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Chiari type I malformations in adults: a morphometric analysis of the posterior cranial fossa

Sabri Aydin, MD[a], Hakan Hanimoglu, MD[a], Taner Tanriverdi, MD[a], Ercument Yentur, MD[b], Mehmet Yasar Kaynar, MD[a,*]

Departments of [a]Neurosurgery and [b]Anesthesiology, Cerrahpasa Medical Faculty, Istanbul University, Istanbul 34301, Turkey

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Abstract

Objective: Chiari type I malformation (CMI) is a congenital disorder characterized by caudal displacement of the cerebellar tonsils through the foramen magnum into the spinal canal. Recent studies suggest that overcrowding in the posterior cranial fossa (PCF) because of underdeveloped bony structures in the intrauterine life is the main cause of this malformation. For this reason, the authors want to contribute to the current literature, which focuses on bone abnormalities in the PCF in patients with CMIs.

Methods: We examined a retrospective cohort of 60 adult patients with CMIs, and multiple measurements were made on magnetic resonance imaging. The results were compared to 30 healthy adult control subjects. Mann-Whitney *U* test was used as a statistical method.

Results: All measurements except mean anteroposterior diameter of the foramen magnum were reduced in patients compared to control. An increase in the anteroposterior mid-sagittal distance of the foramen magnum in patients reached statistically significant difference compared to control. All patients had tonsillar herniation at least 5 mm below the plane of the foramen magnum. Chiari type I malformation in this study was associated with syringomyelia in 46 patients.

Conclusion: This study with a limited number of patients suggests that the bony components of the PCF are not developed fully, supporting the current concept that CMI is a disorder of the para-axial mesoderm.

Keywords: Chiari malformations; Posterior fossa; Syringomyelia

1. Introduction

The Chiari malformations (CMs) are a group of disorders of unknown etiology that has been traditionally described as the downward displacement of the posterior cranial fossa (PCF) contents into the spinal canal. Since the first description by an Austrian pathologist, Hans Chiari, in 1891 [9], CMs have been divided into 2 distinct groups according to the degree of herniation: type I represents herniation of the cerebellar tonsils and type II displays herniation of the fourth ventricle and the medulla oblongata as well as the caudal part of the cerebellar tonsils. However, from a clinical point of view, CMs have also been divided into adult and pediatric types [13]. Adult type is mostly considered as Chiari type I (CMI) malformations and usually presents after the second or third decade of life. The signs and/or symptoms are mainly resulting from the tightness of PCF and associated syringomyelia [21,25,31].

Current evidence trying to explain the pathogenesis of CMI can be attributed to magnetic resonance imaging (MRI) and suggests that overcrowding of the hindbrain by an underdeveloped PCF is the main cause of the CMI and explains the formation of syringomyelia. Recent morphometric studies focusing on the bony part of PCF in adult patients who had CMI have lent support to this hypothesis [21,25,31,35,37,41].

* Corresponding author. P.K: 4, Cerrahpasa, 34301, Istanbul, Turkey. Tel.: +90 212 414 3427; fax: +90 212 414 3427.

E-mail address: tanerato2000@yahoo.com (M.Y. Kaynar).

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In the present study, we measured bony elements that formed in the PCF of adults with CMI and mean to serve as a contribution to the current literature. The measurements taken from the patients were compared to the control group which included healthy subjects, and we discussed our findings in the light of pertinent literature.

2. Patients and methods

2.1. Patients

We retrospectively evaluated 97 adult patients who had been treated in our neurosurgical department due to CMI between January 1992 and January 2003. Sixty patients, whose preoperative radiological studies were available, served as subjects in this study. All patients included in this study had undergone surgical treatment for CMI with or without syringomyelia. Thirty adult individuals who had undergone MRI for the complaints of headache and showed no cranial or intracranial pathology were chosen as the control group.

