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Impact of morphological measurements on symptoms in Chiari malformation type 1

Chiari tip 1 malformasyonunda morfolojik ölçümlerin semptomlar üzerine etkisi

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Abstract

Aim: Chiari malformation Type 1 (CM1) is a pathology resulting from herniation of cerebellar tonsils or tonsils into the spinal canal. We aimed to examine the impact of the cranium's morphological measurements on the symptoms with CM1 patients.

Methods: This research was designed as a retrospective case-control study in a single-center. 2309 patients aged between 18-70 who underwent brain magnetic resonance imaging (MRI) to confirm or exclude the diagnosis of CM1 as a result of clinical and examination findings were evaluated. Cranium's morphological measurements, the amount of herniation, patient's symptoms, and the modified Asgari score were retrospectively assessed.

Results: Patients with a final diagnosis of CM1 after the MRI evaluation were classified as study group (n=212), and the others control group (n=2097). The maximum cranial length, maximum cranial height, supra occiput length, posterior cranial fossa (PCF) anteroposterior length, in the study group were shorter, whereas the sagittal diameter of the foramen magnum and the longest anteroposterior diameter of the cerebrum were longer (P<0.001 for all mentioned comparisons). Tentorium cerebelli slope was found to be significantly lower in the study group (P<0.001). The most prevalent symptoms were a headache (92%). The herniation amount had a negative correlation with maximum cranial length and maximum cranial height (r=-0.184, P=0.07; r=-0.158 and P=0.022, respectively) and a positive correlation with the modified Asgari score (r=0.598; P<0.001).

Conclusion: The cranium's morphological measurements have an impact on the symptoms of patients with CM1.

Keywords: Chiari I malformation, Magnetic resonance imaging, Posterior cranial fossa, Tonsillar herniation

Öz

Amaç: Chiari Tip 1 malformasyonu (CM1) serebellar tonsil veya tonsillerin spinal kanala herniasyonu sonucu ortaya çıkan bir patolojidir. CM1 hastalarında kraniyumun morfolojik ölçümlerinin semptomlar üzerindeki etkisini incelemeyi amaçladık.

Yöntemler: Çalışmamız tek merkezde retrospektif vaka-kontrol çalışması olarak tasarlandı. Klinik ve muayene bulguları sonucunda CM1 olduğu düşünülen tanıyı kesinleştirmek veya dışlamak için beyin manyetik rezonans görüntüleme (MRG) yapılan, yaşları 18-70 arasında 2309 hasta değerlendirildi. Kranium, morfolojik ölçümleri, herniasyon miktarı, hastaların semptomları ve modifiye Asgari skoru retrospektif olarak incelendi.

Bulgular: MRG değerlendirilmesinden sonra kesin CM1 tanısı alanlar çalışma grubu (n=212) ve diğerleri kontrol grubu (n=2097) olarak hastalar sınıflandırıldı. Çalışma grubunda maksimum kranial uzunluk, maksimum kranial yükseklik, supraocciput uzunluğu, posterior kranial fossa (PCF) anteroposterior uzunluğu kısa iken, foramen magnumun sagital çapı ve serebrum'un en uzun ön arka çapı uzun idi. (hepsi için P<0,001). Çalışma grubunda tentorium serebelli eğimi belirgin olarak düşük saptandı (P<0,001). En sık görülen semptom baş ağrısıydı (%92). Herniasyon miktarı maksimum kranial uzunluk ve maksimum kranial yükseklik ile negatif korelasyon (sırası ile r=-0,184, P=0,07; r=-0,158, P=0,022) ve modifiye Asgari skoru ile pozitif korelasyon göstermekte idi (r=0,598; P<0,001).

Sonuç: Kraniyumun morfolojik ölçümleri CM1 hastalarının semptomlarını üzerinde etkilidir.

