

ARAŞTIRMA / RESEARCH

Radiological determination of fossa cranii posterior morphometry in Chiari malformation type I

Chiari malformasyonu Tip-I'de fossa cranii posterior morfometrisinin radyolojik olarak değerlendirilmesi

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Abstract

Amaç:. Bu çalışmada, Chiari Malformasyonu Tip-I hastaları ve kontrol grubuna ait Manyetik Rezonans (MR) görüntülerinde fossa cranii posterior (FCP) ve cranium' a ait morfometrik ölçümler yapılarak oluşan morfolojik değişiklikleri değerlendirmek amaçlandı.

Gereç ve Yöntem: Çalışmamız retrospektif nitelikte olup çalışmaya 21 ve 47 yaş aralığında 32 kişilik kontrol grubu (16 kadın, 16 erkek) ve 12 kişilik CMI' lı hasta grubu (6 erkek, 6 kadın) dahil edildi. CMI' lı hasta ve kontrol grubuna ait T2 ağırlıklı MR görüntüleri üzerinde ölçümler yapıldı.

Bulgular: Maksimum cranial uzunluk, maksimum cranial yükseklik, foramen magnum sagittal çapı, basis cranii uzunluğu, supraocciput uzunluğu, clivus uzunluğu, fossa cranii posterior' un ön-arka çapı, occipital cord uzunluğu, cerebellum yüksekliği, herniasyon miktarı CMI' lı hastalarda sırasıyla 164.13±10.67mm, 134.60±10.71 mm, 36.89±5.28 mm, 108.5±7.9 mm, 38.5±3.7 mm, 36.7±7.1 mm, 84.9±8.8 mm, 79.7±11.5 mm, 53.3±4.2 mm, 7.8±4.9 mm olarak ölçüldü.

Sonuç: Sağlıklı bireylere ait değerlerle karşılaştırıldığında, CMI'lı hastalarda foramen magnum ön-arka çapında ve tonsiller herniasyonda artış; clivus uzunluğu, occipital cord uzunluğu ve supraocciput uzunluğunda azalma olduğu görüldü. CMI'lı hastalara ait FCP morfometrisinin bilinmesinin hastaların takibine ve cerrahisine katkı sağlayacağı düşünüldü.

Key words:. Fossa cranii posterior, tonsiller herniasyon, Chiari malformasyonu

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Öz

Purpose: The aim of this study is to evaluate the morphological changes in Magnetic Resonance Images (MRI) of Chiari Malformation Type-1 (CMI) patients by performing morphometric measurements of fossa cranii posterior (FCP) and cranium.

Materials and Methods: Our study is retrospective and included a control group of 32 (16 females, 16 males) and 12 patients (6 males, 6 females) with CMI between the ages of 21 and 47 years. Measurements were made on T2-weighted cranial MR images of the patient with CMI and the control group.

Results: Maximum cranial length, maximum cranial height, sagittal diameter of foramen magnum, cranium base length, supraocciput, clivus length, anterior-posterior diameter of fossa cranii posterior, occipital cord length, cerebellum height, amount of herniation in CMI were found 164.13±10.67mm, 134.60±10.71mm, 36.89±5.28mm, 108.5±7.9mm, 38.5±3.7 mm, 36.7±7.1mm, 84.9±8.8mm, 79.7±11.5mm, 53.3±4.2mm, 7.8±4.9mm respectively.

Conclusion: In our study, the measurements of the CMI and the control group were compared and a decrease in the length of the supraocciput, the length of the clivus and the length of the occipital cord has been found and an increase in the length of the cranial base and the sagittal diameter of the foramen magnum has been found in CMI patients. As a result of the data obtained in our study, FCP morphometry of patients with CMI can contribute to patient follow-up and surgery.

