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MORPHOMETRIC ANALYSIS OF THE POSTERIOR CRANIAL FOSSA IN HEALTHY ADULT POPULATION AND PATIENTS WITH CHIARI MALFORMATION

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ABSTRACT

Aim: To analyse the mophometric parameters of the posterior cranial fossa along with the brainstem and cerebellum in patients with Chiari malformation. This study is an attempt to analyse the morphology and dimensions of the bony and neural structures of the PCF in the Indian adult population and to study the correlation between the analysed parameters and tonsillar herniation.

Introduction: Chiari malformation is a hindbrain malformation characterized by a downward herniation of the caudal part of the cerebellum (tonsil) and/or medulla oblongata into the spinal canal. This leads to a spectrum of clinical symptoms such as occipital headache, neck pain, along with cerebellar, bulbar, ocular and sensorymotor symptoms.

Materials and Methods: 30 patients with Chiari Malformation and 44 control subjects were enrolled in the study. MRI brain was done in all the patients. Control subjects underwent MRI for unrelated problems such as migraine. Posterior fossa volume, Length of basiocciput, length of the brainstem, length of the cerebellum, tonsillar descent and compression of the CSF cisterns at the level of the foramen magnum were measured and analysed with the aid of SPSS software for Windows (version 11, SPSS, Inc).

Results: Therewas a significant reduction of posterior cranial fossa volume as well as compression of the CSF citerns at the level of the foramen magnum in patients with Chiari Malformation.

Conclusion: This study reestablishes the fact that Chiari Malformation – I is a disorder of the mesoderm and the fundamental problem is a volumetrically small posterior cranial fossa which results in varying degrees of hindbrain overcrowding and results in tonsilar descent and CSF flow abnormalities leading to syringomyelia and hydrocephalus and the resultant symptoms

KEYWORDS : Chiari Malformation, Morphometric, Posterior fossa volume

INTRODUCTION

Chiari malformation is a hindbrain malformation characterized by a downward herniation of the caudal part of the cerebellum (tonsil) and/or medulla oblongata into the spinal canal. This leads to a spectrum of clinical symptoms such as occipital headache, neck pain, along with cerebellar, bulbar, ocular and sensorymotor symptoms. More cases of Chiari malformation (CM-I) are being diagnosed recently with the advent of magnetic resonance imaging (MRI) of brain. Syringomyelia is a common finding in CM-I.

Experimental models[26,27,34] supported by morphometric studies[4,31,32,39,40,42] have shown that the chronic tonsillar herniation(CTH) occurring in classic CM-I mainly results from overcrowding of a normally developing hindbrain within a congenitally small and shallow posterior cranial fossa(PCF) due to occipital bone underdevelopment. The exact pathogenesis of CM-I is not clear. This also is associated with the development of Hydrocephalus and Syringomyelia due to the block in normal CSF circulation Recent morphometric studies focussing on the bony part of the posterior cranial fossa in adult patients with Chiari malformation have lent support to this hypothesis[39,42,45] There has been no morphometric study of the PCF from the Indian adult population.

MATERIALS AND METHODS

In this study an analysis of the morphometry of posterior cranial fossa in 30 patients with Chiari I malformation who were symptomatic and 44 healthy controls was done.

All patients who were referred to the Institute of Neurology between August 2011 and March 2014, where all the three authors were working as Assistant Professors, were enrolled in the study. The control group included 44 patients who underwent brain MR imaging for headaches or migraine in the same period and whose MRI was normal. Control group included patients aged more than 16 years and were enrolled from the outpatient section and the radiology department. All postoperative patinets and those with basilar impression, craniosynostosis were exclude from the study.

METHODS

This study compared the parameters between 30 adult patients with CM-I and 44 healthy subjects. The patient group consisted of 11 women and 19 men, with a mean age of 30 years. The control group consisted of 33 women and 11 men, with a mean age of 36 years.

