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# Clinical Presentation of Cervical Myelopathy at C1–2 Level

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**Study Design:** Single-center retrospective study.
**Purpose:** To clarify the clinical features of cervical myelopathy at the C1–2 level.
**Overview of Literature:** Methods for distinguishing the affected level based on myelomere symptoms or dysfunction of the conducting pathway were established. However, no symptoms have been identified as being specific to the C1–2 level segment.
**Methods:** We evaluated 24 patients with cervical myelopathy due to spinal cord compression at the C1–2 level. Preoperative neurological assessment were investigated and compared with the rate and site of compression of the spinal cord using computed tomography-myelography.
**Results:** Impaired temperature and pain sensation were confirmed in 18 of the 24 patients with that localized to the upper arms (n=3), forearm (n=9), both (n=2), and whole body (n=4). Muscle weakness was observed in 18 patients, muscle weakness extended from the biceps brachii to the abductor digiti minimi in 10 patients, and in the whole body in 8 patients. Deep tendon reflexes were normal in 10 patients, whereas hyperactive deep tendon reflexes were noted in 14 patients. The rate of spinal cord compression was significantly higher in patients with perceptual dysfunction and muscle weakness compared with those with no dysfunction. However, no significant difference in the rate and site of compression was identified in those with dysfunction.
**Conclusions:** Perceptual dysfunction and muscle weakness localized to the upper limbs was observed in 58% and 42% of patients, respectively. Neurological abnormalities, such as perceptual dysfunction and muscle weakness, were visualized in patients with marked compression.

**Keywords:** Cervical spine; Myelopathy; Atlantoaxial joint; False localizing sign; Spondyloarthropathy

## Introduction

Cervical myelopathy at the C1–2 level has a diverse range of clinical presentations. False localizing signs (FLSs) are commonly found, indicating a level of cervical dysfunction that is inconsistent with neurological symptoms. Myelopathy at the C1–2 level can be difficult to diagnose if the patient's chief complaints are hand numbness or perceptual symptoms in the trunk and when compressive lesions are detected between multiple vertebrae. Many studies have compared neurological symptoms of cervical myelopathy at this level with those of that at the subaxial level. In this one report [1], methods for distinguishing the affected level based on myelomere symptoms or dysfunction of the conducting pathway were established. However, few long-tract symptoms are pathognomonic of specific perceptual

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disorders in the upper cervical cord, and no symptoms have been identified as being specific to the C1–2 level segment. Furthermore, although FLSs have been described in lower cervical cord lesions, few reports have examined FLSs at the upper cervical cord level, particularly at the C1–2 level [2-5]. In this report, we describe the clinical features of cervical myelopathy at the C1–2 level.

## Materials and Methods

We evaluated 24 patients (9 males, 15 females) in whom cervical myelopathy due to spinal cord compression at the C1–2 level was diagnosed on imaging and who had undergone cervical decompression and fusion for relief in our department from April 2005 to March 2012 (Table 1). Data were collected prospectively and analyzed retrospectively. This study received Institutional Review Board approval. The mean duration of morbidity was 10.1 months (range, 3–25 months) and the mean age at the time of surgery was 62.7 years (range, 16–84 years). Identified conditions included atlantoaxial subluxation (n=13), retro-odontoid pseudotumors (n=5), spinal cord tumor (n=5), and basilar impression (n=1).

Preoperative neurological assessment of the upper cervical cord revealed defects in perception of temperature and pain sensation, muscle strength, and reflexes. Patients with perceptual and muscular impairments limited to the upper limbs were classified as upper-limb type, and those with symptoms from the neck to the torso and in the lower limbs were classified as whole-body type. The rate and site of compression of the spinal cord were determined using computed tomography-myelography (CTM). The

