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Original Research Article

Morphometric Analysis of the Posterior Cranial Fossa in Chiari Type I Malformation in Adults

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ABSTRACT

Objective: The aim of this study is to measure the bony part of the posterior cranial fossa and correlate our clinical findings with the current literature.
Method: This study was conducted in the first affiliated Hospital of Xi'an Jiaotong University since January 2010 to February 2012. A total of 15 cases with adult Type 1 Chiari malformation (CMI) were included and various bony measurements were undertaken in the sagittal Magnetic Resonance Imaging (MRI). Thirty healthy controls were matched with their respective age and sex. The values of all the parameters measured for the patients and the control groups were assessed by independent sample Student's T tests.
Results: The length of the clivus, anteroposterior diameter of posterior fossa and height of posterior cranial fossa in CMI were reduced in size compared to the control groups and it was statistically significant. (p value <0.001, 0.029 and 0.03, respectively). The anteroposterior diameter of foramen magnum was more (p value <0.001) and the tentorial angle was also steeper (p value<0.001) in CMI. Seven patients with CMI had associated syringomyelia.
Conclusion: Even with limited number of patients we could illustrate that the bony components of the posterior cranial fossa is underdeveloped in CMI. This study supports the current literature stating that CMI is the result of underdeveloped occipital somites and the paraxial mesoderm.

Keywords: Chiari Malformation; Posterior fossa, Syringomyelia

INTRODUCTION

Chiari malformations or hindbrain hernias are group of rare congenital disorders described as the downward displacement of the posterior cranial fossa contents into the spinal canal. It was first described in 1891 by an Austrian Pathologist, Hans Chiari while studying the effects of hydrocephalus on the cerebellum. [1] Though it has been divided into four distinct groups, only two types are more common. Type 1 Chiari malformation (CMI) is characterized by descent of cerebellar tonsils, 5 mm or greater into the cervical canal [2] and type II represents herniation of fourth ventricle and medulla

oblongata as well as the caudal part of the cerebellar tonsil. Type II malformation is more common in pediatric population and is often associated with spinal dysraphism. [3] Adult type is more considered as the Chiari Type 1 and it usually presents after second or third decade of life. The clinical features of CMI can vary from symptoms of cerebellar, brainstem and spinal cord pathology resulting from the tightness of the posterior cranial fossa and the associated syringomyelia with it. [4-6] The definitive treatment of CMI has remained posterior fossa decompression commonly involving the posterior fossa craniectomy, laminectomy and duroplasty. [7,8]

The prevalence of CMI is estimated to be in the range of 1 per 1000 to 1 per 5000 individuals. [9] Even though most of the cases are sporadic, the familial transmission can occur by autosomal recessive inheritance or autosomal dominant inheritance with incomplete penetrance. [10,11] The pathogenesis of CMI and syringomyelia are incompletely understood. [12] However recent evidences to explain the pathogenesis is attributed to the Magnetic Resonance Imaging (MRI). These MRI findings have suggested that the overcrowding of the hindbrain due to underdevelopment of the posterior cranial fossa is the main cause of CMI. [13] Many morphometric studies focusing on the bony part of the posterior cranial fossa have supported this hypothesis. [4,5,14,15]

In the present study we have measured the bony parts of the posterior cranial fossa in adult Chiari cases. The measurements taken from the adult Chiari cases were compared with the control group who were matched with their respective age and sex.

