Cervico-medullary compression ratio: A novel radiological parameter correlating with clinical severity in Chiari type 1 malformation

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ABSTRACT

Objectives: Chiari malformation type 1 (CM-1) is associated with cough headache, intracranial hypertension, cerebellar and spinal cord symptoms/signs. Herniated cerebellar tonsil length (HCTL) is widely used radiological parameter to determine the severity of CM-1, but with limited utility due to its weak correlation with some clinico-radiological findings. In this study, we aimed to evaluate a novel, practical parameter (cervico-medullary compression ratio; "CMCR") for its relationship with clinico-radiological findings in CM-1.

Patients and methods: Thirty-five adult patients (17 F, 18 M) with CM-1 were included in this retrospective study. Head CT and craniospinal MR images were assessed. CMCR was calculated as the ratio of herniated cerebellar tonsil surface area to foramen magnum surface area, and HCTL was measured. These two parameters were correlated with clinical and radiological findings.

Results: The mean CMCR was 0.60 ± 0.15 and mean HCTL was 8.91 ± 3.4 mm with no significant difference between gender and age groups for both parameters. For cough headache (0.64 ± 0.14 vs 0.52 ± 0.15, $p = 0.043$) and syringomyelia (0.67 ± 0.11 vs 0.56 ± 0.16, $p = 0.039$), only CMCR; for intracranial hypertension (CMCR: 0.64 ± 0.14 vs 0.55 ± 0.16, $p = 0.049$; HCTL: 9.66 ± 3.59 mm vs 7.79 ± 3.03 mm; $p = 0.045$) and cerebellar symptoms (CMCR: 0.65 ± 0.14 vs 0.54 ± 0.16, $p = 0.048$; HCTL: 10.4 ± 3.5 mm vs 7.4 ± 2.8 mm, $p = 0.041$), both CMCR and HTCL were significantly different between patients with and without respective findings. However, neither CMCR nor HTCL was different between patients with and without spinal cord symptoms and hydrocephalus.

Conclusion: CMCR is a superior numerical parameter than HCTL for the assessment of clinical severity in CM-1 cases and needs further validation with larger studies.

1. Introduction

Chiari malformation type 1 (CM-1) has been described as the herniation of the cerebellar tonsils from the foramen magnum (FM) to the cervical spinal canal and its prevalence in adult population is as high as 1% [1,2]. It might be associated with congenital pathologies, such as basilar invagination, platybasia, craniosynocytosis, posterior fossa hypoplasia, atresia of foramina of Magendie and Luschka, larger cerebellar structures and clival anomalies [3–5]. Congenital biomechanical deficiencies as well as secondary or iatrogenic factors such as trauma, hydrocephalus, tethered cord and intraspinal hypotension are blamed in the etiology [5–11]. Most cases are usually asymptomatic in the early period. Symptoms begin to emerge between the ages of 25–40, and they could progress from mild headaches and dysesthesias to respiratory failure and tetraplegia [1–4].

Herniated cerebellar tonsil length (HCTL) should be at least 5 mm below the FM for the CM-1 diagnosis [2,7]. However, HCTL for symptomatic CM-1 could be as little as 2–3 mm [12,13]. On the other hand, several studies suggested a correlation between HCTL and the severity of clinical symptoms [12–15]. CM-1 severity has been classified according to HCTL: mild: 5–9 mm, moderate: 9–14 mm, and severe: > 14 mm [2,7,14]. However, this view is still controversial and HCTL is thought to be inadequate to determine neither response to nor timing of the surgical treatment for CM-1 [15–18]. Interestingly, nearly 20% of

cases with HCTL greater than 5 mm, even with HCTL up to 27 mm, might be asymptomatic [15,16]. Inverse relationship between age and tonsil length has also been reported such that the size of the herniation diminishes but the clinical findings progress as the age increases [17]. As a result, there was no consensus about the relationship between the clinical symptoms of CM-1 cases and numerical value of HCTL and hence conquest for a better clinical and/or radiological marker still continues [18].

