Clinical Neurology and Neurosurgery xxx (2011) xxx–xxx

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Posterior cranial fossa morphometry in symptomatic adult Chiari I malformation patients: Comparative clinical and anatomical study

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ARTICLE INFO

Article history:
Received 26 May 2010
Received in revised form 1 November 2010
Accepted 26 December 2010
Available online xxx

Keywords:
Chiari malformation Type I
Occipital bone hypoplasia
Posterior cranial fossa
Morphometric study

ABSTRACT

Background and objectives: The cerebellar tonsillar herniation occurring in Chiari malformation Type I (CMI) mainly results from overcrowding of a normally developing hindbrain within a primary small posterior cranial fossa (PCF) due to an anomaly in the embryological development of the occipital bone. In the present study, the lengths of PCF parameters were studied in adult CMI patients.
Patients and methods: The authors retrospectively examined 15 adult patients with CMI. Multiple measurements were made on magnetic resonance images (MRIs). The results were compared with the findings in 25 controls and 30 dry skulls.
Results: Length of the neural structures did not significantly differ between the CMI and the control groups. The average length of the basiocciput was significantly shorter in the CMI group as compared with the control group. The mean length of the supraocciput was significantly shorter and the average diameter of the foramen magnum was significantly longer in the CMI group when compared to the control group and dry skulls.
Conclusions: The morphometric data suggest that, in CMI, a hypoplastic occipital bone, possibly due to the paraxial mesodermal defect of the parachordal plate, causes overcrowding in PCF, which contains the normally developed neural structures.

1. Introduction

CMI is a congenital disorder consisting of caudal herniation of the cerebellar tonsils through the foramen magnum into the cervical canal often associated with syringomyelia [1,2]. Synonyms for CMI include the adult type Chiari malformation, chronic tonsillar herniation, hidbrain herniation and usually CMI presents after the second or third decade of life [3,4]. CMI is found as an isolated phenomenon and is associated with a great variety of other conditions [3]. In 50–76% of patients, the malformation is associated with hydromyelic cavitation of the spinal cord and medulla oblongata [5–7]. This type of Chiari malformation is often related to bone anomalies in the base of skull, such as basilar invagination, and is less frequently associated with brain abnormalities other than cerebellar tonsillar herniation [4].

CMI is a result of the paraxial mesodermal defect of the parachordal plate or premature stenosis of the sphenooccipital synchondrosis [8,9]. However, the basiocciput derives from the mesodermal cells of the occipital somites and not from the neural crest [10]. Endochondral ossification of the chondrocranium forms the bones in the base of the cranium. The parachordal cartilage is formed around the cranial end of the notochord and fuses with the cartilages derived from the sclerotome regions of the occipital somites. This cartilaginous mass contributes to the base of the occipital bone; later, extensions grow around the cranial end of the spinal cord and form the boundaries of the foramen magnum [11]. Growth of the cranial base after birth continues, especially at the sphenooccipital synchondrosis, until adolescence period. This is responsible for much of the cranial lengthening, mostly at synchondroses between the sphenoid and occipital bones [12].

Clinical and experimental studies have shown that the chronic cerebellar tonsillar herniation occuring in CMI mainly results

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doi:10.1016/j.clineuro.2010.12.020

from overcrowding of a normally developing hindbrain within a primary small and shallow posterior cranial fossa due to an anomaly in the embryological development of the occipital bone [4,8,9,13–18].

The aim of the present study was to investigate appropriately the relationship of the overcrowding of the posterior cranial fossa neural structures to occipital bone in patients with symptomatic adult CMI. The evaluated parameters suggest an underdeveloped occipital bone in adult CMI which comprise the normally developed hindbrain.

2. Patients and methods

We retrospectively investigated the morphometric measurements in 15 patients (6 males and 9 females) with a symptomatic adult CMI as compared with 25 healthy individual controls (10 males and 15 females). In addition, 30 dry skulls were chosen from the archieves of the Department of Anatomy.

