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Original article

Occipito-atlanto-axial Hypermobility : Clinical Features and Dynamic Analysis of Cranial Settling and Posterior Gliding of Occipital Condyle.
Part 2 : Findings in Patients with Post-traumatic Condition

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Abstract

Object : To examine that post-traumatic condition (PTC) can present with lower brain stem symptoms attributable to occipito-atlanto-axial hypermobility and cranial settling, and its relationship to Chiari malformation type I (CMI).

Methods : We defined the condition of continuous brain and neck symptoms after motor vehicle accidents or falling down with loss of consciousness as PTC and 56 patients met to the criteria. Osseous structures comprising the craniocervical junction were investigated morphometrically using reconstructed 2D-CT and plain x-ray images in 50 patients with PTC, and the results were compared to normal controls (n=55).

Results : There were 28 cases (50%) in PTC of CMI. The diagnostic features of PTC with CMI had a greater incidence of lower brain stem symptoms and signs. We performed measurements of the basion-dens interval (BDI), basion-atlas interval (BAI), atlas-dens interval (ADI), dens-atlas interval (DAI), clivus-atlas angle (CAA), clivus-axis angle (CXA), and atlas-axis angle (AXA). They were the same in supine and upright positions in normal controls. In patients with PTC, there was reduction of the BDI (2.7 mm), enlargement of the BAI (2.8 mm), and reduction of the CXA (11.4°), CAA (6.6°, $p<0.001$), and AXA (11.0°) upon assumption of the upright position. These changes were reducible by cervical traction.

Conclusions : Morphometric evidence in this cohort of cranial settling and posterior gliding of the occipital condyles in PTC suggests hypermobility of the atlanto-occipital and atlanto-axial joints. This hypermobility induces greater brain stem symptoms in patients associated with CMI. The patients with CMI have greater hypermobility of occipito-atlanto-axial joints.

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Key words
craniocervical instability, cranial settling, trauma, Chiari malformation, retroodontoid mass lesion

Introduction

We reported the clinical features of patients with hereditary disorders of connective tissue (HDCT) and Ehlers-Danlos syndromes (EDS) and a subset of patients with HDCT・EDS in which varying degrees of craniocervical instability appeared to be related to underlying occipito-atlanto-axial joints hypermobility in previous articles[1].

It is well known that the patients in post-traumatic condition (PTC) have similar symptoms and signs as those in the patients with HDCT・EDS[2]. Beside that, recently some authors reported occipito-atlanto-axial dislocation in traumatic patients who have severe brain stem symptoms and signs[3-12]. According to these evidences, we

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can expect that the patients with PTC will have occipito-atlanto-axial hypermobility and cranial settling. Furthermore it was observed that after severe trauma, some patients with severe brain stem symptoms and signs have Chiari malformation type Ⅰ (CMI) which has been described in relation to the underdevelopment of the occipital bone and para-axial mesodermal insufficiency[13-15].

We examined that PTS could also present with lower brain stem symptoms attributable to occipito-atlanto-axial hypermobility, and cranial settling, posterior gliding and anterior flexion of occipito-atlanto-axial joints as well as HDCT・EDS[1]. We reported that association with CMI made the brain stem symptoms and signs worse, and the patients with CMI have greater hypermobility of occipito-atlanto-axial joints. These evidences suggested that cranio-cervical instability due to occipito-atlanto-axial hypermobility might be closely related with causes of tonsillar herniation[1] and formation of retroodontoid mass lesions which were described by some authors[16-20].

Methods

1 Study population

The subjects of this study were 56 patients who were evaluated consecutively with PTC between January 2006 and December 2007 in the Chiari Institute, North Shore University Hospital (**Table 1**). There were 31 females and 25 males who ranged in age from 19-68 years (mean age=39.8 years±14.4 SD) and in following up periods from 6-32 months (mean months=18.4 months±10.5 SD, **Table 1**).

Table 1 Characteristics of patients

Variable	PTC
No. cases	56
Age (years old)	39.8±14.4
range (years old)	19-68
Sex	
male	25
female	31
Follow up period (months)	18.4±10.5
range (months)	6-32

PTC: post-traumatic condition.

