

## Case Report

### Cruciate paralysis and hemiplegia cruciata: report of three cases

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**Study design:** Report of three cases of cruciate paralysis and hemiplegia cruciata.

**Objective:** To stress the importance of upper cervical spine lesions causing neurological symptoms and signs.

**Setting:** Neuro-orthopedic service, Fukui University Hospital, Japan.

**Results:** Three patients (all females; one with congenital anomaly at the occiput-atlas level, one with assimilation of the atlas, and one with rheumatoid arthritis-related proliferative synovium) had clinical features of cruciate paralysis and hemiplegia cruciata. All three cases underwent decompressive surgeries.

**Conclusion:** Neurological symptoms and signs of cruciate paralysis and hemiplegia cruciata should be carefully assessed, and surgical therapy should be based on the pathological condition.

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**Keywords:** cruciate paralysis; hemiplegia cruciata; Wallenberg syndrome; cervicomedullary junction; pyramidal decussation

## Introduction

The craniocervical junctional area is the site of a variety of diseases as well as neurological symptoms and signs secondary to neural compromise. In 1901, Wallenberg<sup>1</sup> described the complex neuroanatomy of the corticospinal tracts at the cervicomedullary junction. The condition named 'cruciate paralysis' by Bell<sup>2</sup> is an uncommon neurological complication pertaining to mechanical injury, metabolic disorders, or complications with surgery<sup>3–5</sup> at the cervicomedullary junction proximal to the pyramidal decussation. Characteristically, patients with cruciate paralysis present with bilateral paresis or paralysis of the upper extremities without significant involvement of the lower extremities. When the neural compromise occurs predominantly on one side, spastic palsy on the ipsilateral side of the upper extremity is present, which is associated with spasticity on the contralateral side of the lower extremity, described as hemiplegia cruciata.<sup>6–8</sup> Detailed neurological examination and careful neuroimaging will highlight these two types of symptomatology, but delay in the diagnosis and treatment may correlate with unfavorable neurological recovery.

The current communication describes three illustrative cases of hemiplegia cruciata with reference to the possible neuroanatomic compromise.

## Case report

### Case 1

The patient was a 67-year-old woman who presented with progressive tetraparesis following a minor fall 7 months prior to admission. She also reported a slight difficulty in speech. Her past medical record and family history were unremarkable. On presentation in January 1983, neck movements were full in all directions. The only abnormality in the cranial nerve territory was a wasted left side of the tongue and deviation of the tongue to the left. There was marked spasticity particularly involving the left arm and the right leg. Muscle weakness was noted in the left arm and both legs. Pain and thermal sensation deficits were more evident on the right arm and leg, and deficits of touch and vibration were noted on the left arm and leg. Deep tendon reflexes were exaggerated particularly in the left arm and right leg, and pathological reflexes were also positive on the left arm and right leg. The patient also reported slight difficulty in voluntary micturition.

Radiographs showed assimilation of the atlas into the base of the skull and a healed fracture of the odontoid