MATERIALS AND METHODS

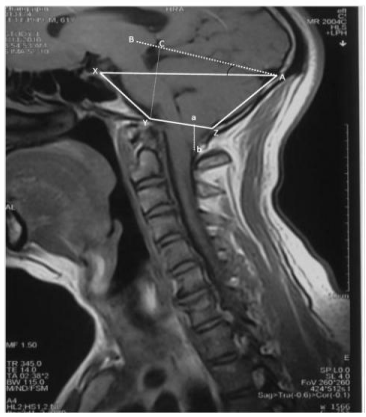
Patient population:

A prospective study was conducted in the first affiliated Hospital of Xi'an Jiaotong University since January 2010 to February 2012. All the patients who were admitted in the department with adult CMI were included in the study. These study group included patients beyond 18 years of age with symptomatic CMI with the tonsillar descent of 5 mm or more on sagittal MRI and 15 patients satisfied the criteria. Thirty adult individuals (with same age group and sex) who had undergone MRI for the complaint of headache and showed no intracranial pathology were chosen as the control group. The age of the control was same as that of the age of the cases. For every one case of 19 years female, there were two control groups of 19 years female and so on. All the cases have written informed consent for the surgical procedure. The study has been approved by institutional ethical committee.

Radiological evaluation:

All patients underwent MRI (Philips Ingenia 1.5 T; Philips Healthcare, Eindhoven, Netherlands). The following measurements were taken in the sagittal MRI from 15 cases and 30 control subjects (figure 1). The length of the clivus was measured from the top of the dorsum sella to the basion (XY).The length of the supraocciput was measured from the center of the internal occipital protuberance to the ophisthion (ZA) and the diameter of foramen magnum was measured from the basion to the opisthion (YZ). [5,13] The height of the posterior cranial fossa was measured from the basion to the peak of tentorium cerebelli (YC). [14] The anteroposterior diameter of posterior cranial fossa was measured from dorsum sella to internal occipital protuberance(XA).The angle of the cerebellar tentorium to the Twinning line(∠BAX) was measured to estimate the steepness of the cerebellum. [5]

Figure 1: Demonstration of measurements of Posterior Cranial Fossa measurement made on T1 weighted Sagittal MRI of case 7.



Cerebellar tonsils:

The degree of herniation of the cerebellar tonsil was measured below the line joining the basion and the opisthion. All the patients had tonsillar herniation more than 5 mm.

Statistical analysis:

The statistical analysis was performed by using SPSS for windows (version 13; SPSS, Inc, Chicago,Ill). Numerical variables were expressed in terms of mean±SD. The values of all the parameters measured for the patients and the control groups were assessed by independent sample Student's T tests. Significance was indicated by a two-tailed p value less than 0.05.

RESULTS

There were 9 (60%) males and 6 females in the study group with a mean age of 32.66 years (range 18 years-61 years). The average duration between the onset of the symptoms and presentation for clinical evaluation was 4.32(range one month-10 years). The commonest clinical presentations were headache and neck pain, upper or lower limb weakness, dizziness and numbness. The clinical features have been summarized in table 1.

Table 1 showing the clinical features of CMI in all the patients:

Case	Age	Sex	Chief complaint	Duration of illness	Hydrocephalus	Syringomyelia	Basilar invagination
1	19	Female	Headache and neck pain	6 years	No	No	No
2	20	Female	Right lower limb weakness	6 months	Yes	No	No
3	25	Female	Headache and neck pain	7 years	No	No	Yes
4	39	Male	Headache and neck pain	10 years	No	Yes(C5-T1)	No
5	29	Male	Headache and dizziness	6 months	Yes	Yes(C1-T1)	No
6	37	Male	Progressive dizziness, and left upper limb numbness	1 month	No	Yes(C2-T2)	No
7	61	Male	Right upper limb weakness with muscle wasting	3 years	No	Yes(C2-7)	No
8	18	Female	Intermittent headache	3 years	No	No	No
9	47	Male	Neck pain and dizziness	5 years	Yes	Yes(T1-T12)	No
10	28	Male	Headache and neck pain	5 years	No	No	No
11	50	Male	Intermittent headache	10 years	No	No	No
12	18	Female	Intermittent headache	4 years	No	No	No
13	21	Female	Right lower limb weakness	8 months	Yes	No	No
14	37	Male	Headache and neck pain	10 years	No	Yes(C3-C7)	No
15	41	Male	Left upper limb numbness	2 months	No	Yes(C2-T1)	No

Note: in patients with syringomyelia, C=cervical spine and T= Thoracic spine

The mean tonsillar descent below the foramen magnum was 10 mm (range 5.71mm- 20 mm). Seven patients (46.6%) in this study had syringomyelia. Out of 7 patients 2 had cervical syrinx, 4 had cervico-thoracic syrinx and 1 had thoracic syrinx. Interestingly none of our female patients had syringomyelia.Four patients also had presence of hydrocephalus. One of the patients with syringomyelia is shown in figure 2. Only one patient (case 3) had features of basilar invagination as shown in figure 3.

