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A Morphometric Evaluation Together with Computed Tomography Examination of Chiari Type 1 Cases with Craniovertebral Junction Anomaly

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ABSTRACT

AIM: To compare the posterior cranial fossa (PCF) dimensions together with the measurements related to basilar invagination and platybasia of craniovertebral junction anomalies (CVJA) in CVJA (+) and CVJA (-) Chiari malformation Type 1 (CM1) patient groups with each other and with healthy control subjects.

MATERIAL and METHODS: The study group was formed of 43 CM1 and 9 tonsillar ectopia (TE) patients.

RESULTS: A decrease was determined in the PCF vertical length (clivus and supraocciput line) and PCF volume and an increase in the transverse length (McRae and Twining line) in the CM1 cases compared to the healthy control group. There was no difference between the CVJA (+) and (-) CM1 groups in respect of the vertical and transverse length and PCF volume values. An increase in the classic and modified skull base angles was observed in the CVJA (+) CM1 group compared to the CVJA (-) CM1 group. The Wackenheim clivus angle was determined to be smaller in the CVJA (+) CM1 group compared to the CVJA (-) CM1 group.

CONCLUSION: The PCF is flattened and smaller in CM1 cases compared to normal control subjects. In the planning of CM1 operations, the angle parameters may be more useful than the PCF and CVJA length parameters between CVJA (+) and (-) CM1 groups. The significant decrease in postoperative recovery in the CVJA (+) CM1 group compared to the CVJA (-) CM1 group supports the need for additional operations and/or a different surgical technique in the treatment of CVJA (+) CM1 patients.

KEYWORDS: Craniovertebral junction anomaly, Chiari malformation Type 1, Posterior cranial fossa, Computed tomography

ABBREVIATIONS: PCF: Posterior cranial fossa, **CVJA:** Craniovertebral junction anomalies, **CM1:** Chiari malformation Type 1, **CSF:** Cerebrospinal fluid, **TE:** Tonsillar ectopia, **CT:** Computed tomography, **n:** Number, **SD:** Standard deviation, **MRI:** Magnetic resonance imaging

INTRODUCTION

Chiari malformation Type 1 (CM1) originates from an embryonic development abnormality in the hindbrain region and is characterised by the caudal migration of the cerebellar tonsils from the foramen magnum by 5 mm or more. This definition is only a radiological definition and in literature there are cases of clinical CM1 with cerebellar tonsils displacement of more than 3 mm (1,8). The prevalence of CM1 has been reported to be approximately 1% of

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the general population (20). In addition to occipital bone dysplasia and often a decrease in the posterior cranial fossa (PCF) dimensions, CM1 is also generally associated with craniovertebral junction anomalies (CVJA) such as clivus concavity, platybasia, and basilar invagination (26).

CM1 is the most mild type of Chiari malformation. It is seen more often in females and as it generally causes symptoms in the third and fourth decades of life it has been named the adult form of Chiari malformations. Although several hypotheses have been proposed, the etiology is still not clear. These hypotheses are based on primary mesodermal failure, hypoplasia of the basilar part of the occipital bone or small PCF associated with platybasia (10,11,14,25).

Neurological signs and symptoms of different severity emerge over time in CM1. These symptoms occur because of entrapment of the cerebellum, brainstem, spinal cord, and cranial nerves, and due to syringomyelia which can form in the spinal cord. Just as these symptoms may be non-specific such as tinnitus, headache, neck pain, dizziness and paresthesia in the extremities, they may also be specific such as glossopharyngeal neuralgia, trigeminal neuralgia, autonomous nerve system findings and hearing loss. If CM1 is not considered in the differential diagnosis, the patient may be misdiagnosed (2,10,11,21).

Flow dynamics of the cerebrospinal fluid (CSF) may be impaired in CM1, and syringomyelia may occur in the cervical region due to CSF flow disorder in approximately 20-72% of CM1 cases. Syringomyelia, which is the most common comorbid disorder of CM1, is the formation of CSF spaces in the spinal cord. Syringomyelia causes paresthesia, hyperesthesia, or anesthesia in a classic cape-like pattern in the neck and shoulders that does not involve a particular dermatome due to spinal cord compression. Enlargement of the central canal causes a loss of dissociation of the sensation of pain and warmth, but the sense of light touch and proprioception are initially preserved. Weakness, non-radicular pain and spasticity may also be observed, predominantly in the upper extremities (19,22,23).

There are studies in literature related to the decrease in PCF dimensions and accompanying platybasia in CM1 patients. However, apart from the Chiari 1000 project, there are few studies comparing PCF dimensions and platybasia measurements in CM1 patients with and without CVJA, as there have been insufficient numbers of patients in these studies (9,13). To the best of our knowledge, this study is only the second in literature to have compared the PCF dimensions, basilar invagination, and platybasia measurements of adult CM1 patients with and without CVJA with each other and with a healthy control group.

