Ann Med Res

Current issue list available at AnnMedRes

Annals of Medical Research

journal page: www.annalsmedres.org

Patients operated on for Chiari malformation type 1: A 10-year evaluation

Yasin Taskin[a,*], Fatih Ersay Deniz[a], Erol Oksuz[a], Ozgur Demir[a], Yunus Emre Kuyucu[b]

[a] *Tokat Gaziosmanpaşa University, Faculty of Medicine, Department of Neurosurgery, Tokat, Türkiye*
[b] *Tokat Gaziosmanpaşa University, Faculty of Medicine, Department of Biostatistics and Medical Informatics, Tokat, Türkiye*

ARTICLE INFO

Keywords:
Arnold chiari
Herniation
Hydrocephalus
Syringomyelia

Received: Sep 12, 2024
Accepted: Nov 18, 2024
Available Online: 29.11.2024

DOI:
10.5455/annalsmedres.2024.09.192

Abstract

Aim: It is aimed to evaluate our patients who were operated for Chiari type 1 malformation with the help of literature and to contribute to the literature.

Materials and Methods: 14 patients who were operated on for Chiari type 1 malformation diagnosis between 2012 and 2022 were retrospectively examined.

Results: The mean age of our 14 patients included in the study was 36.39 (5 months - 59 years). The postoperative intensive care unit stay was 2.57 days, and the total stay until discharge was 10.93 days on average. The mean cerebellar tonsil herniation in our patients was 13.36 mm. The female/male ratio was 1. Syringomyelia was detected in 78.6% of our patients. 13 of the 14 patients were discharged in good health, and 1 patient died in the postoperative period.

Conclusion: In patients with Chiari type 1 malformation, the presence of syringomyelia in addition to tonsillar herniation has been noted as a valuable parameter in the decision to operate. Although the most common condition in symptomatic patients is headache, it should be kept in mind that atypical symptoms may also occur. Patients who have undergone surgery for Chiari type 1 malformation should be closely monitored postoperatively for possible complications.

Introduction

Although its cause remains debated, Chiari malformation type 1 (CM1) is the most prevalent congenital abnormality of the craniovertebral junction (CVJ) [1].

It is marked by the downward displacement of the cerebellar tonsils through the foramen magnum, a condition first described by Hans Chiari in 1896 [2]. Chiari type 1 is the most typical among its subtypes [3]. It is usually symptomatic in the third and fourth decades and found more frequently in women than men [4, 5]. On MRI (Magnetic Resonance Imaging), caudal herniation of 5 mm or more of the cerebellar tonsils is diagnostic [1, 4]. The most common clinical symptom is headache, which is especially aggravated by Valsalva maneuvers [6]. Additionally, symptoms such as diplopia, nystagmus, visual field loss, and blurred vision may develop due to increased intracranial pressure. Cerebellar disorders (ataxia, balance instability, gait disability, coordination disorder, dysdiadochokinesia, dysmetria, dysarthria), signs of cranial nerve damage (dysphagia, voice changes, sleep apnea), symptoms related to syringomyelia (pain, loss of thermal sensation, motor deficit, atrophy, loss of reflex) and various atypical symptoms such as vertigo, nausea, trigeminal neuralgia, radicular pain, burning pain in extremities, numbness may appear [7-12]. It may also be accompanied by syringomyelia, scoliosis, hydrocephalus, vertebral anomalies and many other pathologies [6]. Type 1 Chiari Malformation is associated with Syringomyelia at a rate of 30%-70% and is most commonly seen in the cervical region [6, 8, 13]. Scoliosis has been shown to occur in 30% of patients with Type 1 Chiari Malformation, and in the case of concomitant syringomyelia, around 60% [14]. Moreover, during the diagnostic process, hydrocephalus is seen in around 4-18% of cases with CM1 [15]. Genetic disorders such as Ehlers-Danlos syndrome type 3 and Sickle cell disease may accompany patients diagnosed with CM1 [16]. Conservative treatments can be used for patients regarding their symptoms. Surgery can be planned in appropriate patients according to their follow-up status. The aim of surgery is craniocervical decompression. The standard treatment method is suboccipital decompression and C1 total laminectomy. If necessary, C2 partial laminectomy is performed as well. Additionally, duraplasty or

