S, Goto S, Murakami M, Mochizuki M, Motegi H, Moriya H. Juvenile amyotrophy of the distal upper extremity: pathologic findings of the dura mater and surgical management. *Spine* **22**: 486–492, 1997.
- 8) Schneider LH. Tendon transfer in the upper extremity. in: *Rehabilitation of the Hand: Surgery and Therapy*. 3rd ed. Hunter JM, Schneider LH, Mackin EJ, Callahan AD, Eds. The C.V. Mosby Company, St. Louis, Mo, 1990: 669–675.