MATERIAL and METHODS

The study included 43 CM1 and 9 tonsillar ectopia (TE) patients who had been referred from the Neurosurgery Department to the Radiology Department with a clinical prediagnosis of CM1 between 2007 and 2020 and the diagnosis was confirmed on computed tomography (CT). The patients comprised 34 females and 18 males in the age range of 19-56 years. A control group was formed of 52 age and gender-matched subjects with no diagnosis of CM1. A retrospective evaluation was made of the CT images taken between 2007 and 2020.

Before starting the measurement procedures, approval for the study was granted by the Ethics Committee of Mersin University (decision no: 2021/727). The CM1 patients and control group were scanned with 16 and 64-slice tomography devices (Aquillion 16 and 64, Toshiba Medical Systems, Tokyo, Japan). The slices were of 0.5mm thickness in axial, coronal and sagittal planes, and were reformatted using the Extrempacs imaging program.

For optimal evaluation of the craniovertebral junction, the patients included in the study were those with images including all the bone structures between the sphenoid bone base and the C2 vertebra base. In the CM1 patient group, there was no intracranial space-occupying pathology which could affect the measurements. No control group subjects had any intracranial space-occupying lesion, or congenital or systemic disease related to the craniovertebral junction. Tonsillar herniation length was evaluated in the sagittal plane. In the CM1 patients, the tonsillar herniation extending downwards from the foramen magnum was measured in millimetres (Figure 1). The patients were divided into two groups according to the clinical and radiological diagnosis of TE (3-5mm tonsillar herniation) and CM1 (>5mm tonsillar herniation).

The CM1 patients were also evaluated as those with and without CVJA. The most common of the CVJAs observed were basilar invagination (abnormal localisation of the odontoid process towards the superior and posterior, creating prolapsus to the foramen magnum) (27), occipitalisation of the atlas (the appearance of a transitional vertebra formed from the union of the atlas and occipitus) (27), platybasia (abnormal flattening of the skull base at an angle >143°) (18), and atlantoaxial subluxation (C1-C2 impairment causing impaired neck rotation) (29) (Figure 2).

The cranial length parameters used in the study were the length of the McRae line (the line extending from the basion to the opisthion) (15), the Chamberlain line (the line extending from the posterior end of the hard palate to the opisthion) (15), the length of the clivus (the distance between the inferior end of the dorsum sella and the basion) (15), the length of the Twining line (the line extending from the tuberculum sella to the internal occipital protruberance) (30), the length of the supraocciput line (the line extending from the opisthion to the internal occipital protuberance) (5) and the length of the atlantodental interval (the distance between the C1 anterior arch and the dens axis) (29) (Figures 2, 3).

The cranial angles used in the study were the Welcher classic skull base angle (the angle formed at the junction of the line connecting the center of the hypophysis fossa and the nasion and the line connecting the anterior edge of the foramen magnum with the center of the hypophysis fossa) and the modified skull base angle (the angle formed at the junction of the line extending along the anterior cranial fossa as far as the

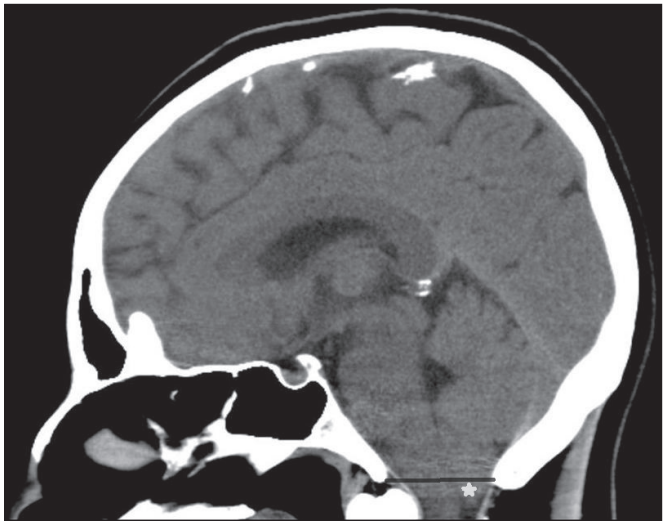

Figure 1: Chiari malformation Type 1 [tonsillar herniation extending inferiorally from the foramen magnum] (*).

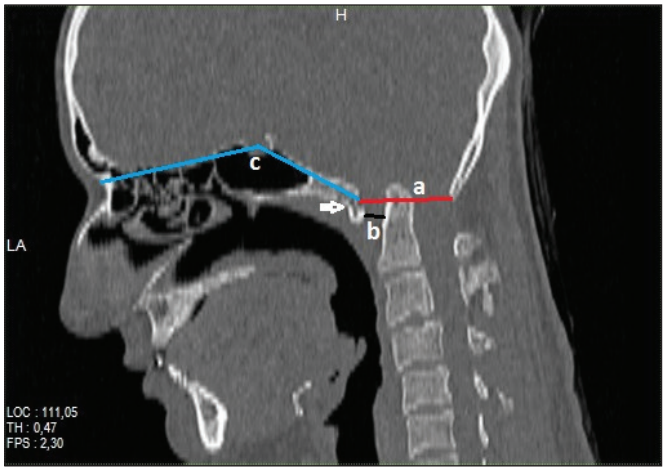

Figure 2: (a) Basilar invagination [extension of the dens axis to the superior of the McRae line], **(b)** atlantoaxial subluxation [atlantodental interval (the distance between the C1 anterior arch and the dens axis) of >5 mm], **(c)** platybasia [abnormal flattening of the skull base at an angle >143°] and (arrow) occipitalisation of the atlas [the appearance of a transitional vertebra formed from the union of the atlas and occipitus]