**Table 1.** Patient characteristics and functional data

| Case | Age (yr) | Sex | Diagnosis | Japanese Orthopaedic Association score (upper/lower extremity/sensory, points) |
|---|---|---|---|---|
| 1 | 32 | Male | Spinal cord tumor | 3/4/3 |
| 2 | 78 | Female | Retro-odontoid pseudotumor | 3/3/5 |
| 3 | 53 | Male | Spinal cord tumor | 3/4/6 |
| 4 | 68 | Female | Basilar impression | 1/1/6 |
| 5 | 74 | Female | Atlantoaxial subluxation | 2/4/6 |
| 6 | 70 | Female | Atlantoaxial subluxation | 2/1/5 |
| 7 | 65 | Female | Atlantoaxial subluxation | 2/4/5 |
| 8 | 84 | Female | Retro-odontoid pseudotumor | 1/0/3 |
| 9 | 67 | Female | Atlantoaxial subluxation | 0/0/5 |
| 10 | 59 | Male | Spinal cord tumor | 2/4/3 |
| 11 | 38 | Male | Retro-odontoid pseudotumor | 2/3/3 |
| 12 | 68 | Male | Atlantoaxial subluxation | 3/4/6 |
| 13 | 58 | Female | Spinal cord tumor | 3/4/5 |
| 14 | 81 | Female | Retro-odontoid pseudotumor | 2/4/5 |
| 15 | 62 | Male | Atlantoaxial subluxation | 4/4/5 |
| 16 | 69 | Female | Atlantoaxial subluxation | 4/4/6 |
| 17 | 72 | Female | Atlantoaxial subluxation | 4/4/5 |
| 18 | 73 | Female | Retro-odontoid pseudotumor | 4/4/5 |
| 19 | 37 | Female | Atlantoaxial subluxation | 4/4/5 |
| 20 | 83 | Male | Atlantoaxial subluxation | 4/4/5 |
| 21 | 63 | Female | Atlantoaxial subluxation | 2/4/5 |
| 22 | 69 | Female | Atlantoaxial subluxation | 3/4/6 |
| 23 | 66 | Male | Atlantoaxial subluxation | 2/1/5 |
| 24 | 16 | Male | Spinal cord tumor | 2/3/5 |

compression rate was calculated from axial CTM images (Prism PACS Enterprise; PRISM Medical Inc., Sapporo, Japan) of the cross-sectional area at C1–2 level compared with that of normal areas. On the same CTM axial images, the transverse diameters of the compression sites were divided into thirds and classified as center type (most compressed site in the midline) or exterior type (lateral compression) (Fig. 1). Statistical analysis was conducted using the Mann–Whitney U test. A $p<0.05$ was considered to be statistically significant.

## Results

Impaired temperature and pain sensation were confirmed in 18 of the 24 patients (75%), localized to the upper arms (n=3), forearm (n=9), or both (n=2) (Fig. 2). Thus, 14 pa-

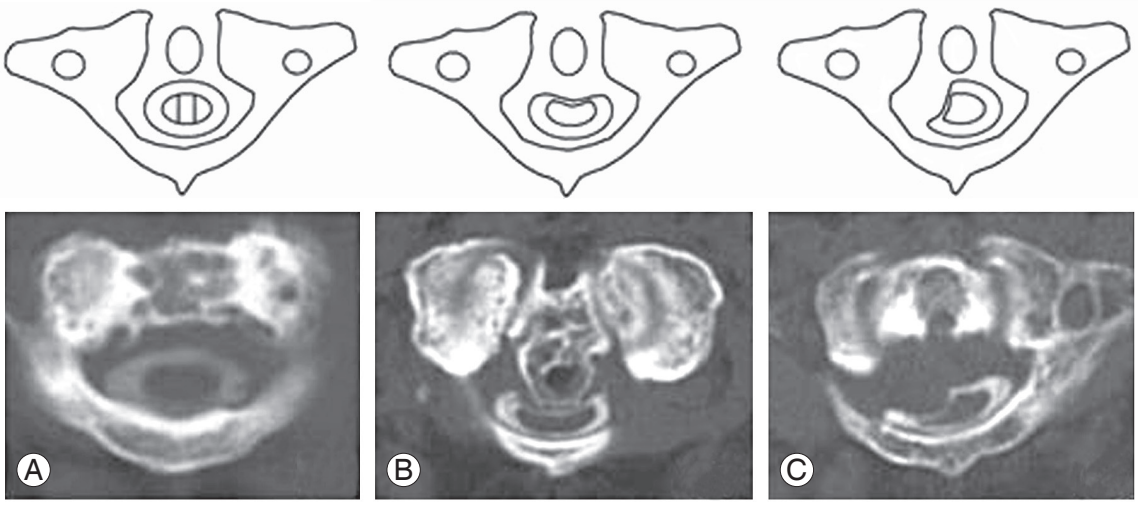


**Fig. 1.** Computed tomography-myelography images. Image at the C1–2 level **(A)**. The transverse diameters of the compression sites were divided into thirds and classified as center type **(B)** or exterior type **(C)**.

