Yoshinori Fujimoto
Shinichi Oka
Nobuhiro Tanaka
Kohichiro Nishikawa
Hiroyuki Kawagoe
Itsushi Baba

Pathophysiology and treatment for cervical flexion myelopathy

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Abstract Previous studies have suggested that spinal cord compression by the vertebral bodies and intervertebral discs during neck flexion cause cervical flexion myelopathy (CFM). However, the exact pathophysiology of CFM is still unknown, and surgical treatment for CFM remains controversial. We examined retrospectively patients with CFM based on studies of the clinical features, neuro-radiological findings, and neurophysiological assessments. The objectives of this paper are to investigate the pathophysiology of CFM, and to examine an optimal surgical treatment. Twenty-three patients (20 male, three female) with age of onset ranging from 11 to 23 years (mean 15.7 years) were examined for the study. All patients were inspected by magnetic resonance imaging (MRI), myelogram, or computed tomographic myelogram (CTM) of the cervical spine. In eight patients, dynamic motor evoked potentials (MEP) studies were performed. Five patients underwent surgical treatment; two patients had cervical duraplasty with laminoplasty, two patients had musculo-tendinous transfer, one patient had both of these procedures, and the remaining 18 patients were treated conservatively. Amyotrophy of the

hand intrinsic and flexor muscle group of the forearm except the brachioradial muscle was observed hemilaterally in 20 patients and bilaterally in three patients. In three patients, T1-weighted MRI with neck flexion showed linear high intensity regions in the epidural space. In all patients, axial MRI/CTM demonstrated flattening of the spinal cord with the posterior surface of the dura mater shifting anteriorly. The amplitude of MEPs decreased after cervical flexion in two patients with progressive muscular atrophy. In three patients, dysesthesia of the upper extremities disappeared following cervical duraplasty. Musculotendinous transfer for three patients significantly improved the performance of their upper extremity. The findings of this study suggest that degenerative changes of the dura mater may be a characteristic pathology of CFM. Cervical duraplasty with laminoplasty is effective for cases at an early stage, and musculotendinous transfer should be selected in patients at a late stage.

Keywords Cervical spine · Flexion myelopathy · Amyotrophy · Duraplasty · Musculotendinous transfer

Y. Fujimoto (✉) · S. Oka · N. Tanaka
K. Nishikawa · H. Kawagoe
Department of Orthopaedic Surgery,
Hiroshima University School of Medicine,
Kasumi 1-2-3, Minami-ku,
Hiroshima, 734-8551 Japan
e-mail: skydog@hiroshima-u.ac.jp,
Tel.: +81-82-2575230,
Fax: +81-82-2575234

I. Baba
Department of Orthopaedic Surgery,
Asa City Hospital, Hiroshima, Japan

Introduction

Reid [43, 44] and Breig and colleagues [3, 4], on the basis of a cadaveric study, hypothesized that spinal cord com-

pression by the vertebral bodies and intervertebral discs during neck flexion caused cervical myelopathy. Later, the pincer mechanism of the cervical spine became emphasised, and neck extension was thought to be the predominant pathology of cervical myelopathy [41]. How-

ever, in recent years, as new information on flexion myelopathy has been obtained, myelopathy due to flexion of the cervical spine has been attracting attention as the pathologic mechanism of Hirayama's disease.

Juvenile muscular atrophy of the unilateral upper extremity (Hirayama's disease), which was first described by Hirayama et al. [13], is characterised by the following conditions:

1. Sporadic occurrence, a low incidence of a family history
2. Predominance in adolescent males aged 14–24 years
3. Unilateral muscle atrophy in the hand intrinsic and flexor muscles of the forearm except for the brachioradial muscle, which showed "oblique amyotrophy" over the forearm
4. Tremor-like movement of the fingers induced by stretching them
5. Cold paresis of the hand and fingers
6. Insidious onset and cessation of the symptoms after a few years of a slow progressive clinical course
7. Neurogenic patterns on electromyography (EMG) in the bilateral arms and hands
8. No sensory disturbance or abnormal deep tendon reflexes

Mukai et al. [32, 33] described lower cervical spinal cord atrophy and anterior shift of the posterior surface of the cervical dura mater during neck flexion on myelography in two patients with Hirayama's disease. Kikuchi et al. [20] named Hirayama's disease "flexion myelopathy". The same condition was also reported to occur in the thoracic spine, called "thoracic flexion myelopathy" [30]. In this paper, flexion myelopathy in the cervical portion of the spine is called a "cervical flexion myelopathy (CFM)".

