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Fourth Ventricle Dimensions Increase in Chiari Malformation Type 1

Halil Can KUCUKYILDIZ, Unal OZUM

Sivas Cumhuriyet University, School of Medicine, Department of Neurosurgery, Sivas, Turkey

Corresponding author: Halil Can KUCUKYILDIZ ✉ drhalilcan@gmail.com

ABSTRACT

AIM: To investigate whether fourth ventricle dimensions and tentorial angulation differ in a healthy control population in our evaluation of patients with CM-1 malformation using MRI.

MATERIAL and METHODS: The radiological and demographic data from 251 patients with CM-1 followed in our clinic between 2014 and 2019 were compared with data from 273 persons in a healthy control group. Fourth ventricle dimensions, amount of cerebellar tonsillar herniation, and tentorium twinning angle were measured. Statistical analysis was performed.

RESULTS: The mean tentorial twinning angle, craniocaudal length, and anteroposterior length of the fourth ventricle were significantly greater than the mean of the same measurements in the healthy control group. In addition, in a subgroup analysis conducted according to treatment modalities of patients with CM-1, the length between the bilateral recesses of the fourth ventricle was found to be statistically significantly greater in the subgroup of patients who underwent surgery compared with those in the nonsurgical subgroup.

CONCLUSION: Fourth ventricle enlargement is a radiographic finding in patients with CM-1. Studies evaluating clinical presentation, severity, and outcome after treatment will be useful in revealing the importance of this entity.

KEYWORDS: Cerebellum, Fourth ventricle, Tonsillar herniation, Tentorium, Posterior fossa

INTRODUCTION

Chiari type 1 malformation (CM-1) is a class of congenital disorders that are characterized by a >5-mm cerebellar tonsillar descent from the foramen magnum toward the spinal canal (1,5). The clinical presentations of CM-1 malformations may vary due to affected adjacent structures such as the cerebellum, brainstem, or spinal cord (6,7).

The fourth ventricle is a structure located in the posterior fossa and surrounded with important anatomical components, particularly the brainstem and cerebellum. In pathologies affecting the posterior fossa, in which there are many vital anatomical structures, the size and shape of the fourth ventricle can also be predicted to be affected (4).

Magnetic resonance imaging (MRI) has an important role in the diagnosis and follow-up of intracranial pathologies. Magnetic resonance imaging can help make it easier to show the amount of tonsillar herniation and tentorial angle changes to improve fourth ventricle visualization, and to measure the dimensions of the structures of the posterior fossa (10).

In this study, we aimed to investigate whether fourth ventricle dimensions and tentorial angulation differ in a healthy control population in our evaluation of patients with CM-1 malformation using MRI.

MATERIAL and METHODS

This study is a retrospective evaluation of radiological and demographic data from patients with CM-1 treated and followed in a single center. The radiological and demographic data from 251 patients with CM-1 followed in our clinic between 2014 and 2019 were compared with data from 273

Halil Can KUCUKYILDIZ : 0000-0003-0922-1750 Unal OZUM : 0000-0003-2065-2033

healthy control persons. Of these 251 patients with CM-1, 82 patients were further assigned to a surgical subgroup and 169 patients to a nonsurgical subgroup. Magnetic resonance images were acquired using the Siemens Aera 1.5 Tesla MRI scanner (Siemens, Erlangen, Germany). Preoperative imaging in the surgical subgroup was compared with imaging obtained at the time of diagnosis in the nonsurgical subgroup. The control group comprised individuals who presented at our hospital with various complaints and underwent brain MRI examination with no pathology findings. All measurements were performed using the Sectra IDS7 Workstation (Linköping, Sweden). Brain MRI measurements in T1 and T2 weighted sequences, were performed in axial, coronal, and sagittal planes in a multiplanar manner by the same researcher. The craniocaudal length, the length between both lateral recesses, the length between the base and the median dorsal recess of the fourth ventricle, the amount of herniation of the cerebellar tonsils from the foramen magnum to the spinal canal, and the tentorium twinning angle were measured on sagittal, coronal axial planes of the MRIs (Figure 1).

During analysis, the general statistics of the data set (descriptive statistics and frequency table) were examined. Whether the dimensions of the fourth ventricle differ in patient–control and nonsurgical patients were examined. First, the normality test of the fourth ventricle dimensions and tentorial twinning angle measurements was investigated using the Shapiro–Wilks test. Because the assumption of normal recovery for all parameters could not be proven ($p<0.05$), the difference analysis was performed with nonparametric test groups. The Mann–Whitney U test was used to analyze the difference. The Spearman correlation test was used to investigate the relationship of fourth ventricle dimensions in patient–control and nonsurgical groups. All analyses were performed using SPSS software, version 22 (Armonk, NY). The significance of the analyses was interpreted according to the 95% confidence level.