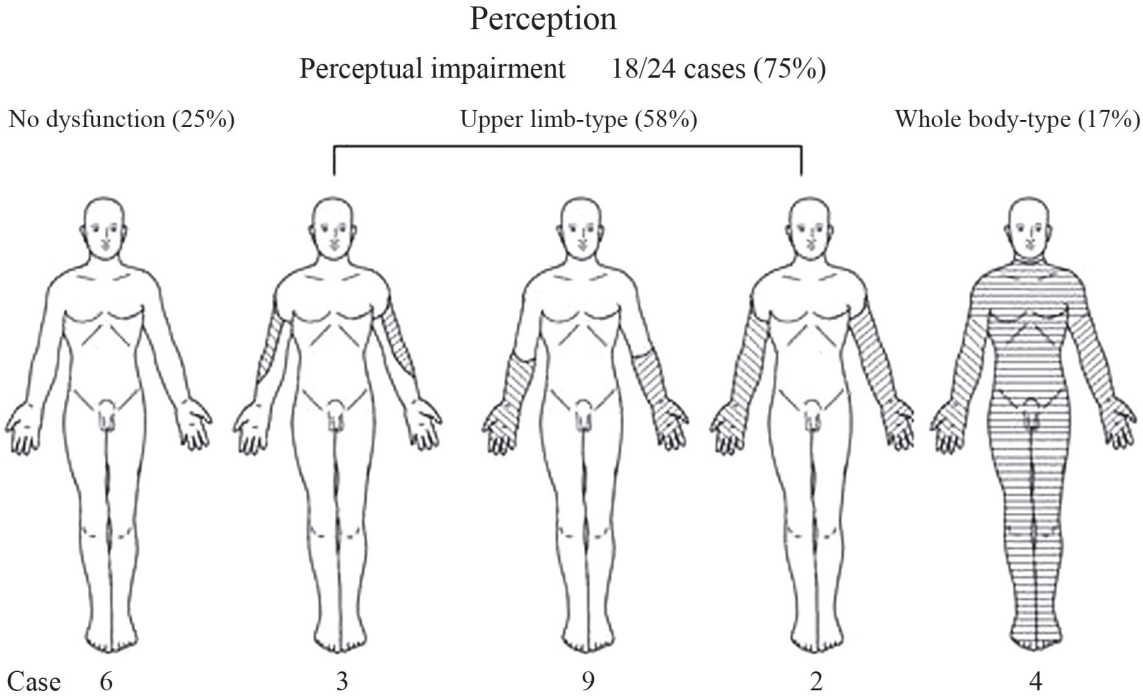


**Fig. 2.** Location of perceptual impairment (diagonal lines).

tients (58%) were classified as upper-limb type with perceptual impairment and 4 patients (17%) with perceptual dysfunction from the neck to the trunk and bilateral lower limbs were classified as whole-body type.

Muscle weakness was observed in 18 patients (75%). In 10 patients (42%) of the upper-limb type, muscle weakness extended from the biceps brachii to the abductor digiti minimi, and in 8 patients (33%) of the whole-body type, muscle weakness extended from below the trapezius to bilateral lower limbs (Fig. 3). After decompression and fusions, 6 cases improved completely, 9 showed gradual improvement, 3 remained the same as their preoperative status of perceptual impairment. Muscle functional recovery completed in 8 cases and 10 patients did not recover to normal levels at a mean follow-up of 21 months (range, 12–31 months). Deep tendon reflexes were normal in 10 patients, whereas hyperactive deep tendon reflexes were noted in 14 patients. Of the patients with hyperactive deep tendon reflexes, a hyperactive scapulohumeral reflex was evident in 7 patients, whereas in the other 7 patients, a hyperactive reflex below the biceps brachii was found, but the scapulohumeral reflex was normal. Pathological reflexes were indicated by a positive Hoffmann's sign in 10 cases and a positive Babinski's sign in 4.