Classical headache of CM-1 is the most common symptom and described as cough headache because it increases during a Valsalva maneuver. This situation is due to the pressure gradient between the cranial and spinal compartments induced by the maneuver [18–20]. Later, progressive pathophysiological process ensues and results in bulbar, cerebellar and spinal cord symptoms and signs including paresthesia, dysesthesia, motor disturbances, trigeminal and glossopharyngeal neuralgia, hemifacial spasm, phonation disorders, sleep apnea, swallowing difficulty, ataxia, dysmetria and dysdiadokokinesia, sensory discrimination, sphincter insufficiency and impotence [6,8,11,21–28]. These findings are thought to be related to ischemia and syringomyelia/syringobulbia, especially at the level of foramen magnum (FM), resulting from tonsillar herniation [8,21]. Progressive nature of CM-1 has also been reflected in recently described Chiari Severity Index [18]. In conclusion, all theories attempting to explain clinical symptomatology in CM-1 patients suggest that the etiopathogenesis is closely related to the pressure on the cervicomedullary compartment at the craniovertebral junction.

In this study, we evaluated a possible relationship between clinico-radiological findings of patients and a novel, simple numerical parameter ("cervicomedullary compression ratio" at the FM level). In addition, the results obtained were compared with HCTL values that had been previously validated and widely used in the literature.

2. Patients and methods

2.1. Study design and data collection

This study has been conducted at Diskapi Yildirim Beyazit Training and Research Hospital, Ankara, Turkey upon the approval by the Ethics Committee for Clinical Investigations (Date: 27/04/2015, No: 22/14). A total of 35 patients who were treated at our institution between April 2013 and April 2015 were included and retrospectively evaluated in this study (17 females, 18 males; mean age: 37, range: 18–65 years). Patients were divided into 5 groups according to their age (18–25, 26–35, 36–45, 46–55, 56–65) (Table 1). Patients with systemic or other congenital pathologies have been excluded from the study. Demographic groups of patients participating in the study were homogeneous. Magnetic resonance imaging (MRI) and computed tomography (CT) data and clinical findings were obtained retrospectively from the archives of the Departments Radiology and Neurosurgery. Demographic characteristics of the patients were given in the Table 1.

2.2. Measurement and calculation of CMCR

Topographic area calculations were performed on axial head CT images of the patients (Toshiba Alexion TSX-034A), at the level of FM opening in the craniovertebral junction. Section properties Pitch factor 0.688, helical pitch value 11. Average 120 kV, 110 mas, rotation time 0.75, FOV 240 were used. Images were obtained from CT images with 1 mm cross-sectional thickness and 0.8 mm cross-section intervals. Foramen magnum surface area (FMSA) and herniated cerebellar tonsil surface area (HCTSA) were measured as cm^2 at the bone and parenchymal windows (General Electric AW volume Share 5 workstation), respectively. The ratio of the HCTSA to the FMSA (HCTSA/FMSA) was calculated and described as the CMCR (Fig. 1A, B) (Table 1).

Table 1
Distribution of radiological measurements according to demographic characteristics.

Demographic characteristics	HCTSA (cm^2)	FMSA (cm^2)	CMCR	HCTL (mm)
Age				
18–25 yo (n = 9)	5.47 ± 1.31	8.47 ± 1.57	0.58 ± 0.14	8.42 ± 3.77
26–35 yo (n = 10)	4.35 ± 1.34	8.51 ± 1.22	0.62 ± 0.12	7.75 ± 2.37
36–45 yo (n = 5)	5.60 ± 1.26	10.30 ± 1.02	0.68 ± 0.16	7.96 ± 1.96
46–55 yo (n = 6)	5.66 ± 1.42	8.19 ± 0.89	0.59 ± 0.11	12.03 ± 4.51
56–65 yo (n = 5)	5.40 ± 1.32	7.89 ± 1.61	0.64 ± 0.15	9.78 ± 3.82
P value:	0.578	0.126	0.432	0.446
Gender				
Female (n = 17)	4.98 ± 1.13	7.90 ± 1.34	0.63 ± 0.13	9.18 ± 3.93
Male (n = 18)	5.40 ± 1.52	9.35 ± 1.26	0.57 ± 0.18	8.63 ± 2.98
P value:	0.378	0.002*	0.302	0.654
TOTAL (n = 35)	5.18 ± 1.35	8.60 ± 1.45	0.60 ± 0.15	8.91 ± 3.68

CMCR: cervico-medullary compression ratio calculated as the ratio of HCTSA to FMSA; FMSA: foramen magnum surface area; HCTL: herniated cerebellar tonsil length; HCTSA: herniated cerebellar tonsil surface area.
* Statistically significant p < 0.05.

