Flexion Myelopathy Mimicking Hirayama Disease - Report of A Case

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Abstract -We report a 22-year-old male with muscular weakness and atrophy predominantly involving the left upper limb. These clinical manifestations resembled to those of Hirayama disease. Cervical cord compression during neck flexion but not on extension was demonstrated on magnetic resonance imaging and could be recognized by using somatosensory and motor evoked potentials techniques. Pathophysiologic mechanism of this condition is considered to be similar to that of Hirayama disease. We suggest the employment of electrophysiological methods for evaluation and the use of cervical collars to prevent neck flexion in such cases.
Key Words : Evoked potentials, Flexion myelopathy, Hirayama disease, Spinal cord compression

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INTRODUCTION

Hirayama disease[1], also called juvenile distal spinal muscular atrophy, is a well-known juvenile disorder manifested by non-progressive muscle atrophy localized to the hand and forearm. This disorder, once regarded as an unusual form of motor neuron disease, has also been attributed to a disturbance of microcirculation in the territory of the anterior spinal artery due to dynamic compression of the cervical cord[2]. On neck flexion, the cervical cord can be pressed against the posterior aspect of the vertebral body by an excessive degree of kyphosis of the cervical vertebrae, a protruding intervertebral disk, or an abnormal alignment such as severe subluxation[3]. This condition was termed as flexion myelopathy. We report the clinical features of a young male patient with flexion myelopathy mimicking those of Hirayama disease.

CASE REPORT

A 22-year-old right-handed man fell on his head at the age of 8 years but sustained only a laceration wound on the scalp and has been subsequently symptomless. However, he began to notice weakness of his left hand when he was 10 years of age. Weakness and muscle atrophy of the left hand and forearm gradually progressed in the following 3 years. He also found a fine tremor in both hands. His condition has remained unchanged thereafter. He denied any pain, numbness, paresthesia, or dysesthesia in the limbs. Neurologic examination revealed marked muscle atrophy and weakness in the left hand intrinsic muscles and to a lesser degree in the left forearm muscles and the right hand intrinsic muscles. Mild weakness was also detected in

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the left biceps brachii. No fasciculations were observed but irregular, fine, and tremulous movements were present in the fingers when the patient attempted to extend them. Tendon reflexes were slightly hyperactive in all limbs and plantar responses were flexor. On sensory testing, there was a mildly impaired vibration sense in the right leg. Temperature, pinprick, touch, and vibration sensations were normal in both upper limbs. Cranial nerves and cerebellar functions were intact.

Motor and sensory peripheral nerve conduction studies revealed normal distal latencies, amplitudes of compound muscle action potential, and F wave latencies. Electromyography (EMG) showed reduction of motor unit recruitment and high-amplitude motor unit potentials in bilateral hand muscles and left forearm muscles. There was no fibrillation or positive sharp waves. Somatosensory evoked potentials (SEP) were evaluated on both upper and lower limbs when neck was in neutral position and on flexion (at an angle of around 40 degrees by placing two pillows beneath the head). Prolongation of scalp latencies from the right peroneal nerve was observed when neck was bent forward (Table 1). On motor evoked potential (MEP) examinations, magnetic stimuli were applied to stimulate the motor cortex and to activate the spinal motor nerve roots. Central motor conduction time (CMCT) to the upper or lower limbs was calculated by subtracting the latency of MEP on cervical or lumbar stimulation from that on cortical stimulation. The patient had prolonged CMCTs to the left hand when his neck was kept both in neutral position and on flexion (Table 2). Plain cervical spine roentgenographic films showed loss of physiologic lordosis (Fig 1). Magnetic resonance imaging (MRI) of the cervical cord taken when the neck was on flexion showed the cervical cord was displaced anteriorly at C5-C6 level and compressed by a bulging intervertebral disk. Cervical cord atrophy was also demonstrated at the same level (Fig 2). As there was no progression of his neurological symptoms for 9 years, we suggested him to wear a cervical collar to prevent excessive neck flexion and follow up regularly.

Table 1: *Scalp SEP latencies with neck in neutral position and on flexion*

	Scalp latencies (msec)		
	Neutral position	Flexion	
Right median n.	18.9	18.7	(18.44 ± 0.80)
Left median n.	18.3	18.2	
Rihgt peroneal n.	41.2	50.0	(39.13 ± 2.45)
Left peroneal n.	38.8	40.0	

Normal values in our laboratory are provided in parentheses

Table 2: *MEP with neck in neural position and on flexion*

	Central motor conduction time (msec)		
	Neutral position	Flexion	
Reihgt abductor digiti minimi	8.0	8.1	(7.04 ± 0.60)
Left abductor digiti minimi	11.0	11.8	
Right tibialis anterior	14.1	14.1	(15.94 ± 1.23)
Left tibialis anterior	14.7	14.7	