Patients were divided into the following three subgroups based on perceptual and muscle strength impairment: no dysfunction, upper-limb type, and whole-body type. No statistically significant differences in sex or age were noted between these groups. The rate of spinal cord compression was significantly higher in patients with perceptual dysfunction (Fig. 4), muscle weakness (Fig. 5)

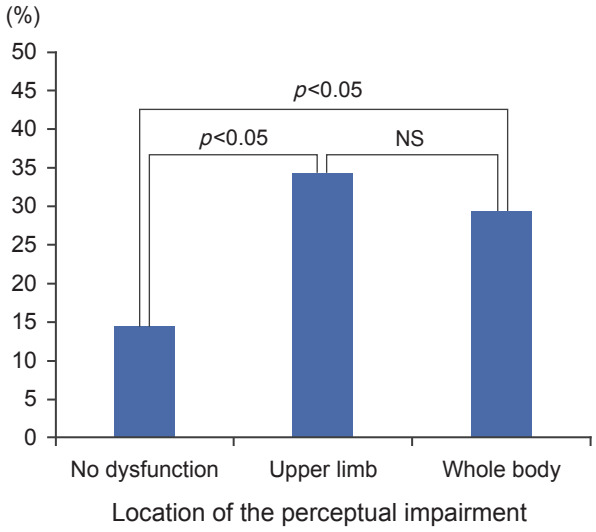


**Fig. 4.** Relationship between perceptual impairment and the rate of spinal cord compression. NS, not significant.

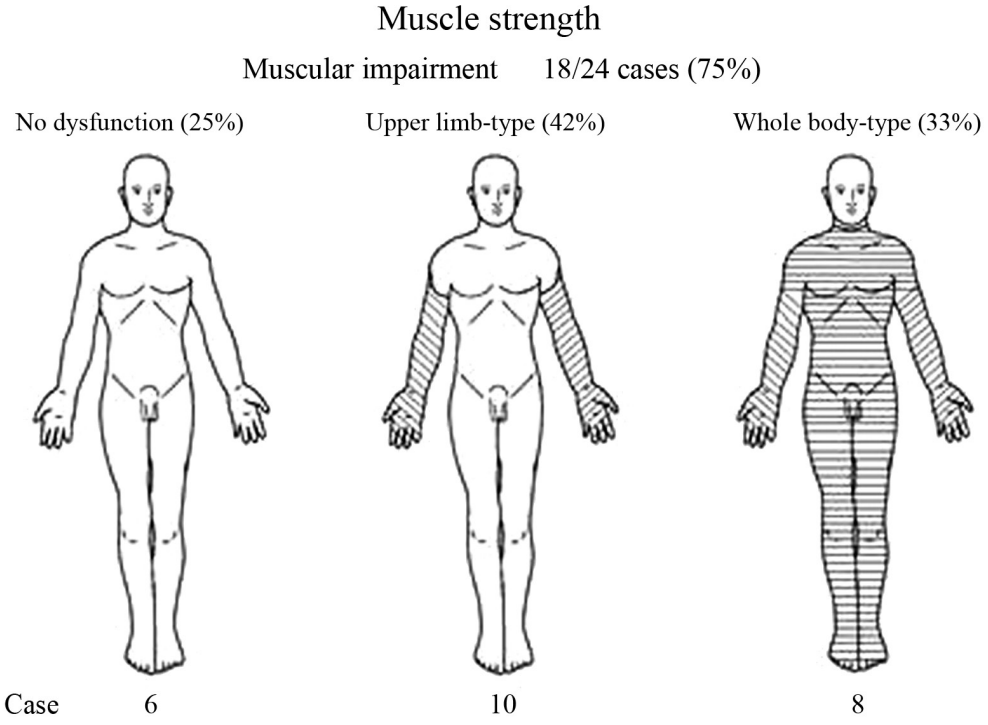


**Fig. 3.** Location of muscular impairment (diagonal lines).

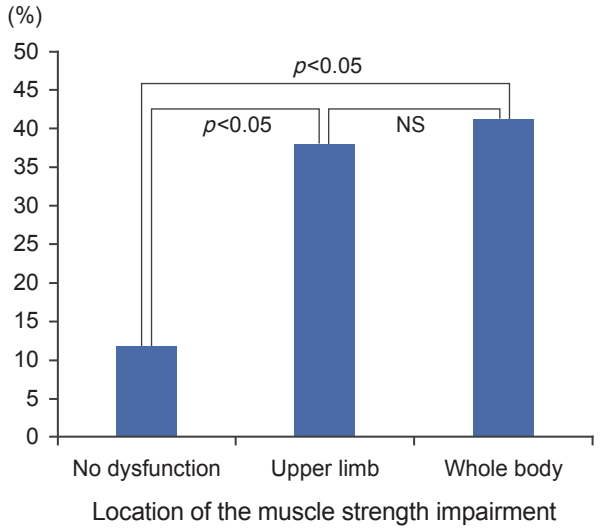


**Fig. 5.** Relationship between muscular impairment and the rate of spinal cord compression. NS, not significant.

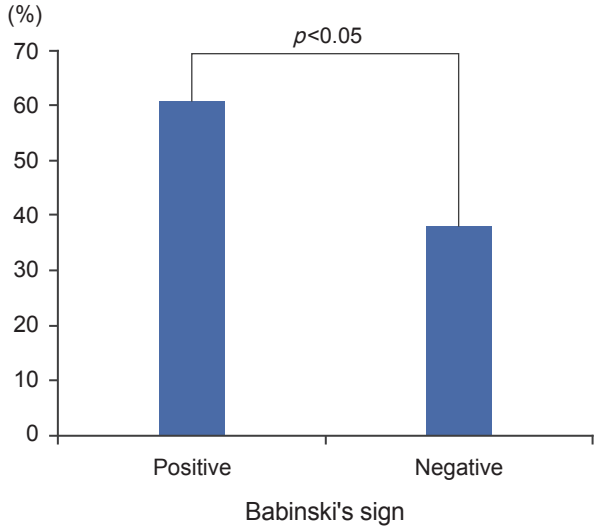


**Fig. 6.** Relationship between Babinski's sign and the rate of spinal cord compression.

and positive Babinski's sign (Fig. 6) compared with those with no dysfunction. However, no significant difference in compression rate was identified between the upper-limb and whole-body type groups.

During a comparison of compression sites, no perceptual dysfunction or muscle weakness was observed in some center-type patients, whereas in all exterior-type patients, some impairment was observed. No relationship was observed in terms of the site of compression between the upper-limb and whole-body type groups (Table 2).

## Discussion

In this study, the clinical presentation of cervical myelopathy at the C1–2 level was investigated. Of the patients with whole-body type dysfunction, which involved perceptual disturbances of the occipital region, including the neck and shoulders, or which involved upper limb muscle weakness below the trapezius, perceptual dysfunction was found in only 4 (17%), whereas weakness of the trapezius muscle was found in 8 (33%). In contrast, upper-limb type dysfunction was common; perceptual dysfunction of the upper arm and/or forearm was identified in 14 patients (58%) and muscle weakness localized to this area was evident in 10 patients (42%). This phenomenon of contradictory findings between the spinal cord compression level (assessed by imaging) and the level indicated by functional neurological symptoms is referred to as FLSs. In patients with cervical myelopathy at the C1–2 level examined in this study, FLSs were present in 42%–58% of patients.

FLSs were first described by Stookey [6] who observed temperature and pain perceptual dysfunction in two to three segments lower than the compression level in patients with cervical disc herniation. Sonstein et al. [4] reported 11 patients with cervical myelopathy at the C3–4