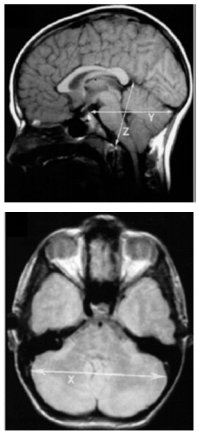
Twelve patients had CM-I only (40%), whereas eighteen patients had CM-I with syringomyelia (60%). Eleven patients presented with a history of paroxysmal occipital headaches. 11 patients with syringomyelia had radicular pains, wasting and thermoalgesic dissociation. 4 patients displayed bipyramidal signs and 4 patients had cerebellar symptoms and signs.

Using midsagittal MR imaging, the following linear measurements of the PCF were made

x - maximum width of bony posterior cranial fossa

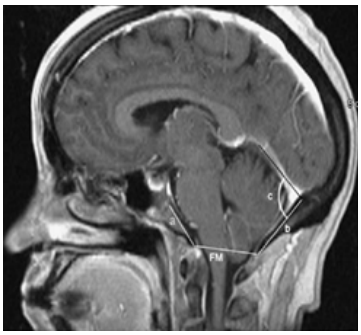
y - distance from posterior clinoid process to torcula heterophili

z - Height of Posterior Cranial fossa - from basion to peak of tentorium cerebelli

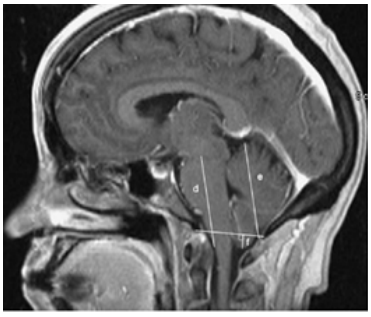


The volume of the posterior cranial fossa was calculated using the formula, **PCF Volume - 4/3 x (x/2 x y/2 x z/2)** The volume of PCF in the controls was calculated in a similar fashion.

The length of the basiocciput **(a)** was measured from the basioccipital synchondrosis to the basion. The basioccipital synchondrosis was clearly visualized in both the groups. The length of supraocciput **(b)** was measured from the internal occipital protuberance to opisthion. The tentorial angle **(c)** was the angle subtended by a line, connecting the internal occipital protuberance to opisthion, with the tentorium.



The brainstem length **(d)** was calculated from midbrain-pons junction to cervicomedullary junction. The cerebellar length **(e)** was the length of the line connecting the most rostral and caudal points of the cerebellar hemisphere along a straight line drawn caudally and parallel to the bottom of the fourth ventricle. The length of tonsillar descent **(f)** was measured by a perpendicular line from the McRae's line to the tip of the tonsil.



Compression of the CSF cisterns posterior and lateral to the cerebellum was identified by the absence of the hyperintense signal at the lowest point of the cerebellar hemisphere on sagittal T2-weighted MR images.

The accuracy of the formula was ascertained using 10 dry skulls, the posterior cranial fossa volume of which was calculated by filling it with sand. A CT was obtained of the dry skull and the above said linear measurements made and the volume calculated using the given formula. There was no statistically significant difference in the volume calculated by either means.

Statistical analysis was performed The mean differences in the linear measurements and volume of the PCF for controls and Chiari group was measured using independent-sample student's *t* tests. Significance was indicated by a two-tailed P value of less than 0.05.

Results:

Volume of Posterior cranial fossa: the mean volume was 239 ±17 cc whereas in the 44 control participants the average volume was 274 ± 23 cc. The volume of the posterior cranial fossa was significantly smaller in patients with a Chiari malformation as compared with normal controls ($p < 0.001$).

There was no significant difference in the width of the posterior cranial fossa or the posterior clinoid - torcular herophili distance; However, the height of the PCF (basion-peak of the tentorium cerebella) was significantly shorter in those with a Chiari malformation (6.58 ± 0.3 cm) as compared with controls (7.13 ± 0.33 cm; $p < 0.001$).