2.2. Morphological features of PCF

The following measurements were made in midline sagittal MRI from 60 patients with CMI and 30 control subjects (Fig. 1): (1) the line from the center of the internal occipital protuberance to the opisthion measures the length of the supraocciput (C,D); (2) the anteroposterior diameter of the foramen magnum was measured from the basion to the opisthion (B,C); (3) the length of the clivus was measured from the top of the dorsum sella to the basion (A,B); (4) the anteroposterior diameter of the PCF was measured along a line parallel to the plane of the foramen magnum, from the top of the dorsum sella to a point 1 cm above the internal occipital protuberance (A,F); (5) the line from the superior border of the PCF (splenium of corpus callosum) perpendicular to the plane of the foramen magnum was accepted as the height of the PCF (G,H).

2.3. Cerebellar tonsils

The degree of the cerebellar tonsils was measured in a manner similar to that used by others [22,32]. Simply, the line between basion and opisthion was accepted as the plane of the foramen magnum. Tonsillar herniation was then evaluated by measuring a line perpendicular from this line to the most inferior aspect of the tonsils. In this study, tonsillar herniation was defined as downward herniation of at least 5 mm below the foramen magnum.

2.4. Statistical analysis

We performed statistical analysis by using SPSS for Windows (version 11; SPSS, Inc, Chicago, Ill). Mean values were presented with their standard deviations. The values of all parameters measured for the patient and the control groups were assessed using the Mann-Whitney *U* test. The result was accepted as statistically significant if the *P* value was less than .005.

3. Results

3.1. Clinical presentations

Table 1 displays the variations in the clinical symptoms in patients with CMI. The patient group consisted of 36 women and 24 men, with a mean age of 35.1 ± 12.7 years (range, 25-65 years). The majority of patients (39 patients) reported histories of lifelong complaints such as headache radiating to the neck. Patients with syringomyelia received diagnoses earlier because of the early presentation of various degrees of paresthesias on the extremities that forced the patient to search for medical attention.

3.2. Clinical syndrome

The majority of patients with CMI had headache that radiated to the neck and shoulders. This symptom was commonly accompanied with disequilibrium. The headache was accentuated by physical exertion and sudden changes in posture. One patient showed ocular disturbance as diplopia

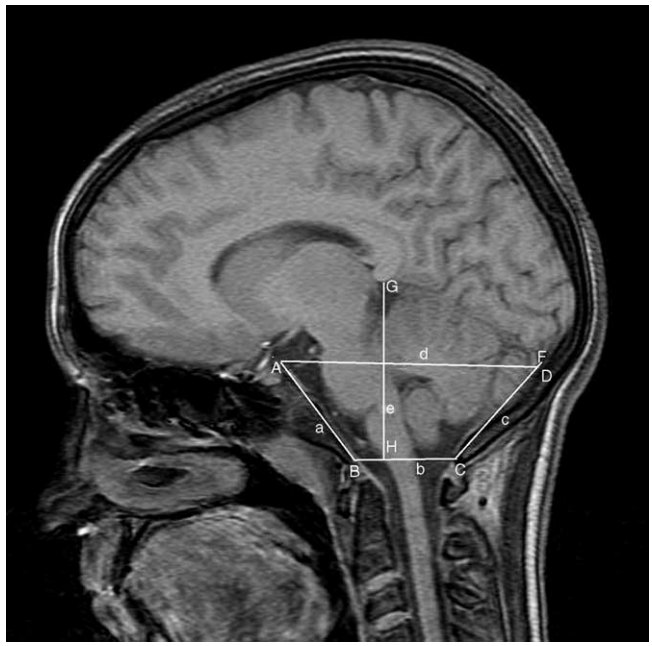

Fig. 1. Demonstration of the measurements of PCF made on T1-weighted mid-sagittal MRI of healthy subjects. The length of the clivus (a) was measured along a line drawn from the top of the dorsum sella (A) to the basion (B). The anteroposterior diameter of the foramen magnum (b) was measured from the basion (B) to the opisthion (C). The length of the supraocciput (c) was measured along a line drawn from the opisthion (C) to the center of the internal occipital protuberance (D). The anteroposterior diameter of the PCF (d) was measured along a line parallel to the plane of the foramen magnum, from the top of the dorsum sella (A) to a point 1 cm above the internal occipital protuberance (F). The height of PCF (e) was measured with a line drawn from the inferior aspect of the splenium (G) perpendicular to the plane of the foramen magnum (H).

and one showed difficulty in swallowing in addition to quadriparesia.