2 Assessments

All patients underwent a physical examination, a complete neurological examination, cervical MRI, and measurement of articular mobility. Additional information was provided in some patients by whole neuraxis MRI, computed tomography (CT) of the head with 2D and 3D reconstruction, CINE-MRI, standup MRI, flexion and extension x-rays of the cervical spine, CT of the spine. The clinical disability of each patient was measured using the Karnofsky performance scale (KPS) of 0 to 100[21].

1. Post-Traumatic Condition (PTC)

In this report, with the condition defined as continuous brain and neck symptoms after motor vehicle accidents or falling down as PTC, 56 patients met to the criteria. Inclusion criteria of PTC for this cohort were derived as follows:

1) loss of consciousness to 13 points or less of Glasgow Coma Scale at the accident or injury or

2) continuous neurological signs of brain stem, cerebellum and upper cervical cord since the accident or injury, which had been evaluated by neurologist or

3) continuous brain fog, dizziness and neck pain, repeated nausea, abnormal sensation of extremities, abnormalities of vision since accident or injury, which have been affecting the daily life (KPS 70 or less than 70: KPS 70 means that the patient is able to care of him/herself, but unable to carry on normal activities)[21],

4) ruling out fibromyalgia, polymyaly rheumatica, polymyosis, and other connective tissue diseases by neurologist and rheumatologist and

5) ruling out psychiatric problems (depression, generalized anxiety disorder, post-traumatic stress disorder, panic disorder, somatization disorder and malingering etc.) by psychologist. Exclusion criteria were derived for this cohort as follows:

1) evacuated intracranial mass lesion

2) previous surgery for spine, skull or neuroaxis

3) fracture or subluxation compressing to the brain or spinal cord

4) spondylotic changes or herniated disc compressing to the spinal cord except for the occipito-atlanto-axial region.

2. Morphometric analysis of the craniocervical junction and invasive cervical traction

Using reconstructed 2D-CT sagittal images and plain X-rays, osseous components of the CCJ were measured in 50 patients with PTC and 55 age and sex-matched nor-

Table 2 Clinical and imaging comparison of PTC patients with and without Chiari malformation type Ⅰ

Variable	PTC	
	PTC/CMI	PTC
Total no.	28	28
Symptoms		
nausea	20 (71)	21 (75)
dysthesia or numbness of extremities	18 (64) †	7 (33)
sleep apnea	17 (61) †	4 (14)
palpitations	17 (61) †	6 (21)
double vision	12 (43)	11 (39)
Diagnostic findings (neurological abnormalities)		
motor weakness of extremities	11 (39) †	4 (14)
sensory disturbance of extremities	12 (43) †	4 (14)
downward nystagmus	7 (25)	2 (7)
ataxia	7 (25)	2 (7)
orthostatic hypotension	10 (36)	2 (7)
postural orthostatic tachycardia syndrome	87 (70)	3 (11)
Radiologic findings		
cervical disc disease	10 (32)	11 (35)
cervical spine subluxation	4 (14)	4 (14)
scoliosis	7 (25)	7 (25)
Karnofsky performance score	61.7±7.8 †	71.8±10.7

Number in parentheses denote percentages. Mean values are expressed as±standard deviations.
CMI：Chiari malformation type Ⅰ, PTC：post-traumatic condition, †：significant differences as compared to patients without CMI ($p<0.01$).

mal controls. Morphometric measurements in the two cohorts were compared in the supine, upright (sitting), and distractive positions. We described the details about morphometric analysis in craniocervical junction and invasive cervical traction in the previous article (pp168-175).

3 Statistical analysis

Statistical analyses of clinical data were performed with SPSS for Windows (version 15.0；SPSS, Inc., Chicago, IL). Mean values are presented with their standard deviations. The incidence of associated abnormalities was analyzed using the chi-square test. Demographic differences between patients and normal controls were tested with nonparametric Mann-Whitney *U*-test. Comparisons of data with patients in the supine, sitting, and dis traction positions were tested with rANOVA. The distribution of the data was analyzed using the *F*-test. Significance was indicated by a two-tailed p value of less than 0.01.