Tentorial angle: there was a significant difference (p - 0.001) in the mean angle of the cerebellar tentorium against Twining's line, which was 85.11^0 in the Chiari group and 84.32^0 in the control group. This indicated that the cerebellar tentorium was significantly steeper in the Chiari group than in the control group.

Length of the occiput: Although the mean length of the Basiocciput in the Chiari group was shorter than that in the control group (2.69 cm in the Chiari group and 2.73 cm in the control group), it was not statistically significant ($p > 0.05$).

Length of the supraocciput: No significant difference ($p > 0.05$) was found in the length of the supraocciput (between the internal occipital protuberance and the opisthion), which measured 40.9 mm(mean) in the Chiari group and 40.8 mm(mean) in the control group.

Length of the brainstem: no significant difference was found in the length of the brainstem from the midbrain-pons junction to the cervicomedullary junction (mean - 4.93 cm in the Chiari group and 4.87 cm in the control group) or in the long axial length of the cerebellar hemisphere (mean - 4.91 cm in the Chiari group and 4.94 cm in the control group).

Compression of the retrocerebellar CSF spaces of the cisterna magna was seen in 100% of our patients.

The mean values of the eight parameters measured for each group are shown in this Table :

In Chiari patients the mean value of tonsillar descent was 8.1 mm. In controls the tonsil was above the McRae's line. On analysing the relationship between the volume of PCF and the extent of tonsilllar

	Group Statistics				
	Group	**N**	**Mean**	**Std. Deviation**	**Std. Error Mean**
Volume	Control	44	274.03	23.44	3.53
	Chiari	30	239.65	17.61	3.22
Basiocciput	Control	44	2.73	0.40	0.06
	Chiari	30	2.69	0.31	0.06
Foramen Magnum	Control	44	3.41	0.33	0.05
	Chiari	30	3.45	0.27	0.05
Supraocciput	Control	44	4.08	0.12	0.02
	Chiari	30	4.09	0.13	0.02
Tentorial Angle	Control	44	84.32	0.85	0.13
	Chiari	30	85.11	1.11	0.20
Brainstem Length	Control	44	4.87	0.16	0.02
	Chiari	30	4.93	0.15	0.03
Cerebellar Length	Control	44	4.94	0.14	0.02
	Chiari	30	4.91	0.13	0.02
Height	Control	44	7.13	0.33	0.05
	Chiari	30	6.58	0.30	0.05

descent there was no relationship between the two parameters.

DISCUSSION

Various theories have been postulated to explain the pathogenesis of Chiari 1 malformation. Most accepted theory is a primarily small posterior cranial fossa. To establish this in the Indian population, an attempt has been made and various parameters have been analysed.

The length of the supraocciput was 4.08 cm in the control group and 4.09 cm in the patient group and was statistically insignificant on a gross comparative analysis and further gender analysis. These values are consistent with the ones reported , in Noudels study[38].

The present study also showed that the cerebellar tentorium in the Chiari group was significantly steeper than in the control group. The mean angle of the cerebellar tentorium was 85.11^0 in the Chiari group and 84.32^0 in the control group and the tentorium was significantly steeper on further gender analysis.

The mean **anteroposterior diameter of the foramen magnum** was 3.45 cm in the Chiari group and 3.41 cm in the control group and were statistically insignificant. These values compare well

with the AP diameter of foramen magnum in Noudel's study[38]. Aydin et al[4] have documented a AP diameter of 3.17 cm and 2.52 cm in Chiari group and Control group respectively. These were statistically significant. It has been shown that caudal displacement of the hindbrain can enlarge the foramen magnum .

In the present study **the mean axial lengths of the brainstem and cerebellum** were 4.87 cm and 4.94 cm in the control group. In the Chiari group the values were 4.93 cm and 4.91 cm respectively. These values compare well with the values documented by previous studies.