peak of the dorsum sella and the line drawn along the edge of the clivus arch) (18), and the Wackenheim clivus angle (the angle between the line drawn from the peak of the dorsum sella to the basion and the line extending from the basion to the posterior surface of the C2-3 vertebrae) (15) (Figures 3, 4).

PCF volume was measured using the Cavalieri method. In order to obtain an unbiased volume calculation with the Cavalieri method, the sum of the cross-sectional surface areas of the slices or sections along the structure is multiplied by the distance between the sections, that is, by the section thickness (7).

The coexistence of syringomyelia in the cervical spinal cord was examined in the TE, CVJA (+) and (-) CM1 groups

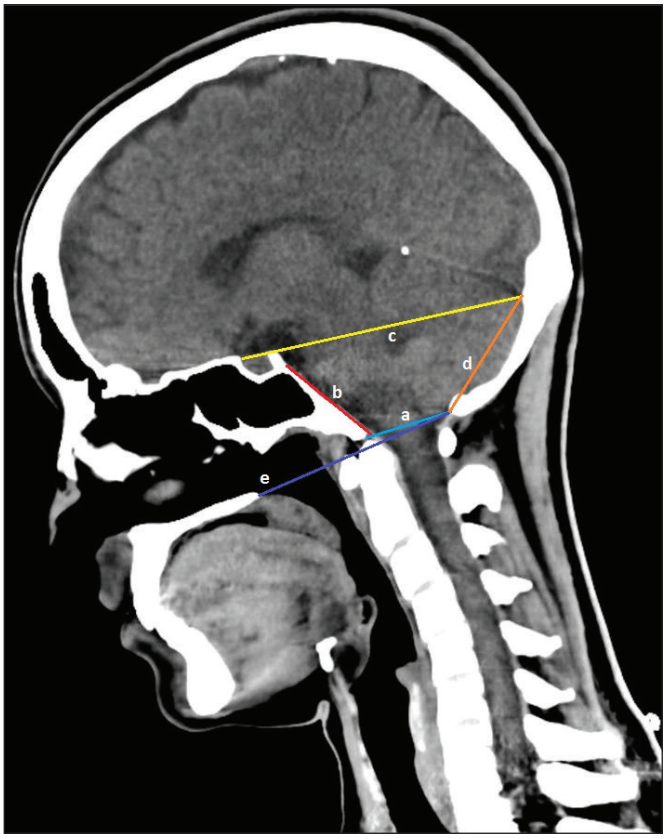

Figure 3: (a) The McRae line [the line extending from the basion to the opisthion], **(b)** the clivus [the distance between the inferior end of the dorsum sella and the basion], **(c)** the Twining line [the line extending from the tuberculum sella to the internal occipital protuberance], **(d)** the supraocciput line [the line extending from the opisthion to the internal occipital protuberance], and **(e)** the Chamberlain line [the line extending from the posterior end of the hard palate to the opisthion].

(Figure 5). Possible relationships between the presence of syringomyelia and the previously measured tonsillar herniation length were examined.

The conformity to normal distribution of the measurements obtained from the groups and subgroups was assessed with the Shapiro Wilk test. Categorical data were stated as number (n) and percentage (%). Continuous variables were found to show normal distribution and were stated as mean ± standard deviation (SD) values. The Chi-square test was used to determine relationships between two groups of categorical variables. In the comparisons of more than two groups, the Anova test was applied. Paired groups were compared against the control group using the Duncan test.

RESULTS

The distribution of males and females in the TE, CM1 and control groups was determined to be homogenous (p=0.676). Females constituted 77.8% of the TE group and 62.8% of the CM1 group (Table I). The mean age was determined to be homogenous in the TE, CM1 and control groups (p=0.943).