Anahtar kelimeler: Fossa cranii posterior, tonsillary herniation, Chiari malformation

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INTRODUCTION

The fossa cranii posterior (FCP) is the largest of the three cranial fossa and lodges the cerebellum, pons and medulla oblongata¹. Many developmental disorders, including Chiari Malformations, affect the diameters and volume of the FCP2-5. Chiari Malformations describes an increased degree of herniation of the structures of the hindbrain from the foramen magnum⁶. Moreover, Chiari Malformation Type-1 is defined as a herniation of the cerebellar tonsil(s) into the spinal canal > 3-5 mm beyond the basion-opisthion line (McRae's line)7-9. Although the etiology of CMI is not fully known, it is thought to be multifactorial¹⁰⁻¹¹. Furthermore, spinal and head base abnormalities of bone structure such as scoliosis, kyphosis, cervical canal enlargement, atlantoaxial assimilation, C1 incomplet ossification, Klippel-Feil Syndrome are often seen in patients with CMI and structural brain abnormalities are not observed in patients with CMI7,12,13,14. Syringomyelia is frequently found in patients with CMI.

Moreover, MRI is the best imaging method for diagnosing CMI4. Clinical signs and symptoms in patients with CMI are related to structures that are suppressed and develop loss of function with caudal displacement of the cerebellar tonsils through the foramen magnum. According to Milhorat et al, they were classified these clinical manifestations in five main groups: Suboccipital headache, ocular disorders, neurological disorders, clinical manifestations related to the brainsytem-cerebellum-lower cranial nerves and spinal cord disorders. The most common symptom is headache that spreads especially to the occipital and upper cervical region and is triggered or exacerbated by the valsalva maneuver⁹. Furthermore, in symptomatic CMI cases, each patient is treated according to the current neurological findings and the presence of additional pathologies. Surgical methods such as craniovertebral junction decompression, duroplasty, ventriculoperitoneal shunt can be used for treatment^{15,16}.

The aim of the present study was to evaluate the morphological changes in FCP using MR images of CMI patients by performing morphometric measurements of FCP and cranium.

MATERIALS AND METHODS

Study group

The study was performed on cranial MRI in the

Radiology Department of Adana Medline Hospital. Patients (12 patients) with tonsillar herniation greater than 3 mm between the ages of 21-47 years between January 2020 and April 2021, were included in the group with CMI.

Cranial MR images of 32 individuals (16 male, 16 famale) aged 21-47 years who did not undergo intracranial surgery, who did not have any disease affecting the bones, and who did not have a history of skull fracture or intracranial tumor mass were randomly selected in the control group. This retrospective study is approved by the Cukurova University Faculty of Medicine Non-Interventional Clinical Research Ethics Committee (Date: 10.09.2021, Decision No: 41; Number of Meetings: 114). The measurements were made by an expert radiologist.

MR Protocol

Brain MRI protocol including sagittal T2-weighted spin echo (TR:3600, TE: 87 ms; slice thickness 5 mm; gap 1.5 mm) was used. The measurements were performed from digital MR images using caliper function with x2 magnification.



Figure 1. Demonstration of morphometric measurements performed on the midsagittal line on T2-weighted Cranial MRI.

L1: Maximum cranial length, L2: Maximum cranial height, L4: Cranium base length, L8: Occipital cord length.

Morphometric measurement parameters

Using the midsagittal T2-weighted spin echo image, the following parameters of posterior cranial fossa

Cilt/Volume 47 Yıl/Year 2022

dimensions were evaluated ^{4,6,7,9,17,19} (Figure 1, Figure 2).

 Maximum cranial length (L1): The distance between the glabella and the opisthocranion.



Figure 2. Demonstration of morphometric measurements performed on the midsagittal line on T2-weighted Cranial MRI

L3: Sagittal diameter of Foramen magnum, L5: Supraocciput length, L6: Clivus length, L7: FCP anteroposterior length, CH: Cerebellum Height, TH: Tonsiller Herniation

- Maximum cranial height (L2): The distance between the basion and the vertex
- Foramen magnum sagittal diameter (L3): The distance between the basion and the opisthion
- Cranium base length (L4): The distance between the nasion and the basion.
- Supraocciput length (L5): The distance between the opisthion and the protuberentia occipitalis interna
- Clivus length (L6): The distance between the basion and the dorsum sellae top edge
- FCP anteroposterior length (L7): The distance between the dorsum sellae and the protuberentia occipitalis interna
- Occipital cord length (L8): The distance between the opisthion and the lambda
- Cerebellar height (CH): The distance between the lowest and highest points of the cerebellum.

