Original Article

JASI

Association of Chiari Type 1 Malformation and Cervical Spine Curve Changes

Abstract

Introduction: In this study, we aimed to examine the association of cervical spine curve abnormalities (loss of cervical lordosis or reversal of cervical curve) with Chiari Type 1 malformation (CM1). Further, a possible relation of syrinx formation in the cervical spinal cord and disc protrusion with CM1 was analyzed. **Material and Methods:** Cervical spinal magnetic resonance imagings of 998 patients were retrospectively screened for the presence of CM1. The frequency rates of syrinx formation within the spinal cord, cervical spinal curve changes, and cervical disc herniation among CM1+ and CM1− patients were compared. **Results:** Patients with CM1 have significantly higher rate of loss of cervical lordosis when compared with those who have not CM1. The syrinx formation rate was also found lower in the CM1+ patients with loss of cervical lordosis than in CM1+ patients with either normal cervical lordosis or reversed cervical curve. No significant difference was detected between CM1+ and CM1− patients regarding cervical disc herniation rate. **Discussion and Conclusion:** As the loss of cervical lordosis rate is higher in CM1, the patients with lateral X-ray findings of cervical lordosis flattening may be evaluated regarding typical neurological symptoms of syringomyelia.

Keywords: *Cervical lordosis, Chiari Type 1 malformation, magnetic resonance imaging, syrinx*

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Introduction

Chiari Type 1 malformation (CM1) is a cerebellar tonsil herniation of the foramen magnum that results in compression of the posterior cranial fossa structures or pathologic blockage of fourth ventricle cerebrospinal fluid (CSF) flow.[1] CM1 can lead to a syrinx formation within the spinal cord due to abnormal flow of the CSF at the foramen magnum.[2]

Cervical spinal curve changes including loss of cervical lordosis can cause neck pain which can be shown by various radiological modalities such as X-ray and magnetic resonance imaging (MRI).[3]

In this study, we aimed to examine the frequency of cervical curve changes in patients with CM1 by MRI. Further, association of the degree of tonsillar herniation and the frequency of syrinx formation were investigated.

Material and Methods

Patients

Medical records and radiological images of the patients ≥18 years old who admitted to our hospital with various complaints and underwent a nonenhanced cervical spine MRI between February 2018 and October 2018 were retrospectively screened on the hospital database. The patients with previous neck or cranial surgery history or rheumatic disease or previous severe trauma anamnesis or also congenital–neoplastic cranial or spinal cord diseases or craniocervical junction disorders were excluded from the study. The study was approved by the university research ethic committee with a number of 2018-18/157 dated October 9, 2018. The procedures followed in accordance with the ethical standards of the responsible committee and with the Helsinki Declaration of 1975, as revised in 2000.

Magnetic resonance imaging

The MRIs were acquired using a 1.5 T MRI system (GE Signa EXCITE™ 1.5 T MRI) with a field of view: 200, matrix: 256 × 256, FA 60°. Fast spin echo sequences were used to obtain T1- and T2-weighted images in sagittal and T2-weighted in axial planes. The images were transferred to a workstation and interpreted by two radiologists who have 5 and 6 years of experience of cervical spinal imaging.

Article Info

Received: 27 April 2020
Accepted: 25 July 2021
Available online: 23 September 2021

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Access this article online

Website: www.jasi.org.in

DOI: 10.4103/JASI.JASI_67_20

Quick Response Code:

How to cite this article: Alpaslan M, Özkaçmaz S, Dadalı Y, Uçar İ. Association of chiari type 1 malformation and cervical spine curve changes. J Anat Soc India 2021;70:162-7.