2.3. Measurement of the HCTL

Herniated cerebellar tonsil length (HCTL) below the FM was obtained from mid-sagittal T2W MRI images. All MRI was performed on 1.5 T scanner (Philips Intera, Philips Medical Systems, Best, Netherlands) with a multi-channel cervical-thoracic-lumbar spine coil and included the following sequence: sagittal T2-weighted image (TR/TE = 3000–4000 ms/70–80 ms, FOV = 24 cm, matrix 480 × 480, section thickness/gap = 4/1 mm). Measurements were made using mid-sagittal MRI sections on the PACS system (K-PACS, Kuratorium OFFIS e.V. Healthcare Information and Communication Systems, Oldenburg/Germany). The McRae line (from basion to opisthion at the level of the FM) was considered as the reference point. HCTL was measured on a direct line from McRae line to the tip of herniated tonsil and determined as mm (Fig. 2A). According to the severity of HCTL, patients were divided into 4 groups: 2–5 mm (suspicious), 5–9 mm (mild), 10–14 mm (moderate), and more than 14 mm (severe) (Table 2) [8].

2.4. Clinical evaluation of symptom and signs

The presence of headache aggravated by cough was regarded as cardinal symptom of CM-1. Triad of cough headache, nausea/vomiting and papillary edema were accepted as increased intracranial pressure syndrome (IIPS). The presence of any of the findings of dysmetria, dysdiadokokinesia, and nystagmus was determined as cerebellar involvement (ataxia was excluded because it may also be due to syringomyelia). Pain-temperature discrimination of the extremities and/or presence of motor weakness were considered as spinal cord symptoms. Clinical evaluation results were marked as either present or absent, and the patients were grouped accordingly. Clinical signs were correlated with CMCR and HCTL (Table 3).

2.5. Radiological evaluation of hydrocephalus and syringomyelia

The presence of hydrocephalus and syringomyelia were evaluated on the parenchymal axial head CT or MR images (for hydrocephalus; temporal horn width ≥ 2 mm or Evan's ratio > 30%) and on sagittal T2 cervical-thoracic MRI sections (for syringomyelia) (Fig. 2B). The results were recorded as either present or absent, and the patients were
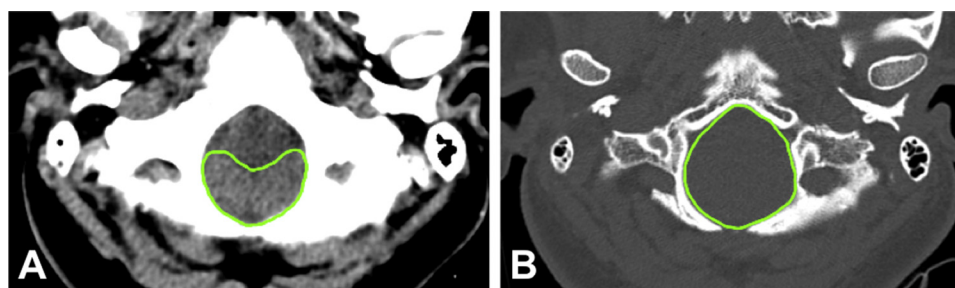
Fig. 1. Radiological measurements at the level of foramen magnum on axial CT images. A: Herniated cerebellar tonsil surface area measured on parenchymal window of head CT; B: Foramen magnum surface area measured on bone window of head CT.

grouped accordingly. Whether the results were correlated with CMCR and HCTL was investigated (Table 3).

2.6. Statistical analysis

After all data were obtained, IBM SPSS Statistics Version 22 for Windows was used for analysis. Frequency analysis was used for demographic analysis. The descriptive statistics were analyzed as mean ± S.D., minimum, maximum and percentage (%). The Kolmogorov–Smirnov test was used to determine the normalization of data. One-way ANOVA and Kruskal–Wallis tests were used for comparisons between multiple groups while Student *t*-test and Mann–Whitney U tests were used to compare the means of two groups with parametric and non-parametric variables, respectively. Spearman's correlation test was used to compare the correlation between continuous numerical variables and grouping variables. The value of $p < 0.05$ was considered to be statistically significant.