 Tonsillar herniation (TH): The distance between McRae line and the lowest point of tonsilla cerebelli.

Statistical analysis

Statistical Package for the Social Sciences (SPSS 21.0) was used to analyze the measurement results. The independent sample t-test was used to compare the measurement parameters L1, L2, L4, L5, L6, L7, L8, CH and The Mann-Whitney U test was used to compare the L3 and TH parameters. The mean \pm standard deviation and P values of the measurements were recorded. The statistical significance level was accepted as 0.05 for all tests.

RESULTS

In our study, the average age of patients with CMI was found to be 30.6, and the average age of individuals in the control group was found to be 36.4.

In the morphometric measurements, mean foramen magnum sagittal diameter (CMI: 36.89 ± 5.28 mm, control: 32.41 ± 3.61 mm) and cranium base length (CMI: 108.5 ± 7.9 mm, control: 99.8 ± 6.6 mm) were found to be statistically significantly higher in CMI group, than the measurements of the control group (Table 1).

However, maximum cranial length (CMI: 164.13 ± 10.67 mm, control: 180.19 ± 8.51 mm), supraocciput length (CMI: 38.5 ± 3.7 mm, control: 50.5 ± 3.9 mm), clivus length (CMI: 36.7 ± 7.1 mm, control): 44.1 ± 3.7 mm) and occipital cord length (CMI: 79.7 ± 11.5 mm, control: 94.9 ± 5.7 mm) were found to be statistically significantly lower in patients with CMI than in the control group (Table 1).

There were no statistically significant difference between the patients with CMI and the control group in terms of maximum cranial height (CMI: 134.60 \pm 10.71 mm, control: 139.75 \pm 5.09 mm), anteriorposterior diameter of fossa cranii posterior (CMI: 84.9 \pm 8.8 mm, control: 83.9 \pm 4.7 mm) and cerebellum height (CMI: 53.3 \pm 4.2 mm, control: 55.4 \pm 4.2 mm) (Table1). The mean tonsillar herniation was 7.8 \pm 4.9 mm in the group with CMI and 0.3 \pm 1.8 mm in the control group (Table 1). Morphometric measurement results of FCP and cranium in T2-weighted MR images of patients with Chiari Type-1 malformation and control group are shown in the table1.

Table 1. Comparison of the mean morphometric measurements of the MR images of the Patient and Control Groups

| | CMI (n:12) | Control (n:32) | |
|----------------------------------|-------------|----------------|----------|
| Morphometric measurements (mm) | (mean±sd) | (mean±sd) | P-value |
| Maximum cranial length | 164.1±10.7 | 180.2±8.5 | < 0.001* |
| Maximum cranial height | 134.6 ±10.7 | 139.8±5.1 | 0.134 |
| Foramen magnum sagittal diameter | 36.9±5.3 | 32.4±3.6 | 0.008* |
| Cranium base length | 108.5±7.9 | 99.8±6.6 | 0.001* |
| Supraocciput length | 38.5±3.7 | 50.5±3.9 | < 0.001* |
| Clivus length | 36.7±7.1 | 44.1±3.7 | 0.004* |
| FCP anteroposterior length | 84.9±8.8 | 83.9±4.7 | 0.725 |
| Occipital cord length | 79.7±11.5 | 94.9±5.7 | 0.001* |
| Cerebellar height | 53.3±4.2 | 55.4±4.2 | 0.154 |
| Tonsiller herniation | 7.8±4.9 | 0.3±1.8 | < 0.001* |

CMI: Chiari Malformation Type-1, FCP:Fossa cranii posterior SD:Standart deviation, (*) Statistically significant