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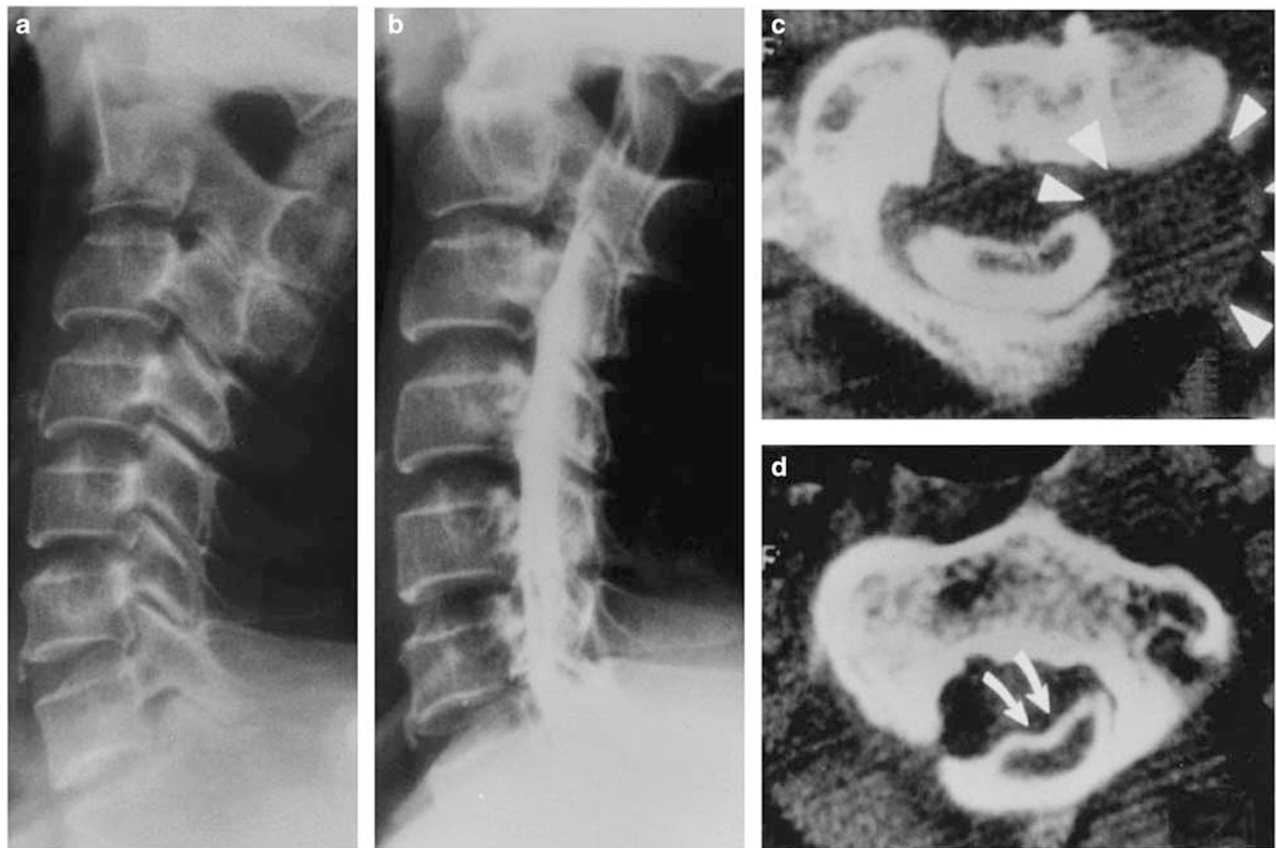
base. Myelography demonstrated a displaced cervical cord posteriorly at the level of foramen magnum to C3 vertebra (Figure 1a, b). Computed tomography myelography showed severe flattening and displacement of the spinal cord by an abnormal space-occupying mass (cystic) lesion extending extraspinally through the left-sided interlaminar space (Figure 1c, d).

With a provisional diagnosis of a compressive lesion most likely a hematoma, cyst or an epidural tumor, the patient underwent one-session anterior and posterolateral decompression with autologous bone grafting. In the first, the odontoid process was resected via the transoral approach followed by iliac bone grafting between the clivus and the axis. An old hematoma was found anterior to the dura mater and this was curetted and drained. Next, after partial resection of the left side of the posterior arch of the atlas, the 'Blutwurst' – like hematoma was fairly completely removed posterolaterally. However, even after extensive decompression, the dura mater did not expand with vigorous pulsation. Autologous iliac bone grafting was then pursued between the occiput and the C3 vertebra. The post-operative period was uneventful with no complications. The autologous bone graft appeared fused at the time of