Anahtar kelimeler: Chiari I malformasyonu, Manyetik rezonans görüntüleme, Posterior kranial fossa, Tonsiller herniasyon

Introduction

Chiari malformation Type 1 (CM1) is a clinical condition that results from 5-mm herniation of one or 3–5 mm herniation of two cerebellar tonsils into the spinal canal [1-4]. Neurologic symptoms and findings may differ depending on herniation level and disruption of cerebrospinal fluid (CSF) circulation [5-7]. The etiology of the disease is unknown, although genetic factors, trauma, factors secondary to an infection, and deformations during the organogenesis period are suspected [5,8]. This pathology may occur alone or with other pathologies such as syringomyelia, scoliosis, odontoid retroflexion, basilar invagination, occipitalization of the atlas, and caudal migration of the brain stem. Symptoms may differ in the presence of these additional pathologies [5]. While the prevalence of the disease remains unclear, it has been reported to be 0.5% –0.9% [1,6,9,10]. Moreover, asymptomatic course of the disease in certain cases and mild symptoms observed in others hamper the clear determination of its prevalence [5].

CM1 is diagnosed with magnetic resonance imaging (MRI) following initial suspicion noted in the clinic. MRI evaluates posterior cranial fossa (PCF) along with the whole brain tissue and CSF flow, [1-4]. Neurologic symptoms and findings may differ depending on herniation level and disruption of CSF circulation [5-7]. The etiology of the disease is unknown, although genetic factors, trauma, factors secondary to an infection, and deformations during the organogenesis period are suspected [5,8]. This pathology may occur alone or with other pathologies such as syringomyelia, scoliosis, odontoid retroflexion, basilar invagination, occipitalization of the atlas, and caudal migration of the brain stem. Symptoms may differ in the presence of these additional pathologies [5]. While the prevalence of the disease remains unclear, it has been reported to be 0.5%–0.9% [1,6,9,10]. Moreover, asymptomatic course of the disease in certain cases and mild symptoms observed in others hamper the clear determination of its prevalence [5].

CM1 is diagnosed with MRI following initial suspicion noted in the clinic. MRI evaluates PCF along with the whole brain tissue and CSF flow, craniometric measurements. It enables the evaluation of angles, lines of intracranial structures, osteo-neural topographic images, cisterns, PCF pentagon, and localization of cerebellar tonsils [6].

Conservative treatment is recommended in mildly symptomatic or asymptomatic cases with less than 5 mm of tonsil ectopia, whereas surgical treatment is recommended in symptomatic cases [6].

The aim of the study was to analyze the impact of the cranium's morphological measurements in patients with CM1 on symptoms.

Materials and methods

Patient population and study design

Between January 1, 2015 and October 1, 2018, 2309 patients aged between 18-70 admitted to the neurosurgery clinic with various symptoms and referred to the radiology department for brain MRI with suspicion of CM1 after clinical and examination findings were evaluated retrospectively.

The local ethics committee of University of Health Sciences, Dışkapı Yıldırım Beyazıt Training and Research Hospital approved the study that was prepared according to ethical standards of 1975 Helsinki Declaration's Human Experiment Committee which was revised in 2000.

Exclusion criteria from the patients for the study were; having had a diagnosis of CMI, had a history of trauma, had brain surgery before, having increased intracranial pressure for some reasons, had a history of congenital craniocervical junction malformations and having pathological findings on the cranium structure.

Patients with a final diagnosis of CM1 after the MRI evaluation were classified as the study group (n=212), whereas the other patients were classified as the control group (n=2097). Age, sex, cranium, cerebrum and cerebellum morphological measurements, amount of herniation, the study group symptoms, and the modified Asgari score were evaluated.

