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 paresis 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 paresis 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
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 0.8 kg). 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-
Operative Reconstruction in Hirayama Disease

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