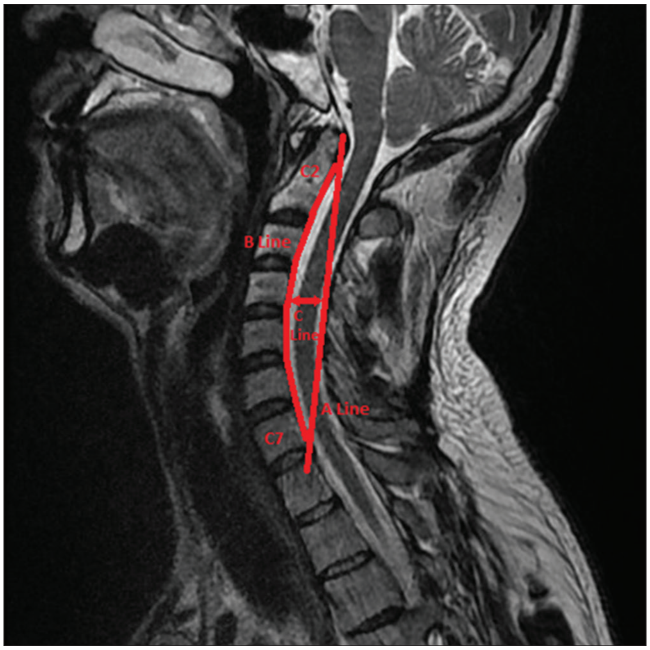

Figure 1: Measurement of C distance on sagittal T2‑weighted image

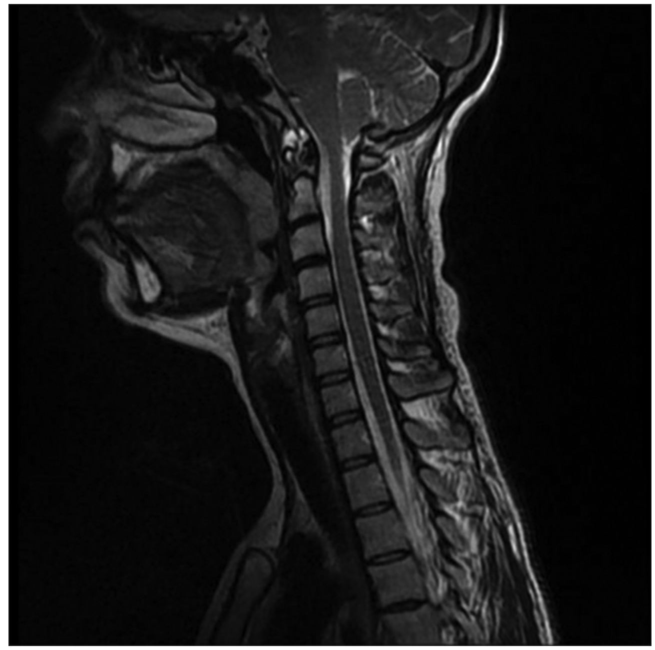

Figure 2: Chiari Type 1 malformation with loss of cervical lordosis (C distance is zero)

Figure 3: Chiari Type 1 malformation with reversed curve (C distance is negative)

Assessment of Chiari Type 1 malformation

For the diagnosis of CM1, measurement of how far cerebellar tonsils protrude below the inferior margins of foramen magnum was used. First, a line was drawn from anterior (basion) to posterior (opisthion) inner margins of the foramen magnum on sagittal T2‑weighted images. Inferior part of the cerebellar tonsils protruded below this line was also measured. The patient with a protruded tonsil part ≥5 mm was diagnosed as CM1 and the ones with <5 mm were identified as normal.[4] For the patients with different herniation sizes in two sides (right and left tonsils), the measurement of most herniated side was recorded.

Evaluation of cervical lordosis

A straight line (A) was drawn from the superior‑posterior aspect of the C2 odontoid process to the posterior‑inferior aspect of the corpus of the C7 vertebra. Second line (B) was traced along the posterior aspect of the intervening cervical vertebral bodies. A third line (C) intersects perpendicularly at the point of greatest distance between A and B. The length of C recorded in millimeters is the depth of the cervical lordosis [Figure 1]. When the C value was zero, it was termed as loss of cervical lordosis (cervical flattening) [Figure 2]. Positive C value measurements suggest normal lordosis [Figure 1] and negative values are identified as evidence of reversal of the lordotic curve [Figure 3].[5] We classified our patients to three groups according to the status of lordosis as patients with normal lordosis, with loss of cervical curve, and with reversed cervical curve.

Assessment of herniation

Discal herniation is defined for a localized displacement of disc material beyond the limits of the intervertebral disc space.[6]

According to this description, the patients were classified as without herniation, with only one‑level herniation, with two‑levels herniation, and with three or more levels of herniation by evaluating cervical spinal MRIs [Figure 4].