Results

1 Clinical presentation of patients

Variations in the clinical presentation of PTC with or without CMI are given in **Table 2**. Compared to patients without CMI, those with the combined disorder experienced a greater incidence of lower brain stem symptoms including dysesthesia or numbness, sleep apnea and palpitations ($p<0.01$). Compared to patients without CMI, diagnostic distinctions in patients with CMI included an increased incidence of motor weakness of extremities and sensory disturbance of extremities ($p<0.01$). The KPS of PTC with CMI cohort (mean=61.7±7.8 SD) was lower than that of PTC without CMI cohort (mean=71.8±10.7 SD, $p<0.01$).

2 Morphometric analyses in supine, sitting, and traction positions

Morphometric measurements of the CCJ in the supine position demonstrated no significant differences in the BDI, BAI, ADI, DAI, CAA, CXA and AXA in patients with PTC and normal controls (**Table 3**). In patients with PTC, however, assumption of the upright (sitting) position resulted in a reduction of the BDI from a mean of 6.4 mm to 3.7 mm ($p<0.001$), posterior gliding of the occipital condyles with an increase of the BAI from a mean of 2.8 mm to 5.6 mm ($p<0.001$) (**Fig. 1**), anterior flexion of the atlanto-occipital joint, as demonstrated by a decrease of the CAA, CXA and AXA by a mean of 6.6° ($p<0.001$), 11.4° ($p<0.001$), and 11.0° ($p<0.001$, **Fig. 2**). These abnormalities were reducible by traction (**Fig. 1, 2**). Morphometric measurements in sitting position and traction demonstrated no significant differences in the ADI and DAI in patients with HDCT・EDS, and normal controls, compared to these in the supine position (**Table 3**).

In patients with CMI, assumption of the upright (sitting) position resulted in a reduction of the BDI from a mean of 6.5 mm to 3.0 mm ($p<0.001$, **Fig. 1**), posterior gliding of the occipital condyles with an increase of the BAI from a mean of 2.9 mm to 5.5 mm ($p<0.001$, **Fig. 1**), anterior flexion of the atlanto-occipital joint, as demonstrated by a decrease of the CAA, CXA and AXA by a mean of 6.4° ($p<0.001$), 13.1° ($p<0.001$), and 11.1° ($p<0.001$), respectively (**Fig. 2**). These abnormalities were reducible by traction (**Fig. 1, 2**). In patients without CMI, assumption of the upright (sitting) position resulted in a

Table 3　Results of morphometric measurements at the cranio-cervical junction

Variable	Normal controls	PTC Total	PTC/CMI	PTC
No. of patients	55	50	28（56）	22（44）
Basion-top of dens（BDI）（mm）				
supine	7.4±1.58	6.4±2.52	6.5±2.50	6.3±2.55
sitting	7.2±1.59	3.7±1.96 † ‡	3.0±2.04 † ‡ §	4.6±1.87 † ‡
traction	NA	7.8±2.34	7.8±2.52	7.8±2.24
Basion-anterior arch of atlas（BAI）（mm）				
supine	1.8±1.21	2.8±1.15	2.9±1.14	2.7±1.17
sitting	2.1±1.88	5.6±1.72 † ‡	5.5±1.69 † ‡	5.7±1.77 † ‡
traction	NA	2.5±1.23	2.2±1.21	2.7±1.25
Top of dens-arch of atlas（DAI）（mm）				
supine	12.3±2.17	12.4±2.72	12.6±2.57	12.1±2.87
sitting	12.5±2.00	12.7±2.52	13.2±3.12	12.0±3.12
traction	NA	11.9±2.87	11.6±2.78	12.2±2.96
Anterior arch of atlas-dens（ADI）（mm）				
supine	1.5±0.55	1.7±0.54	1.6±0.56	1.8±0.53
sitting	1.5±0.57	1.8±0.61	2.1±0.70	1.4±0.51
traction	NA	1.5±0.57	2.0±0.61	0.9±0.54
Clivus-atlas angle（CAA）（°）				
supine	37.9±7.14	41.3±6.34	41.7±6.26	40.8±6.44
sitting	36.5±7.52	34.7±6.22 †	35.3±6.21 † ‡	34.1±6.23 † ‡
traction	NA	44.1±6.87	44.2±6.91	44.0±6.82
Clivus-axial angle（CXA）（°）				
supine	147.6±6.61	145.4±6.35	143.2±6.28	148.5±6.43
sitting	147.8±6.00	134.0±6.41 † ‡	130.1±6.37 † ‡ §	139.0±6.45 † ‡
traction	NA	167.0±5.87 ‡	164.2±5.81	170.8±5.92 ‡
Atlas-axial angle（AXA）（°）				
supine	64.8±4.71	67.4±6.42	67.0±6.44	67.9±6.39
sitting	62.0±4.88	56.4±6.13 † ‡	55.9±6.21	57.0±6.03 † ‡
traction	NA	66.2±5.57	65.6±5.62	67.0±5.51