In recent years, as the treatment for CFM, conservative cervical collar therapy [14, 50] or surgical arthrodesis of the cervical spine [22, 24, 25, 26, 29, 31, 35, 36, 37, 48] have been reported and obtained satisfactory results. However, in cervical collar therapy, the collar is required to be worn for several years, and is frequently troublesome. Surgical treatments carry a risk of operative complications and degenerative change at unfused levels. Surgical indication is still controversial. The authors have surgically treated patients with CFM by cervical duraplasty with laminoplasty or musculotendinous transfer using the brachioradial muscle. In the present study, the surgical indication and outcomes are reported. A pathophysiology of CFM is discussed, based on the studies of clinical features, neuroradiological findings, and neurophysiological assessments.

Materials and methods

Twenty-three patients (20 male and three female) with CFM were studied. The items examined were as follows.

Clinical features

Data on age at onset, distribution and progression period of muscular atrophy, neurological findings, and severity of active daily living (ADL) disability were obtained.

Neuroradiological studies

Neuroradiological studies were conducted with plain cervical radiography, myelogram, computed tomography assisted with myelogram (CTM), and magnetic resonance (MR) imaging.

Neurophysiological assessments

Neurophysiological assessments were carried out with electromyogram (EMG), nerve conduction studies, dynamic motor-evoked potentials (MEPs), and evoked spinal cord potentials (ESCPs) during surgery in patients who underwent cervical duraplasty. Dynamic MEPs were examined at the neutral/flexed position of the cervical spine, and central motor conduction time (CMCT) was calculated from the latencies of MEPs of the abductor digiti minimi (ADM) and abductor hallucis (AH) muscles elicited by transcranial magnetic stimulation using Magstim model 200 (Magstim Co., Ltd, Wales, UK). ESCPs studies were performed intraoperatively using three ESCPs: ascending, descending, and segmental. Ascending ESCPs were elicited by direct stimulation of the lower thoracic spinal cord using a catheter electrode that was inserted into the subarachnoid space before surgery under a local anesthesia. Descending ESCPs were elicited by transcranial electrical stimulation, and segmental ESCPs were elicited by stimulation of surface electrodes on the ulnar or median nerve. Averaged responses were recorded from the platinum needle electrodes inserted into each of the ligamenta flava in the surgical field. Square-wave pulses of 0.2 ms in duration were delivered to the ulnar or median nerve at three times the motor threshold, to the lower thoracic spinal cord at 10 mA, and to transcranial electrical stimulation at 100 mA, respectively. The responses were averaged, stored, and reproduced at least once (Nicolet Viking IV, Nicolet Biomedical, Inc., Madison, Wis.).

Surgical procedures

Cervical duraplasty with laminoplasty was indicated for the patients at the progression stage of muscular atrophy, and musculotendinous transfer using the brachioradial muscle was performed for the patients in whom the progression of muscular atrophy has already ceased. They were investigated based on the surgical outcomes in four patients who underwent cervical duraplasty or musculotendinous transfer of the brachioradial muscle, and one patient who underwent musculotendinous transfer secondary to cervical

Table 1 Severity of activities of daily living (ADL) disability, according to the Tokumaru's classification [49]

Severity I	Grip strength of the affected hand is kept at 50% or more of that of the unaffected side, or muscular atrophy is mild, causing almost no disability in daily living
Severity II	Grip strength of the affected hand is decreased to less than 50% but more than 30% of that of the unaffected side, or muscular atrophy is moderate, causing moderate disability in daily living
Severity III	Grip strength of the affected hand is decreased to below 30% of that of the unaffected side, or muscular atrophy is severe, causing serious problems in daily living

Table 2 Clinical features and neurological findings in 23 patients with cervical flexion myelopathy (*DTR* deep tendon reflex, *L/E* lower extremity, *U/E* upper extremity, *D-plasty* Duraplasty, *MTT* musculotendinous transfer)