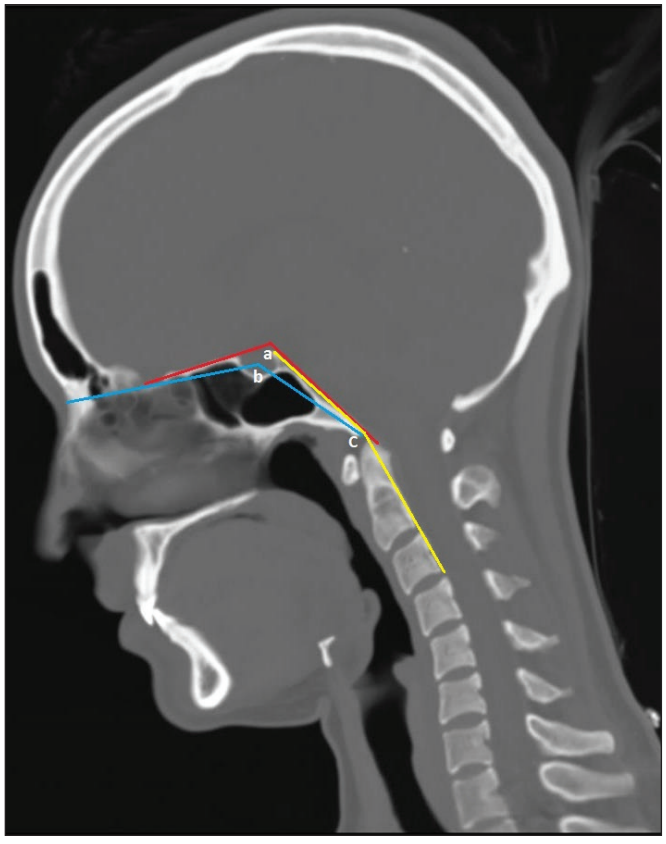

Figure 4: (a) The modified skull base angle [the angle formed at the junction of the line extending along the anterior cranial fossa as far as the peak of the dorsum sella and the line drawn along the edge of the clivus arch], **(b)** the Welcher classic skull base angle [the angle formed at the junction of the line connecting the center of the hypophysis fossa and the nasion and the line connecting the anterior edge of the foramen magnum with the center of the hypophysis fossa], **(c)** the Wackenham clivus angle [the angle between the line drawn from the peak of the dorsum sella to the basion and the line extending from the basion to the posterior surface of the C2-3 vertebrae].

Table I: Distribution of Patient Gender

		Group			
		Control n (%)	TE n (%)	CM1 n (%)	p
Gender	Female	34 (65.7)	7 (77.8)	27 (62.8)	0.676
	Male	18 (34.6)	2 (22.2)	16 (37.2)	

The distribution of males and females was homogenous in the groups (p=0.676).

The mean age was 36.89 ± 10.25 years in the TE group and 38.12 ± 9.72 years in the CM1 group (Table II).

The mean length of the clivus was shorter in the TE group than in the control group, but the difference was not statistically significant (p=0.089). The mean length of the clivus was significantly shorter in the CM1 group than in the control group (p<0.001) (Table II). Within the CM1 group, the mean length of the clivus was shorter in the CVJA (+) group than in the CVJA (-) group, but the difference was not statistically significant (p=0.205) (Table IV).

The mean length of the McRae line showed no significant difference between the TE group and the control group (p=0.067), and was statistically significantly longer in the CM1 group than in the control group (p<0.001) (Table II). No statistically significant difference was determined between the CVJA (+) and CVJA (-) CM1 groups in respect of the mean length of the McRae line (p=0.830).

The mean length of the supraocciput line was shorter in the TE group than in the control group, but the difference was not statistically significant (p=0.17). The mean length of the supraocciput line was statistically significantly shorter in the CM1 group than in the control group (p=0.03) (Table II). No statistically significant difference was determined between the CVJA (+) and CVJA (-) CM1 groups in respect of the mean length of the supraocciput line (p=0.171).

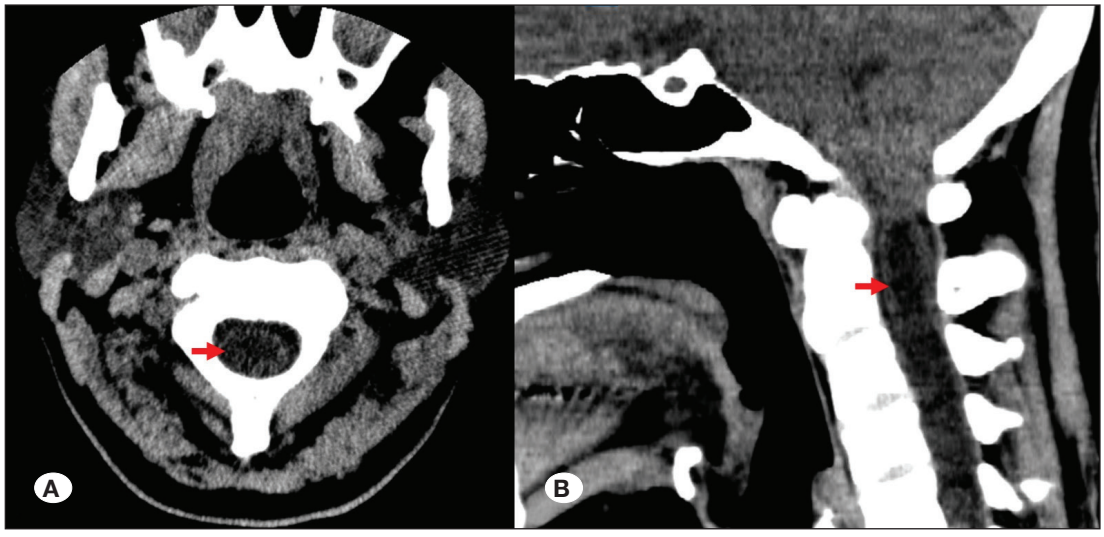

Figure 5: Syringomyelia on axial **(A)** and sagittal **(B)** images of the cervical spinal cord.