The diagnosis of CMI was based on the determination of a cerebellar tonsillar herniation of at least 5 mm below the foramen magnum on MRI assessment. The average age of patients with CMI was 35.8 years (22–59 years), and that of the controls was 41.7 years (18–67 years). The mean duration of the symptoms of our patients was 10 months ranging between 2 and 36 months. All patients had been treated in our department between 2007 and 2010. Six patients had CMI only (40%), however 9 patients had CMI with syringomyelia (60%). All patients presented with headache, radicular pain, neurootological signs, and cerebellar symptoms. The control group included 25 patients who underwent brain MRI evaluation for headache between 2007 and 2010. The MRI results of our control cases were reported as normal.

The structures of the brain and base of the skull were investigated using MR imaging (Signa 1.5-tesla; TR: 625 ms, TE: 10–20 ms, Matrix: 224 × 256; General Electric, Medical Systems, Milwaukee, Wisconsin). Using midline sagittal T1-weighted MRI, we measured the basiocciput part of the clivus (a: from the spheno-occipital synchondrosis to the basion), the basisphenoid part of clivus (b: from the spheno-occipital synchondrosis to the top of the dorsum sellae), clivus (c: from the basion to the top of the dorsum sellae; [a + b]), the supraocciput (d: from the internal occipital protuberance to the opisthion), the anteroposterior diameter of the foramen magnum (e: from the distance of the McRae line between the basion and opisthion), the brainstem (f: pons and medulla oblongata, between the midbrain-pons junction and the cervicomedullary junction), the long axial length of cerebellar hemisphere (g: from the highest to the lowest point of the cerebellar hemisphere) on MRI. Besides, the angle (α) between the tentorium and a line connecting the internal occipital protuberance to the opisthion was measured to estimate the steepness of the cerebellar tentorium. In addition, the degree of tonsillar herniation was measured for the CMI group (h: the distance between the tip of the cerebellar tonsils and the McRae line) (Fig. 1A and B) [4,8,11,15,19].

Three parameters were analyzed on 30 dry skulls. The measured parameters were: the length of clivus (c^1), the supraocciput (d^1), and the anteroposterior diameter of the foramen magnum (e^1), (Fig. 2A and B). In addition, these parameters were also measured on computed tomography (CT) and mentioned as c^2, d^2, and e^2 (Fig. 3A and B). The basi and sphenoocciput were not evaluated separately due to the difficulty in deciding the demarcation zone.

Statistical analysis was performed with SPSS. The values of all morphometric parameters were comparatively assessed within the CMI group, dry skull group and the control group by using the parametric student t test and nonparametric Mann–Whitney U-test. A p value < 0.05 was considered as statistically significant.