Forty-six patients (76.6%) with syringomyelia disclosed disturbances in spinal cord function that led to the symptoms typical for syringomyelia [6,7,18,21,25,26,31]. The severity of symptoms in this group varied with the extension of the syrinx cavity. In 14 patients without syringomyelia, the most common symptoms were muscular weakness, paresthesia/hyperesthesia, and nonradicular segmental pain.

3.3. MRI findings

The most common radiological finding was the compression of the cerebrospinal fluid (CSF) spaces due to herniation of the cerebellar tonsils. All patients in this study showed various degrees of tonsil herniation, measuring 5 to 38 mm. Other findings included reduced length of the clivus, reduced length of the supraocciput, reduced height of the PCF, reduced anteroposterior diameter of the PCF, and increased anteroposterior diameter of the foramen magnum. Minor findings included hydrocephalus and basilar invagination. Spinal abnormalities associated with CMI included syringomyelia (46 patients), cervical kyphosis (1 patient), Dandy-Walker syndrome (1 patient).

3.4. Statistical results

This study compared the parameters between 60 adult patients with CMI and 30 healthy subjects. The patient group consisted of 36 women and 24 men, with a mean age of 35.1 ± 12.7 years. The control group consisted of 15 women and 15 men, with a mean age of 52.2 ± 18.2 years. There is statistically significant difference between 2 groups regarding the age (P = .00001). There was no significant difference when regarding sex (P = .4). Table 2 summarizes statistical results of the parameters evaluated in this study. For patients with CMI, the following statistically significant abnormalities were demonstrated: reduced mean length of the clivus (P = .00001), reduced mean anteroposterior diameter of the PCF (P = .00001), reduced mean length of the PCF (P = .00001), reduced mean height of the supraocciput with no significant difference was found (P = .008), and increased mean anteroposterior diameter foramen magnum (P = .00001).

Table 1
Characteristics of the patients and clinical presentations

Variable	CMI (n = 60)
Sex (M/F)	24/36
Age (y)	35.1 ± 12.7
Clinical presentation	
Lifelong complaints	39
Age of onset	22.5 ± 10.3
Precipitating factors	
None	50
Trauma	5
Coughing or sneezing	3
Pregnancy	2

Values are means ± SDs.

Table 2
Summary of the statistical results of measurements made on MRI

Variable	Patients (n = 60)	Control (n = 30)	P
Age (y)	35.1 ± 12.7	52.2 ± 18.2	<.001
Sex (M/F)	24/36	15/15	.4
Measurements (mm)			
Supraocciput	42.1 ± 9.3	46.7 ± 4.3	.008
Foramen magnum[a]	31.7 ± 6.1	25.2 ± 3.8	<.001
Clivus	39.0 ± 7.7	48.4 ± 4.9	<.001
Posterior fossa[b]	60.4 ± 10.6	74.7 ± 3.5	<.001
Height[c]	124.7 ± 15.7	141.2 ± 6.8	<.001

Values are means ± SDs.
P values are for comparisons between patients and control (Mann-Whitney U test).
[a] Anteroposterior diameter of the foramen magnum.
[b] Anteroposterior diameter of the posterior fossa.
[c] Height of the posterior fossa.

4. Discussion

The overcrowding of PCF was estimated by some bone measurements on MRI in this study, and we supported the current notion that hypoplasia of the bony structures involved in the formation of PCF is the main cause of CMs in adults with no neural structural anomalies were noted. We found 2 embryological parts of the occipital bone, namely, supraocciput and exocciput, that were shorter than that of control. The height of PCF was also shorter. The bony structures that underdeveloped in the intrauterine life lead to downward herniation of the PCF contents as the caudal hindbrain develops normally. This, in turn, serves to broaden the anteroposterior diameter of the foramen magnum. It can be inferred that herniation of the cerebellar tonsils into the spinal canal through the foramen magnum obstructs the normal anatomical path by which CSF circulates. As a result, the formation of syringomyelia is likely, and in this study, in the majority of the patients, CMIs were associated with various degrees of syringomyelia. Studies measuring the flow of CSF at the foramen magnum have demonstrated that blockage of CSF at this gate between the intracranial and intraspinal subarachnoid space is the most important contributing factor for the formation of syringomyelia [4,11,14,33].