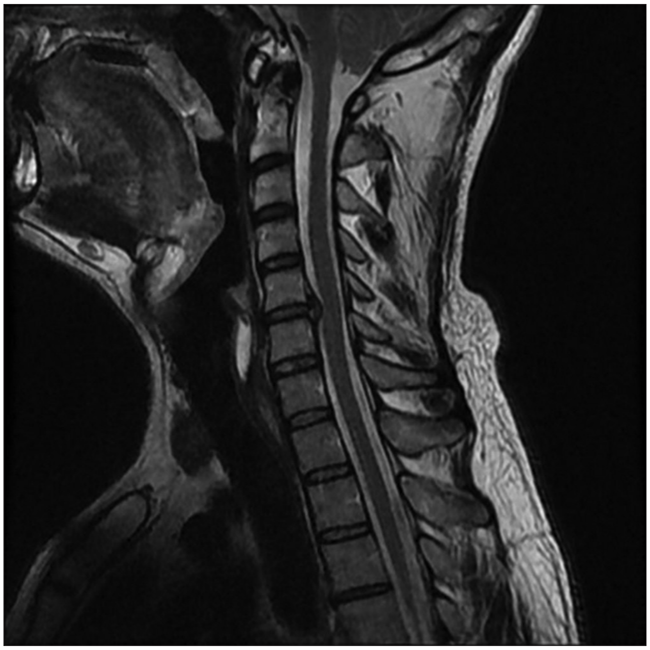

Figure 4: Chiari 1 malformation with one-level discal herniation

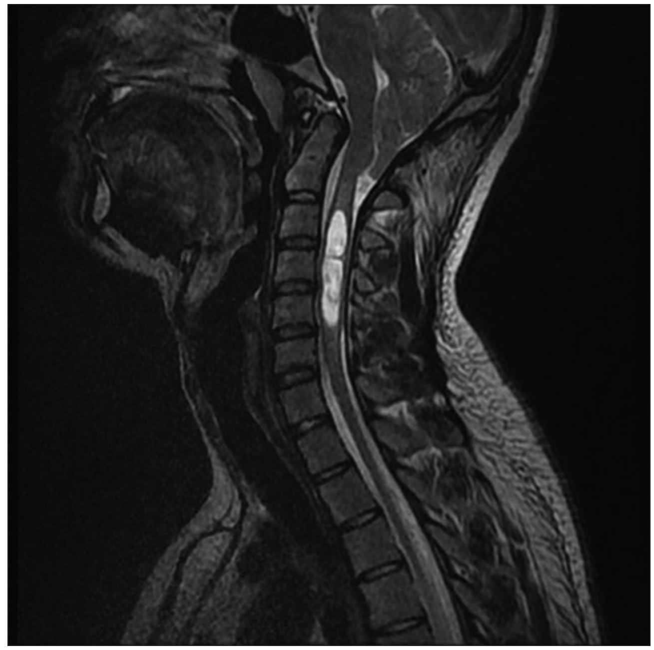

Figure 5: Chiari 1 malformation with syrinx formation

Table 1: Distribution of the patients to the groups according to gender

	Male		Female		Total	
	n	Rate (%)	*n*	Rate (%)	*n*	Rate (%)
Chiari Type 1 malformation +	13	4.58	22	3.08	35	3.51
Chiari Type 1 malformation −	271	95.42	692	96.91	963	96.49
Total	284	28.46	714	71.54	998	100

Table 2: Distribution of the patients to the groups according to cervical curve status

	Loss of cervical lordosis		Reversed cervical curve		Normal cervical lordosis		Total
	n	Rate (%)	*n*	Rate (%)	*n*	Rate (%)	
Chiari Type 1 malformation +	18	51.43	6	17.14	11	31.43	35
Chiari Type 1 malformation −	293	30.43	263	27.31	407	42.26	963
Total	311	31.16	269	26.95	418	41.88	998

Assessment of syrinx formation

A syrinx applies to a cavity within the spinal cord that may or may not communicate with the central canal.[7] It may be congenital (90%) or secondary (posttraumatic, postinflammatory, associated with spinal cord tumors and vascular insuffiency).[7] Patients who have a cavity within cervical spinal cord with a diameter of >2 mm[8] were classified as having syrinx cavity, while remained ones were identified as with no syrinx on MRIs [Figure 5].