In Chiari patients the mean value **of tonsillar descent** was 8.1 mm . The mean value of tonsillar descent was 9.8 mm in Milhorat's study[31]. In the control group the tonsil was above the McRae's line. This is in discordance with Milhorat's study[31] in which the mean tonsillar descent was 2.1 mm in the Control group. The degree of tonsillar herniation does not seem to be representative of the cranioencephalic disproportion. In Nishikawa's study[33] no significant relationship was found between the volume of the herniated brain below the foramen magnum and the PCF volume or between the volume of the herniated brain below the foramen magnum and the volume ratio.

Our data failed to show any significant correlation between the extent of tonsillar ectopia and any other measured parameters of the PCF, as has Vega's[45] and Noudels[38] studies. However, Schady[39] et al have found an inverse relationship between the size of the PCF and the degree of cerebellar herniation, whereas Stovner et al.[42] have shown a strong positive correlation between the two parameters.

Furthermore, our results confirm that the most consistent MR imaging finding is **compression of the retrocerebellar CSF spaces of the cisterna magna** by the herniated tonsils, which provides substantial evidence of overcrowding. This finding has been documented by Milhorat et al[31], Aydin et al[4] and Noudel et al[38] .

In the present study, MRI findings of reduced height of the PCF and increased slope of the tentorium,are consistent with a defect of the para-axial mesoderm.

Milhorat et al. and Nishikawa et al.[31,32,33] have postulated that the fundamental pathogenic entity in CMI is most likely underdevelopment of the para-axial mesoderm resulting in posterior fossa hypoplasia with CSF flow abnormalities.

Since the volume of PCF showed a significant correlation it indicates that decompression of the posterior cranial fossa may relieve overcrowding of the posterior cranial fossa. It can be inferred that overcrowding of the posterior cranial fossa induces remodeling of neural structures as the cerebellar tentorium shifts upward and the cerebellar tonsils herniate to accommodate the growing brain, rather than remodeling the cranium.

Consequently, most clinical symptoms result from displacement of newly formed CSF from the subarachnoid spaces of the PCF into available spaces within the supratentorial and spinal compartments. Current evidence[38] suggests that hindbrain-related syringomyelia, observed in 60% of our patients, is also a complication resulting from obstructed CSF flow between the cranial and spinal compartments.

The considerable delay in the occurrence of neurological symptoms could be explained by the relatively late, mainly postnatal, growth spurt of the cerebellum within a small and inadequate PCF.

Thus, when paraxial mesodermal insufficiency is regarded as the pathogenesis of Chiari malformation, adult-type Chiari malformation can be considered a mild form and the pediatric type a severe form.

Badie et al. have demonstrated that patients with smaller posterior fossa volumes presented at a younger age and had a better response to surgery. Cardiac-gated phase-contrast cine MR can be a valuable tool in identifying patients who are less likely to respond to suboccipital decompression for CMI.

Morphometric and volumetric studies are useful tools in increasing the understanding of the pathophysiological conditions at play in the development of CTH. Morphometric parameters of the posterior cranial fossa could influence the natural history of patients with CMI as well as their prognosis after surgical treatment and could reduce the risk of postoperative complications such as cerebellar ptosis.

Further studies are needed to identify preoperatively the steps of the surgical treatment and the bone resection needed for decompression of the posterior fossa in the individual. This is the first morphometric study of the PCF in the Indian adult population. A more extensive study, using repeatable measures with a larger number of patients is necessary to confirm the results of this study and evaluate their further application.

However this is only a preliminary study with a relatively small sample size and a larger study with more number of patients will throw more light into the pathogenesis of CM-I. This study has been done only in adult population. A separate study is needed for pediatric population with CM-I.

CONCLUSION

This study reestablishes the fact that Chiari Malformation – I is a disorder of the mesoderm and the fundamental problem is a volumetrically small posterior cranial fossa which results in varying degrees of hindbrain overcrowding and results in tonsilar descent and CSF flow abnormalities leading to syringomyelia and hydrocephalus and the related symptoms.