Table II: Comparison of the Age, Lengths, and Angle Parameters of the Control Group and TE and CM1 Patients

	Group			
	Control (n=52)	**TE (n=9)**	**CM1 (n=43)**	
	mean ± SD	**mean ± SD**	**mean ± SD**	**p**
McRae line length	31.10 ± 2.98	33.27 ± 2.28	33.94 ± 2.70[a]	**<0.001**
Atlantodental interval length	1.47 ± 0.29	2.12 ± 1.36[a]	1.62 ± 0.90	**0.048**
Clivus length	36.95 ± 3.13	34.77 ± 1.48	34.56 ± 3.05[a]	**0.001**
Chamberlain line length	74.68 ± 5.27	74.41 ± 3.69	74.88 ± 4.56	0.958
Supraocciput line length	40.67 ± 4.18	38.43 ± 2.99	38.82 ± 2.94[a]	**0.029**
Twining line length	88.55 ± 4.38	90.47 ± 3.42	91.00 ± 4.43[a]	**0.023**
Classic skull base angle	130.24 ± 4.73	135.17 ± 7.29[a]	132.52 ± 6.52	**0.028**
Modified skull base angle	116.66 ± 6.23	117.41 ± 7.29	115.90 ± 7.80	0.790
Wackenheim clivus angle	158.21 ± 8.89	148.42 ± 14.70[a]	147.03 ± 11.9[a]	**<0.001**
Age (years)	37.90 ± 9.72	36.89 ± 10.25	38.12 ± 9.72	0.943
Posterior Cranial Fossa Volume	156.61 ± 14.59	154.08 ± 26.12	138.96 ± 16.34[a]	**<0.001**

a: *difference shown compared to the control group.*

In the current study, the mean length of the Twining line was longer in the TE group than in the control group, but the difference was not statistically significant (p=0.389). The mean length of the Twining line was longer in the CM1 group than in the control group (p=0.014) (Table II). No statistically significant difference was determined between the CVJA (+) and CVJA (-) CM1 groups in respect of the mean length of the Twining line (p=0.401).

In addition to the PCF borders, the cranial length parameter of the Chamberlain line was evaluated. No statistically signficant difference was determined in the mean length of the Chamberlain line between the control group and the TE and the CM1 groups, or between the CVJA (+) and CVJA (-) CM1 groups (Table II, Table IV, respectively).

CVJA was observed in 1 (11.1%) of the TE group and in 9 (20.9%) of the CM1 group (p<0.001) (Table III). Of these 10 patients, basilar invagination was observed in 9, occipitalisation of the atlas in all 10, platybasia in 3, and atlantoaxial dislocation in 3.

Basilar invagination was observed in 8 (18.6%) patients in the CM1 group. The mean Wackenheim clivus angle was determined to be significantly lower in the TE group and the CM1 group than in the control group (p=0.027, p<0.001, respectively) (Table II). The mean Wackenham clivus angle was determined to be smaller in the CVJA (+) CM1 group than in the CVJA (-) CM1 group (p=0.001) (Table IV). Occipitalisation of the atlas was observed in all the CM1 cases with basilar invagination.

Both the classic skull base angle and the modified skull base angle were evaluated using MRI. Higher values were observed in the CVJA (+) CM1 group compared to the control group in respect of both the classic and modified skull base angle (p<0.001, p=0.02, respectively) (Table IV). The mean classic skull base angle was greater in the CM1 group than in the control group, but the difference was not statistically significant (p=0.109). The mean classic skull base angle was greater in the TE group than in the control group (p=0.039) (Table II). The classic skull base angle was determined to be greater in the CVJA (+) CM1 group than in the CVJA (-) CM1 group (p=0.001) (Table IV).

Table III: Distribution of Skull Base Anomaly in the Groups

		Group			
		Control n (%)	**TE n (%)**	**CM1 n (%)**	**p**
Skull Base Anomaly	Absent	52 (100)	8 (88.9)	34 (79.1)	<0.001
	Present	0 (0)	1 (11.1)	9 (20.9)	

A relationship was determined between the groups and skull base anomaly (p<0.001).

The modified skull base angle measurements showed no statistically significant difference between the control group, the TE group and the CM1 group (p=0.790) (Table II). The modified skull base angle was determined to be significantly greater in the CVJA (+) CM1 group than in the CVJA (-) CM1 group (p=0.001) (Table IV).

The mean atlantodental interval was longer in the TE group (p=0.029) (Table II) and the CVJA (+) CM1 group compared to the control group (p=0.008) (Table IV). The mean atlantointerval length was longer in the CVJA (+) CM1 group than in the CVJA (-) CM1 group (p=0.011) (Table IV).