Numbers in parentheses denote percentages. Mean value expressed as±standard deviatioins.
PTC：post-traumatic condition, CMI：Chiari malformation type Ⅰ, NA：not applicable.
†：significant differences as compared to patients with normal controls（$p<0.001$）.
‡：significant differences as compared to supine position（$p<0.001$）.
§：significant differences compared to patints without CMI（$p<0.01$）.

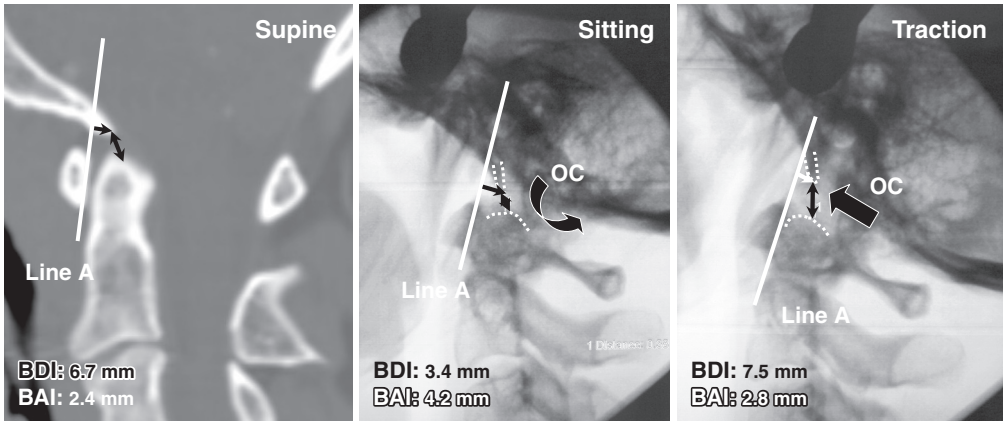

Fig. 1
Evidence of functional cranial settling and posterior gliding of the occipital condyle in a 35 year old male with PTC. Midsagittal reconstructed 2D-CT image in supine position（left）shows baseline BDI of 6.7 mm and BAI of 2.4 mm. Plain X-ray images（center and right）show a decrease of the BDI（3.4 mm）and increase of BAI（4.2 mm）in the sitting position and an increase of the BDI（7.5 mm）and decrease of BAI（2.8 mm）during traction with 10 kg weight.
Line A：plane of the posterior surface of the anterior arch of atlas, BDI：interval between basion and top of dens, BAI：interval between basion and Line A, OC：occipital condyle.