Morphometric studies have shown clearly that significantly smaller volume, or underdeveloped bony parts of the PCF in adult [21,25,31,35,37,41] and in pediatric patients [40,44], are the main causes for the downward herniation of the normally developing caudal hindbrain. Furthermore, a smaller upper part of the PCF has also been demonstrated in adult patients with CMs [31].

When we looked into the incidence of syrinx and basilar invagination in patients with adult-type CMIs, we found that 46 had syrinx cavity with a variety of degree and 6 had

basilar invagination. The width of the syrinx cavity ranged from 2 to 18 mm. The majority of syrinx cavity was found in the cervicothoracic region (24 patients) of the spinal cord.

Other MRI findings provided substantial evidence of PCF overcrowding. The most constant finding was compression of the CSF spaces, posterior and lateral to the cerebellum (60 patients, 100%). Tonsillar herniation, at least 5 mm below the plane of the foramen magnum, was also found in all patients. The magnitude of tonsillar ectopia ranged from 5 to 38 mm (mean, 12.6 mm).

Knowledge regarding the epidemiological features of CMI is limited; however, it has been defined as a rare congenital disorder [30]. The literature says female subjects outnumber male subjects by a wide margin as in this study [25,31,34]. The most common precipitating factor has been said to be whiplash injuries and direct blow to the head and neck [19,25], and we cannot state that trauma is the most common precipitating factor in this study because the majority of patients (50 patients) had no precipitating factors.

The bulk of the papers in the literature have documented the complex symptom patterns for CMI, headache being the most common as in this report [7,8,10,15,16,20,21,23,25,27,31,38,39,42]. There is evidence that a large number of the symptoms in patients with CMI are CSF-related [2,3,17]. Our study, in consistency with the literature, illustrated that the most obvious CSF-related symptoms were those attributed to the presence of syringomyelia [14,24,25,43]. This is because syringomyelia causes stretching and distension of the neural tissue of dissection of the central canal cavities into the parenchyma of the spinal cord. Current evidence suggests that formation of the syringomyelia is a direct result of the obstruction of CSF flow at the foramen magnum, which causes an exacerbation of the pulsatile systolic pulse wave in the spinal subarachnoid space and drives CSF through perivascular and interstitial space into the central canal of the spinal cord [28,29,33,36]. All patients (46 patients, 76.6%) with syringomyelia in this study had spinal cord disturbances. Our observation also showed that patients with tonsillar herniation can be asymptomatic; therefore, the severity of the symptoms is not directly related to the degree of tonsillar herniation, and this finding could not confirm some reports [1,5,38] because tonsillar herniation of at least 5 mm can be encountered as an incidental finding in asymptomatic patients with CMI [12,21].

5. Conclusions

Our results led us to conclude that underdevelopment of occipital somite originating from the paraxial mesoderm during the intrauterine life is the main cause of CMs. Downward herniation of normal-sized hindbrain is secondarily induced by overcrowding in the PCF. Displacement of CSF probably contributes to symptoms, and formation of the syringomyelia is directly a result of the obstruction at the foramen magnum. Nevertheless, our understanding of the pathophysiology of this disease remains frustratingly incomplete. Considerable effort should be spent to understand the pathogenesis and progression with a large patient series in the future.

