CASE REPORT

Scapular winging as a symptom of cervical flexion myelopathy

Hiroaki Yaguchi, Ikuko Takahashi, Jun Tashiro, Sachiko Tsuji, Ichiro Yabe and Hidenao Sasaki

Department of Neurology, Hokkaido University Graduate School of Medicine, Kita-15 Nishi-7, Kita-ku, Sapporo 060-8638, Japan

Corresponding; Ichiro Yabe,

Department of Neurology, Hokkaido University Graduate School of Medicine, Kita-15 Nishi-7, Kita-ku, Sapporo 060-8638, Japan

Tel; +81-11-706-6028

Fax; +81-11-700-5356

e-mail; yabe@med.hokudai.ac.jp

Key words; flexion myelopathy, Hirayama disease, scapular winging, juvenile muscular atrophy of distal upper extremity, JMADUE

Abstract

A 23-year-old man complained of weakness of the right arm that he first noted six years prior to his visit. Neurological examination revealed atrophy and weakness of the triceps and serratus anterior muscle on the right side, which resulted in scapular winging on that side. MRI with neck flexion revealed compression of the cervical cord enabling a diagnosis of flexion myelopathy. Proximal muscle weakness and atrophy in flexion myelopathies including Hirayama disease are extremely rare. Here, we report a case of unilateral, proximal upper limb atrophy with scapular winging, attributed to middle cervical flexion myelopathy.

Introduction

“Flexion Myelopathy” is caused by a compression of the spinal cord during flexor movements and has been attributed to a combination of many factors. The forward movement of the dural canal in flexion and no dural folding in extension are considered to be important factors (1). Among the flexion myelopathies, juvenile muscular atrophy of distal upper extremity (JMADUE, Hirayama disease), which was first reported in 12 patients by Hirayama et al. in 1959 (2), is now a well-known entity in Japan, however, similar cases have also been described in various countries (3). The pathomechanism of Hirayama disease has been attributed to cervical flexion myelopathy involving the lower cervical cord, presenting a predominantly unilateral hand and forearm involvement (1,3). Recent neuroradiological evaluation using MRI revealed atrophy, flattening and forward displacement of the lower cervical cord, and forward displacement of the posterior dura with expansion of the posterior epidural spaces in a large number of cases (3). However, proximal muscle weakness and atrophy in flexion myelopathies including Hirayama disease are extremely rare (3). Here, we report a case of unilateral, proximal upper limb atrophy with scapular winging,

attributed to middle cervical flexion myelopathy.

Case Report

The patient was a 23-year-old man who complained of weakness of the right arm that he first noted six years prior to his visit. His past history included temporal lobe epilepsy, for which he was taking sodium valproate (600 mg / day). He had not let the neck and shoulder muscles exercise intensely. Neurological examination revealed atrophy and weakness of the triceps, serratus anterior muscle on the right side which resulted in scapular winging on that side (Fig. 1A), without any sensory disturbances. In addition, mild weakness of the right common digital extensor muscle without atrophy was also observed. The oblique amyotrophy of the forearm, characteristic of Hirayama disease, was not present. The deep tendon reflexes, except for mild hyporeflex of the right triceps, were normal in both upper and lower limbs without Babinski sign. He was not aware of “cold paresis” but trembling of the fingers was noted.

The results of laboratory tests were within normal limits, including creatine kinase, aldolase, myoglobin, sodium, potassium, calcium, c-reactive protein, endocrine and

metabolic parameters. In neck and thoracic CT, abnormality that caused the long thoracic nerve palsy was not detected. Cerebrospinal fluid examination results were also normal, including the IgG index.

He showed no signs of conduction block with a conventional motor nerve conduction study of the median and ulnar nerves. Median somatosensory evoked potentials were normal. Electromyogram in the right triceps muscle showed polyphase and high amplitude motor unit potentials and reduced recruitment, indicative of neurogenic change. Because he was undergoing medical treatment for his temporal lobe epilepsy, a magnetic evoked potential could not be recorded.