3. Results

3.1. Measurements of HCTSA and FMSA and calculation of CMCR

The average HCTSA was $5.18 \pm 1.35 \, \text{cm}^2$. No statistically significant difference was found between genders (F: 4.98 ± 1.13 vs M: 5.40 ± 1.52, p = 0.378). In addition, there was no statistically significant difference between age groups (p = 0.578) (Table 1). The average FMSA was found to be $8.60 \pm 1.45 \, \text{cm}^2$. When genders are compared, it was found that FMSA in males was significantly higher than females (F: 7.90 ± 1.34 vs M: 9.35 ± 1.26), p = 0.002). However, there was no significant difference between age groups (p = 0.126) (Table 1). When the CMCR (HCTSA/FMSA ratio) was examined, the mean value was 0.60 ± 0.15, there was no statistically significant difference between gender and age groups (F:0.63 ± 0,13vs M:0.57 ± 0.18, p = 0.302) (For age groups; p = 0,432) (Table 1).

Table 2
Cervico-medullary compression ratio in different cerebellar tonsil herniation severity groups based on HCTL. Although there is a trend of increase in CMCR with the extent of herniation (severity), difference among severity groups is not statistically significant (p = 0.435).

Severity (HCTL)	CMCR
Suspicious (2–5 mm)	0.57 ± 0.13
Mild (5–9 mm)	0.59 ± 0.14
Moderate (10–14 mm)	0.63 ± 0.15
Severe (≥14 mm)	0.63 ± 0.14

CMCR: cervico-medullary compression ratio; HCTL: herniated cerebellar tonsil length.

3.2. Measurement of HCTL

The average HCTL was 8.91 ± 3.4 mm. There was no statistical difference between male and female patients in terms of HCTL (F: 9.18 ± 3.93 vs M:8.63 ± 2.98, p = 0.654). When age groups were compared for the HCTL, it was found that there was no statistically significant difference (p = 0.446). As the severity of HCTL increased, the CMCR also increased gradually, but the difference between groups were not significant (p = 0.435) (Table 2).

3.3. Clinical evaluation of symptoms and signs

Mean CMCR of patients with cough headache (n = 24) was significantly higher than patients without headache (n = 11) (0.64 ± 0.14 vs 0.52 ± 0.15, p = 0.043). However, when the HCTL was examined, there was no statistically significant difference between the patients with and without cough headache (mean values for HCTL; 8.99 ± 3.66 mm vs 8.74 ± 3.06 mm, respectively; p = 0.835). There were significant differences in terms of both CMCRs and HCTLs of 21 patients who had IIPS, and 14 patients without IIPS (mean values for CMCR: 0.64 ± 0.14 vs 0.55 ± 0.16, p = 0.049, respectively; mean values for HCTL were 9.66 ± 3.59 mm vs 7.79 ± 3.03 mm,
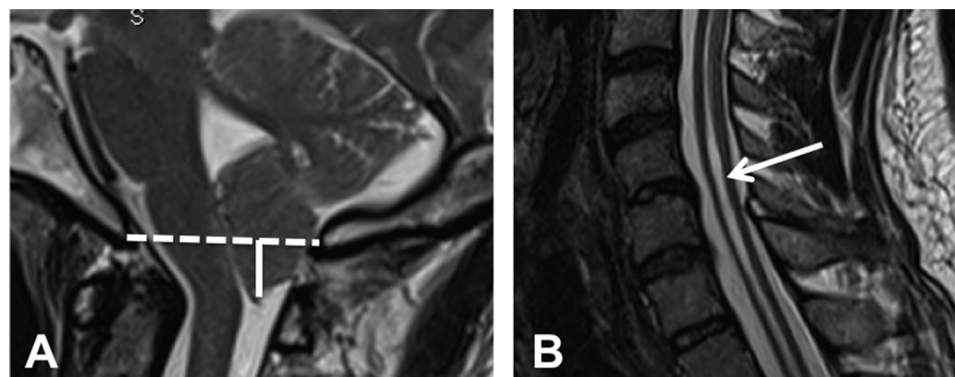

Fig. 2. Radiological measurements and syringomyelia on mid-sagittal T2W MR images. A: McRae line from basion to opisthion (dashed line) and herniated cerebellar tonsil length measured between McRae line and the tip of herniated tonsil (uninterrupted line); B: Arrow shows syringomyelia in cervical mid-sagittal T2W MR image.