BIBLIOGRAPHY

1. Ali MM, Russell N, Awada A, McLean D: A cranio-cervical malformation presenting as acute respiratory failure. J Emerg Med 14:569-572, 1996.
2. Alvarez D, Requena I, Arias M, Valdes L, Pereiro I, De la Torre R: Acute respiratory failure as the first sign of Arnold-Chiari malformation associated with syringomyelia. Eur Respir J 8:661-663, 1995.
3. Arcaya J, Cacho J, Del Campo F, Grande J, Maillo A: Arnold-Chiari malformation associated with sleep apnea and central dysregulation of arterial pressure. Acta Neurol Scand 88:224-226, 1993.
4. Aydin S, Hanimoglu H, Tanriverdi T, Yentur E, Kaynar MY: Banerji NK, Millar JHD: Chiari malformation presenting in adult life. Brain 97:157-168, 1974.
5. Barry A, Patten BM, Stewart BH: Possible factors in the development of the Arnold-Chiari malformation. J Neurosurg 14:285-301,1957.
6. Barton JJ, Sharpe JA: Oscillopsia and horizontal nystagmus with accelerating slow phases following lumbar puncture in the Arnold-Chiari malformation. Ann Neurol 33:418-421, 1993.
7. Batzdorf U: Chiari I malformation with syringomyelia. Evaluation of surgical therapy by magnetic resonance imaging. J Neurosurg 68:726-730,1988.
8. Bindal AK, Dunsker SB, Tew JM Jr: Chiari I malformation: classification and management.Neurosurgery 37:1069-1074,1995.
9. Birns JW: An unusual form of laryngeal paralysis associated with Arnold-Chiari malformations. Ann Otol Rhinol Laryngol 93:447-451, 1984.
10. Chiari H: Concerning alterations in the cerebellum resulting from cerebral hydrocephalus. PediatrNeurosci 13:38,1987.
11. Chiari type I malformations in adult: a morphometric analysis of the posterior cranial fossa. Surg Neurol 64:237–241,2005.
12. Da Silva JA, Brito JC, Da Nobrega PV: Autonomic nervous system disorders in 230 cases of basilar impression and Arnold-Chiari deformity. Neurochirurgia 35:183-188, 1992.
13. Du Boulay G, Shah SH, Currie JC, et al: The mechanism of hydromyelia in Chiari type 1 malformations. Br J Radiol 47:579-587,1974.
14. Dyste GN, Menezes AH, VanGilder JC: Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. J Neurosurg 71:159-168,1989.
15. Dyste GN, Menezes AH, VanGilder JC: Symptomatic Chiari malformations: An analysis of presentation, management, and long-term outcome. J Neurosurg 71:159-168, 1989.
16. Friede RL, Roessmann U: Chronic tonsillar herniation. An attempt at classifying chronic herniations at the foramen magnum. Acta Neuropathol 34:219-235,1976.
17. Gamache FW, Ducker TB: Syringomyelia: A neurological and surgical spectrum. J Spinal Disord 3:293-298, 1990.
18. Gardner WJ, Goodall RJ: The surgical treatment of Arnold-Chiari malformation in adults. An explanation of its mechanism and importance of encephalography in diagnosis. J Neurosurg 7:199-206,1950.
19. Gingold SI, Winfield JA: Oscillopsia and primary cerebellar ectopia: Case report and review of the literatureNeurosurgery 29:932-936, 1992.
20. Hendrix RA, Bacon CK, Sclafani AP: Chiari I malformation associated with asymmetric sensorineural hearing loss. J Otolaryngol 21:102-107, 1992.
21. Jeremy Greenlee, P. Charles Garell, Nicholas Stence, Arnold H. Menezes. Comprehensive approach to Chiari malformation in pediatric Patients . Neurosurg Focus Article 4, 1999.
22. Johnson GD, Harbaugh RE, Lenz SB: Surgical decompression of Chiari I malformation for isolated progressive sensorineural hearing loss. Am J Otol 15:634-638, 1994.
23. Khurana RK: Headache spectrum in Arnold-Chiari malformation. Headache 31:151-155, 1991.
24. Levy WJ, Mason L, Hahn JF: Chiari malformation presenting in adults: a surgical experience in 127 cases. Neurosurgery 12:377-390,1983.
25. Lewis AR, Kline LB, Sharpe JA: Acquired esotropia due to Arnold-Chiari I malformation. J Neuroophthalmol 16:49-54, 1996.
26. Marin-Padilla M, Marin-Padilla TM: Morphogenesis of experimentally induced Arnold-Chiari malformation. J NeurolSci 50:29–55, 1981.
27. Marin-Padilla M: Cephalic axial skeletal-neural dysraphic disorders: embryology and pathology. Can J Neurol Sci 18:153–169, 1991.
28. Maroun FB, Jacob JC, Mangan M: The Chiari malformation in adults. Can J Neurol Sci 2:115-120, 1975.
29. McLone DG, Knepper PA: The cause of Chiari II malformation: a unified theory. Pediatr Neurosci15:112,1989.
30. Melissa A. Parisid , William B. Dobyns Human malformations of the midbrain and hindbrain: review and proposed classification scheme Molecular Genetics and

