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## **Cervical flexion myelopathy in a patient showing apparent long tract signs: a severe form of Hirayama disease**

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## Summary

We describe an 18-year-old male with cervical flexion myelopathy with Hirayama disease-like features who showed apparent long tract signs. He first experienced insidious-onset hand muscle weakness and atrophy at the age of 15. Subsequently, he developed sensory disturbance in his lower limb. Neurological examination revealed atrophy and weakness in the right hand and forearm, pyramidal signs in the right lower extremity, and disturbance of superficial sensation in the lower left half of the body. Cervical magnetic resonance images and computed tomographic myelography revealed anterior displacement with compression of the cervical cord in flexion that was more apparent in the right side. The right side of the cervical cord showed severe atrophy. The mechanisms of myelopathy in our patient appeared to be same as that of “tight dural canal in flexion,” which has been reported to be the mechanism of juvenile muscular atrophy of the unilateral upper extremity (Hirayama disease). Patients with Hirayama disease generally show minimal sensory signs and no pyramidal signs. An autopsy case of Hirayama disease revealed confined necrosis of the cervical anterior horn without obvious changes in the white matter. Our patient’s disease progression suggests that

cervical flexion myelopathy patients with severe cervical cord compression in flexion

may develop extensive cervical cord injury beyond the anterior horn.

(210 words)

Key words: flexion myelopathy; Hirayama disease; MRI; pyramidal sign

## Introduction

Cervical flexion myelopathy (CFM) is a condition characterized as spinal cord injury caused by neck flexion [1,2]. In patients with CFM several mechanisms of myelopathy have been proposed such as follows: (1) spinal cord compression is caused by the anterior elements, including posterior surface of the vertebral body and intervertebral architectures [3]; (2) the spinal cord is stretched in an up-down direction; (3) the spinal cord is compressed by a narrow dural canal caused by forward migration of the lower cervical posterior dura mater “tight dural canal in flexion” [1,2]. The last mechanism could account for the disease entity of juvenile muscular atrophy of the unilateral upper extremity (Hirayama disease) [1,2,4]. Hirayama disease affects younger males and it is characterized by muscle atrophy in the unilateral hand and ulnar side of the forearm, cessation of symptoms after several years of insidious progression, no apparent sensory disturbance or pyramidal signs, and forward migration of spinal cord and dural sac in flexion [4,5,6]. An autopsy case study of Hirayama disease described confined necrosis of the cervical anterior horn without obvious changes in the white matter [7]. Although several CFM and atypical Hirayama disease patients have shown sensory disturbance in

the upper extremities and long tract signs [8], these features have not been previously described in detail. In this case report, we describe a CFM patient with pathomechanisms same as that of Hirayama disease and who showed features of unilateral spinal cord injury such as spasticity, hyperreflexia and positive plantar reflex in the ipsilateral lower extremity, and sensory disturbance of contralateral trunk and lower extremity.

### **Case report**

An 18-year-old Japanese male was admitted to our hospital. He had first experienced insidious-onset right hand muscle weakness and atrophy at the age of 15. One year later, he developed disturbance of pain and temperature sensations in his left lower extremity. His sensory disturbance extended to his left lower body. On admission, he showed atrophy and weakness in his right-hand and forearm muscles. His forearm atrophy was more prominent in the ulnar side. No fasciculations were observed. His muscle tonus showed no abnormality. There was no hypermobility syndrome. His right triceps reflex was decreased. Right knee and ankle jerks were brisk, and right plantar reflex was positive. A hyposensation of pain and temperature was observed below the level of Th11