Table 3
Association of clinical and radiological findings with CMCR and HCTL.

Clinical symptom and signs	CMCR		P value	HCTL (mm)		P value
	present	absent		present	absent	
Cough Headache	0.64 ± 0.12	0.52 ± 0.12	0.043[*]	8.99 ± 2.95	8.74 ± 3.12	0.835
IIPS	0.64 ± 0.14	0.55 ± 0.16	0.049[*]	9.66 ± 3.59	7.79 ± 3.03	0.045[*]
Cerebellar symptom and signs	0.65 ± 0.14	0.54 ± 0.16	0.040[*]	10.40 ± 3.50	7.46 ± 2.80	0.040[*]
Spinal cord symptom and signs	0.65 ± 0.13	0.56 ± 0.16	0.080	9.07 ± 3.20	8.78 ± 3.70	0.830
Radiological findings	CMCR		P value	HCTL (mm)		P value
	present	absent		present	absent	
Hydrocephalus	0.66 ± 0.15	0.58 ± 0.15	0.170	10.47 ± 3.34	8.37 ± 3.40	0.128
Syringomyelia	0.67 ± 0.11	0.56 ± 0.16	0.039[*]	9.45 ± 3.34	8.55 ± 3.57	0.454

CMCR: cervico-medullary compression ratio; HCTL: herniated cerebellar tonsil length; IIPS: increased intracranial pressure syndrome.

* Statistically significant p < 0.05.

respectively; p = 0.045). Similarly, both parameters were found to have statistically higher values in patients with cerebellar symptoms (n = 19) compared to patients without cerebellar symptoms (n = 16) (mean values for CMCR: 0.65 ± 0.14 vs 0.54 ± 0.16, p = 0.048, and mean values for HCTL: 10.4 ± 3.5 mm vs 7.4 ± 2.8 mm, p = 0.041, respectively). Although CMCR was more sensitive compared to HCTL measurement in patients with (n = 17) and without (n = 18) spinal cord symptoms, this sensitivity was not found to be statistically significant (mean CMCR: 0.65 ± 0.13 vs 0.56 ± 0.16, p = 0.126; mean HCTL: 9.07 ± 3.2 mm vs 8.78 ± 3.7 mm, p = 0.83, respectively) (Table 3).

3.4. Radiological evaluation of hydrocephalus and syringomyelia

The mean CMCR of patients with hydrocephalus (n = 9) was 0.66 ± 0.15, while the mean value of those without hydrocephalus (n = 26) was 0.58 ± 0.15, with no statistically significant difference between the two groups (p = 0.17). Similarly, there was no statistically significant difference in HCTL (mean HCTL of patients with and without hydrocephalus was 10.47 ± 3.34 mm vs 8.37 ± 3.4 mm, p = 0.128). The mean CMCR of patients with syringomyelia (n = 14) was 0.67 ± 0.11, while those without syringomyelia (n = 21) were found to be 0.56 ± 0.16, p = 0.039). However, there was no statistically significant difference between the two groups in terms of HCTL (mean HCTL of patients with and without syringomyelia was 9.45 ± 3.4 mm vs 8.55 ± 3.5 mm, p = 0.454) (Table 3).

4. Discussion

In this study, we calculated the ratio of herniated cerebellar tonsil surface area to foramen magnum surface area (cervico-medullary compression ratio, CMCR) on computed tomography images of Chiari malformation type 1 (CM-1) and evaluated its correlation with clinical symptoms and signs of CM-1 patients. We also critically analyzed clinical and radiological results with CMCR in comparison to widely used, albeit controversial, herniated cerebellar tonsil length on midsagittal T2 weighted magnetic resonance images.

Foramen magnum (FM), which connects cranial cavity and vertebral canal, is composed of the posterior part of the skull, basilar, lateral and squamous parts of the occipital bone [29,30]. FM is the largest of foramina at the skull base (mean diameter; from anterior to posterior: 30–40 mm, transverse: 30–35 mm), which is bordered by basion in the front, opisthion in the posterior and occipital condyles on both sides [29–32]. Medulla oblongata (MO), vertebral arteries, anterior and posterior spinal arteries, and spinal roots of nervus accessorius pass through the FM [29,31]. Even if it has different shapes and sizes, it has a standard average effective diameter for critical structures within

itself such as MO. In a classical study, it was reported that the effective diameter of FM should be at least 19 mm [33]. In the presence of pathologies that reduce the effective diameter such as basilar invagination, tumour, infection, tonsillar herniation, foraminal stenosis and craniodiafical dysplasia, neurological deficits occur due to compression caused by these pathologies to MO [33–41]. For these pathologies, surgical strategies have been developed such as transoral odontoidectomy (for anteriorly compressing pathologies) or FM posterior wall resection (for posteriorly compressing pathologies) aimed at alleviating the stress on the brain stem to counteract the etiopathogenesis [35–39].