Metabolism p.36-53 Vol.80 2003.
31. Milhorat TH, Chou MW, Trinitad EM, Kula RW, Mandell M,Wolpert C, et al: Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 44:1005–1017, 1999.
32. Milhorat TH: Chiari and hindbrain-related syringomyelia. Br J Neurosurg 21:465–466, 2007.
33. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y:Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg 86:40–47, 1997.
34. O'Rahilly R, Müller F: The early development of hypoglossal nerve and occipital somites in human staged embryos. Am J Anat 169:237–257, 1984.
35. Paul KS, Lye RH, Strang FA, et al: Arnold-Chiari malformation. Review of 71 cases. J Neurosurg 58:183-187,1983.
36. 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 28:639-645, 1991.
37. R. Shane Tubbs, Mark Hill, Marios Loukas, Mohammadali M. Shoja,W. Jerry Oakes Volumetric analysis of the posterior cranial fossa in a family with four generations of the Chiari malformation Type I J Neurosurg January 2008 Volume 1.
38. Rémy Noudel, Nicolas Jovenin,Cristophe Eap,Bern ard Scherpereel, Laurent Pierot,Pascal Rousseaux. Incidence of basioccipital hypoplasia in Chiari malformation Type I: comparative morphometric study of the posterior cranial fossa J Neurosurg 111:1046–1052, 2009.
39. Schady W, Metcalfe RA, Butler P: The incidence of craniocervical bony anomalies in the adult Chiari malformation. J Neurol Sci 82:193–203, 1987.
40. Sekula RF Jr, Janetta PJ, Casey KF, Marchan EM, Sekula LK,McCrady CS: Dimensions of the posterior fossa in patients symptomatic for Chiari I malformation but without cerebellar tonsillar descent. Cerebrospinal Fluid Res 2:11, 2005.
41. Spooner JW, Baloh RW: Arnold-Chiari malformation: Improvement in eye movements after surgical treatment. Brain 104:51-60, 1981.
42. Stovner LJ, Bergan U, Nilsen G, Sjaastad O: Posterior cranial fossa dimensions in the Chiari I malformation: relation to pathogenesis and clinical presentation. Neuroradiology 35:113–118, 1993.
43. Sunil V. Furtado, Darpan J. Thakre, Prasanna K. Venkatesh, Kalyan Reddy,A. S. Hegde Morphometric analysis of foramen magnum dimensions and intracranial volume in pediatric Chiari I malformation Acta Neurochirurgica Volume 152, February, 2010.
44. Tubbs, R. Shane; Griessenauer, Christoph J. Loukas, Marios Shoja, Mohammadali M. Cohen-Gadol, Aaron A. Morphometric Analysis of the Foramen Magnum: An Anatomic Study Neurosurgery: p 385–388 Volume 66 February 2010.
45. Vega A, Quintana F, Berciano J: Basichondrocranium anomalies in adult Chiari type I malformation:a morphometric study. J Neurol Sci 9:137-145,1990.