Case no.	Age at onset (years)	Sex	Affected side	Grip strength (kg)		Sensory disturbance	Sudo-resis of the hand	Dysesthesia on neck flexion	DTR of L/E	Method of surgical treatment	Progression of muscle atrophy (months)	Follow-up period (months)	Follow-up (severity) (Preop.→Postop.)
				Right	Left								
1	17	M	Right	0	36	No	No	No	Normal	MTT	45	148	III→II
2	13	M	Bilateral (left<right)	17	27	No	No	No	Hyper		50	41	I
3	17	M	Right	28	60	Right C8	No	No	Normal		84	132	II
4	14	F	Right	6	20	Right C8	Right, yes	Bilateral U/E	Normal		24	36	III
5	19	M	Right	36	56	No	No	No	Normal		36	60	I
6	17	M	Right	28	53	No	No	Neck pain	Hyper	MTT	48	60	III→II
7	17	M	Left	58	0	Left C7,C8,T1	No	Bilateral U/E, backache	Hyper	D-plasty	24, recurred	48	III→II
8	13	M	Right	17	47	No	No	Back, U/E, L/E	Hyper		48	65	III
9	14	F	Left	39	25	No	No	No	Normal		12	312	I
10	17	M	Right	32	44	No	Right, yes	No	Hyper		84	108	I
11	16	M	Bilateral (left<right)	17	18	No	No	No	Normal		24	50	I
12	12	M	Right	39	53	No	Bilateral, yes	No	Normal		12	29	II
13	11	M	Bilateral (left<right)	15	15	No	Bilateral, yes	No	Hyper		60	63	?
14	17	M	Right	17	42	No	No	No	Hyper		36	36	III
15	12	M	Left	25	1	No	Left, yes	Bilateral U/E	Normal		24	50	III
16	19	M	Left	37	5	No	No	Left U/E	Hyper		6	9	III
17	18	M	Left	25	19	Left C7,8	No	Left U/E, right thigh	Hyper	D-plasty	6	36	III→I
18	23	M	Left	43	23	No	No	No	Normal		12	7	I
19	15	M	Left	47	28	No	No	No	Normal		15	12	II
20	17	M	Left	0	36	No	No	Bilateral U/E, backache	Normal	D-plasty, MTT	6, recurred	38	III→II
21	17	M	Right	12	35	Right C8	No	Bilateral U/E	Hyper		15	29	II

Table 2 (continued)

Case no.	Age at onset (years)	Sex	Affected side	Grip strength (kg)		Sensory disturbance	Sudo-resis of the hand	Dysesthesia on neck flexion	DTR of L/E	Method of surgical treatment	Progression of muscle atrophy (months)	Follow-up period (months)	Follow-up (severity) (Preop.→Postop.)
				Right	Left								
22	11	F	Right	14	17	No	No	Bilateral U/E	Hyper		36	18	II
23	15	M	Left	39	19	No	No	Back pain, U/E	Hyper		40	15	II
Minimum	11										6	7	
Maximum	23										84	312	
Average	15.7										34.1	61.0	

duraplasty. The follow-up periods after cervical duraplasty and musculotendinous transfer were 36–48 months (mean 41 months) and 35–148 months (mean 81 months), respectively.

Outcome of the conservative and surgical treatment

The conservative treatment included muscle exercises for affected extremities and neck immobilization with braces. The severity of ADL disability after the treatment was classified into three degrees according to the Tokumaru's classification [49] (Table 1).

Operative procedures

Cervical duraplasty

The patient was placed in a prone position with the head supported in a neck neutral position by a Mayfield skull fixator. Modified expansive open-door laminoplasty [16] from C3 to C7 was performed. The dura mater was transversely dissected preserving the arachnoid membrane. The dura mater spontaneously contracted in the vertical direction after its transection because of its hypertonic change. The defect of the dura mater was repaired watertight with the autogenous tensor fascia lata. The extradural venous plexus was so engorged as to cause extensive hemorrhage, and then hemostasis was carried out successfully using an irrigation bipolar coagulator.

Musculotendinous transfer

Typical muscular atrophy of the unilateral upper extremities is recognized in the hand and flexor muscles of the forearm (C7–T1 myotomes), whereas the brachioradial muscle (C6 myotome) is usually spared. Therefore, the brachioradial muscle was used as the source of muscular force of the musculotendinous transfer for surgical reconstruction of the hand and fingers.

Diagnosis of CFM

In this study, according to Kikuchi et al. [20], patients who had clinical features similar to Hirayama's disease with abnormal findings of myelogram, CTM, and MR imaging, such as an anterior shift of the posterior surface of the lower cervical dura in the neck-flexed position and lower cervical spinal cord atrophy, were diagnosed as having CFM. Entrapment neuropathy (e.g. cubital tunnel syndrome, thoracic outlet syndrome), cervical spine disorders, syringomyelia, motor neuron disease, and chronic inflammatory demyelinating polyneuropathy, which develop muscular atrophy of the upper extremities similar to CFM, were excluded in cooperation with neurologists and hand surgeons.