Table IV: Comparisons of the Mean Lengths and Angle Parameters of the CM1 Patients with and without CVJA

	Group			
	Control (n=52)	**CVJA (-) CM1 (n=34)**	**CVJA (+) CM1 (n=9)**	
	mean ± SD	**mean ± SD**	**mean ± SD**	**p**
McRae line length	31.10 ± 2.98	33.85 ± 2.80[a]	34.31 ± 2.42[a]	**<0.001**
Atlantodental interval length	1.47 ± 0.29	1.49 ± 0.38	2.13 ± 1.82[a]	**0.013**
Clivus length	36.95 ± 3.13	34,92 ± 2.98[a]	33.21 ± 3.13[a]	**0.001**
Chamberlain line length	74.68 ± 5.27	74.14 ± 4.24	77.70 ± 4.86	0.154
Supraocciput line length	40.67 ± 4.18	38.37 ± 2.88[a]	40.53 ± 2.63	**0.017**
Twining line length	88.55 ± 4.38	91.37 ± 4.54[a]	89.62 ± 3.97	**0.018**
Classic skull base angle	130.24 ± 4.73	131.04 ± 5.46	138.13 ± 7.46[a]	**<0.001**
Modified skull base angle	116.66 ± 6.23	114.05 ± 6.84	122.89 ± 7.56[a]	0.002
Wackenheim clivus angle	158.21 ± 8.89	149.87 ± 10.08[a]	136.30 ± 12.99[a]	**<0.001**
Posterior Cranial Fossa Volume	156.61 ± 14.59	139.17 ± 17.72[a]	138.18 ± 10.30[a]	<0.001
Posterior Cranial Fossa Volume	156.61 ± 14.59	154.08 ± 26.12	138.96 ± 16.34[a]	**<0.001**

a: *difference shown compared to the control group.*

Table V: Comparison of the Patient Conditions in Groups

		Group			
		TE	**CVJA (-) CM1**	**CVJA (+) CM1**	
		n (%)	**n (%)**	**n (%)**	**p**
Patient condition	Postoperative follow-up	3 (33.3)	19 (55.9)	8 (88.9)	0.102
	Non-operative follow-up	5 (55.6)	10 (29.4)	1 (11.1)	
	Asymptomatic patient	1 (11.1)	5 (14.7)	0 (0)	
	Total	9 (100)	34 (100)	9 (100)	

p<0.05: statistical significance value.

The mean volume of the PCF was less in the TE group than in the control group, but the difference was not statistically significant (p=0.888). The mean volume of the PCF was significantly less in the CM1 group than in the control group (p<0.001) (Table II). The mean volume of the PCF was less in the CVJA (+) CM1 group than in the CVJA (-) CM1 group, but the difference was not statistically significant (p=0.984) (Table IV).

In the TE group, the total number of patients followed up without surgery and asymptomatic patients was greater than the number of patients who underwent surgery. In the CVJA (-) CM1 group, the total number of patients followed up without surgery and asymptomatic patients was lower than the number of patients who underwent surgery, but the difference was not statistically significant. While no asymptomatic patients were observed in the CVJA (+) CM1 group, the number of patients who underwent surgery was significantly greater than the number of patients without surgery (Table V).

None of the patient complaints improved after the operation in the CVJA (+) CM1 group, and the complaints improved in approximately half of the patients in the CVJA (-) CM1 group after the operation (p=0.009) (Table VI).

There was no significant difference between the TE, CVJA (+) and (-) CM1 groups in terms of cervical syringomyelia coexistence (p=0.739) (Table VII). The minimum-maximum and mean tonsillar herniation length values of the cases with cervical syringomyelia in the groups were 3.3-3.5 (3.4 ± 0.14) mm in TE, 5.3-13.2 (8.7 ± 2.87) mm in CVJA (-) CM1, and 6.2-13.1 (9.65 ± 4.88) mm in CVJA (+) CM1. The minimum-maximum and mean tonsillar herniation length values of the cases without cervical syringomyelia in the groups were 3.2-4.3 (3.56 ± 0.41) mm in TE, 5.1-23.3 (8.1 ± 3.91) mm in CVJA (-) CM1, and 5.3-15.3 (9.6 ± 4.22) mm in CVJA (+) CM1.

Table VI: Comparison of the Postoperative Complaints in Groups

		Group			
		TE	CVJA (-) CM1	CVJA (+) CM1	
		n (%)	n (%)	n (%)	p
Postoperative complaint	(-)	1 (33.3)	10 (52.6)	0 (0)	0.009
	(+)	2 (66.7)	9 (47.4)	8 (100)	
	Total	3 (100)	19 (100)	8 (100)	

p<0.05: statistical significance value.

Table VII: Comparison of the Presence of Cervical Syringomyelia in Groups

		Group			
		TE	CVJA (-) CM1	CVJA (+) CM1	
		n (%)	n (%)	n (%)	p
Syringomyelia	(-)	2 (22.2)	11 (32.4)	2 (22.2)	0.739
	(+)	7 (77.8)	23 (67.6)	7 (77.8)	
	Total	9 (100)	34 (100)	9 (100)	

p<0.05: statistical significance value.