DISCUSSION

There are many studies supporting that insufficient development of the paraxial mesoderm is responsible for the pathogenesis of CMI7,9,17. Inadequate development of the paraxial mesoderm causes hypoplasia of the occipital bone and small FCP18. Thus, an abnormality in any of the bone structures that make up the FCP can cause widening of the FM diameter^{5,19,20}. Therefore, Chiari Malformations describe an increased degree of herniation of the hindbrain structures from the foramen magnum. In this investigation, FM anteroposterior diameter was found to be higher in the group with CMI (36.9 ± 5.3) mm) than in the control group $(32.4\pm3.6 \text{ mm})$. Similarly, some researchers also found the foramen magnum AP diameter to be statistically significantly higher in the CMI group than in the control group^{19,2,6}. However, Sekula et al. (CMI: 43.55±55.9mm, control: 42.52±5.9mm), Noudel et al. (CMI: 37.1 mm, control: 35.4mm) and Urbizu et al. (CMI: 43.5mm, control: 42.5 mm) did not find a statistically significant difference between the measurements of the CMI and control group7,21,4

In a prospective study conducted by Milhorat and colleagues, the mean supraocciput length was measured as 37.7 ± 5.9 mm in the group with CMI and 41.8 ± 5.2 mm in the control group that found statistically significant⁹. On the other hand, Nishikawa et al. did not estimate a statistically significant difference between the group with CMI and the control group in terms of supraocciput length in Japanese people¹⁷. Additionally, this value was

measured as 34.29 mm in CMI patients and 40.18 mm in the control group in Koreans but the difference was also not found statistically different²². Conversely, in our morphometric study this value was found to be lower in the group with CMI $(38.5\pm3.7 \text{ mm})$ than in the control group $(50.5\pm3.9 \text{ mm})$. We think that these diversities may be due to age, race and individual differences.

Moreover, it was concluded that clivus length was the most important variable of PCF measurements in radiologically distinguishing between CMI and control groups²³. Data supporting this result were obtained from our study and from many studies conducted in different populations^{2,4,6,7,9,19}. In our study, the clivus length was found to be lower in the group with CMI (36.7 ± 7.1 mm) compared to the control group (44.1 ± 3.7 mm).

Mikulis et al separated the patients according to age groups and they found that there was a decrease in tonsillar herniation in the advanced age group²⁴. In our study, the amount of TH was found to be 7.8 ± 4.9 mm in patients with CMI. Since our patient group was small in number, the correlation between age and TH could not be examined by grouping according to age.

The cerebellar hemisphere height was found to be statistically significantly higher in both Taştemur et al.⁶ (CMI: 59.13 mm, Control: 53.32 mm) and Hwang et al.²² (CMI: 61.51 mm Control: 53.89 mm) CMI groups. However, in our investigation, there is no statistically significant difference was found between the CMI (53.3 ± 4.2 mm) and the control group

(55.4±4.2 mm) in terms of cerebellar hemisphere height. Additionally, in this paper, maximum cranial length in patients with CMI (164.1±10.7 mm) compared to the control group (180.2±8.5 mm) was statistically lower in the group CMI. In our study, unlike the study conducted by Taştemur and his colleagues the length of the basis cranii was found to be statistically significantly longer in the group with CMI (108.5 ± 7.9 mm) than in the control group (99.8±6.6 mm)⁶.

There are many studies showing that insufficient development of the paraxial mesoderm is responsible for the pathogenesis of CMI, and in these studies, the occipital bone was found to be hypoplasic and the clivus shorter than normal^{7,9,17,12}. The results of our study support this hypothesis for the pathogenesis of the CMI. The limitation of our study is that we have a small number of cases due to the fact that CMI is a rare malformation.

In our study, when the measurement values of the CMI and control groups were compared, a decrease in supraocciput length, clivus length and occipital cord length, an increase in the length of the basis cranii, and diameter of the foramen magnum were observed in the CMI group. In conclusion, we think that this study on FCP morphometry of patients with CMI will contribute to the follow-up and surgery of patients.

Yazar Katkıları: Çalışma konsepti/Tasarımı: DV, MÖ; Veri toplama: MÖ, DV; Veri analizi ve yorumlama: DV, MÖ; Yazı taslağı: DV, MÖ; İçeriğin eleştirel incelenmesi: DV, MÖ; Son onay ve sorumluluk: DV, MÖ; Teknik ve malzeme desteği: DV, MÖ; Süpervizyon: DV, MÖ; Fon sağlama (mevcut ise): yok.

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Vurallı and Öksüzler

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