Results

Clinical features

The clinical features are summarized in Table 2. The age at onset of symptoms was 11–23 years (mean 15.7 years) and the progression period of muscular atrophy ranged between 6 months and 84 months (mean 34 months), in seven patients, the progression of muscular atrophy had already halted by the first visit to our hospital. In case 7 and 20, muscular atrophy had temporarily stopped, but it had recurred 10 years later [18]. The severity of ADL dis-

Fig. 1 A T1-weighted sagittal magnetic resonance (MR) image in neck flexion showing posterior epidural high intensity (arrow). B,C T2-weighted sagittal (B) and axial (C) MR image in neck flexion showing intramedullary high intensity at the C5-C6 level (arrows)

ability after cessation of muscular atrophy was classified into severity I in seven patients, II in six, and III in 10.

The affected side of muscular atrophy was right in 11, left in nine, and bilateral (right side predominant) in three. Muscular atrophy was observed in the hand intrinsic muscles and flexor muscles of the forearm in all patients. Two patients also had muscular atrophy of the extensor muscles of the forearm. Other symptoms of the hand included tremor-like movement of the fingers ($n=16$), cold paresis of the fingers ($n=11$), claw hand deformity due to atrophy of the interosseous muscles ($n=10$), sudoresis in the hand ($n=5$), and mild sensory disturbance in the affected hand to the ulnar forearm ($n=5$). Dysesthesia of the upper extremities or back pain appeared upon cervical flexion in 11 patients. Abnormal deep tendon reflexes (DTRs) in the upper extremities were recognized in 11 patients, and hyperactive DTRs in the lower extremities in 12 patients, one of whom showed spastic gait.

Neuroradiological studies

In 17 patients, cervical plain radiographs revealed straight alignment, kyphotic, or sigmoid deformity with a diameter of the spinal canal ranging from 14.5 to 19.0 mm (mean 16.8 mm). No developmental cervical canal stenosis was identified. Lower cervical cord atrophy of the affected side was demonstrated in 15 of the patients who underwent MRI. Posterior epidural high-signal intensity was evident on T1-weighted images in three of five patients in whom neck-flexed cervical MRI was obtained, and intramedullary high intensity at the C5-C6 level on T2-weighted imaging was demonstrated in three patients

Fig. 2 Myelogram (A) and computed tomographic myelogram (CTM) at C5-6 (B) and C6-7 (C), showing anterior shift of the posterior surface of the lower cervical dura on the neck-flexed position (arrows)

(Fig. 1). All 11 patients who underwent myelography showed anterior shift of the posterior surface of the lower cervical dura in the neck-flexed position (Fig. 2). Deformities of the dural tube and cord compression from C4 to C7 during neck flexion corresponded to the distribution of the muscle atrophy. Postoperative myelogram and CTM revealed restoration of the dural tube and arachnoid space even in the neck-flexed position.

Fig. 3 Waveforms of motor evoked potentials (MEPs) in neutral and flexed positions

Fig. 4 Intraoperative ascending and descending evoked spinal cord potentials (ESCPs), showing a decrease at the C4–C6 level

Neurophysiological assessments

EMG monitoring of the atrophied muscles revealed the denervation pattern such as high-amplitude action potentials in association with a reduction of the motor-unit voltage during the maximum voluntary contraction. Motor and sensory conduction velocity of the median and ulnar nerves was within the normal range.

Eight patients underwent dynamic MEPs studies. CMCT was normal in only one patient, and CMCT in the four extremities was prolonged in a patient with spastic paralysis of the lower extremities. In two patients with progressive muscular atrophy, the amplitude of MEPs of AH muscle decreased 15 min after cervical flexion, while no changes were observed in patients in whom the progression of muscular atrophy had already stopped (Fig. 3). Regarding ESCPs during duraplasty, ascending and descending ESCPs revealed normal wave forms, but the amplitude of the N2 potential of segmental ESCPs elicited by the ulnar nerve stimulation decreased in two patients who had no symptoms of the white matter. In a patient with spastic gait, amplitudes of the ascending and descending ESCPs decreased or disappeared at the C4–C6 level (Fig. 4).