DISCUSSION

In literature, CM1 has been reported more in females (10,11,14,25). The findings of this study were consistent with literature as there was a predominance of female patients and the symptoms of CM1 were generally seen in the third and fourth decades of life (10,11,14,25).

The major differences between MRI and CT imaging modalities are that MRI images demonstrate superior soft-tissue contrast compared to CT scans, whereas CT scans provide better images of bony material and blood vessels. MR images are subject to unique artifacts that must be recognized. CT should always be the first imaging method selected as it is the best tool for the imaging of bone changes following trauma, for the evaluation of bone structures, and for visualising the relationships between different bone elements, or the imaging of soft tissue calcifications. There are a few studies in literature showing a decrease in PCF dimensions in CM1 patients. The PCF borders are formed of the clivus, the McRae line, the supraocciput line, and the Twining line (3,4,15,28).

In studies by Karagöz et al. and Alkoç et al., the mean length of the clivus was found to be statistically significantly shorter in the CM1 group than in the control group (p=0.007, p=0.001, respectively) (4,15). Al-Habib et al. found the mean length of the clivus to be shorter in the CVJA (+) CM1 group than in the CVJA (-) CM1 group (p=0.001) (3). The reason for the shorter clivus length in the CVJA (+) CM1 group was stated to be due to CVJA (+) criteria including a short clivus.

In the study by Karagöz et al., the McRae line was determined to be longer in the CM1 group than in the control group but not at a statistically significant level (p=0.43) (15). Al-Habib et al. found no statistically significant difference between the CVJA (+) and (-) CM1 groups in respect of the mean length of the McRae line (p=0.365) (3).

In studies by Karagöz et al. and Alkoç et al., the mean length of the supraocciput was found to be statistically significantly shorter in the CM1 group than in the control group (p<0.001, p=0.001, respectively) (4,15). Al-Habib et al. found no statistically significant difference between the CVJA (+) and CVJA (-) CM1 groups in respect of the mean length of the supraocciput line (p=0.757) (3).

Karagöz et al. found the mean length of the Twining line to be longer in the CM1 group than in the control group (p<0.016) (15). Al-Habib et al. found no statistically significant difference between the CVJA (+) and CVJA (-) CM1 groups in respect of the mean length of the Twining line (p=0.993) (3).

To the best of our knowledge, no other study in literature has compared the mean length of the Chamberlain line between CM1 patients and a control group. Consistent with the findings of the current study, Al-Habib et al. found no statistically significant difference between the CVJA (+) and CVJA (-) CM1 groups in respect of the mean length of the Chamberlain line (p=0.435) (3).

By differentiating between normal and abnormal bone, CT can be helpful in deciding whether CVJA is congenital or acquired (30). In the current study, evaluation was made of the most frequently observed CVJAs, such as platybasia, basilar invagination, atlantoaxial dislocation and occipitalisation of the atlas. Basilar invagination is diagnosed radiologically with the McRae line or the Chamberlain line passing the peak of the odontoid process by >5mm (16,17). The combination of basilar invagination with CM1 has been reported as 12% and 14.2% (16,17). Xu et al. determined a significant decrease in the mean Wackenheim clivus angle in basilar invagination, and stated that this angle was a new method in the determination of basilar invagination (31). In the study by Alkoç et al., the mean Wackenheim clivus angle was found to be smaller in the CM1 group than in the control group but the difference was not statistically significant (p>0.05) (4). The absence of a statistically significant difference may have been due to the lack of anomalies in addition to basilar invagination in a sufficient number of CM1 patients. That there was the additional anomaly of basilar invagination in 8 of the 9 patients in the CVJA (+) CM1 group and in 1 of the TE group may have been the reason for the more evident decrease in the mean Wackenheim clivus angle in the CVJA (+) CM1 group. Unlike the current study, Al-Habib et al. found no statistically significant difference between the CVJA (+) and CVJA (-) CM1 groups in respect of the mean Wackenheim clivus angle (p=0.058) (3). The reason for this lack of significant difference may be that the Al-Habib study did not include as many cases with basilar invagination as the current study. One of the limitations of this study was the inability to evaluate radiographic instability, which may partially affect the measurements related to basilar invagination. Occipitalisation of the atlas is often seen in CM1 cases with basilar invagination (6).

The skull base angle is used in the diagnosis of platybasia and basilar kyphosis. There are several different techniques that can be used on sagittal images obtained from CT or magnetic resonance imaging (MRI). In the past, skull base angle measurements were traditionally based on direct radiographs of the skull. According to the classic skull base angle normal values, platybasia is evaluated as 125°-143°, and basilar kyphosis as <125° (24). MRI techniques yield lower basal angles than the values reported using traditional radiographs. Therefore, Koenigsbert et al. defined the modified skull base angle method for MRI which provides consistent and repeatable measurements (18). In reports in literature that have collated the MRI measurements of several studies, platybasia is evaluated as skull base angle >129° (12).