MRI with neck flexion revealed compression of the cervical cord at the level of the C4/5 vertebrae, with predominant flattening of the right side in the axial view (Fig. 1C,D), compatible with the neurological findings. This image suggested a diagnosis of flexion myelopathy. In this case, weakness of the right upper extremity was not progressive. We decided to conduct follow ups without an operation.

Discussion

Tashiro et al. described the clinical requirements for the diagnosis of Hirayama disease (3). One of the diagnostic criteria is distal dominant muscle weakness and atrophy in the forearm and hand, which means the peak in the flexion position is generally at the C6 level. Maximal tension is distributed from the C7 to Th1 vertebral level, as it takes the shortest route through the posteriorly convex spinal canal, and muscle weakness is distributed from the C7 to Th1 myelomere. Therefore, our case was not initially diagnosed as Hirayama disease. In Hirayama disease, spinal MRI in cervical flexion shows forward displacement of the dural sac and compressive flattening of the lower cervical cord with widely opened epidural spaces, suggestive of the venous plexus with a flow void (1). In this case, the peak in the flexion position was at the C4/5 level on cervical MRI and muscle weakness from the C5 to C7 myelomere was apparent. Thus, by cervical MRI this case seemed to be a flexion myelopathy, with the same pathomechanism as Hirayama disease. Similar cases have been reported in various countries (3).

In the Japanese literature, two cases with the same mechanism of flexion myelopathy, presenting muscular atrophy of proximal upper limbs, unilaterally dominant or unilateral, have been reported (4, 5) and in both cases the peak in the flexion position was at the C4 level on the neuroradiological image, but no scapular winging has been documented.

Hence our case could be classified as a specific type of flexion myelopathy, almost identical to Hirayama disease, but the presentation of scapular winging and this particular level of cord involvement have not been previously reported for this disorder. This suggests that patients presenting with scapular winging must be carefully distinguished from those with flexion myelopathy.

References

1 Kikuchi S, Tashiro K. Juvenile Muscular Atrophy of Distal Upper Extremity (Hirayama's Disease). In: Jones HR Jr, De Vivo DC, eds. *Neuromuscular Disorder of Infancy, Childhood, and Adolescence. A Clinician's Approach.* Philadelphia: Butterworth Heinemann, 2003 :167-81.

2. Hirayama K, Toyokura Y, Tsubaki T. Juvenile muscular atrophy of unilateral upper extremity; a new clinical entity. Psychiatr Neurol Jpn 1959; 61: 2190–7. (In Japanese, abstract in English.)

3. Tashiro K, Kikuchi S, Itoyama Y, et al. Nationwide survey of juvenile muscular atrophy of distal upper extremity (Hirayama disease) in Japan. Amyotrophic Lateral Sclerosis 2006; 7: 38-45.

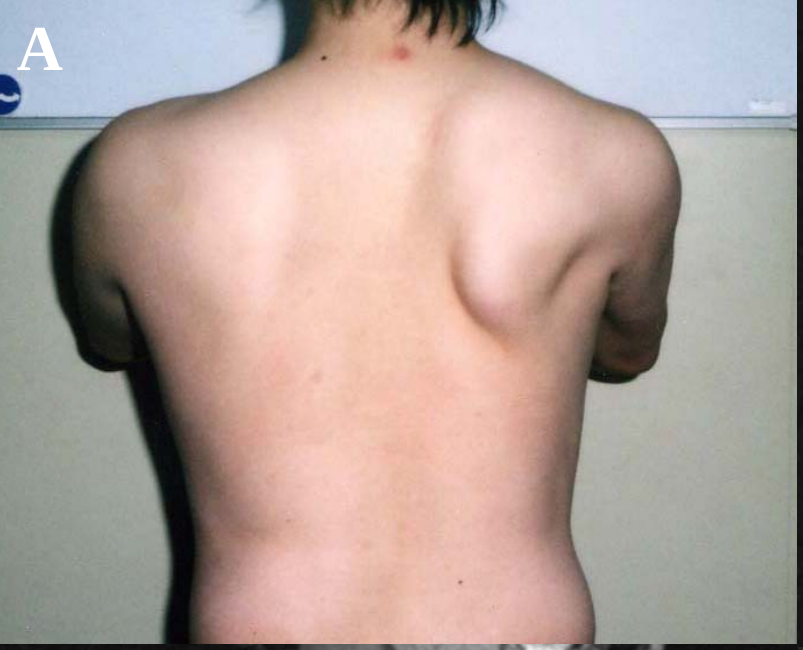
4. Masaki T, Hashida H, Sakuta M, et al. A case of flexion myelopathy presenting juvenile segmental muscular atrophy of upper extremities - a successful treatment by cervical spine immobilization. Rinsho Shinkeigaku 1990; 30: 625-9. (in Japanese, abstract in English.)