MRI protocol and measurements

MRI was performed using two 1.5T MRI imaging scanners (Magnetom, Aera, Siemens, Erlangen, Germany) and Philips Achieva (Philips Medical Systems, Eindhoven, The Netherlands) with a standard head coil. The imaging protocol constituted the following five routine sequences: axial T1-weighted [repetition time (TR)/echo time (TE): 348/8.9 ms, voxel size: 0.7x0.7x0.5 mm, field of view (FOV): 23x23 cm, slice thickness: 5 mm]; axial T2-weighted (TR/TE: 4160/102 ms, voxel size: 0.6x0.6x5.0 mm, FOV: 23x23 cm, slice thickness:5 mm); axial FLAIR (TR/TE: 8000/86 ms, voxel size: 0.7x0.7x5.0 mm, FOV:23x23 cm, slice thickness: 5 mm); coronal T2-weighted (TR/TE: 4730/94 ms, voxel size:0.6x0.6x5.0 mm, FOV: 22x22 cm, slice thickness: 5 mm), and sagittal FLAIR (TR/TE: 9000/87 ms, voxel size: 0.4x0.4x5.0 mm, FOV: 23x23 cm, slice thickness: 5 mm).

The study group and the control group measurements were evaluated on MRI via Extreme Picture Archiving and Communications System (PACS) system (Ankara, Turkey).

Patients whose cerebellar tonsil(s) into the spinal canal 3 mm beyond the basion-opisthion line was accepted as the study group.

We used Houston et al. criteria to determine the mid-sagittal plane in the MRI sections, at least three of the following four structures should be seen in a single sagittal view; the genu of the corpus callosum, the splenium of the corpus callosum, the pituitary infundibulum and the cerebral aqueduct. Measurements were taken on the mid-sagittal FLAIR images. The standard measurement methods stated in the literature were used [11-14]. Length measurements in millimeters and angle measurement were made in degrees.

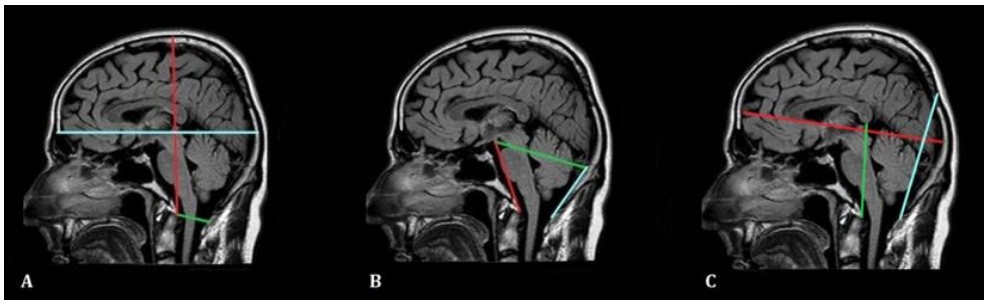

Figure 1: Sagittal FLAIR (A, B, C), an image demonstrating the distance measurements. (A) Maximum cranial length is shown by a blue line, maximum cranial height is depicted by a red line, foramen magnum sagittal diameter is shown by a green line. (B) Supra occiput length shown by a blue line, clivus length is depicted by a red line, posterior cranial fossa anteroposterior length is shown by a green line. (C) The occipital cord length is shown by a blue line, longest anteroposterior diameter of the cerebrum depicted by a red line, posterior height of the cranial fossa is shown by a green line.

Maximum cranial length (C1): is the line between glabella-opisthocranion, Maximum cranial height (C2): is the line between basion-vertex, Foramen magnum sagittal diameter (C3): is the line between basion-opisthion (Figure 1A), Supra occiput length (C4): is the line between opisthion-protuberentia occipitalis interna, Clivus length (C5): is the line between basion-dorsum sellae top edge, PCF anteroposterior length (C6): is the line between dorsum sellae-protuberentia occipitalis interna (Figure 1B), Occipital cord length (C7): is the line between opisthion-lambda, Longest anteroposterior diameter of the cerebrum(C8): is the line between polus frontalis-polus occipitalis, Posterior height of the cranial fossa (C9): is the perpendicular line between inferior of the corpus callosum splenium to foramen magnum plane (Figure 1C).

Tentorium cerebelli slope (A): is the angle between the cerebellum tentorium and the supra occiput length (Figure 2).

Tonsillar herniation (TH): is the distance between the tip of the cerebellar tonsils and opisthion - basion line (Figure 3) [14].