Surgical findings and outcomes

The patients graded as ADL disability severity I or II accommodated themselves well to muscle atrophy with insidious onset and gradual development. Surgical indication was decided on the basis of information regarding the clinical features and natural course of CFM, especially regarding whether or not the symptoms ceased after a few years of a slow progressive clinical course. A limited number of the patients suffering from severe dysesthesia of the extremities and backache in a neck-flexed position, massive muscular atrophy with severity III of ADL disability, or spastic gait were selected as candidates for surgical intervention.

Cervical duraplasty

The tonus of the dura mater was so high that the dural defect occurred automatically in the vertical direction after its transection (Fig. 5). Histology of the resected dura mater showed widespread hyaline-like degeneration with dispersion of enlarged elastic fibers in amorphous degenerated tissue, with the disappearance of collagen fibers (Fig. 6). In case 7, dysesthesia of the upper extremities and back pain in the neck-flexed position resolved, and grip strength was improved postoperatively (Fig. 7). In case 17, with spastic gait, motor dysfunction of the lower extremities was improved after surgery. However, in patient 20, with severe muscular atrophy, grip strength was not improved, and the patient underwent musculotendinous transfer secondary to cervical duraplasty.

Fig. 5 Cervical duraplasty with laminoplasty

Fig. 6 Histology of the dura mater showing a marked hyaline-like degeneration

Musculotendinous transfer

The brachioradial muscle maintained the normal muscular color, but the atrophied muscles had decreased color intensity. Their histology demonstrated neurogenic patterns of muscle atrophy, such as a grouped atrophy (Fig. 8). Grip strength was significantly recovered, and the effect of musculotendinous transfer persisted for a long-term follow-up even at 10 years or more after surgery.

Fig. 7A–C Case 7. **A** postoperative myelogram and **B,C** CTM reveal widening of the dural tube and restoration of the subarachnoid space even in the neck-flexed position

Fig. 8 Histology of the atrophied muscle showing neurogenic patterns of muscle atrophy, such as grouped atrophy

Discussion

Pathophysiology of CFM

In the first half-term report on juvenile muscular atrophy of the unilateral upper extremity (Hirayama's disease), no abnormalities were pointed out in the cervical spine on neuroradiological studies [9, 10, 11, 13, 14]. Later, abnormal findings were noted on neuroradiological examination of the cervical spine [49]:

1. Dural tightness: an anterior shift of the posterior surface of the dura mater of the lower cervical spine on

myelography or MR images in flexed-neck position [17, 20]

2. An engorged internal vertebral venous plexus: high signal intensity changes in the epidural space on T1-weighted MR images in flexed-neck position [34, 38]
3. Atrophy of the anterior horns: atrophy of the lower cervical spinal cord on MR images and CTM [2, 27]

In light of autopsy findings of two patients with this disease, no pathological changes were observed in the cervical white matter, but a decrease in and necrosis of the anterior horn cells were noted [1, 15]. Hirayama et al. hypothesized that the pathogenic mechanism of this disease is ischemic necrosis of the anterior horn of the spinal cord accompanied by an anterior shift of the posterior surface of the lower cervical dura in the neck-flexed position [15]. Repeated neck flexion causes ischemia of the spinal cord and impairs the anterior horn cells most vulnerable to ischemia. However, according to neurophysiological studies using somatosensory evoked potentials, a prolongation of latency between the N11 and N13 peaks and a decrease in the amplitude of N13 in neck-flexed position were detected [28]. These results indicated that the spinal cord lesion presented in the posterior horn as well as the anterior horn. Furthermore, ESCPs during duraplasty showed the conduction block spread to the white matter as the clinical symptoms became severe. Other case reports described that many patients demonstrated white matter lesions that manifest sensory disturbance, long tract sign or accentuated deep tendon reflexes of the lower extremities [6, 19, 29]. Therefore, Hirayama's disease is considered to be a selective syndrome of the lower cervical anterior horn cells, and can be included in CFM. In contrast, CFM can manifest white matter lesions as well as the anterior horn syndrome.