Normal values in Chinese are provided in parentheses(4)

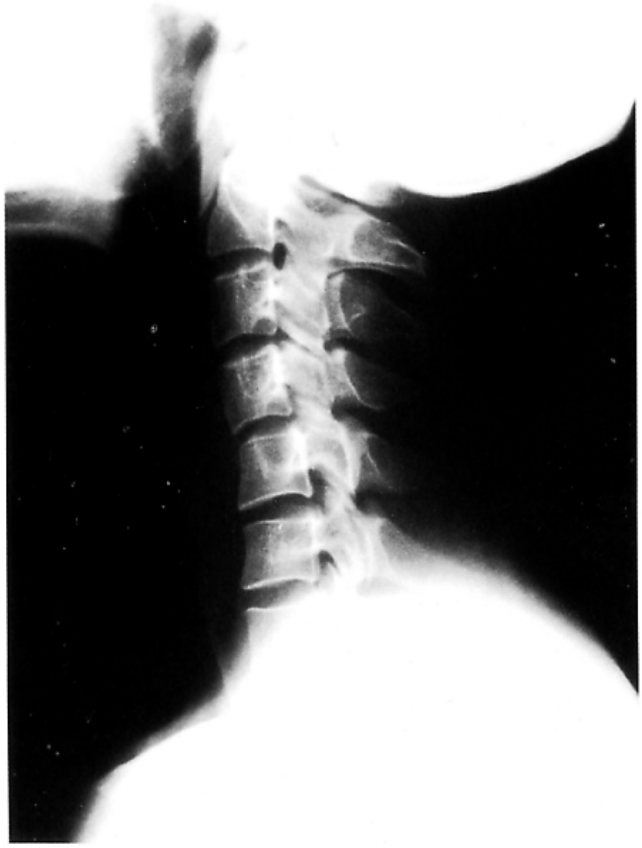

Fig 1. Lateral radiograph of the neck showing loss of physiologic lordosis.

DISCUSSION

In 1959, Hirayama et al[5] reported 12 young patients with unilateral weakness and atrophy of the upper limb. Since then, about 150 cases have been reported in Japan[1,6,7], 47 in India[8,9], 27 in Taiwan[10], 19 in Malaysia[11], and a few cases in other parts of the world[12]. This disease is manifested by insidious fatigability, weakness, and atrophy in one limb, usually limited to the hand and forearm muscles. Hyperreflexia has been observed in 10-20% of the affected patients, but pathologic reflex is uncommon. Most patients lack sensory disturbances. Weakness exaggerated by cold has been described in approximately 22%, while a fine and irregular tremor upon finger extension in 8%. It predominates on one side, but not necessarily on the side of dominant hand. Muscle biopsy and EMG study show evidence of denervation[13,14]. No abnormal findings are found in nerve conduction velocity, cerebrospinal fluid, and SEP studies. Atrophy of the lower cervical cord could frequently be demonstrated on computed tomographic myelography (CTM).

The clinical and EMG features of our patient closely resembled to those of Hirayama disease except for a preceding history of head trauma and mild asymmetry of vibration sensibility in the lower extremities. When the neck flexed, the absolute latencies of scalp SEP from the right peroneal nerve lengthened. This phenomenon was in good accordance with his mildly impaired vibration sense in the right foot. CMCT to the left hand was prolonged when neck was kept either in neutral position or on flexion. These electrophysiological findings indicated that the disease process in our patient involved not only anterior horn cells but also corticospinal tract and posterior column. MRI further demonstrated anterior compression of the cervical cord in the flexion position.

Al-Mefty et al[15] studied the pathophysiologic mechanisms in chronic compression myelopathy at the cervical level. The cervical cord is suddenly pinched with every neck movement, producing compression forces to the cord and interfering the microcirculation. These intermittent effects result in ischemia or hypoxia primarily within the watershed area of the cord, sparing the posterior half of the posterior horn[16]. Large motor neurons and intermediate gray matter are more vulnerable to such intermittent tissue hypoxia. Pathological changes can extend to the level below that of the compression. Involvement of the white matter is usually minimal, but changes may be seen either in the ventral inner portion of the dorsal column or in the lateral columns bordering the gray matter[17]. These sites of lesion are similar to those in Hirayama disease [18]. Moreover, a similar dynamic compression of the cervical cord in Hirayama disease has been demonstrated on CTM and MRI[19]. Tokumara and Hirayama found an anterior shift of lower cervical dural canal during neck flexion, particularly of its posterior wall at the 6th vertebral level, resulting in an anteroposterior compression of the cord segment from