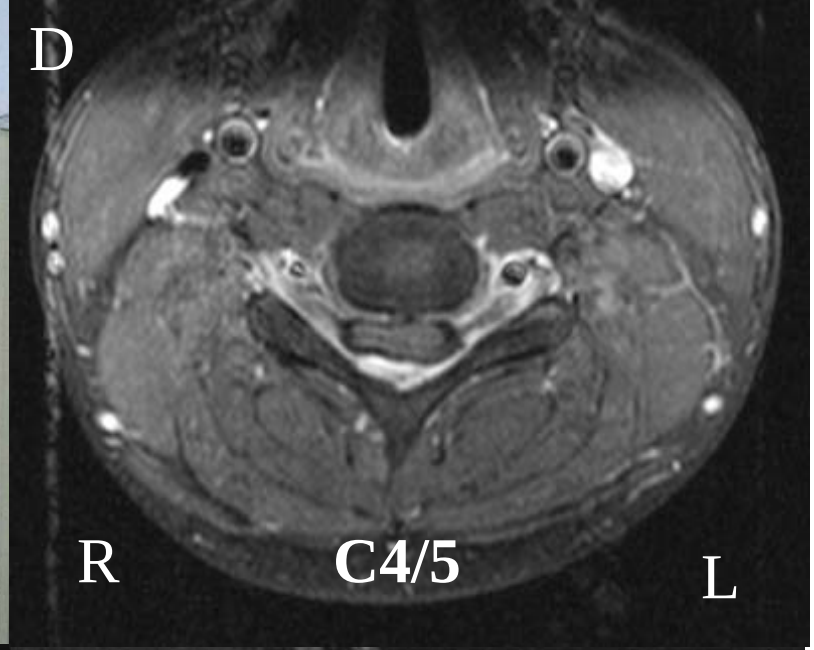
5. Ando T, Fukatsu H, Kameyama T, et al. A case of flexion myelopathy presenting with