Statistical analysis

Statistical Package for the Social Sciences version 21.0 software for Windows (IBM SPSS Statistics for Windows, Version 21.0. Armonk, NY: IBM Corp., USA) was used for statistical analysis. Descriptive statistics for variables were expressed as mean ± standard deviation and frequency (*n*, %). Statistical significance between groups was examined by Chi-square test. A $P \leq 0.05$ was considered statistically significant.

Results

A total of 998 patients (18–88 years old, mean age 47.7 ± 18.7 years) were included in this study. Among 714 females, 22 had CM1 malformation (3.08%), while 13 of 284 males (4.58%) CM1 was detected. Overall CM1 malformation rate was 3.51%. The CM1 malformation rate of males was mildly higher than females, but the difference was not statistically significant [Table 1].

Loss of cervical lordosis was detected in 18 (51.43%) and reversed cervical curve in 6 patients (17.14%) among 35 patients with CM1 (Mean age 44.1 years). Normal cervical lordosis was seen in remaining 11 patients (31.43%).

Loss of cervical lordosis was detected in 293 (30.43%) and reversed cervical curve in 263 patients (27.31%) among 963 patients without CM1 (Mean age 47.8 years).Normal cervical lordosis was seen in remaining 407 ones (42.26%).

Patients with CM1 had statistically significant higher

Table 3: Distribution of the patients to the groups according to discal protrusion

	No protrusion		Protrusion in one level		Protrusion in two levels		Protrusion in three or more levels		Total
	n	Rate (%)	*n*	Rate (%)	*n*	Rate (%)	*n*	Rate (%)	
Chiari Type 1 malformation +	25	71.43	8	22.86	1	2.86	1	2.86	35
Chiari Type 1 malformation −	669	69.47	175	18.17	83	8.62	36	3.72	963
Total	694	69.54	183	18.34	84	8.42	37	3.71	998

loss of cervical lordosis rate when compared with patients without CM1 (51.43% vs. 30.43%, respectively, $P < 0.05$) [Table 2].

Patients with CM1 had lower reversed cervical curve rate when compared with patients without CM1 (17.4% vs. 27.31%, respectively, $P > 0.05$) [Table 2].

Patients with CM1 had lower normal cervical lordosis rate when compared with patients without CM1 (31.43% vs. 42.26%, respectively, $P > 0.05$) [Table 2].

In 25 of 35 (71.43%) patients with CM1, no any cervical discal protrusion was detected, while in 669 of 963 (69.74%) patients without CM1, protrusion was not seen. The rates regarding having protrusion of these two groups were similar ($P > 0.05$).

Further, the rates of protrusion in one-level and protrusion in three or more levels were similar too. A marked difference between two groups regarding the protrusion in two levels is seemed to be due to small number of CM1+ patients with protrusion in two levels (n: 1) [Table 3].

A syrinx cavity formation was found to be as 20% of the CM1+ and 0.83% of the CM1− patients as expected. The difference between two groups was statistically significant ($P < 0.05$) [Table 4].

The syrinx cavity rate was similar in the CM1 + patients with reversed cervical curve and with normal cervical lordosis (27.3% vs. 33.3%). However, when compared with both the two subgroups above, CM1+ patients with loss of cervical lordosis group had lower syrinx rate (11.11%, $P > 0.05$) [Table 5].

In 26 of 35 CM1+ patients, caudal extension of herniation was between 5 and 10 mm. In 6 patients, the extension was between 11 and 15 mm, while in 3 patients, it was >15 mm.

The syrinx rate of the CM1+ with extension more than 15 mm was higher than the rate of CM1+ with extension between 11 and 15 mm (66.7% vs. 50%, $P > 0.05$). Further, the syrinx rate of CM1+ with extension between 11 and 15 mm was statistically higher than the rate of CM1+ with extension between 5 and 10 mm (50% vs. 7.69%, $P < 0.05$) [Table 6].