in the left side. His touch, vibration, and position sensations were normal. He had no ataxia, his gait was not unusual, and Romberg's sign was negative. Autonomic failure was not observed. His complete blood counts, blood chemistry analysis, and urinary analysis revealed nothing unusual. His cerebrospinal fluid revealed mildly elevated protein levels (49 mg/dl) without pleocytosis. The cytology and antibodies against poliovirus were negative. No myelin basic proteins or oligoclonal bands were detected. Nerve conduction studies and sensory evoked potentials in the extremities were normal. In electromyogram studies, high amplitude units and reduced interference patterns, indicating chronic denervation, were observed in the atrophied muscles (right brachioradialis muscle, right triceps muscle, and right interosseus dorsalis muscle); however, the right biceps brachii muscle was normal. Cervical MRI showed atrophy and hyperintense lesions on T2-weighted images, apparent in the right side, in the C5 and C6 vertebral levels (Fig. 1A-C). In the anterior flexion, anterior displacement of the cervical posterior wall of the cervical dural canal was evident (Fig. 1B). A hyperintense epidural mass was noted on the T2-weighted image (Fig. 1B). No gadolinium-enhanced lesions were observed. The brain, thoracic, and lumbar MRIs showed no abnormality. Computed

tomographic myelography revealed anterior displacement and stenosis, more apparent in the right side, of the spinal cord in anterior flexion of the neck (Fig. 2A,B). Four months later, therapeutic surgical posterior fusion was performed. Four months after the operation, his neurological findings had not deteriorated.

## **Discussion**

The neurological findings—atrophy and weakness in the right upper limb muscles, pyramidal signs in the right lower extremity, and disturbance of superficial sensation in the left lower half of the body—indicated injury of the right-side cervical cord, except for the dorsal column. The results of the electromyogram and cervical MRI were compatible with cervical cord disturbance in the C5 and C6 vertebral levels. The findings of anterior displacement of the cervical cord and dural sac in flexion and subsequent cervical cord compression were identical to those reported in CFM and Hirayama disease characterized by the mechanisms of “tight dural canal in flexion” [1,2,4].

Hirayama disease is characterized by atrophy of unilateral distal upper

extremity. Among the possible pathomechanisms of Hirayama disease, it has been proposed that biomechanical factors acting on the cervical cord and dural sac during neck flexion cause a circulation disturbance leading to necrosis in the cervical anterior horns [5,7]. Although most patients with Hirayama disease show no or minimal sensory disturbance and long tract signs [5], some patients have been reported to have long tract signs [8]. In a nationwide survey of Hirayama disease in Japan, 7 of 287 patients showed Babinski sign [6]. However, there have been no reports of patients with apparent long tract signs (pyramidal signs and sensory disturbance) of the lower limb, and “tight dural canal in flexion” as observed in Hirayama disease.

Regarding the pathomechanism of the long tract sign and sensory disturbance in CFM, a different stress distribution in the white and gray matter was suggested in experimental models of cervical flexion myelopathy [9] and cervical spondylotic myelopathy [10] using a finite element method. Studies that were conducted with these experimental models revealed that mild cord compression resulted in stress being concentrated in only the gray matter; however, increased cord compression caused higher stress in both the gray and white matter [9,10]. Our patient showed more severe canal

stenosis and cervical cord compression in the right side than the left side. It has been reported that the severity of the anterior displacement of cervical cord in flexion is unequal on both side [11]. Uneven cervical cord atrophy is a common feature of Hirayama disease [12]. A patient with Hirayama disease-like features and severe uneven atrophy of cervical cord showing edge-shaped compression of the cervical cord has also been reported [13]. In that case, the dura of the cervical cord thickened with no inflammatory cell infiltrations or granuloma formations, suggesting that the dura mater impairments could be a primary pathomechanism of CFM [13]. Another CFM patient showed a marked hyaline-like degeneration of the dura mater [8]. Our experience with the patient described in this case report indicates that severe cervical canal stenosis in anterior flexion and subsequent cervical cord injury could result in necrosis of the white matter beyond the anterior horn. In patients with Hirayama disease, the anterior shift of the cervical cord could be decreased in the late clinical course. The neck collar therapy has been recommended [5], and surgical therapies have been recommended for patient demonstrating no improvement under neck collar therapy [14]. For severe cord compression such as that found in our patient, surgical therapies such as cervical spine

fusion could be recommended in the early clinical course. In conclusion, we described a CFM patient with severe cervical cord compression in anterior flexion showing obvious long tract signs.