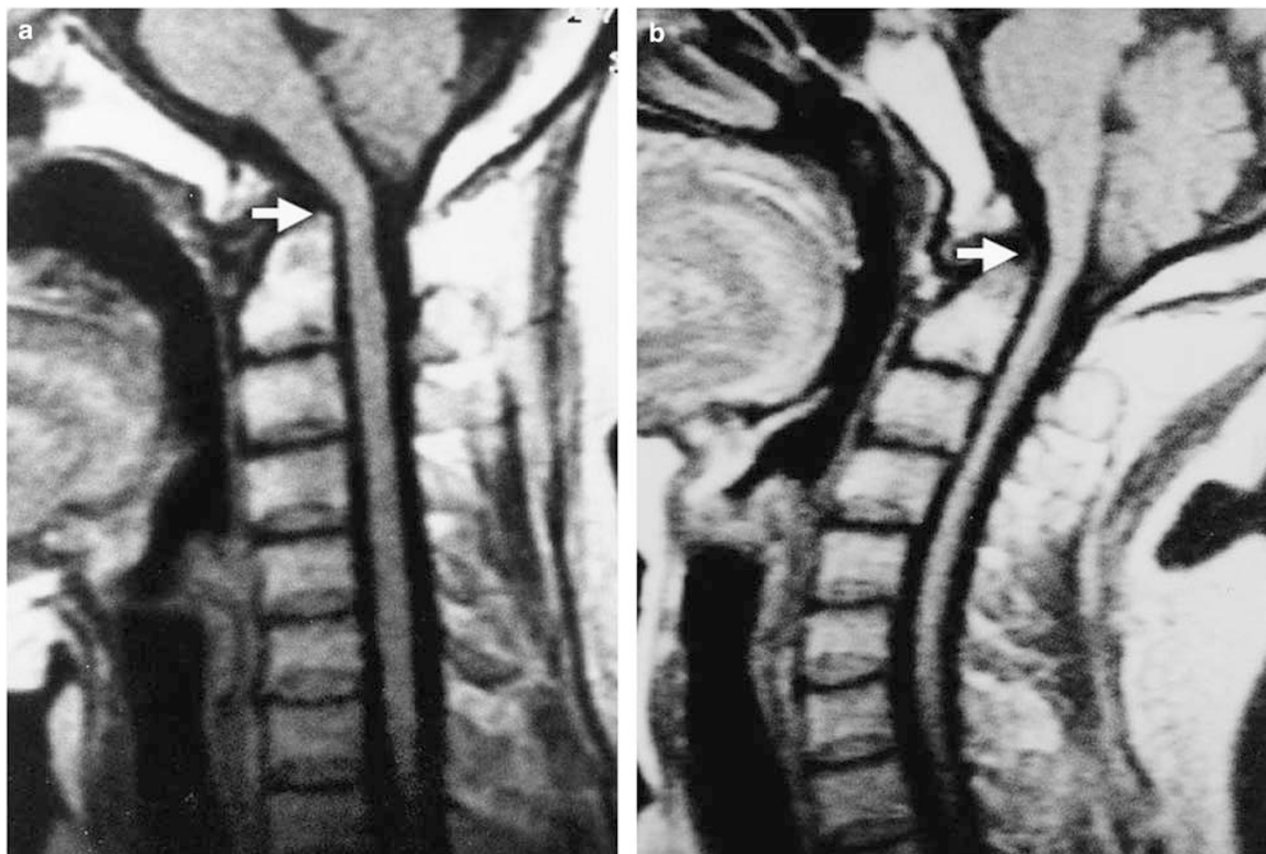
discharge 4 months after surgery. However, no neurological improvement was noted even 2.3 years after surgery. No further follow-up could be provided.

#### Case 2

A 49-year-old woman was admitted to our University Hospital in 1988 complaining of difficulty in walking. In 1976, she noticed 'dullness' in both arms, and numbness that gradually extended to both arms and legs and culminated in inability to use the right hand for writing. In 1983, she underwent transoral resection of the odontoid process at another hospital, but bone grafting between the atlas (presumably the anterolateral part of the arch) and axis subsequently failed. A year prior to referral to our hospital, she could not walk without support. The past history and family history were unremarkable. On presentation, cranial nerve examination was unremarkable. There was marked spasticity predominantly in the right arm and left leg with a significant unsteady and ataxic gait. Muscle weakness was noted in the right arm and both legs. Pain and thermal sensation disturbances were noted mainly in the right arm and leg, and touch and vibration senses were



**Figure 1** Case 1. (a) Plain radiography showing assimilation of the atlas. (b) Myelography showing displaced cervical cord at the level of foramen magnum to C3 vertebra. (c, d) Computed tomography myelography showing displacement of the cord at the atlantoaxial level by a space-occupying mass (white arrow)



**Figure 2** Case 2. MRI showing compression of the spinal cord anteriorly by the odontoid process (white arrow) (a; flexion, b; extension)

markedly diminished in the extremities except for the left leg. Deep tendon reflexes in both arms and legs were exaggerated, but no pathological reflexes could be elicited. The patient complained of progressive dysurination (retardation).

Radiologically, the posterior arch of the atlas was assimilated with the occiput and the space available for the cord at the atlas level measured 12 and 15 mm in neck flexion and extension, respectively, indicating mechanical instability. The odontoid process and the body of the axis had moved cephaladly (Ranawat scale, 11 mm; Redlund-Johnell scale, 26 mm). Magnetic resonance imaging (MRI) demonstrated considerable anterior impingement of the spinal cord by the superoposterior tip of the odontoid process (Figure 2a, b).

In December 1988, the patient underwent suboccipital craniotomy for posterior decompression, and posterior fusion (Newman's procedure) between the occiput and C3 vertebra in neck-extended position in order to relieve as much as possible the cervical cord from anterior compression. Solid bony union was observed 4 months later. At 7-year follow-up, the patient was ambulatory with only very occasional use of a cane and was doing well as a housewife, even though recovery of sensory deficits was insignificant.

### Case 3

A 67-year-old woman with a 15-year history of rheumatoid arthritis developed clumsiness of both hands, predominantly on the right, 2 years before presentation. On examination in December 1998, the gait was normal and cranial nerves examination was unremarkable. Spasticity was noted in the right arm and left leg. Muscle weakness was evident in both legs, mainly on the left side. Pain and temperature sensation were diminished particularly on the left upper and lower extremities, while decreased sense of touch was symmetric. Deep tendon reflexes were insignificantly positive on the right arm and left leg. Hoffman's and Wartenberg's reflexes were positive especially on the right side, while Babinski's and Chaddock's reflexes were positive on the contralateral side. A urodynamic study showed a neuropathic bladder (pollakiuria, retardation).