Discussion

CM1 is characterized with ≥5 mm descent of the caudal tip of cerebellar tonsils past the foramen magnum, which occurs in approximately 1 in 1000 births.[9,10] It can cause neurological symptoms by either direct compression to the neurological structures located in foramen magnum and spinal canal or causing syrinx formation within spinal cord. The obstruction of CSF outflow finally leads to syrinx development within spinal cord or brain stem, which results in neurological symptoms as cavity expands.[11] Association of CM1 and scoliosis is well known;[12,13] however, in literature, there is no any study about such relationship between CM1 and cervical curve changes including reversal of cervical curve–loss of cervical lordosis.

In the present study, we detected lower rates of reversed cervical curve and normal lordosis in the patients with CM1 when compared with patients without CM1 patients. Further, the loss of cervical curve rate was found to be higher in the CM1+ patients when compared with the CM1− ones. The difference was statistically significant. We think that this may be due to a possible compensatory

Table 4: Distribution of the patients to the groups according to presence of syrinx formation

	Syrinx+		Syrinx−		Total
	n	Rate (%)	*n*	Rate (%)	
Chiari Type 1 malformation +	7	20.0	28	80.0	35
Chiari Type 1 malformation −	8	0.83	955	99.17	963
Total	15	1.50	983	98.50	998

Table 5: Distribution of the Chiari malformation Type I patients according to presence of syrinx cavity and cervical curve status

	Syrinx+	
	n	Rate (%)
CM+ + reversed cervical curve	3/11	27.27
CM+ + normal cervical lordosis	2/6	33.33
CM+ + loss of cervical lordosis	2/18	11.11
Total	7/35	20

CM: Chiari malformation

Table 6: The distribution of the syrinx formation to the groups according to the extension of herniation

	5-10 mm		11-15 mm		>15 mm	
	n	Rate (%)	*n*	Rate (%)	*n*	Rate (%)
Syrinx+	2	7.69	3	50	2	66.67
Syrinx−	24	92.31	3	50	1	33.33
Total	26	74.29	6	17.14	3	8.56

mechanism which decreases the compression of spinal cord. Flexion or extension of occipito–cervical junction may increase the compression of the spinal cord by cerebellar tonsils at the level of foramen magnum, while the loss of cervical curve can minimize this compression. As there are no any data about this issue, such possible association must be further evaluated with large series.

Cervical syrinx cavity occurs due to congenital (myelomeningocele, CM1, CM2, Dandy-Walker malformation, Klippel–Feil syndrome) or acquired (cervical canal stenosis, postinflammatory, secondary to a spinal cord tumor, secondary to a hemorrhage, due to vascular insufficiency) conditions.[7]

In a study, syringohydromyelia was detected in 39 (36.1%) of 108 symptomatic CM1 patients. They suggested craniocervical junction osseous anomalies (platybasia, retroverted odontoid, short hypoplastic clivus, basilar invagination) as most predictive of syrinx formation that can create a point of mechanical stress and tension, leading to anterior brain stem compression.[14] Our syrinx frequency in the CM1+ patients was significantly higher than the frequency in CM1− patients as expected. (20% vs. 0.83%, respectively). However, the syrinx frequency in the CM1+ patients with loss of cervical lordosis was lower than both in CM1+ patients with reversed cervical curve or with normal cervical lordosis (11.1% vs. 27,3% and 33.3%, respectively). We think that this lower frequency may be due to a vertical position of proximal medulla spinalis segments, while anteflexion or retroflexion of spinal cord can contribute the compression of medulla spinalis in the level of foramen magnum and also leads to the CSF flow blockage in subarachnoid space, which finally results in syrinx formation. This possible association must be further examined with larger series.

In the literature, there are some studies which suggest that increased amount of tonsillar herniation was associated with a greater likelihood of an associated spinal syrinx,[4,15,16] while a study did not detect any association of syrinx formation and degree of tonsillar herniation.[14] Our results also supported that the extension size of the tonsils would affects the syrinx cavity formation. The syrinx frequency in CM1+ patients with a tonsil herniation between 5 and 10 mm was significantly lower than in the patients with a herniation between 11 and 15 mm and also with a herniation of >15 mm (7.69% vs. 50% and 66.67%, respectively). As herniation extension size increase, the syrinx cavity frequency increases significantly may suggest that a longer compressed segment of the spinal cord can be associated with increased risk of syrinx formation.