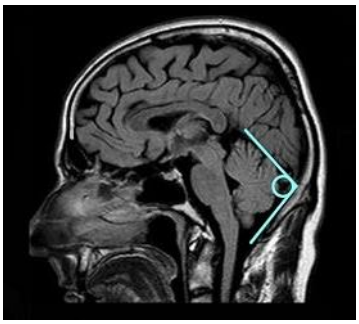

Figure 2: Sagittal FLAIR image demonstrating the tentorium cerebelli slope is shown as the angle between the blue lines

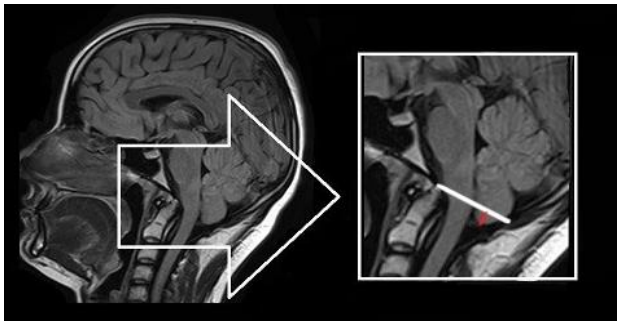

Figure 3: Sagittal FLAIR image demonstrating basion-opisthion line in white and tonsillar herniation measurement as a red line

All measurements were performed by the same observer. To test the intra-observer reliability, the same radiologist repeated all measurements three months after the first assessment. To test the inter-observer reliability, another radiologist blinded performed quantitative and semi-quantitative measurements of randomly selected patients and control groups 1000 brains among 2309 brains.

Symptoms of the study group were evaluated. The modified Asgari score, which is determined prior to surgeries, was used to evaluate the severity of the lesions. The relationship among the modified Asgari score, measurements, and age were evaluated.

Statistical analysis

The SPSS package (Statistical Package for the Social Sciences for Windows, Version 22.0, SPSS Inc., Chicago, IL, U.S.A.) was used for data analysis in our study. Kolmogorov–Smirnov test was used for evaluating the distribution of variables, whereas Pearson's Chi-Square test was used for the analysis of qualitative data. Spearman's correlation test was used in the analysis of quantitative data, whereas the student's t-test was used in the comparison of qualitative data and quantitative data. $P<0.05$ was accepted as the level of statistical significance. The intra- and interobserver reliability of MRI examinations were tested using Kappa statistics.

Results

CM1 was detected in 212 (9.2%) out of 2309 patients included in the study. The mean age of the patients was 39.3 (13) years and there was no difference between the study and the control groups in terms of age ($P=0.543$). Of the patients, 53.3% were male, and the number of males in the study group was significantly higher ($P=0.007$). While the C1, C2, C4, C5, C6, and C7 lengths were significantly shorter in the study group, it was found that the C3 length was significantly longer ($p < 0.001$ for all mentioned comparisons). C8 and C9 lengths were significantly shorter in the study group ($P<0.001$). The slope of tentorium cerebelli was found to be significantly smaller in the study group ($P<0.001$). Herniation length of the study group was determined as 8 (1.6) mm, and the modified Asgari score was determined as 4.8 (1.9) (Table 1).

The most prevalent symptoms were a headache (92%) and neck ache (35.4%) (Figure 4).

In the study group, it was observed that age had a positive correlation with C1, C3, C6 (r=0.515, $P<0.001$; r=0.467, $P<0.001$; r=0.572, $P<0.001$, respectively) and a negative correlation with C4, C8, C9, and tentorium cerebelli slope (r=-0.390, $P<0.001$; r=-0.212, $P=0.002$; r=-0.446, $P<0.001$, respectively) (Table 2). The herniation amount had a negative correlation with C1 and C2 (r=-0.184, $P=0.07$; r=-0.158, $P=0.022$, respectively) and a positive correlation with the modified Asgari score (r=0.598, $P<0.001$) (Table 2).

Intra and interobserver intraclass correlation coefficients for all quantitative and semi-quantitative measurements were 0.81 and 0.87, respectively.