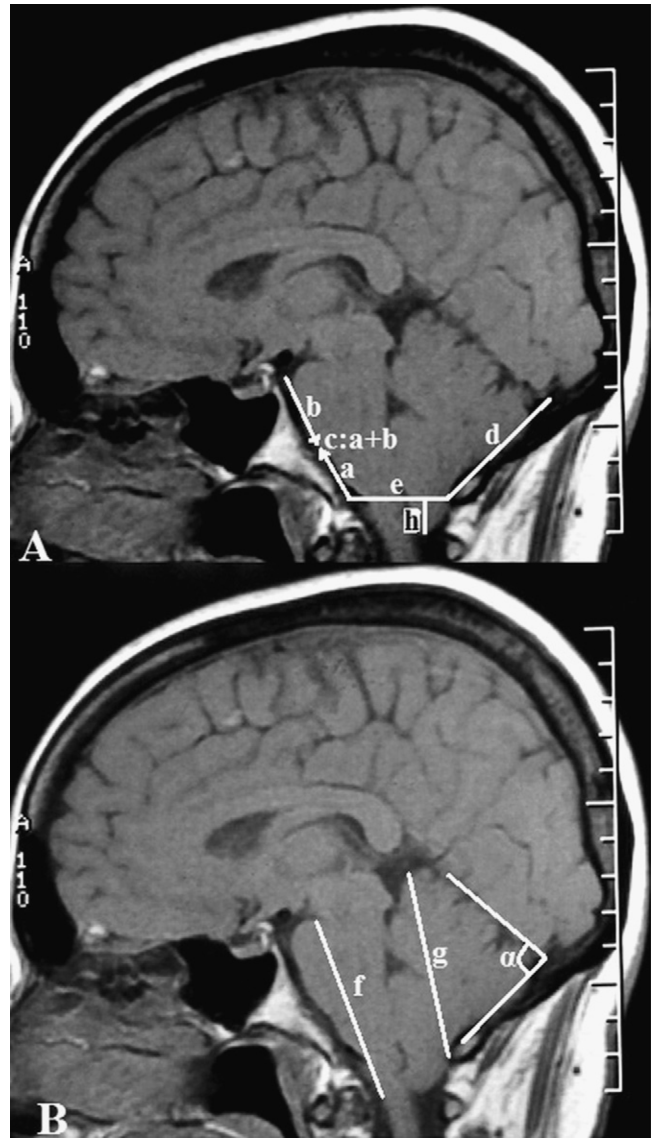

Fig. 1. T1-weighted sagittal MR image showing the midline structures of the posterior cranial fossa and the brainstem and cerebellum. (A) a: Length of the basiocciput between the sphenooccipital synchondrosis and the basion; b: length of the basisphenoid between the top of the dorsum sellae and the sphenooccipital synchondrosis of the clivus; c: length of the clivus (a + b); d: length of the supraocciput between the internal occipital protuberance and the opisthion; e: the anteroposterior diameter of the foramen magnum from the distance of the McRae line between the basion and opisthion; h: degree of tonsillar descent from the McRae line; (B) f: length of the hindbrain midbrain-pons junction and the medullocervical junction; g: length of the cerebellar hemisphere; α: angle of the cerebellar tentorium.

3. Results

Posterior cranial fossa measurements on MRI for the 15 patients and 25 controls are shown in Table 1. The average length of the basiocciput and of the supraocciput were significantly shorter in the CMI group (basiocciput: 19.1 mm, range 14.9–27.4 mm and supraocciput: 33.9 mm, range 20.2–39.9 mm) when compared to the controls (basiocciput:24.6 mm, range 19.7–27.9 mm and supraocciput: 40.8 mm, range 36.5–44.4 mm) (P: 0.001). The average length of the foramen magnum and basisphenoid were significantly longer in the CMI group (foramen magnum: 40.1 mm, range 35.8–43.6 mm and basisphenoid: 21.7 mm, range 17–26 mm) when compared to the controls (foramen magnum: 32.4 mm, range 27.7–35.5 mm and basisphenoid: 17.8 mm, range 15–21.5 mm) (P: 0.001 and P: 0.003). The average length of the clivus was shorter