The cervical discal protrusion incidences of the CM1+ and CM1− patients were similar with no statistically significant difference which suggests that CM1 or syrinx cavity are not associated with cervical discal herniation. In the literature, resolution of cervical syrinx cavity after a surgical treatment of discal hernia was reported in a patient without CM1, which can suggest a possible association of discal protrusion with syrinx formation, leading to spinal stenosis or direct compression of spinal cord.[17] For this reason, a cervical syrinx cavity in a CM1+ patients, in whom cervical discal herniation is detected must be carefully evaluated before a surgery for accurate identification of the cause of syrinx cavity.

The major limitations of the study are the retrospective design and small patient number. Especially in some subgroups, small patient number did not allow an optimal comparison. Further, the results were not correlated with clinical symptoms and signs.

Conclusion

CM1 must be kept in mind for the patients with cervical flattening seen on X-ray and computed tomographic images, especially when symptoms and signs associated with syrinx formation are present.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Nash J, Cheng JS, Meyer GA, Remler BF. Chiari type I malformation: Overview of diagnosis and treatment. WMJ 2002;101:35-40.
2. Armonda RA, Citrin CM, Foley KT, Ellenbogen RG. Quantitative cine-mode magnetic resonance ımaging of Chiari I malformations: An analysis of cerebrospinal fluid dynamics. Neurosurgery 1994;35:214-24.
3. Lippa L, Lippa L, Cacciola F. Loss of cervical lordosis: What is the prognosis? J Craniovertebr Junction Spine 2017;8:9-14.
4. Elster AD, Chen MY. Chiari I malformations: Clinical and radiologic reappraisal. Radiology 1992;183:347-53.
5. Borden AG, Rechtman AM, Gershon-Cohen J. The normal cervical lordosis. Radiology 1960;74:806-9.
6. Fardon DF, Williams AL, Dohring EJ, Murtagh FR, Gabriel Rothman SL, Sze GK. Lumbar disc nomenclature: version 2.0: Recommendations of the combined task forces of the North American Spine Society, the American Society of Spine Radiology and the American Society of Neuroradiology. Spine J. 2014;14(11):2525-2545. doi:10.1016/j.spinee.2014.04.022.
7. Batnitzky S, Price HI, Gaughan MJ, Hall PV, Rosenthal SJ. The radiology of syringohydromyelia. RadioGraphics 1983;3:585-611.
8. Sherman JL, Barkovich AJ, Citrin CM. The MR Appearan'ce of Syringomyelia: New Observations. AJNR 1986;7:985-95.
9. Hidalgo JA, Tork CA, Varacallo M. Arnold chiari malformation. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2020. Available from: https://www.ncbi.nlm.nih.gov/books/NBK431076/.
10. Jayamanne C, Fernando L, Mettananda S. Chiari malformation type 1 presenting as unilateral progressive foot drop: A case report and review of literature. BMC Pediatr 2018;18:34.

11. Sabba MF, Renor BS, Ghizoni E, Tedeschi H, Joaquim AF. Posterior fossa decompression with duraplasty in Chiari surgery: A technical note. Rev Assoc Med Bras (1992) 2017;63:946-9.
12. Kelly MP, Guillaume TJ, Lenke LG. Spinal deformity associated with chiari malformation. Neurosurg Clin N Am 2015;26:579-85.
13. Brockmeyer DL. Editorial. Chiari malformation Type I and scoliosis: The complexity of curves. J Neurosurg Pediatr 2011;7:22-3.
14. Gad KA, Yousem DM. Syringohydromyelia in patients with chiari I malformation: A retrospective analysis. AJNR Am J Neuroradiol 2017;38:1833-8.
15. 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;28:639-645.
16. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO. Chiari malformation Type I and syrinx in children undergoing magnetic resonance imaging. J Neurosurg Pediatr 2011;8:205-13.
17. Yaman ME, Eylen A, Ayberk G. Resolution of isolated syringomyelia after treatment of cervical disc herniation: Association or coincidence? Bratisl Med J 2012;113:500-2.