Atlantoaxial subluxation was evident on radiographs. Cervical MRIs showed flattening of the cervical cord anteroposteriorly and an abnormal lesion suggestive of synovial hypertrophy, with hyperintense signal intensity impinging on the cord anteriorly at vertebral level of the atlas (Figure 3a, b). A hypertrophied pannus posterior to the odontoid process was suspected, which was treated by medication, including steroids, and transoral



**Figure 3** Case 3. MRI T2 weighted images showing abnormal region compressing the spinal cord anteriorly at the atlas level (white arrow) (a; sagittal image, b; axial image). (c) Postoperative radiograph showing posterior atlantoaxial fusion

anterior surgery was excluded. However, in January 2001, the patient underwent posterior atlantoaxial fusion (Brooks and Jenkins's procedure) and the post-operative course was uneventful (Figure 3c). Bony union was evident 3 months later, and the patient attained almost full neurological recovery at 6-month follow-up.

## Discussion

Wallenberg<sup>1</sup> was the first to describe in detail the complex neuroanatomical disorder of the corticospinal tracts at the cervicomedullary junction, and suggested that the neurological presentation pertaining to the neural compromise in this particular region may vary according to the location of the lesions involving the descending pyramidal tracts. Various pathologies can result in the compression of this particular area of the spinal cord.<sup>9–15</sup> Relief of the cervicomedullary junction from compression followed by restructuring the uppermost part of the vertebrae can result in neurological improvement in some cases.

The corticospinal tract of the upper extremity descends more medially and anteriorly relative to that of the lower extremity.<sup>2</sup> Furthermore, the spinal cord segment level of the pyramidal decussation of the

corticospinal tract of the upper extremity is located proximal to the level of the foramen magnum, which is approximately one cord segment proximal to that of the lower extremity.<sup>16–19</sup> This anatomic and topographic difference sometimes causes unusual clinical manifestations such as cruciate paralysis and hemiplegia cruciata, which is often different from that of cervical myelopathy or typical stroke. Modern imaging techniques, including 18F-2-fluoro-deoxy-D-glucose-positron emission tomography, have enhanced the detection of mechanically compressive lesion(s) to the cord at the cranio-vertebral junction, and thus early diagnosis is possible.<sup>20,21</sup> However, the diagnosis itself may infrequently be delayed because of the tremendous increase in the incidence of brain stroke and/or cervical myelopathy, and thus a careful neurological examination is always essential (Table 1).

Compressive lesions at the medullo-cervical junction can cause preferential paralysis of the upper extremity and cranial nerve dysfunction. Lesions affecting the proximal portion of the pyramidal decussation could also affect associated brain-stem structures, such as IX, X, XI, and XII cranial nerves. In fact, transient respiratory insufficiency, urinary retention, and lower cranial nerve palsies have been described together with,

**Table 1** Clinical presentation, treatment, and outcome in three patients with cruciate paralysis

	Case 1	Case 2	Case 3
Age (years)	67	49	67
Sex	F	F	F
Diagnosis	Old hematoma	Atlas assimilation	Rheumatoid arthritis
Cranial nerve deficits	XII	Normal	Normal
Spasticity	Bil upper extremities (rt < lt), rt lower extremity	rt upper extremity, lt lower extremity	Bil upper extremities (rt > lt), lt lower extremity
Treatment	Transoral odontoid resection, drainage, C0–3 fusion	Suboccipital craniotomy, posterior fusion	Atlanto–axial fusion
Clinical outcome	Spastic gait  Bil upper extremities spasticity	rt upper extremity hypesthesia	Complete recovery

Bil: bilateral, rt: right, lt: left.

though less frequently, hypoglossal, vagal, and glosso-pharyngeal dysfunction.<sup>7,10,12,19</sup> In our cases, hypoglossal nerve injury was identified in one patient and urinary dysfunction was noted in all patients.

The presence of a mechanically compressive lesion warrants surgical therapy, which is another important issue that needs careful consideration. In most cases with this uncommon condition, the lesion is located anterior to the spinal cord rather than posteriorly. For optimal decompression effects, anterior decompression may be ideal and essential. However, the technical intricacy of the trans-oral anterior approach to the spinal cord at the cranio-vertebral junction may not always result in optimal decompression.<sup>22–25</sup> Thus, an additional posterior decompression, either through one or two sessions, may be required, as in our cases. However, long-term conservative treatment without surgery together with prolonged palsy should have a negative effect on neurological outcome. Neurological improvement after surgery depends on neuronal plasticity, early diagnosis, and appropriate surgical therapy.

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