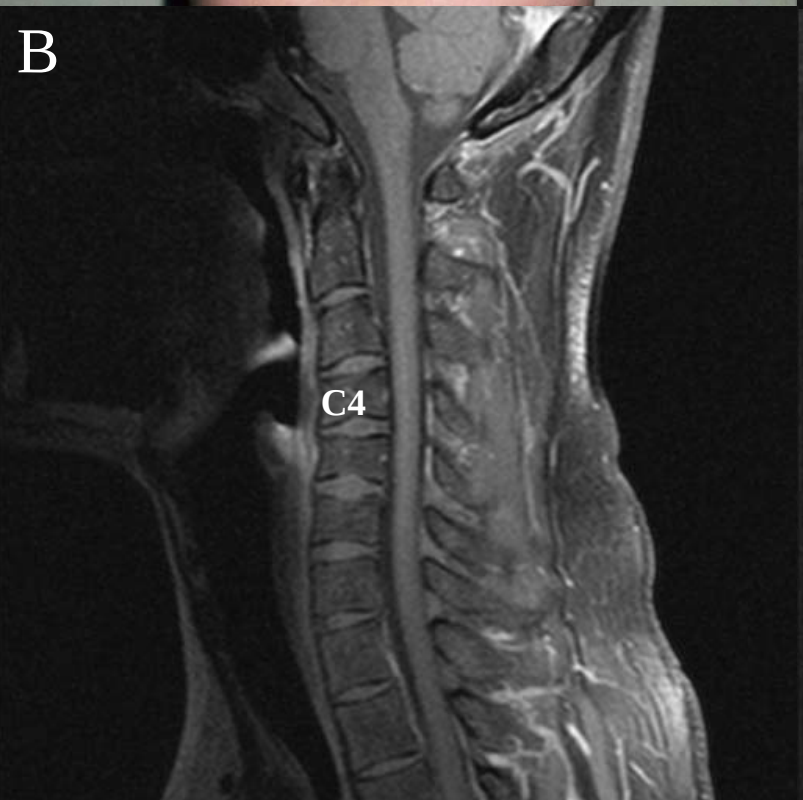
reversible muscular weakness and atrophy of unilateral proximal upper limb. Rinsho Shinkeigaku 1993; 33: 575-8. (in Japanese, abstract in English.)

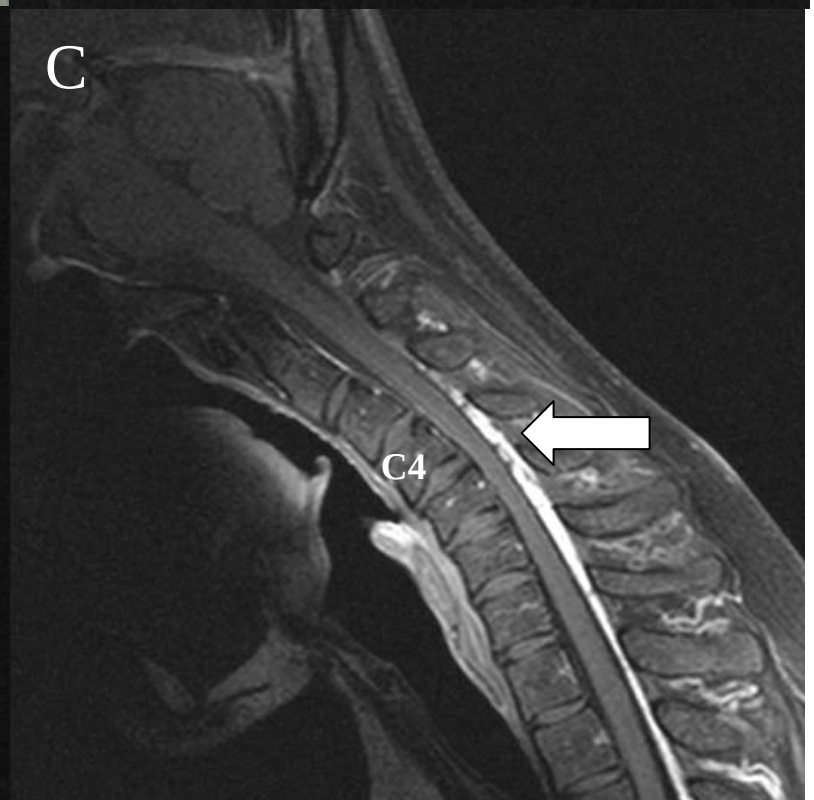
Figure Legend

(A) Scapular winging. (B) Cervical fat-suppressed and Gd-enhanced T1-weighted MRI scan in a neutral neck position shows the posterior dural wall near the spinal canal. (C, D) With neck flexion, a forward displacement of the posterior wall of the dura with an expanded venous plexus is visualized in the epidural spaces, resulting in an asymmetrical compression of the cervical cord.


A


D
R
C4/5
L


B
C4


C
C4