Figure 2: Demonstration of Syringomyelia in a T2 weighted Sagittal MRI of Case 4.

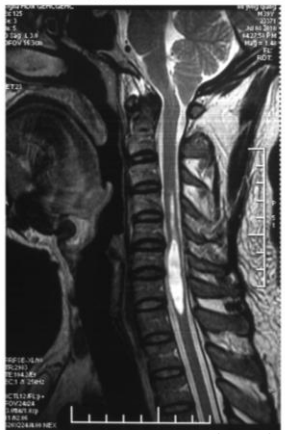


Figure 2: Syringomyelia is seen extending from fourth cervical vertebrae to first thoracic vertebrae level.

Linear measurement of posterior fossa

The mean length of the clivus in the patients was 30.14±4.74 mm, the mean length of supraocciput was 45.98±6.12 and the mean anteroposterior diameter of foramen magnum was 28.61±4.0 mm. The mean anteroposterior diameter of the posterior fossa was 65.35±5.70mm, the height of posterior fossa was 47.28±7.0 mm and the tentorial angle in patients was 19.4±1.94 degree.

Figure 3: Demonstration of basilar invagination in sagittal computed tomography in case 3.

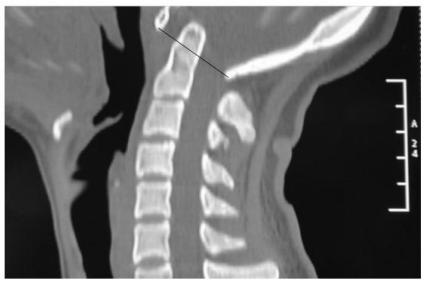


Figure 3: The black line is MC Rae's line and the significant protrusion of dens above this line confirms basilar invagination.

The clival length, the anteroposterior diameter of the posterior cranial fossa and height of posterior fossa were significantly shorter in CMI than in the control groups.(table 2)

Table 2: Statistical results of measurement made on MRI (Magnetic resonance Imaging)

Measurements(mm)	Patients(n=15)	Control(n=30)	p value	Confidence Interval
Clival length	30.14± 4.74	48.62±4.92	<0.001**	(11.87,18.19)
Supraocciput	45.98±6.12	51.21±9.28	0.082	(-0.332, 10.78)
Foramen magnum	28.61±4.0	20.05±3.59	<0.001**	(-12.41, -7.02)
A-P diameter of Posterior fossa	65.35±5.70	75.63±11.02	0.029*	(-11.41-1.21)
Height of posterior fossa	47.28±7.0	54.11±5.45	0.03*	(1.19, 10.64)
Tentorial angle(degree)	19.4±1.94	32.2±1.43	<0.001**	(11.59-14.18)

Values are Mean ±SD(standard deviation),A-P=Anteroposterior
** p value is significant.*
*** p value is highly significant.*

The tentorial angle was also significantly less in CMI.The Anteroposterior diameter of foramen magnum was more in CMI than in Control group. (p value<0.01).Even though the length of supraocciput was lower in case of CMI compared to control, it was not statistically significant. The shorter clival length, smaller tentorial angle and larger foramen magnum compared to control group were highly significant.(p value<0.001)