(1,186 words)

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### **Conflicts of interest**

There are no conflicts of interest related to the manuscript.

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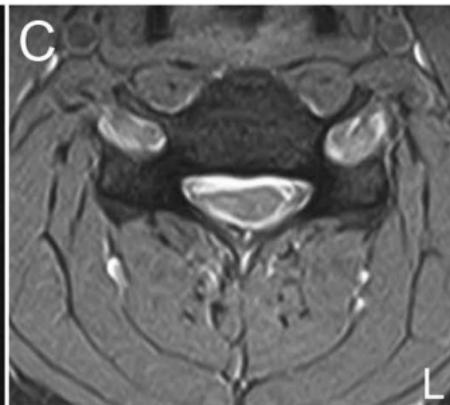
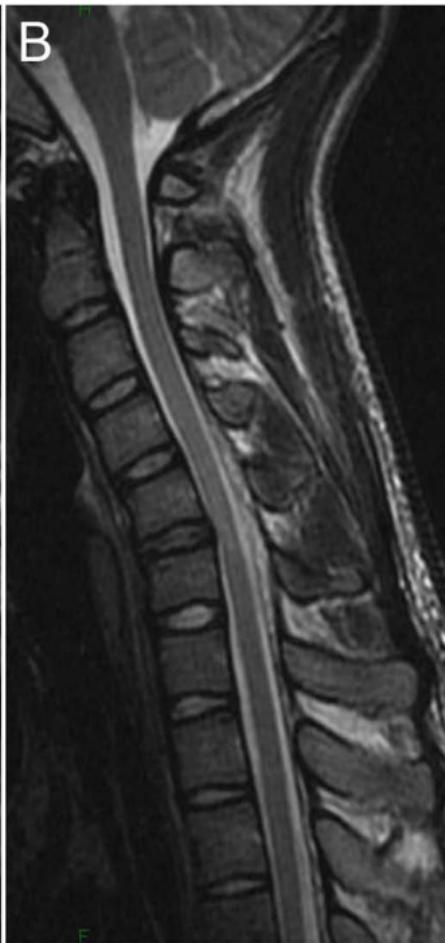
### **Figure legends**

Figure 1: Magnetic resonance image (MRI) in nonflexion (A) and anterior flexion (B)

T2-weighted sagittal images shows anterior displacement of the cervical cord and dural canal with development of a hyperintense epidural mass in flexion (B). T2-weighted axial image at the C5 vertebral level (C) reveals atrophy and hyperintense lesions in the right-side cervical cord.

Figure 2: Computed tomographic myelography at the C6 vertebral level in nonflexion (A)

and anterior flexion (B) shows flattening of the spinal cord in anterior flexion, more apparent in the right side (B).

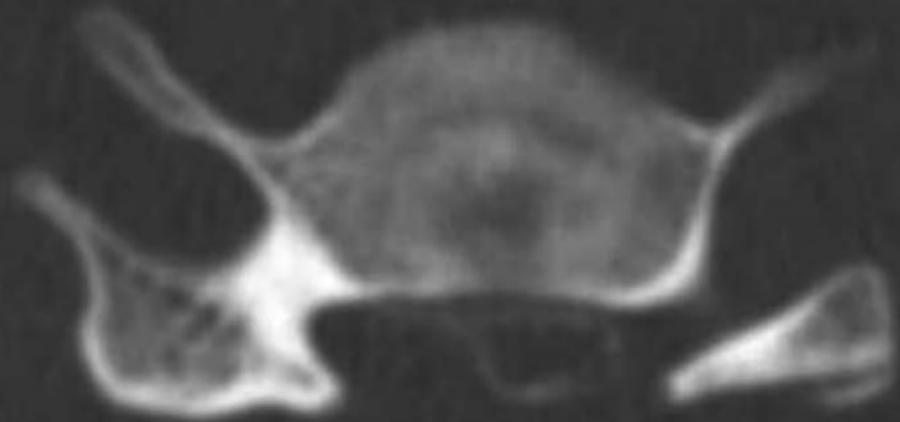


A



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B



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