This study was conducted according to the principles stated in the Declaration of Helsinki and approved by the human research ethics committee of Sivas Cumhuriyet University (Registry No: 2020/05-14).

RESULTS

In total, 251 symptomatic patients with CM-1 were analyzed and compared with 273 healthy control subjects. There was no statistically significant difference in mean age between the groups. However, the female population was 76.9% (n=193) in the CM-1 group and 57.1% (n=156) in the control group. When the CM-1 group cathegorized by age; adult group was 86.9% (n=218), and pediatric group was 13.1% (n=33) of all. These ratios in healthy control group were 78.4% (n=214) and 21.6% (n=59) respectively. Of the 251 patients with CM-1, 82 had been assigned to the surgically-treated subgroup (Table I). Syringomyelia was present in 40 (52.6%) of 76 adult patients and 4 (66.6%) of 6 pediatric patients who were surgically treated on.

The mean tentorial twinning angle, craniocaudal length, and anteroposterior length of the fourth ventricle were significantly greater than the mean of the same measurements in the healthy controls (Table II). In addition, in the subgroup analysis

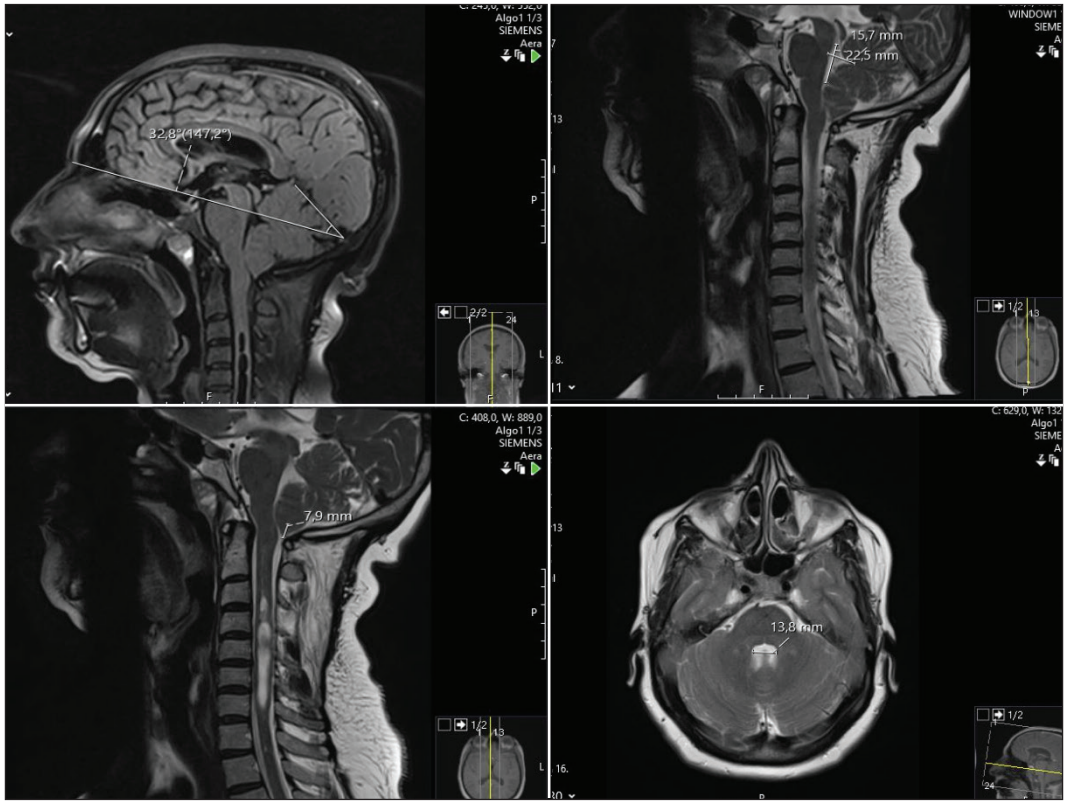

Figure 1: Measurements on magnetic resonance imaging sequences in patients with Chiari type 1 malformation.

according to treatment modalities of patients with CM-1, the length between the bilateral recesses of the fourth ventricle was found to be statistically significantly greater in in the subgroup of patients who underwent surgery compared with the nonsurgical subgroup (Table III).