DISCUSSION

In this study we have estimated the overcrowding of the posterior fossa by its bony measurement in the MRI. This study also supports that the hypoplasia of the bony structures responsible for the formation of posterior cranial fossa is the main cause of the CMI in adults. Many studies have revealed that the CMI is most likely produced by the underdevelopment of the occipital enchondrium, possibly due to the

underdevelopment of occipital somites originating from the paraxial mesoderm. [5,11,16,17] This hypolasia of posterior fossa results in herniation of the normal sized hindbrain and it is actually not the anomaly of the neural structures involved. This herniation of the normal sized hindbrain results in the broadening of the anterioposterior diameter of the foramen magnum. The shorter lengths of the clivus, the supraocciput and height of posterior fossa along with broader foramen magnum in CMI have been also shown by other studies. Sabri Aydin et al have also revealed similar findings in their studies. The shorter length of supraocciput in the CMI was also not statistically significant in their studies as well.[13] However Nishikawa et al have reported that the length of the supraocciput was significantly shorter in their pediatric study group than the control group, however the length of the clivus was not statistically shorter than the control groups. [5]

The lesser angle formed by the tentorium with the twinning line also reveals that there is a steeper tentorium. This steepness is a result of upward shifting of the contents due to overcrowding of posterior fossa along with downward shifting of the hindbrain. Nishikawa et al have hypothesized that the overcrowding of the posterior cranial fossa induces the remodeling of the neural structures as the cerebellum tentorium shifts upward and the cerebellar tonsils herniated to accommodate the growing brain rather than remodeling the cranium. [5]

The herniation of the cerebellar tonsils from the foramen magnum into the spinal canal results in the obstruction of the anatomical pathway of the cerebrospinal fluid (CSF) circulation. This can result in the development of hydrocephalus. Four patients (27%) of our cases primarily presented with features of hydrocephalus. Furthermore it has been also demonstrated that the blockade of the intracranial and the intraspinal subarachnoid space is the most important cause for the development of syringomyelia. [3,18-20] Heiss et al have shown that during each systolic pulse the downwardly displaced tonsil act as a piston on partially isolated spinal CSF producing a pressure wave that acts on the surface of spinal cord. [21] This results in the progression of the syringomyelia and compression of the cord. In our study 7 patients (46.6%) had syringomyelia. In the study by Sunil et al, 71.4% of their cases had syringomyelia. [14] Mattew et al in their series of 44 patients with CMI have stated that one third of their patients had normal CSF flow, one third had decreased CSF flow in the dorsal region and remaining one third had combined ventral and dorsal CSF flow pathology. [3] They have further correlated that the presence of decreased CSF flow in both dorsal and ventral compartment predicted improved response to surgery.

Another important causal factor for syringomyelia is the basilar invagination which also causes the reduction in the posterior fossa volume. [22,23] The radiological feature of basilar invagination can be confirmed when there is any protrusion of the odontoid above the McRae's line. [24] Basilar invagination has been reported to be present in 25-50% of CMI in adults. [24-26] Only one patient(6.7%) in our study had basilar invagination. The surgical planning also differs significantly if basilar invagination is the main cause of CMI and syringomyelia requiring anterior decompression as well. [24]

Many literatures have revealed that female patients outnumber the male patients. [4,5,27] However in our studies male patients (60%) predominates the female patients. Interestingly we have found that the age of presentation for female patients were comparatively less(ranging from18-25years) compared to male patients(ranging from 37-

61years).Additionally, none of the female patients in our study had syringomyelia. We could not correlate such clinical finding on the basis of female gender but we would like to see whether such correlation really exist in larger number of patients.

CONCLUSION

Chiari malformation being a congenital rare disorder, we have limited number of patients in this study. Even with such limited number of patient we could well demonstrate the significant difference in the linear measurement of posterior fossa; further concluding the underdevelopment of the occipital somite resulting in the herniation of the normally developed hindbrain. As the patients with CMI can present with various diversity of symptoms, vigilant workup for clinical diagnosis is required in every cases. Further large scale clinical trials should be needed to understand the pathogenesis of CMI.

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