When the etiopathogenesis of pathologic findings in Chiari malformation is examined, one encounters a chain of problems particularly caused by spatial compression at the level of FM rather than vertical elongation of cerebellar tonsils per se. However, in most studies conducted until now, findings in relation to anterior or posterior compression have only been evaluated with single dimension (i.e. diameter) but not two dimensions (i.e. area) [33]. For this reason, in our study regarding CM-1 cases, we aimed to investigate a numerical value (i.e. CMCR) which better explains the severity of the compression to brainstem. We anticipated that these values should be correlated with the clinical and radiological findings in CM-1 cases. Furthermore, we evaluated whether CMCR is correlated with HTCL, a commonly used measure in CM-1 studies.

In the past, FMSA was calculated in people from different ethnic backgrounds in different countries using radiographic images such as MR and CT or data obtained from cadaver and skull, and similar values were obtained in the case of cross-sectional differences. In males and females, mean FMSA was found to be 829–909 mm^2 and 781–819 mm^2, respectively [2,41–45]. In our study, the FMSA results were 935 ± 120 mm^2 for males and 760 ± 130 mm^2 for females (total average: 860 ± 145 mm^2), similar to literature findings. In order to eliminate discrepancies between studies, HCTSA/FMSA ratio, termed as "cervico-medullary compression ratio (CMCR)", which is independent of these variables, was used in our study. In addition, it was examined whether this radiological value was independent of developmental, sexual and genetic differences for the reliability of the study and it was found that there were no statistically significant differences between the age and gender groups (Table 1). To the best of our knowledge, there are no other studies in the English literature where the HCTSA measurement is performed and the HCTSA/FMSA ratio is calculated.

Patients with CM-1 suffer from a number of symptoms that result from the pressure of the MO, cervical spinal cord and cranial nerves [11,18]. In fact, the most common accompanying symptom in both congenital and acquired CM-1 is cough headache which is exacerbated by Valsalva maneuvers or physical exertions and felt in the suboccipital area [11,19,20]. Various authors suggested that headaches occurred

due to sudden increase in CSF pressure and the tension of the veins or meninges which include pain-sensitive nerve dendrites induced by displacement of the tonsils during a cough or Valsalva action [11,20,45,46]. In our study, the CMCR of cases which have cough headache was statistically significantly higher than the cases without cough headache (0.64 ± 0.14 vs 0.52 ± 0.14, respectively; p = 0.043). On the other hand, HCTL failed to distinguish the patients with and without cough headache (8.99 ± 3.66 mm vs 8.74 ± 3.06 mm, respectively; p = 0.835). Our results suggest that, contrary to what Pascual et al. [19] reported, the cough headache is not directly correlated with HCTL. A herniated cerebellar tonsil may indeed increase intracranial pressure that leads to cough headache before severe ischemia and resultant atrophy develop. One can assume that as the CMCR, rather than HCTL, increases, the CSF flow obstruction should become more severe.

In our study, headache, nausea-vomiting attacks and papillary edema were accepted as IIPS. Even if hydrocephalus was reported occasionally (10–20%) in the CM-1 cases, it is argued to be one of most important etiopathogenetic factors of headache, whose prevalence is much higher [22,27,33]. However, it is known that CM-1 could produce idiopathic intracranial hypertension due to diffuse intracerebral edema without radiologically diagnosed hydrocephalus [46,47]. The theory was supported in CM-1 cases by acetazolamide treatment which leads to decrease in the degree of tonsillar herniation [47]. In our study, it was found that the CMCR of the patients who were diagnosed with IIPS was significantly higher than the results of patients with no IIPS (0.64 ± 0.14 vs 0.55 ± 0.16, respectively; p = 0.049). However, there was no statistically significant difference for hydrocephalus (0.66 ± 0.15 vs 0.58 ± 0.15, respectively; p = 0.173). Same results were obtained for the HCTL values (HCTL for IIPS: 9.66 ± 3.59 mm vs 7.79 ± 3.03 mm; p = 0.045, HCTL for hydrocephalus: 10.47 ± 3.34 mm vs 8.37 ± 3.4 mm; p = 0.128). According to our results, the CMCR might be used as an efficient numerical factor for the decision of surgical treatment in CM-1 cases regardless of the presence of hydrocephalus.