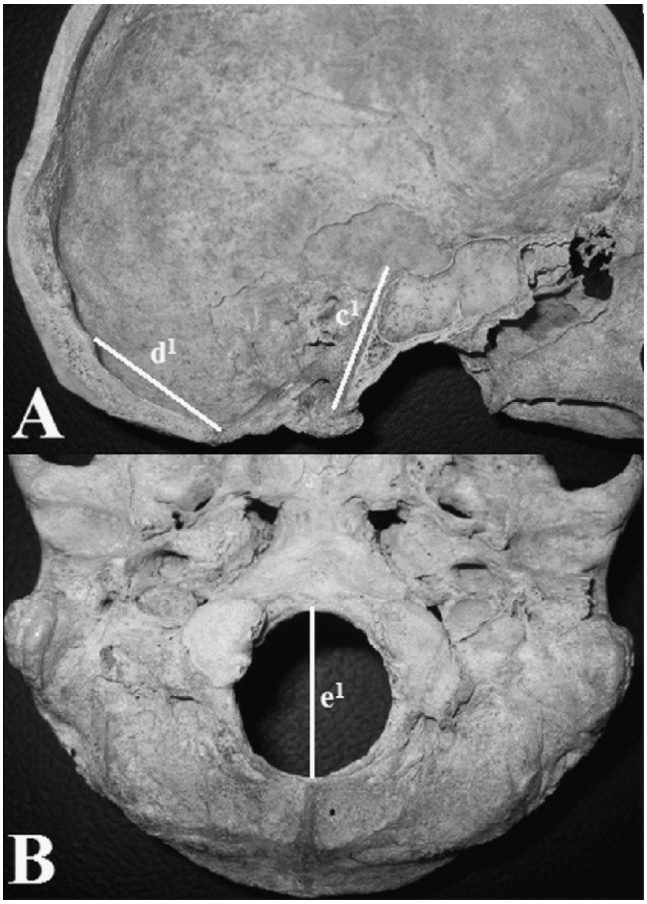

Fig. 2. Posterior cranial fossa measurements on 30 dry skulls; (A) c^1: length of the clivus, d^1: length of the supraocciput; (B) e^1: the anteroposterior diameter of the foramen magnum.

in the CMI group (40.8 mm, range 31.9–44.9 mm) compared to the control group (42.4 mm, range 35.9–46.8 mm), although the difference failed to reach statistical significance ($p = 0.279$).

Significant difference was not found in the dimensions of the measured neural structures, hindbrain and cerebellar hemisphere between the control and CMI group. The average length of tonsil herniation was found 7.4 mm in CMI group (range 5.1–12.1 mm).

Posterior cranial fossa measurements on 30 dry skulls are shown in Tables 2 and 3. The average lengths of clivus, supraocciput and foramen magnum were found as 39.7 mm (range 32.6–43.9 mm), 40.3 mm (range 36.8–45.9 mm), 34.5 mm (range 26.1–41.6 mm), respectively with direct measurement using a caliper; 39.4 mm (range 33.6–42.9 mm), 40.2 mm (range 35.1–45.4 mm), 34.3 mm (range 26–38.9 mm), respectively on CT examination. The average length of the foramen magnum was significantly longer in the CMI group when compared to the dry skulls (*P*: 0.001). The average length of the supraocciput was significantly shorter in the CMI group as compared with dry skulls (*P*: 0.001).

Table 1
Comparison of mean values for measured parameters on MRI between patients with CMI and control group.

Parameter	CMI	Control	*P*
Number of patients	15	25	
Basiocciput (a) (mm)	19.1	24.6	0.001
Basisphenoid (b) (mm)	21.7	17.8	0.003
Clivus (c: a + b) (mm)	40.8	42.4	0.279
Supraocciput (d) (mm)	33.9	40.8	0.001
Foramen magnum (e) (mm)	40.1	32.4	0.001
Hindbrain (f) (mm)	42.9	44.6	0.291
Cerebellum (g) (mm)	50.9	47.7	0.243
Tentorial angle (α) (°)	86.1	84.4	0.174

Table 2
Measured anatomical and radiological parameters on dry skulls.

Dry skulls (*n*: 30)	Anatomical	Radiological (CT)	*P*
Clivus (mm)	c^1: 39.7	c^2: 39.4	0.462
Supraocciput (mm)	d^1: 40.3	d^2: 40.2	0.850
Foramen magnum (mm)	e^1: 34.5	e^2: 34.3	0.610

4. Discussion

The evaluation of the measured parameters in this study contributes to the explanation that abnormality of the occipital bone might be the cause of CMI which may lead to various neurological symptoms including headache, neck pain, ataxia, lower cranial nerve palsies, and sensory deficit [20]. This abnormality results in cranioencephalic discordance between the underdeveloped occipital bone and normal developed hindbrain structures

Table 3
Comparison of mean values for the assessed parameters between MRI measurements of CMI patients and dry skulls (radiological-CT).