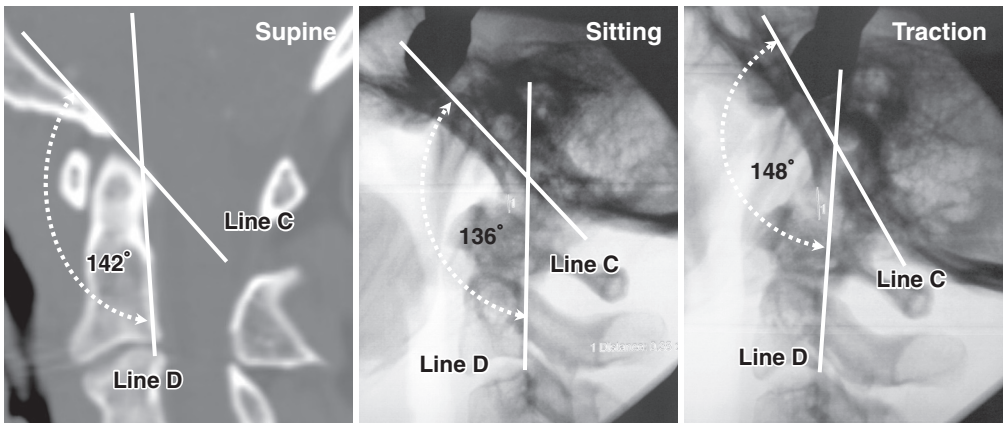

Fig. 2
Evidence of anterior flexion of occipital condyles in the same patient with PTC. Midsagittal reconstructed 2D-CT image in supine position (left) shows baseline CXA of (142°). Plain X-ray images (center and right) show a decrease of the CXA (136°) in the sitting position and an increase of the CXA (148°) during traction with 10 kg weight.
Line D : plane of the posterior surface of dens, Line C : superior plane of clivus, CXA : angle between clivus (Line C) and axis (Line D).

reduction of the BDI from a mean of 6.3 mm to 4.6 mm ($p<0.001$) (**Fig. 1**), posterior gliding of the occipital condyles with an increase of the BAI from a mean of 2.7 mm to 5.7 mm ($p<0.001$) (**Fig. 1**), anterior flexion of the atlanto-occipital joint, as demonstrated by a decrease of the CAA, CXA and AXA by a mean of 6.7° ($p<0.001$), 9.5° ($p<0.001$), and 10.9° ($p<0.001$), respectively (**Fig. 2**). These abnormalities were reducible by traction (**Fig. 1, 2**). Reductions of BDI and CXA in the upright (sitting) position are significantly larger in the CMI patients compared to those without CMI.

Discussion

1 Cranial settling, posterior gliding of occipito-atlantal joint and hypermobility of atlanto-axial joint

As shown in **Table 3**, there were no significant differences in the 5 baseline measurements of patients with PTC and normal controls in supine position. Baseline measurements of the BDI, ADI, and CXA were consistent with previously published values[4,5,9,22-24]. In patients with PTC, assumption of the upright (sitting) position resulted in hypermobility of the occipito-atlanto-axial complex and cranial settling. Reduction of the BDI in the upright position (mean 2.7 mm, $p<0.001$) established the diagnosis of functional cranial settling. Hypermobility of the atlanto-occipital joint was evidenced by posterior gliding of the occipital condyles, as demonstrated by an increase of the BAI (mean 2.8 mm, $p<0.001$) and by anterior flexion of the atlanto-occipital joint, as demonstrated by a reduction of the CAA and CXA by a mean of 6.6° ($p<0.001$) and 11.4° ($p<0.001$), respectively. Hypermobility of the atlanto-axial joint was demonstrated by anterior flexion of the atlas over the axis, reflected by the reduction of AXA (mean 11.0°, $p<0.001$). These results suggested that in patients with PTC, hypermobility of occipito-atlanto-axial joints could be confirmed as well as in patients with HDCT・EDS. In patients with PTC, occipito-atlanto-axial hypermobility might increase and prolong symptoms due to compression and injury to the brain stem and the upper cervical cord.

In patients with PTC, functional cranial settling, posterior gliding and anterior flexion is probably due to one or both of the following mechanisms :

1) laxity of ligaments supporting the atlanto-occipital joint and/or

2) generalized laxity of connective tissue structures at the CCJ including the cruciate ligament, the tectorial membrane, and supporting ligaments of the atlanto-axial joint[7,16,18-20,25].