References

[1] Aboulezz AO, Sartor K, Geyer CA, et al. Position of cerebellar tonsils in the normal population and in patients with Chiari malformation: a quantitative approach with MR imaging. J Comput Assist Tomogr 1985;9:1033-6.
[2] Alford B. Meniere's disease: criteria for diagnosis and evaluation of therapy for reporting—report of subcommittee on equilibrium and its measurement. Trans Am Acad Ophthalmol Otolaryngol 1972;76:1462-4.
[3] Alvarez D, Requena I, Arias M, et al. Acute respiratory failure as the first sign of Arnold-Chiari malformation associated with syringomyelia. Eur Respir J 1995;8:661-3.
[4] Ball MJ, Dayan AD. Pathogenesis of syringomyelia. Lancet 1972;2:799-801.
[5] Ball WS, Crone KR. Chiari I malformation: from Dr Chiari to MR imaging. Radiology 1995;195:602-4.
[6] Bejjani GK. Definition of the adult Chiari malformation: a brief historical overview. Neurosurg Focus 2001;11:1-8.
[7] Bindal AK, Dunsker SB, Tew Jr JM. Chiari I malformation: classification and management. Neurosurgery 1995;37:1069-74.
[8] Birns JW. An unusual form of laryngeal paralysis associated Arnold-Chiari malformations. Ann Otol Rhinol Laryngol 1984;93:447-51.
[9] Chiari H. Concerning alterations in the cerebellum resulting from cerebral hydrocephalus. Pediatr Neurosci 1987;13:3-8.
[10] Da Silva JA, Brito JC, Da Nobrega PV. Autonomic nervous system disorders in 230 cases of basilar impression and Arnold-Chiari deformity. Neurochirurgia 1992;35:183-8.
[11] Du Boulay G, Shah SH, Currie JC, et al. The mechanism of hydromyelia in Chiari type I malformations. Br J Radiol 1974;47:579-87.
[12] Elster AD, Chen MY. Chiari I malformations: clinical and radiologic reappraisal. Radiology 1992;183:347-53.
[13] Friede RL, Roessmann U. Chronic tonsillar herniation. An attempt at classifying chronic herniations at the foramen magnum. Acta Neuropathol 1976;34:219-35.
[14] Gardner WJ, Goodall RJ. The surgical management of Arnold-Chiari malformation in adults. An explanation of its mechanism and importance of encephalography in diagnosis. J Neurosurg 1950;7:199-206.
[15] Gingold SI, Winfield JA. Oscillopsia and primary cerebellar ectopia: case report and review of the literature. Neurosurgery 1992;29:932-6.
[16] Hendrix RA, Bacon CK, Sclafani AP. Chiari I malformation associated with asymmetric sensorineural hearing loss. J Otolaryngol 1992;21:102-7.
[17] House WF. Meniere's disease: management and theory. Otolaryngol Clin North Am 1975;8:515-35.
[18] Levy WJ, Mason L, Hahn JF. Chiari malformation presenting in adults: a surgical experience in 127 cases. Neurosurgery 1983;12:377-90.
[19] Mampalam TJ, Andrews BT, Gelb D, et al. Presentation of type I Chiari malformation after head trauma. Neurosurgery 1988;23:783-98.
[20] Maroun FB, Jacob JC, Mangan M. The Chiari malformation in adults. Can J Neurol Sci 1975;2:115-20.
[21] Meadows J, Kraut M, Guarnieri M, et al. Asymptomatic Chiari type I malformations identified on magnetic resonance imaging. J Neurosurg 2000;92:920-6.
[22] Mikulis DJ, Diaz O, Egglin TK, et al. Variance of the position of the cerebellar tonsils with age: preliminary report. Radiology 1992;183:725-8.