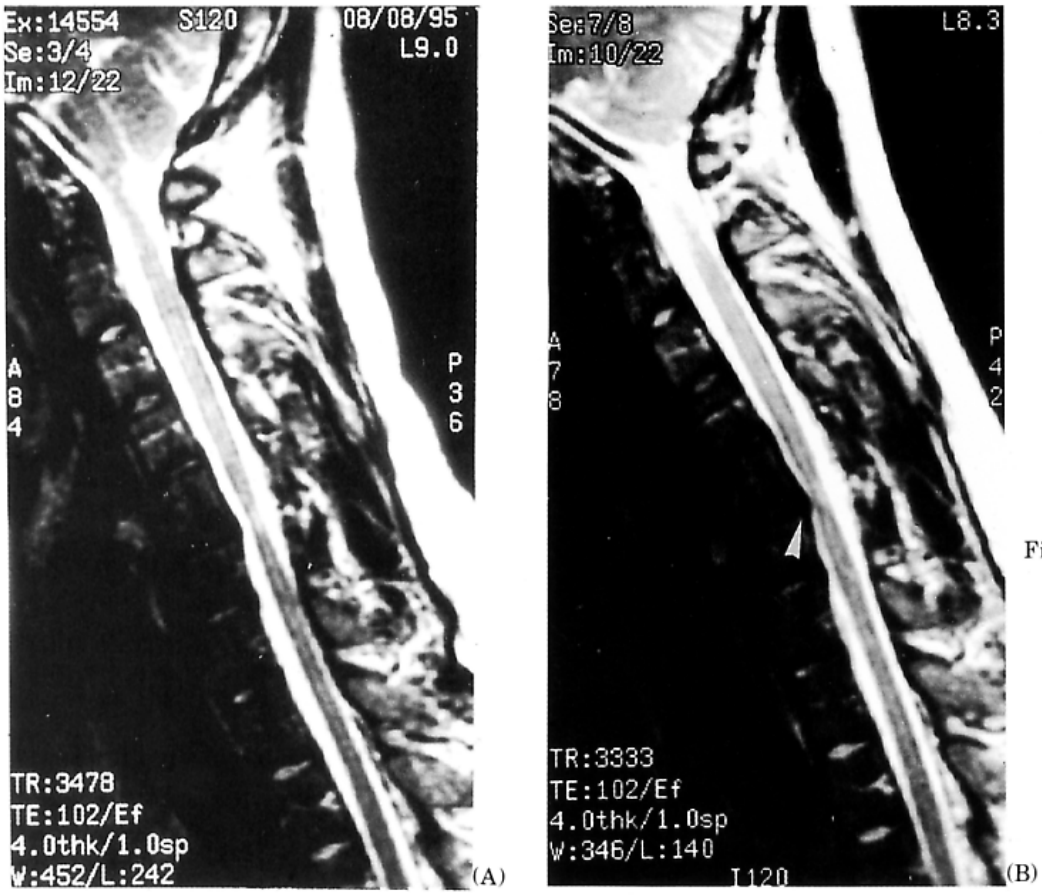

Fig 2. (A), T2-weighted MR image, in the neutral position. The cervical cord is atrophic at the C5-C6 level, but not compressed. (B), T2-weighted MR image, in the flexion position. The cervical cord is compressed anteriorly at C5-C6 level (arrowhead).

C7 to C8. The rates of anterior shift was inversely proportional to the duration of illness and was considered to be a reason why Hirayama disease was self-limited. Such a neck flexion-related compression of the cervical cord was first described as flexion myelopathy by Matsuura and Tashiro[20] in 1989. Since then, dynamic studies to see any functional compression of the cervical cord during neck flexion have been applied in patients with Hirayama disease[21,22].

We propose that Hirayama disease may merely represent the mildest form of compression myelopathy, in which disturbed microcirculation due to dynamic compression is the main pathogenesis. On the other hand, severe compression myelopathy due to spondylosis, narrow spinal canals, opacification of posterior longitudinal ligament, etc., results from a long-lasting mechanical stress on the cervical cord as well as on the blood vessels. Our patient lies between these two extremes because the clinical manifestations indicated that his disease was not limited to the anterior horn, as usually seen in Hirayama disease, but also involved the adjacent lateral column and ventral inner portion of the contralateral posterior column.

Cervical collar has been used for prevention of neck flexion in patients in the early stage of Hirayama disease[23]. Either improvement of the muscle power or shortened duration of deterioration was achieved. Anterior spinal fusion to prevent neck flexion could also result in improvement of the muscle power in some patients[24]. Although our patient maintained a stationary non-progressive course in the past 9 years, we still advised him to wear a cervical collar for both SEP and MEP studies had revealed evidence of dynamic functional impairment. Regular electrophysiological follow-up for early detection of subclinical deterioration, especially MEP for assessment of the motor pathway function[25], is mandatory and may

serve as a good criterion for further surgical intervention.

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