Parameter	CMI	Dry skulls (radiological-CT)	*P*
Clivus (mm)	c: 40.8	c^1: 39.4	0.045
Supraocciput (mm)	d: 33.9	d^1: 40.2	0.001
Foramen magnum (mm)	e: 40.1	e^1: 34.3	0.001

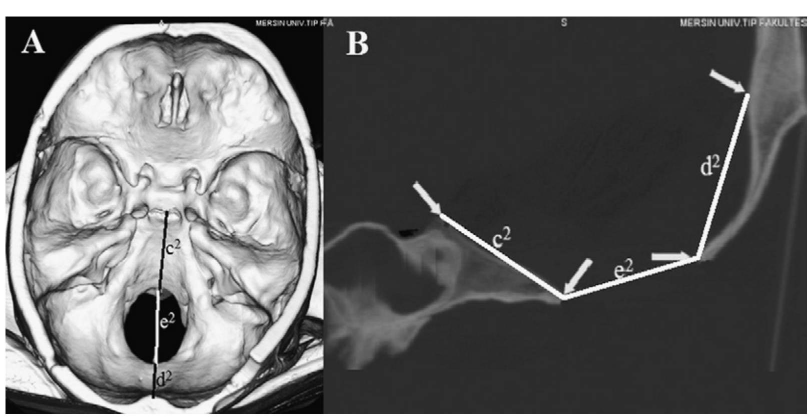

Fig. 3. Parameters were measured on CT for the 30 dry skulls. (A) Three-dimensional reconstruction CT scan c^2: length of the clivus, d^2: length of the supraocciput, e^2: the anteroposterior diameter of the foramen magnum; (B): sagittal CT scan showing showing c^2, d^2, and e^2.

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Table 4
Comparison of mean values for the assessed parameters between MRI measurements of the control group and dry skulls (radiological-CT).

Parameter	Control group	Dry skulls (radiological-CT)	*P*
Clivus (mm)	c: 42.4	c^1: 39.4	0.243
Supraocciput (mm)	d: 40.8	d^1: 40.2	0.365
Foramen magnum (mm)	e: 32.4	e^1: 34.3	0.279

[4,8,13–15,19]. Our study suggests that the posterior cranial fossa is smaller in patients with CMI than in the general population due to significant shortness of the basiocciput and supraocciput.

The authors want to point out the statistical significance regarding the shortness of the basiocciput in CMI when compared to control group. The fact that the sphenooccipital synchondrosis may not persist after the age of 13, a considerable difficulty in the determination of the exact and precise demarcation point for the junction of the occipital and the sphenoid bones should be taken into consideration.

Although basiocciput and sphenoocciput are both measured and evaluated as separate parameters in this study, the sum of the two, which gives the length of the clivus, is thought to be sufficient as a reliable parameter. In contrast, the shortness of basi and supraocciput is worth emphasizing in CMI patients.

Another dilemma is the probable difficulty for the very precise measurement of bony structures by the MRI. Although it may give acceptable results, these findings may not be as precise as the CT evaluation. Another point may be the probable difference in the establishment of the real dimensions through radiological evaluation compared to the morphometric measurement. Therefore, dry skulls were evaluated through direct anatomic, morphometric measurement by the electronic caliper and CT with the comparison of each other. Statistical significance was not found between the anatomical and radiological measurements on dry skulls (Table 2). Moreover, difference failed to reach statistical significance between the dry skull CT measurements and the MRI of the controls (Table 4). However, statistical significance was found regarding the supraocciput and the foramen magnum between the CT findings of the skulls and MRI of the CMI group (Table 3). All of the above mentioned data might encourage to use MRI assessment for the dura mater lined, cerebrospinal fluid filled spaces faced with hypointense cortical bone signal. It would be superior to compare the anatomical and MRI evaluation of the cases with the postmortem cadaveric evaluation of the same people, if possible. However, as might be easily guessed, the aim to reach such a condition is really difficult. Another point is the probable radiation induced future deleterious effects of CT application [21–24]. CT application may be thought to be the first choice in case of pathologies with bony involvement.