[23] Milhorat TH, Capocelli Jr AL, Anzil AP, et al. Clinicopathological correlations in syringomyelia using axial magnetic resonance imaging. Neurosurgery 1995;37:206-13.
[24] Milhorat TH, Capocelli Jr AL, Anzil AP. Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. J Neurosurg 1995;82:802-12.
[25] Milhorat TH, Chou MW, Trinidad EM, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 1999;44:1005-17.
[26] Milhorat TH, Kotzen RM, Anzil AP. Stenosis of central canal of spinal cord in man: incidence and pathological findings in 232 autopsy cases. J Neurosurg 1994;80:716-22.
[27] Milhorat TH, Kotzen RM, Mu HTM, et al. Dysesthetic pain in patients with syringomyelia. Neurosurgery 1996;38:940-7.
[28] Milhorat TH, Nobandegani F, Miller JI, et al. Noncommunicating syringomyelia following occlusion of the central canal in rats: experimental model and histological findings. J Neurosurg 1993; 78:274-9.
[29] Milhorat TH, Nakamura S, Heger I, et al. Ultrastructural evidence of sink function of central canal of spinal cord as demonstrated by clearance of horseradish peroxidase. Electron Microsc Soc Am Proc 1992;50:700-1.
[30] National Organization of Rare Diseases. Directory. New Fairfield (Conn): National Organization of Rare Diseases; 1997.
[31] Nishikawa M, Sakamoto H, Hakuba A, et al. Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg 1997;86:40-7.
[32] O'Connor S, du Boulay G, Logue V. The normal position of the cerebellar tonsils as demonstrated by myelography. J Neurosurg 1973;39:387-9.
[33] Oldfield EH, Muraszko K, Shawker TH, et al. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. J Neurosurg 1994; 80:3-15.
[34] Pillay PK, Awad IA, Little JR, Hahn JF. Symptomatic Chiari malformation in adults: a new classification based on magnetic resonance imaging with clinical and prognostic significance. Neurosurgery 1991;28:639-45.
[35] Schady W, Medcalfe RA, Butler F. The incidence of craniocervical bony anomalies in the adult Chiari malformation. J Neurosurg 1987; 82:193-203.
[36] Stoodley MA, Brown SA, Brown CJ, et al. Arterial pulsation-dependent perivascular cerebrospinal fluid flow into the central canal in the sheep spinal cord. J Neurosurg 1997;86:686-93.
[37] Stovner LJ, Bergan U, Nilsen G, et al. Posterior cranial fossa dimensions in the Chiari I malformation: relation to pathogenesis and clinical presentation. Neuroradiology 1993;35:113-8.
[38] Stovner LJ, Kruszewski P, Shen JM. Sinus arrhythmia and pupil size in Chiari I malformation: evidence of autonomic dysfunction. Funct Neurol 1993;8:251-7.
[39] Sullivan LP, Stears JC, Ringel SP. Resolution of syringomyelia and Chiari I malformation by ventriculoatrial shunting in patient with pseudotumor cerebri and a lumboperitoneal shunt. Neurosurgery 1988;22:744-7.
[40] Tubbs RS, Elton S, Grabb P, et al. Analysis of the posterior fossa in children with the Chiari 0 malformation. Neurosurgery 2001;48: 1050-5.
[41] Vega A, Quintana F, Berciano J. Basichondrocranium anomalies in adult Chiari type I malformation: a morphometric study. J Neurol Sci 1990;99:137-45.
[42] Weig PC, Buckthal PE, Choi SK, et al. Recurrent syncope as the presenting symptom of Arnold-Chiari malformation. Neurology 1991;41:1673-4.
[43] Willams B. On the pathogenesis of syringomyelia: a review. J R Soc Med 1980;73:798-806.
[44] Wu YW, Chin CT, Chan KM, et al. Pediatric Chiari I malformations: do clinical and radiological features correlate? Neurology 1999; 53:1271-6.

Commentary

There is accumulating evidence that CMI is a disorder of the para-axial mesoderm that results in underdevelopment of the PCF and overcrowding of the normally developed hindbrain. In this study, the authors compared linear measurements of the PCF, as determined by MRI, in 60 adult patients with tonsillar herniation of at least 5 mm and 30 adult control subjects. Patients with CMI were found to have the following distinctive findings: reduced mean length of the clivus ($P < .001$), reduced mean length of the supraocciput ($P < .008$), reduced mean height of the PCF ($P < .001$), reduced mean anteroposterior diameter of the PCF ($P < .001$), and increased mean anteroposterior diameter of the foramen magnum ($P < .001$). The most constant radiological finding was compression of the CSF spaces posterior and lateral to the cerebellum (60/60 patients, 100%).

In general, the findings in the current study are consistent with previously published data and support the concept that osseous dysplasia, volumetric reduction of the PCF, and hindbrain overcrowding are fundamental features of CMI. This study is a useful contribution to the literature.

Thomas H. Milhorat, MD
Department of Neurosurgery
North Shore University Hospital
Manhasset, NY 11030, USA