Table 1: Comparison of cranium, cerebrum and cerebellum morphological measurements of study group and control groups

		Study group (n=212)	Control group (n=2097)	*P*-value
Age mean (SD)		39.3 (13)	38.7 (12)	0.543*
Sex. n (%)	Male	113 (53.3)	915 (43.6)	0.007**
	Female	99 (46.7)	1182 (56.4)	
		mean (SD)	mean (SD)	
C1		171.4 (2.7)	174.3 (1.7)	<0.001*
C2		129.6 (1.9)	134.5 (1.6)	<0.001*
C3		35.6 (2.3)	33.9 (2.3)	<0.001*
C4		42.6 (2.3)	44.9 (17.7)	<0.001*
C5		39.2 (1.9)	42.3 (2.1)	<0.001*
C6		76.8 (2.5)	77.8 (3.6)	<0.001*
C7		90.7 (1.9)	92.6 (4)	<0.001*
C8		151 (2.4)	151.7 (2.4)	<0.001*
C9		56.9 (2.2)	59.2 (2.3)	<0.001*
A		88.4 (1.9)	89.6 (1.8)	<0.001*
TH		8 (1.6)		
Modified Asgari score		4.8 (1.9)		

SD: Standard deviation, C1: Maximum cranial length, C2: Maximum cranial height, C3: Foramen magnum sagittal diameter, C4: Supra occiput length, C5: Clivus length, C6: Posterior cranial fossa anteroposterior length, C7: Occipital cord length, C8: Longest anteroposterior diameter of the cerebrum, C9: Posterior height of the cranial, A: Tentorium cerebelli slope, TH: Tonsillar herniation length, * Student's t-test, ** Chi-Square test

Table 2: Comparison of cranium, cerebrum and cerebellum morphological measurements with study group's age, herniation, and the modified Asgari score (Spearman's correlation test)

	Age		TH		MAS	
	r	*P*-value	r	*P*-value	r	*P*-value
C1	0.515	<0.001	-0.184	0.007	-0.104	0.130
C2	0.039	0.574	-0.158	0.022	-0.141	0.040
C3	0.467	<0.001	-0.020	0.767	0.026	0.705
C4	-0.390	<0.001	-0.037	0.588	-0.040	0.558
C5	-0.131	0.057	-0.110	0.110	-0.089	0.197
C6	0.572	<0.001	-0.027	0.699	0.038	0.584
C7	-0.064	0.354	0.100	0.148	-0.004	0.956
C8	-0.212	0.002	0.078	0.255	-0.021	0.758
C9	-0.446	<0.001	0.006	0.936	-0.085	0.218
A	-0.178	0.009	0.129	0.061	0.120	0.081
TH	-0.127	0.065		1.000	0.598	<0.001
MAS	-0.122	0.077	0.598	<0.001		1.000

C1: Maximum cranial length, C2: Maximum cranial height, C3: Foramen magnum sagittal diameter, C4: Supra occiput length, C5: Clivus length, C6: Posterior cranial fossa anteroposterior length, C7: Occipital cord length, C8: Longest anteroposterior diameter of the cerebrum, C9: Posterior height of the cranial, A: Tentorium cerebelli slope, TH: Tonsillar herniation length, MAS: Modified Asgari score

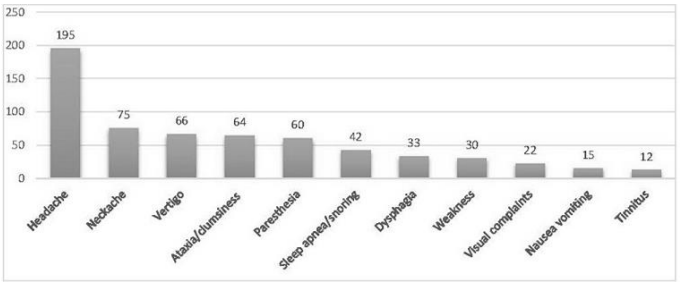

Figure 4: Symptoms in the study group

Discussion

Although genetic defects and defects during the embryologic period are considered to cause CM1, it has been reported that CM1 can develop during the period after birth and that the existing disease can progress or regress over time. Moreover, etiology of CM1 includes gene mutations occurring during the embryologic period, teratogenicity, and abnormal mechanical force(s) which can result in shape defects in the skull [6]. To prevent the progression of CM1, surgical treatment is recommended as soon as possible for patients with CM1 exhibiting progressive or persistent symptoms and/or widening syringomyelia. Moreover, it has been reported that the symptoms may regress after the application of this treatment [8].