Recently, morphometric studies have shown clearly the significantly smaller volume, or underdeveloped bony parts of the posterior cranial fossa in adults [4,8,13–15,19,25,26]. Noudel et al. [8] have demonstrated a significant smaller basiocciput in CMI group but, no significant difference was found in the length of the supraocciput and foramen magnum. Nishikawa et al. [4] reported no significant difference in the length of the clivus, basiocciput and spheniocciput between the CMI and the control group. However, they have shown a significant smaller supraocciput in CMI group. Aydin et al. [13] have reported a significant smaller length of clivus and supraocciput in CMI group. They have demonstrated significant longer foramen magnum in CMI group. Our study has demonstrated that the average lengths of the basiocciput and supraocciput were significantly shorter in the CMI group as compared with the control group. The average lengths of the foramen magnum and basisphenoid were significantly longer in the CMI group as compared with the control group. Basiocciput/basisphenoid ratio was found to be 1.38 (24.6/17.8) in the control group. This ratio was found as 0.88 (19.1/21.7) in CMI group. These ratios indicate the importance of the underdevelopment of the occipital bone showing that the patients with CMI had various degrees of occipital bone hypoplasia. However, no significant difference was found in the dimensions of the neural structures in the posterior cranial fossa. These morphometric parameters suggest that, in CMI, a hypoplasic occipital bone, possibly due to the paraxial mesodermal defect of the parachordal plate, causes overcrowding in the posterior cranial fossa, which contains the normally developed hindbrain and cerebellum.

The average amount of tonsillar herniation was found to be 7.4 mm (range 4.9–12.1 mm) in CMI group. No significant correlation was found between the extent of tonsillar herniation and any other measured parameters of the posterior cranial fossa. Noudel et al. [8] and Nishikawa et al. [4] have found no relationship between the size of the measured posterior cranial fossa parameters and the degree of cerebellar tonsillar herniation.

A large number of clinical syndromes are associated with CMI, and the radiologic appearance characteristic of the CMI may be found as an apparently incidental finding in patients undergoing MRI examinations for other reasons [3]. Classic presentations for the adult CMI include neurological symptoms such as headache, neck pain, ataxia, lower cranial nerve palsies, and sensory deficit [4,27].

The most frequent pathology associated with CMI is syringomyelia. In 50–76% of patients, the CMI is associated with hydromyelic cavitation of the spinal cord and medulla oblongata [5–7]. The pathophysiology of syringomyelia in the context of the CMI is extremly controversial [3]. Oi et al. [28] reported that this pathology was probably analogous to hydrocephalus, except that it occured within central canal rather than within the ventricles of the brain. The cervical segmet is the most frequently involved, but the dilation may extend cranially to the brainstem or caudally to the thoracic or lumbar regions [29]. The syringomyelia is now treatable by different types of surgical intervention, such as drainage of cerebrospinal fluid or decompression of the foramen magnum with or without plugging the central canal [4]. Nine of our fifteen patients with CMI had syringomyelia (60%) which is in accordance with the related literature. All patients with syringomyelia underwent decompression surgery with or without duraplasty.

5. Conclusions

The present study revealed a smaller occipital bone in CMI patients when compared to both the controls and the randomly chosen dry skulls. In CMI patients, both the basi and supraoocciput were found to be hypoplasic. However, a similar hypoplasia was not found through measurements of basisphenoid of CMI group in comparison to the controls. This fact may emphasize the embryological impacts on the occipital bone rather than a closure pathology of the sphenooccipital synchondrosis indicating that CMI might be a result of the paraxial mesodermal defect of the parachordal plate.

The MRI examination seems to be sufficient in the measurement of the related parameters discussed in our study. The advancements in MRI technology gives an opportunity to have more precise evaluation in the cerebrospinal fluid filled dura mater and bone intersection surfaces. However, CT may be complementary if it has already been performed. Computed tomography may be chosen as the first choice of evaluation for the lesions with bony involvement and/or destruction. CT assessment should not be done just for academical or investigational basis for its probable future deleterious effects regarding its radiation exposure.

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