In the literature, different morphometric measurements have been performed on radiological imaging of CM-1 cases for determining prognosis and survival of cases [48–53]. Dufton et al. reported that the clivus length was shorter and the FM was wider in CM-1 cases than normal healthy subjects [48]. In similar studies, increased tentorial angle, expanded basal opening, decreased depth of posterior fossa and smaller diameter of posterior fossa [49–52]. It has been thought that while cerebellar tissues naturally develop, the bony structures of posterior fossa could not comply with this development [50,50,51,52]. Interestingly, a recent study found no clinically useful 2D or 3D radiological measurements to reliably distinguish symptomatic and asymptomatic CM-1 cases in children [53]. However, none of these studies have focused on cerebellar impairment which is one of the main sources of clinical symptoms resulting from compressive and ischemic damage following tonsillar herniation. In our study, both CMCR and HCTL of cases were significantly higher in patients with impaired cerebellar tests than those without (For CMCR: 0.65 ± 0.14 vs 0.54 ± 0.16, respectively, p = 0.048; for HCTL: 10.4 ± 3.5 mm vs 7.4 ± 2.8 mm, respectively, p = 0.041). It is possible to say according to our results that the disruption of cerebellar functions in CM-1 cases is not only due to cerebellar compression caused by small posterior fossa, but may also be due to compression on spinocerebellar or cerebellospinal tracts at the craniovertebral junction.

It is stated that syringomyelia causes scoliosis, pain, sensation disorder (especially pain-temperature discrimination), weakness and ataxia in CM-1 cases [8,21,54]. Recent studies have demonstrated with cine-mode MR images that compliance at both spinal and intracranial CSF sites decreases in the presence of syringomyelia in CM-1 cases [55–57]. As a result, increased CSF pressure will cause posterior spinal venous pressure to increase, which in turn disturbs the absorption of CSF to the intravenous compartment. The inhibition of CSF flow results in the rupture of the central canal and formation of syrinx. Various authors believe that this condition is triggered by restriction of the subarachnoid space by herniated cerebellar tonsils at the craniovertebral junction [56–58]. However, to the best of our knowledge, there are no quantitative studies in the literature investigating severity of restricted subarachnoid space at the level of FM. In our study, it was observed that the CMCR was more sensitive than HCTL measurement in CM-1 cases with spinal cord symptoms, but there was no statistically significant difference in terms of both parameters. However, when CMCR of patients with and without syringomyelia were examined, it was found that there was a significant difference between the two groups; the higher the ratio is, the more commonly syringomyelia is seen (Table 3). In addition, there was no statistically significant difference between the groups despite the fact that patients with syringomyelia had higher HCTLs than those without (Table 3). While syringomyelia is diagnosed in a case whose HTCL values is 3.16 mm, it was not diagnosed in two other patients whose HTCL values were 16.3 and 13.8 mm [12–15]. Our study demonstrated on the contrary to what Milhorat et al. [14], Stovner et al. [45] and Godzik et al. [54] claimed that there is not a certain relation between syringomyelia and HTCL values, and CMCR ratio is a more sensitive value.

Due to retrospective design of the study as well as limited study population with only surgically treated cases, this study needs further confirmation. Future studies with larger cohorts and repeated CMCR measurements during longer follow-up periods are needed. By superimposing CT and MRI findings, CMCR measurements can be refined and thus radiological findings can be better correlated with disease specific symptoms and CSF flow perturbation in future studies.

5. Conclusions

Our study results show that HCTSA/FMSA ratio or CMCR is a superior numerical value than HCTL for clinical severity in CM-1 cases. CMCR successfully distinguished patients with cough headache, increased intracranial pressure syndrome, cerebellar signs and syringomyelia. CMCR is a promising radiological marker that can possibly be used for determining prognosis and selecting patients for surgical treatment.

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