The prevalence of CH1 was reported as 0.5%–0.9% in various populations [1,6,9,10]. In our study, we found the prevalence as 9.2% on brain MRI, much higher compared to other studies in the literature. The patients in our study group were suspected of CM1 after clinical and examination findings.

In studies conducted on adult patients, the mean age has been reported to be between 36 and 43 years [14-16]. The mean age of the study group in our study was 39.3 (13) years, which is similar to that reported in the literature. We believe that CM1 is prevalently detected in young adult patients as a result of MRI scans performed to identify the etiology of symptoms such as long-term headaches.

Various studies describe that CM1 has a higher prevalence among women than among men [6,14,16]. Arnautovic et al. [15] reported that CM1 is more prevalent among women, and suggested that the cause for this is the coincidental identification of lesions in women while determining the cause of headaches. In contrast with the literature, the prevalence was higher among male patients in our study group, and this may be related to the differences between societies and the number of male patients admitted.

It is considered that the defect in the cranial structures in CM1 occurs during the intrauterine period as a result of the disruption of the spinal migration [9]. It has been stated that patients with CM1 have larger foramen magnum and smaller PCF than control patients [9,17]. Houston et al. observed that fastigium height, pons height, corpus callosum height, clivus length, and PCF height are shorter in patients with CM1, McRae line length are higher whereas the PCF volume is similar to those in the control group [11]. In the same study, it was emphasized that the condition leading to CM1 was due to abnormalities at many points of the cranium [11].

In their study, Taştemur et al. [14] stated that in individuals with tonsillar herniation, foramen magnum sagittal diameter, cerebellum height, and cerebellum sagittal diameter increases, whereas the maximum cranial height, supraocciput length, clivus length, and fossa cranii posterior height decreases. It has been stated that the cranium would bend forward and the tonsils would be lower in the case of short villus and hypoplastic condyle [6]. It was found in our study that the bone structures of the study group were smaller and foramen magnum was larger. Microcephaly or early closing of sutures for any reason (such as genetic factors, trauma, and injective agents) during organogenesis and following periods may have caused the head to be smaller. This condition may have led to the movement of the parenchymal tissues toward the space/gap (spinal canal) into which they could move. Moreover, this herniation may have caused the widening of the foramen magnum. Houston et al. reported that the structures within PCF are lower in patients with CM1 than the control group [11]. Taştemur et al. [14] stated that the cerebellar structures in patients with CM1 were larger than in those in the control group in their study. Poretti et al. [6] reported that herniation occurs when there is excess parenchymal tissue within PCF. In our study, it was observed in the control group that the cerebral tissue in the parenchyma was larger, it grew in the vertical axis, and it was narrower in other axes. We believe that parenchymal tissue did not fit inside the cranium owing to its excessive amount, and the overflowing area pressured the cerebellar tissue and caused herniation of the cerebellar tissue into the spinal canal. Parenchymal tissues may thus have the tendency to grow in the vertical plane.

Houston et al. [11] and Taştemur et al. [14] stated that although the slope of the tentorium cerebelli is smaller in the

CM1 group than the control group, there is no significant difference. In our study, the tentorium cerebelli slope was found to be significantly lower in the study group. We believe that the cerebellar tissue was pushed forward owing to the narrowness of the angle. The cerebellum was thus longer in the vertical axis, and the cerebellum that was unable to fit into the PCF shifted toward the spinal canal.