In the current study, the reason for the difference in the classic skull base angle and the modified skull base angle can be attributed to there being no control group subjects with platybasia anomaly, whereas it was present in 3 of the 9 patients in the CVJA (+) CM1 group. In the study by Alkoç et al., the Welcher skull base angle was found to be greater in the CM1 group than in the control group (p=0.04) (4). Al-Habib et al. found the classic skull base angle to be greater in the CVJA (+) CM1 group than in the CVJA (-) CM1 group (p<0.001) (3).

Alkoç et al. also found no statistically significant difference in the modified skull base angle values of the control and CM1 groups (p>0.05) (4). The modified skull base angle comparison between CVJA (+) and (-) groups was not made in the study by Alkoç et al. (4). Al-Habib et al. compared the CVJA (+) and (-) groups in respect of the classic skull base angle but not in respect of the modified angle (3). Similar to the current study, Eppelheimer et al. found that the mean modified skull base angle was increased in CM1 cases with the CVJA of craniocervical instability, compared to all CM1 cases (p>0.05) (9).

On CVJ sagittal images, the atlantodental interval measured as >5mm in patients aged <9 years, and >3mm in patients aged >9 years is diagnostic for atlantoaxial dislocation. In the current study, the mean atlantodental interval was longer in the TE group and the CVJA (+) CM1 group compared to the control group. This could have been due to the presence of the additional anomaly of atlantoaxial dislocation in 1 patient in the TE group and in 2 patients in the CVJA (+) CM1 group. Behari et al. published a series of 39 cases showing the combination of CM1 and atlantoaxial dislocation (6). To the best of our knowledge, no other study in literature has compared CVJA (+) and (-) groups in respect of the mean length of the atlantodental interval.

In studies by Alkoç et al., the mean volume of the PCF was found to be statistically significantly less in the CM1 group than in the control group (p=0.009) (4). To the best of our knowledge, the current study is the first study in literature to have compared CVJA (+) and (-) groups in respect of the mean volume of the PCF.

The results of the current study demonstrated that the symptoms are more severe in all CM1 groups, especially in the CVJA (+) CM1 group, than in the TE group. While none of the patient complaints improved after the operation in the CVJA (+) CM1 group, the complaints improved in approximately half of the patients in the CVJA (-) CM1 group after the operation. The main statistical differences in measurements between the CVJA (-) and (+) CM1 groups were determined in the Classic and Modified skull base angles and the Wackenheim Clivus angle. Based on this, it can be said that the Classic and Modified skull base angles and the Wackenheim Clivus angle have an important role in determining the severity of CM1 symptoms and predicting improvement after surgery.

CONCLUSION

There was no significant difference between the TE, CVJA (+) and (-) CM1 groups in terms of cervical syringomyelia coexistence. Although there are many hypotheses regarding the mechanism of formation of syringomyelia, the etiology remains unclear. Further more comprehensive studies are required to clarify the association of CM1 and syringomyelia.

The results of this study demonstrated that the PCF vertical length (clivus and supraocciput line) was shorter in CM1 cases than in the control group, and an increase was observed in transverse length (McRae and Twining lines). According to these findings, the PCF is flattened in CM1 cases compared to normal control subjects. The volume of the PCF was significantly less in the CM1 group than in the control group, but no significant difference was determined in PCF volume between the CVJA (+) and (-) CM1 groups.

The PCF vertical length (clivus and supraocciput line) was also seen to be shorter in TE cases than in the control group, and an increase was observed in transverse length (McRae and Twining lines) although not statistically significant. The volume of the PCF was less in the TE group than in the control group, but not statistically significant. Therefore, TE can be considered a transitional form between CM1 and normal cases when PCF volume and the lengths of the PCF margins are compared between CM1, TE and normal cases.

No difference was determined between the CM1 group, the TE group and the control group in respect of the classic and modified skull base angles, while an increase was determined in the skull base angles of the CVJA (+) CM1 group compared to the CVJA (-) CM1 group. The Wackenheim clivus angle was seen to be smaller in the CM1 group and TE group than in the control group and in the CVJA (+) CM1 group compared to the CVJA (-) CM1 group.

In the planning of CM1 operations, the angle parameters may be more useful than the PCF and CVJA length and volume parameters to differentiate between CVJA (+) and (-) CM1 groups. The significant decrease in postoperative recovery in the CVJA (+) CM1 group compared to the CVJA (-) CM1 group supports the need for additional operations and/or a different surgical technique in the treatment of CVJA (+) CM1 patients.

AUTHORSHIP CONTRIBUTION

Study conception and design: BT, CB, SKK

Data collection: BT, CB, HHY

Analysis and interpretation of results: BT, CB, GT

Draft manuscript preparation: BT, SKK, HHY

Critical revision of the article: BT, CB, GT

Other (study supervision, fundings, materials, etc...): BT, CB

All authors (BT, CB, HHY, GT, SKK) reviewed the results and approved the final version of the manuscript.

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