Comparison of Clinical Symptoms in Chiari Malformation type 1 and Chiari 1.5 Malformation related with the Position of Brainstem

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Introduction

Chiari 1.5 malformation (CM-1.5) is defined as a Chiari malformation type 1 (CM-1) with downward displacement of the obex. Radiological definition is clear but the difference of clinical symptoms between CM-1.5 and CM-1 patients is obscure. The purpose of this study is to clarify the clinical and other radiological differences in CM-1.5 and CM-1.

Methods

forty-five patients with CM-1 aged from 14 to 66 years (mean: 38.9 years) who were treated between 2000 and 2013 were retrospectively analyzed. Their clinical symptoms and the following measurements were analyzed: the distances from the basion-opisthion (BO) to tip of the cerebellar tonsil, from BO to the obex, and from basion to opisthion, the clivo-axial angle, and the length of the clivus using T1-weighted sagittal images of the cervical spine.

Figure 1

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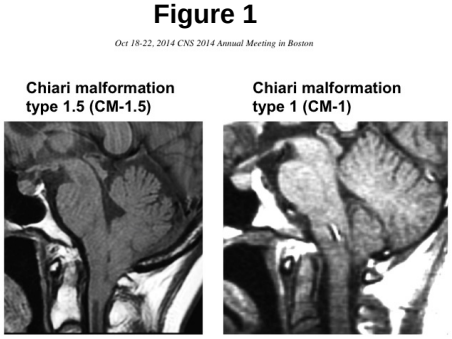

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Figure 2

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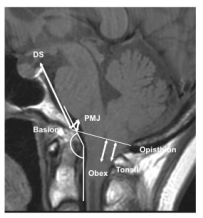

PMJ-Ba is the distance between the pontomedullary junction (PMJ) and the basion.
PMJ-BO is the distance between the PMJ and the basion-opisthion line (BO, white line).
BO-Tonsil is the distance from the basion-opisthion line to the tip of the tonsil.
BO-Obex is the distance from the basion-opisthion line to the obex.
The measured values are expressed as negative if the tip of the tonsillar is positioned below the foramen magnum.
The length of the clivus is measured from the top of the dorsum sella (DS) to the basion.

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Each measurements about the position of pontomedullary junction and cerebellar tonsil, length of clivus, clivo-axial angle

Results

There were 11 men and 34 women. In whom 22 patients (48.9%) with obex below the foramen magnum were diagnosed as Chiari 1.5 malformation (CM-1.5). Twenty-three patients had syringomyelia, 15 of them (65.2%) were in CM-1.5. In 23 CM-1 patients, headache and neck pain, motor weakness, sensory disturbance, and brainstem syndromes were 73.9%, 69.6%, 60.9%, and 34.8%, respectively. In CM-1.5, these symptoms were 86.4%, 72.7%, 63.6%, and 27.3%, respectively. The mean value of clivo-axial angle and the positions of both cerebellar tonsil and obex in CM-1.5 patients were lower than in CM-1 patients significantly. The length of the clivus and the distance between basion and opisthion were similar in both patients.

Table 1

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	CM – 1.5 (n=22)	CM – 1 (n=23)
Sex (Man:Woman)	5:17	6:17
Age	41.1 ± 14.5 (14-66)	36.9 ± 13.5 (14-64)
Syringomyelia	68.2% (15/22)	34.8% (8/23)
Headache, neck pain	86.4% (19/22)	73.9% (17/23)
Motor weakness	72.7% (16/22)	69.6% (16/23)
Sensory disturbance	63.6% (14/22)	60.9% (14/23)
Brainstem syndromes	27.3% (6/22)	34.8% (8/23)

CM, chiari malformation

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Summary of clinical symptoms

Table 2

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(mm)

	CM – 1.5 (n = 22)	CM – 1 (n = 23)	*p* value
PMJ-BO	8.37 ± 3.09	11.0 ± 2.82	< 0.05
PMJ-Basion	8.99 ± 2.76	11.1 ± 2.85	< 0.05
Tonsil-BO	− 14.14 ± 5.28	− 7.59 ± 3.49	< 0.05
Obex-BO	− 7.84 ± 4.89	− 7.04 ± 3.32	< 0.05
Clivus	41.1 ± 4.05	42.9 ± 3.30	ns
Clivo-axial angle	140.0 ± 12.6	148.9 ± 8.92	< 0.05
Basion-Opisthion	34.8 ± 2.89	34.8 ± 2.97	ns

CM, chiari malformation; PMJ, pontomedullary junction; BO, basion-opisthion; ns, no significance

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Summary of radiological measurements

Discussion

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The widespread use of MR imaging has led to CM-1 being diagnosed with increasing frequency. Despite advances in neuroimaging, many questions about pathophysiology of CM-1 remain unanswered. The downward herniation of the obex from the level of foramen magnum (basion – opisthion) with CM-1 seems to be used relatively sensitive as a method of diagnosis of CM-1.5, but not an absolute value. When treating patients with CM-1 or CM-1.5, should the treatment be changed?

Patient characteristics in both groups were similar such as sex and age. However the age in CM-1 patients was younger than in CM-1.5 without significance because some patients in CM-1 group were diagnosed incidentally with mild symptoms such as headache, vertigo. The clinical symptoms were similar in both patients, but headache and neck pain, brainstem syndromes were more frequency in CM-1.5, CM-1, respectively because the CM-1.5 patients tended to have a syringomyelia and brainstem syndromes included vertigo, dizziness, diplopia, dysarthria, dysphagia.

From our results of measurements, the distances from ponto-medullary junction to the basion and foramen magnum in CM-1.5 patients were shorter significantly than those in CM-1 patients. Considering this fact, the position of brainstem is lower in CM-1.5 patients than in CM-1, and it means there is the brainstem ptosis in CM-1.5 and CM-1 patients.

The length of the clivus and the distance between basion and opisthion were similar in both patients, but in CM-1.5 patients, the mean value of clivo-axial angle was smaller than those in CM-1 patients. Small clivo-axial angle is considered to may lead narrower posterior cranial fossa and the ptosis of brainstem and cerebellar tonsil in chiari malformation patients. The volume of the posterior cranial fossa was considered to be associated with the downward herniation of cerebellar tonsil and obex, and compression of brainstem. These may lead their clinical symptoms and syringomyelia.

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Conclusions

From our study, the positions of cerebellar tonsil and obex were significant different, however, the patients in CM-1.5 and CM-1 had similar clinical symptoms except headache and neck pain and brainstem syndromes. The precise differential diagnosis between Chiari malformation type 1 and Chiari 1.5 malformation may be controversial.

Learning Objectives

By the conclusion of this session, participants should be able to: 1) Describe the importance of both position of cerebellar tonsil and brainstem in Chiari malformation type 1., 2) Discuss, in small groups, about the relationship between clinical symptoms and radiological features. 3) Identify an effective treatment for Chiari malformation type 1.