Cervical Spondylotic Amyotrophy Associated with Hirayama’s Disease

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A 47-year-old man with Hirayama’s disease who developed cervical spondylotic amyotrophy (CSA) is presented. The patient had noted weakness and atrophy of hand and forearm muscles bilaterally at the age of 16. At the age of 40, he developed proximal muscle atrophy and weakness bilaterally after 20 years of a non-progressive state. Myelography and computed tomography (CT)-myelography revealed that ventral cord compression at multiple levels of C4-7 vertebral bodies was increased when the neck was extended. The clinical diagnosis was CSA associated with Hirayama’s disease. To our knowledge, this is the first such case to be reported. (Internal Medicine 36: 647–650, 1997)

Key words: cervical spondylosis, spinal canal stenosis, ischemic myelopathy

Introduction

A new clinical disease entity, juvenile muscular atrophy of unilateral upper limb (Hirayama’s disease), was initially described by Hirayama et al (1). Cervical spondylotic amyotrophy (CSA) is characterized by muscular atrophy in the upper extremity with absent or insignificant sensory deficit (2). For both Hirayama’s disease and CSA, the responsible lesion is thought to be in the anterior horn, but the two differ in age of onset and distribution of muscular atrophy. However, the pathogeneses of these two disorders are controversial. Here, we present a case of CSA preceded by Hirayama’s disease.

Case Report

A 47-year-old right-handed man was admitted to our hospital in 1994. His past and family history were non-contributory. In 1963, he first noticed weakness of the right fingers followed by weakness of the left hand two months later. He had also noted aggravation of the motor weakness of his hands in cold weather. X-ray studies of the cervical spine had revealed no spondylotic change, dislocation, or vertebral canal stenosis. The progression of atrophy of his hands and forearm muscles had been relatively rapid during the period of one year after onset, and he had lost all grasping power in his hands. The progression of symptoms was arrested within 4 years after onset. His condition remained unchanged without treatment from 1967 until 1987, when he developed proximal muscle atrophy and weakness bilaterally in the upper extremities.

On admission, mental status and cranial nerve examinations were normal. He had severe weakness and atrophy of the distal muscles innervated by the C7 through T1 spinal segments predominantly on the right side, and mild weakness and atrophy of the proximal muscles innervated by the C5 and C6 segments bilaterally in the upper extremities (Fig. 1). The lower extremities were not impaired. No atrophy of the face, neck, trunk, or leg muscles was noted. Loss of deep tendon reflexes of the triceps on both sides was observed. Hyperreflexia and Babinski’s sign were observed in the right lower extremity. No fasciculation was observed in the atrophied muscles. No ataxia, extrapyramidal signs, sensory disturbance, Horner’s sign, or abnormalities in sweating and urination were observed.

Blood chemistry, peripheral blood, and cerebrospinal fluid were normal. Motor nerve conduction studies showed no response bilaterally in the ulnar and median nerves, and normal findings for the lower extremities. Sensory nerve conduction velocities of the four extremities were normal. Electromyography revealed neurogenic changes in the atrophic muscles innervated by the C5 through T1 spinal segments. A cervical magnetic resonance imaging (MRI) revealed localized atrophy of the spinal cord at the level of the C4-7 vertebral bodies (Fig. 2). Myelography and computed tomography (CT)-myelography revealed a focal bony spur, disc herniation, and ventral cord flattening at multiple levels of the C4-7 vertebral bodies on neck extension. However, no anterior shift of the spinal cord and the posterior wall of the dural canal was observed on neck flexion.
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Figure 1. A) Atrophy of both upper limbs, with relative sparing of the brachioradial muscles. B) Bilateral muscular atrophy of the hand and forearm predominantly on the right side.

(Figs. 3, 4).

Discussion

In the present case, plain cervical spine radiographs revealed no cervical spondylosis or disc herniation 20 years ago. The stationary course along with localized muscle atrophy and age at onset make it possible to differentiate this patient’s disease from other diseases accompanied by muscle atrophy. His clinical features were compatible with Hirayama’s disease.

Brain et al (3) reported cases of cervical spondylosis with muscle atrophy of the upper extremities without sensory disturbance or pyramidal signs. The dissociated motor loss syndrome in cervical spondylosis was reported by Keegan, and the etiology of this syndrome was thought to be selective damage by bony spurs of the motor roots (4). However, Yanagi et al (2) stated that the cause of CSA might be anterior horn damage; they summarized the main clinical features of CSA. The presence of

Figure 2. A proton density sequence sagittal MRI with the neck in neutral position. Localized atrophy of the spinal cord at the level of the C4-7 vertebral bodies is observed.
sensory disturbance, older age at onset, and results of radiological studies differentiate this disease from motor neuron disease and Hirayama’s disease. The onset of CSA is thought to occur at the age of 40 or older, and the responsible lesion is thought to be in the anterior horn, based on the neuroradiologic finding of cord atrophy.

Disease onset in the present case was insidious, and progression was relatively rapid in the early stage from the age of 16 years, but was slow from the age of 40. Interestingly, he exhibited no sensory impairment at all over 30 years, and no pathological changes involved the ascending tracts in the cervical cord, as demonstrated by cortical somatosensory evoked potential recording. It is assumed that he had lesions at the age of 47 involving not only the anterior horns but also the adjacent white matter, producing a right long tract sign.

In Hirayama’s disease, radiological examination (myelography) with CT and MRI reveals dynamic compression of the spinal cord. In neck flexion, the posterior wall of the dural canal shifts anteriorly leaving the vertebral arch around the sixth cervical vertebra, resulting in antero-posterior compression of the cord segment from C7 to C8. The degree of the anterior shift is inversely correlated to the duration from onset (5). This might be a reason why anterior shift was not found at the age of 47 in our patient. Lapresle (6) stated that the anterior horn, particularly the neurons in its central portion, was the most sensitive to spinal circulatory disorders. Compression may cause microcirculatory disturbances in the territory of the anterior spinal artery or in the anterior portion of the spinal cord. Chronic circulatory disturbance resulting from repeated flexion or sustained flexed posture of the neck may produce necrosis of the anterior horns, which are most vulnerable to ischemia. The first autopsy study of Hirayama’s disease was reported by Hirayama et al (7). The lesion was addressed to the bilateral anterior horns with a side-preponderance from the cord segment C5 to T1, mostly evident at C7 and C8, where necrotic changes presumably due to local circulatory failure (ischemic myelopathy) were found. Likewise, in our patient, subclinical lesions bilaterally in the anterior horns from cord segment C5 to C6 due to Hirayama’s disease were suggested, which might have readily caused atrophy of the proximal muscles bilaterally in the upper limbs by CSA at the age of 40. This is the first report of CSA followed by Hirayama’s disease.

In conclusion, it is likely that Hirayama’s disease and CSA occurred coincidentally in the present patient. Although the prognosis of Hirayama’s disease is assumed to be better, affected patients must be followed up carefully for complications of CSA during middle age.

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References
Figure 4. CT with intrathecal contrast medium. This CT-myelogram demonstrates typical findings of cervical spondylosis and cord impingement due to bony spurring. No anterior shift of the posterior wall of the dural canal is observed during neck flexion.