Yada and colleagues have pointed out a rostral shift of the posterior root entry zone of the middle and lower cervical spinal cord on neck-flexed myelogram in nine cases of CFM. They hypothesized that the spinal cord could be stretched in an up-down direction due to growth imbalance between a long cervical spine and a short spinal cord (overstretch mechanism) [21, 51, 52]. Tachibana et al. conducted an experimental study on a canine model of overstretching spinal cord [46, 47]. They measured the evoked spinal cord potentials and the spinal cord internal pressure, and concluded that overstretching of the spinal cord during neck flexion could lead to anterior horn impairment by disturbing the intramedullary circulation.

Iwasaki et al. [17] and Kikuchi et al. [20] proposed that the cervical spinal cord may be compressed by an anterior shift of the posterior surface of the dura mater in the flexed-neck position (tight dural canal mechanism). Mochizuki et al. [31] and Konno et al. [25] observed a decrease in the amount of elastin and a straightening of the wavy structure that is characteristic of normal elastic fiber in the dura mater excised during duraplasty. Kondoh [23] pointed out an immature collagen on analysis of muscle

and skin tissue and suggested an involvement of abnormal collagen metabolism in this disease. In our study, marked elongation of the dural stump in the vertical direction was observed immediately after transection of the dura mater during duraplasty, and a marked hyaline-like degeneration of the dura mater was observed in its histology. Hyper-tonic change due to organic abnormality of the cervical dura mater is considered to be involved in a tight dural canal mechanism.

Surgical indication and method

Until recently, the only treatments attempted for CFM were conservative cervical orthosis [12, 50], surgical arthrodesis of the cervical spine [22, 24, 26, 29, 35, 36, 37, 48], arthrodesis with duraplasty [25, 31], or duraplasty only [6]. Tokumaru and Hirayama [50] placed their patients in a cervical orthosis for 3–4 years, because the progression period of this disease usually lasts for several years, and they obtained good results in patients in the early period after the onset. Wearing a rigid collar for several years may be a troublesome problem for the patients. Surgical treatments have not been established. Mii et al. [29], Okamoto et al. [37], and Ogasa et al. [35] performed anterior cervical fusion, and they reported the symptoms were diminished. Kondoh et al. [24], Ohnari et al. [36], and Masaki et al. [26] described a posterior fusion procedure, and reported that the symptoms improved. Konno et al. [25] and Mochizuki et al. [31] reported the effectiveness of cervical posterior fusion with duraplasty. However, for adolescent patients with CFM, arthrodesis of spinal segments leads to excessive stress at unfused adjacent levels, and poses a problem. Hilibrand et al. [8] reported that 25–89% of patients who underwent anterior cervical arthrodesis and were followed for a long period had degenerative changes at adjacent levels. Our study showed that duraplasty with laminoplasty obtained good results without complicating postoperative kyphosis and pseudomeningocele. Duraplasty only with laminoplasty is considered to be an appropriate surgical procedure to resolve the pathology of a tight dural canal mechanism.

Mii et al. [29] have described the need for early surgery. It is ideal to perform surgery of the cervical spine during the progressive period of muscular atrophy. However, accurate judgement of the pathological activity of CFM has not been established. Hirayama et al. [11] reported that the frequency of F-wave in the neck-flexed position and the jitter phenomenon on single-fiber EMG increase during the progression stage. In studies of dynamic MEPs, changes in the latency and amplitude of MEPs in the neck-flexed position were useful not only for evaluation of the spinal cord white matter, but also for predicting the progression/non-progression of muscular atrophy.

The surgical outcome is poor in patients whose muscular atrophy is already complete. It is difficult to obtain good recovery through surgery for the cervical spine, be-

cause of loss of the anterior horn cells [29]. Fortunately, typical muscular atrophy is recognized in the hand and flexor muscles of the forearm (C7–T1 myotomes), whereas the brachioradial muscle (C6 myotome) is usually spared. Therefore, musculotendinous transfer using the brachioradial muscle is a useful method to reconstruct paralyzed hands in CFM. We have applied musculotendinous transfer to patients with persisting severe muscular atrophy, and obtained good long-term outcomes. Musculotendinous transfer can be considered as one surgical therapy option for patients in whom muscular atrophy is already complete.

Previous case reports of CFM emanate predominantly from Japan; reports of non-Japanese patients are very rare [2, 5, 7, 39, 40, 42, 45]. However, CFM can be inappropriately treated as entrapment neuropathy, such as cubital tunnel syndrome, mainly due to muscular atrophy of the C7–T1 myotomes, or as a motor neuron disease due to the absence of sensory disturbance. Increased knowledge of and interest in this disease can lead to earlier diagnosis and appropriate treatment.

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