Approximately 37% of pediatric patients with CM1 are asymptomatic and such patients are diagnosed by coincidence [6]. Yarbrough et al. [18] stated that the clinical findings and symptoms of patients with CM1 may vary, and the results of these findings/symptoms can also differ among patients. It has been suggested that the pressure caused by the soft tissue and bone structures on PCF and spinal canal result in the development of symptoms [9]. It has been reported that the most prevalent symptoms in CM1 are a headache, neck ache, poor balance, and cognitive function disorder [6,8,19]. Gilmer et al. stated that the most commonly encountered symptoms among the patients in their study were a headache (93.1%) and neck ache (47.9%) [20]. Lei et al. [16] reported that the most prevalent symptoms among patients are headache and neck ache (73%), sensory disorder (58.9%), and motor weakness (41%). In our study, the most prevalent symptoms were determined as a headache and neck ache which is consistent with the literature. We believe that headache and neck ache is the most prevalent indications of increased intracranial pressure secondary to the disrupted CSF circulation.

In addition, we believe that the other symptoms emerge in parallel to the rate at which the centers in the herniated area are affected.

CM1 herniation can be as small as 3 mm and as large as 25 mm [11]. Taştemur et al. [14] reported the average herniation amount in individuals with tonsillar herniation of 4.85 (3.09) mm. Lei et al. [16] reported the amount of tonsil ectopia in patients to be 13.6 mm. CM1 causes the increase in the amount of tissue within foramen magnum and this disrupts the CSF circulation. As the amount of tissue increases, CSF circulation becomes more disrupted and in parallel with this, the patient's symptoms worsen [18]. Moreover, there are studies stating that there is no relationship between the level of herniation and the severity of symptoms [10,20]. In our study, it was detected that the amount of herniation was 8 (1.6) mm, and it had a positive correlation with the modified Asgari score. We believe that the increase in the amount of herniation would cause further disruption in CSF pressure and/or result in an increase in the areas being subjected to pressure, making the symptoms become even worse.

Gilmer et al. [20] stated in their study that the symptoms of patients in advanced ages are less and that there is a negative correlation between Chicago Chiari Outcome Scale and age. In this study, it was stated that although CM1 is a congenital abnormality, the patients can become symptomatic at any age and the symptoms can change over time. In the anamneses obtained from symptomatic geriatric patients, it was noted that CM1 actually existed congenitally or since childhood and became worse [20]. In our study, it was found that in parallel with the increase in age, bone structures grew with the decrease in parenchyma tissues. However, there was no relationship noted between age and clinic. We believe that bones continue to grow with age and the formed bone structure is osteoporotic; moreover, there is a decrease in the parenchyma tissue simultaneously owing to atrophy. We believe that the space within the cranial cavity increases with age but the herniated tissue does not go back to its previous location, and the symptoms do not change due to the permanence in the bone changes related to CM1 (such as the widening of the foramen magnum, and clivus shortness).

Previous studies show that the narrowness of PCF or the excess in parenchyma plays a role in herniation [6,11,14]. An earlier study stated that a 2-mm change in the intracranial distance can cause considerable differences in the patient's symptoms [11]. In our study, it was found that the amount of herniation and the modified Asgari score had a negative correlation with the size of cerebral structures and a positive correlation with the length of the cerebellar structure. We believe that the amount of herniated tissue increases largely owing to the decreased volume in the PCF and the excessive parenchymal tissue. Longer cerebral structures may be an indicator of an increased amount of herniation. Thus, the patient's symptoms may have been worsened in relation to the amount of herniation. Its negative relationship with the cranial length may have resulted in a possible excess of parenchyma, causing the parenchyma tissue to shift not into the spinal canal but into the cranium.

Our study has certain limitations. Our study had a retrospective design, and it was based on the patient file analyses. Further studies showing the correlation of CM1 symptoms with radiological measurements are needed.

Conclusion

The bone structures forming the cranium have an effect on the symptoms of patients with CM1. It was particularly determined that the pathologies causing the narrowing of the volume of the posterior fossa were related to tonsillar herniation.

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