**Table 2.** The relationship between the site of compression and dysfunction type groups

| | No dysfunction | Upper limb-type | Whole body-type |
|---|---|---|---|
| The perceptual impairment and the site of compression | | | |
| Center-type | 6 | 10 | 2 |
| Exterior-type | 0 | 5 | 1 |
| The muscle strength impairment and the site of compression | | | |
| Center-type | 6 | 6 | 5 |
| Exterior-type | 0 | 5 | 2 |

level, a lower cervical level dysfunction, particularly characterized by atrophy of intrinsic muscles of the hand and numbness in the hands and fingers. FLSs are rare in lower cervical myelopathies, in which systematic segmental symptoms and conduction pathway disturbances contradict the diagnosis of a higher level lesion. In contrast, FLSs was observed in a majority of cervical myelopathy patients at the C1–2 level examined in this study. The area of perceptual dysfunction for these lesions (the occiput and the area from the neck to the shoulders) was only affected in four patients (17%). In the remaining patients, highly variable clinical presentations involved perceptual disturbances in the upper limb. These perceptual FLSs symptoms (upper-limb type) were observed in 14 patients (58%). In addition, in 10 patients (42%), FLSs were associated with biceps brachii or the area below triceps brachii muscle weakness.

Cranial nerve symptoms may be present because of specific anatomical features in the bulbospinal transition area and accessory nerves at the C1–2 level. Cranial nerve symptoms occur infrequently and typically manifest only after myelopathy has progressed considerably. One case of dysphagia due to glossopharyngeal, vagal, and hypoglossal nerve dysfunction was noted in this study. In this case, dysphagia was associated with perceptual dysfunction from the neck to bilateral lower limbs and muscle weakness below the trapezius muscle. Although systemic symptoms may manifest in severe patients, no relationship was found in this study between the rate of compression and the range of dysfunction (Figs. 4–6).

The mechanisms underlying FLSs remain unclear, but several factors have been implicated, including vascular factors. In monkeys, Taylor and Byrnes [7] found that spinal cord compression at the C2–3 level led to congestion of the spinal venous plexus, degeneration of the ventral horn cell, and posterior horn hemorrhage at the C8–Th1 level. Ochiai et al. [5] also suggested a relationship between FLSs and ischemia in the anterior spinal artery due to decreased superficial perception, but normal deep sensation in patients of severe compression from the ventral side of the spinal cord. They also reported that compression from the ventral side was significantly stronger in the FLSs group (upper-limb type) than in the non-FLSs group (whole-body type). In the present study, no significant differences in the compression rate or site were found between the FLSs and non-FLSs groups (Table 2) (Figs. 4, 5). Because the anterior spinal artery originates at the C1–2 level, it may account for the large proportion of FLSs patients in the current study of C1–2-level lesions.

Hirabayashi et al. [8] reported that from an anatomical viewpoint, the appearance of FLSs may be related to the proximity of the ventral and posterior horns because at the functional level, the posterior horn is approximately one segment higher than the ventral horn. However, this cannot account for the findings of the present study. In patients with FLSs, perceptual dysfunction and muscle weakness were localized four to five segments lower than the spinal cord compression level, as assessed by imaging. These results are in accordance with those of other studies of FLSs [2-4,9].

Ichihara et al. [10] proposed a novel mechanism underlying the development of spinal compressive lesions based on spinal biomechanics. They suggested that sudden posterior spinal compression causes axonopathy of the posterior and lateral funiculi, and subsequent severe compression leads to hemorrhage and softening of the gray and white matter in the center of the spinal cord. Considering this central spinal cord dysfunction from the viewpoint of cervical cord cross-sectional alignment, Tsuzuki [11] proposed a mechanism whereby the upper limb descending fibers are selectively impaired. Similarly, Forester [12] suggested that motor dysfunction is significantly more common in the upper limbs because the descending fibers of the corticospinal tract are deeper in the spinal cord. Based on spinal biomechanics, the relatively greater size of the subarachnoid space at the C1–2 level compared with that at other levels may lead to diversity in the range of dysfunction and symptoms from lesions at the C1–2 level.

The limitations of this study include the small sample size and the fact that all patients had undergone surgery. In addition, differences in primary disease and associated neurological symptoms were not investigated in this study. The mechanisms behind FLSs remain unclear, and many of the clinical features of cervical compressive myelopathy at the C1–2 level are not understood. Large-scale studies are required to solve these problems.

## Conclusions

We examined the clinical presentation of 24 patients with C1–2 level cervical myelopathy. Perceptual dysfunction localized to the upper limbs was observed in 58% of cases and muscle weakness localized to the upper limbs was observed in 42% of cases. Cases with marked compression,

as observed on imaging, had neurological abnormalities, such as perceptual dysfunction and muscle weakness.

## Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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