Table I: Demographic Properties of Patients with CM-1 Patients and Healthy Controls

	CM-1	Healthy controls
Mean age	35 (0-79)	32 (0-66)
Adults (≥18 age)	218 (86.9%)	214 (78.4%)
Female gender	193 (76.9%)	156 (57.1%)
Treatment method		
Surgical	82 (32.6%)	-
Conservative	169 (67.4%)	-

DISCUSSION

CM-1 is a congenital disease with cerebellar tonsillar herniation that is diagnosed radiologically and supported by associated clinical findings. Researchers have carried out many studies thus far with the prediction that CM-1 is not limited to cerebellar tonsillar herniation only. Studies published to date have shown that CM-1 is accompanied by malformations of other anatomical structures in the posterior fossa in addition to cerebellar tonsillar herniation. For example, Guvenc et al. and Shrestha et al. have reported that occipital bone hypoplasia that forms the bony roof of the posterior fossa accompanies CM-1 (2,10). Marin-Padilla et al have reported that the amount of tonsillar herniation into the spinal canal increased due to

Table II: Radiographic Measurements in Patients with CM-1 and Healthy Controls

		Minimum	Maximum	Mean	SD	p
Tentorial twinning angle	Control	2.50	59.90	40.81	6.69	**0.001**
	CM-1	23.94	72.08	43.55	8.33	
Craniocaudal length of the fourth ventricle	Control	8.20	27.80	17.01	2.85	**0.001**
	CM-1	5.34	47.11	18.71	5.56	
Anteroposterior length of the fourth ventricle	Control	5.10	13.80	9.14	1.49	**<0.001**
	CM-1	3.32	23.47	10.51	2.84	
Bilateral length of the fourth ventricle	Control	6.80	20.00	12.78	1.86	0.183
	CM-1	4.29	36.64	13.48	3.83	

SD: *Standard deviation. Bold values highlight significance (p<0.05).*

Table III: Radiographic Measurements in Patients with CM-1 in Surgical and Nonsurgical Subgroups

		Mean	SD	p
Age	Conservative	34	16	0.079
	Surgical	38	13	
Tentorial twinning angle	Conservative	43.41	8.54	0.451
	Surgical	43.82	7.93	
Craniocaudal length of the fourth ventricle	Conservative	18.42	5.05	0.76
	Surgical	19.29	6.48	
Anteroposterior length of the fourth ventricle	Conservative	10.45	2.81	0.484
	Surgical	10.61	2.91	
Bilateral length of the fourth ventricle	Conservative	12.73	3.55	**<0.001**
	Surgical	15.03	3.92	
Amount of tonsillar herniation	Conservative	8.74	3.73	**<0.001**
	Surgical	16.53	4.94	

SD: *Standard deviation. Bold values highlight significance (p<0.05).*

posterior fossa hypoplasia in 2 separate studies (3,4). There are other studies reporting that the depth and surface of the posterior fossa are reduced in patients with CM-1 compared with these measures in healthy subjects (2,8,9,11).

In a study evaluating the craniovertebral junction pathologies and supratentorial–infratentorial angulations accompanying CM-1, it was reported that the tentorium twinning angle increased and the association of CM-1 with basilar invagination was reported (3). In our study, we also founded the twinning line significantly wider patients with CM-1.

In this study, we predicted that the dimensions of the fourth ventricle, which is an important anatomical structure of the posterior fossa, may be affected in patients with CM-1. When the literature was examined, it was seen that there was only 1 known study focusing on this subject. In our study, we found that the length measurements of the fourth ventricle in the craniocaudal, bilateral, and anteroposterior planes in patients with CM-1 increased statistically significantly compared with these measurements in the healthy study group. In the study by Scott et al, in which the fourth ventricle volume was calculated in patients with CM-1, as similarly calculated in our study, it was reported that the fourth ventricle size increased compared with the fourth ventricle size the in healthy populations (12).

It was an advantage that our study was conducted in larger patient and control populations compared with those in similar studies. We believe that the enlarged fourth ventricle outcome we obtained in this large group may be due to an obstruction at the distal of fourth ventricle in a shrunken posterior fossa.

The limitations of our study are its retrospective design, lack of clinical information, and use of radiographic measurements. However, we think that the diagnosis, follow-up, treatment, and evaluation of large patient and control groups conducted at a single center by the same team constitute the strength of the study.

CONCLUSION

In the patients with CM-1, the fourth ventricle size was found to significantly increase compared with that in the healthy control participants. As a physiopathological mechanism, it can be suggested that the fourth ventricle size may affect the amount of tonsillar herniation and the amount of herniation may affect the size of the fourth ventricle. This determination is not yet clear. However, it would be useful to consider fourth ventricle dimensions in the evaluation of patients with CM-1. In determining the importance of fourth ventricular dimensions in CM-1, a wider range of studies evaluating clinical severity, accompanying brainstem findings, symptoms that cannot be explained by tonsillar herniation alone, and postsurgical outcome are needed.

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AUTHORSHIP CONTRIBUTION

Study conception and design: HCK, UO

Data collection: HCK

Analysis and interpretation of results: HCK, UO

Draft manuscript preparation: HCK

Critical revision of the article: UO

Other (study supervision, fundings, materials, etc...): N/A

All authors (HCK, UO) reviewed the results and approved the final version of the manuscript.

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