2 Clinical features and relationship between PTC and Chiari malformation type Ⅰ

Main brain stem symptoms in this cohort were nau-

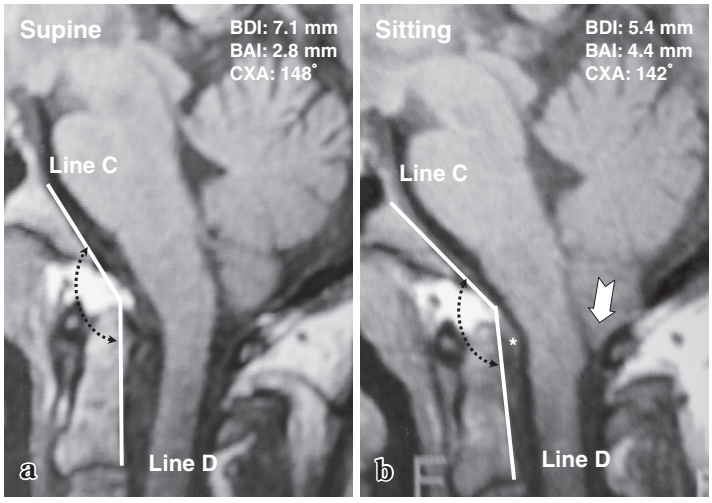

Fig. 3
Illustrative MRI images showing cranial settling and posterior gliding of the occipital condyle.
Arrow：tonsillar herniation, ＊：retroodontoid mass lesion.
BDI reduced on sitting position（7.1 mm）compared with that on supine position（5.4 mm）. BAI enlarged on sitting（4.4 mm）compared with that on supine position（2.8 mm）. CXA reduced on sitting position（142°）compared with that on supine position（148°）.
a：midsagittal T1 weighted MR image in supine position shows tight foramen magnum.
b：midsagittal T1 weighted MR image in sitting position shows compression of brain stem and tonsillar herniation induced by cranial settling, posterior gliding and anterior flexion of atlanto-occipital joint.

sea, dysesthesia or numbness of extremities, sleep apnea, palpitations and double vision. These symptoms were similar in both patients groups with HDCT・EDS and PTC. These findings, associated with CMI, had high incidence of the brain stem symptoms and suggested that occipito-atlantal hypermobility combined with CMI made brain stem symptoms and signs worse. Both occipito-atlanto-axial hypermobility and CMI in PTC affect onset and deterioration of the brain stem symptoms[1,15]. Reports describing that minor head or neck trauma precede the onset of symptoms in Chiari malformation, strongly support our data[2,26]. Functional cranial settling and anterior flexion of occipito-atlanto-axial joints were greater in patients with than without CMI. Both occipito-atlanto-axial hypermobility and CMI in PTC affect the onset and deterioration of the brain stem symptoms[1,15,27]. Although occipito-atlanto-axial hypermobility can be one of causes of CMI（tonsillar herniation）, at this point, we cannot confirm or exclude that hypermobility of occipito-atlanto-axial joints by trauma had caused CMI（tonsillar herniation）or the patients have been having CMI all along. In that context, we have some cases who had a greater descent of cerebellar tonsils in upright position（stand up MRI）compared to that in supine position and in those cases the compression to the brain stem was greater in upright position（stand up MRI）compared to that in supine position（**Fig. 3**）.

2 Retroodontoid mass lesion

A retroodontoid mass formation is a common finding in patients with rheumatoid arthritis[25], and has been reported in association with chronic atlanto-axial subluxation[17,18,20,28]. Occipito-atlanto-axial hypermobility might be one of etiologies of retroodontoid mass formation. Although the mechanism of retroodontoid mass formation has yet to be elucidated, reports of retroodontoid mass reduction or disappearance following atlanto-axial fusion provide an indirect evidence that instability of the atlanto-axial joint is a possible cause of retroodontoid mass formation[19]. Retroodontoid mass is also one of factors which cause compression to the brain stem or the upper cervical spinal cord, causing symptoms（**Fig. 3**）.

Conclusions

We have described a previously unrecognized association of occipito-atlanto-axial hypermobility in patients with PTC and with HDCT・EDS. Evidence is presented that patients with the combined disorder exhibit varying degrees of occipitoatlantoaxial hypermobility, resulting in functional cranial settling and posterior gliding of occipital condyles. Occipito-atlantal hypermobility combined with CMI made brain stem symptoms and signs of worse. In patients with CMI, functional settling and anterior flexion of occipitto-atlato-axial joints were greater than in these without CMI.

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