*Corresponding author:
Email address: dryasintaskin@gmail.com (Yasin Taskin)

duratomy can be performed [15, 17]. After surgery, complications such as cerebrospinal fluid (CSF) fistulae, pseudomeningocele, cerebellar sag, aseptic meningitis, surgical site infection, and even death may occur. Therefore, close monitoring is absolutely crucial in the treatment and follow-up of patients [3, 18-22]. We also presented our experiences in patients who underwent surgery for Type 1 Chiari Malformation in our clinic, comparing them with the literature.

Materials and Methods

After receiving the approval of the local ethics committee (Tokat Gaziosmanpaşa University Clinical Research Ethics Committee) with the decision dated June 27. 2024 and numbered 24-KAEK-207 for conducting the study. The records and radiological images of patients who underwent surgery for Chiari type 1 malformation between 2012 and 2022 at the Tokat Gaziosmanpaşa University Faculty of Medicine Hospital, Department of Brain and Nerve Surgery were retrospectively examined on the automation system. The sample size consists of patients who underwent surgery due to CM1 between 2012 and 2022. Before the surgery, additional imaging was performed to detect concomitant conditions such as syringomyelia and hydrocephalus. Postoperative complications were evaluated. Patients who were diagnosed with Chiari type 1 malformation and underwent surgery were included in the study. Patients other than this were excluded from the study.

Statistical analysis

Descriptive statistics were utilized to detail the general characteristics of the study groups. Data for variables are defined using mean and standard deviation. The calculations were performed using commercial statistical software (IBM SPSS Statistics 22, SPSS Inc., an IBM Company, Somers, NY).

Results

Fourteen patients were involved in the study. The mean age of our patients (5 months - 59 years) was 36.39. Head and neck pain were the most frequent presenting complaint, followed by paresis-paresthesia and dizziness. The postoperative length of intensive care unit stay was 2.57 days, and the hospital length of stay until discharge was 10.93 days. The mean amount of cerebellar tonsil herniation in our patients was 13.36 mm. The female-to-male ratio was 1. Thirteen patients were discharged in good condition, and one died in the postoperative period. Syringomyelia was detected in 78.6% of our patients. None of the patients had any revision or instability during the postoperative period. Two patients (14.3%) developed postoperative incisional discharge. Moreover, two developed postoperative hydrocephalus, and a ventriculoperitoneal shunt was inserted. Suboccipital decompression + duraplasty + C1 laminectomy was performed in 85.7% of the patients, while suboccipital decompression + duraplasty + C1 total and C2 partial laminectomy were performed in 14.3% of the patients (Tables 1, 2, 3).

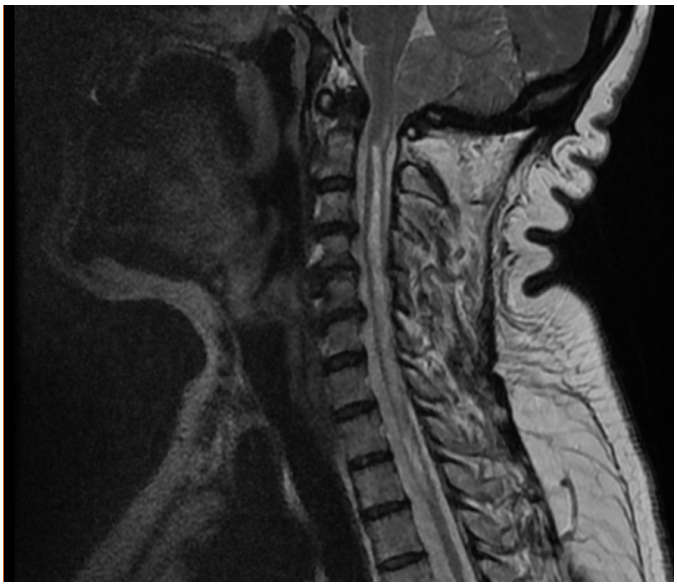

Figure 1. Preoperative Chiari type 1 malformation MRI.

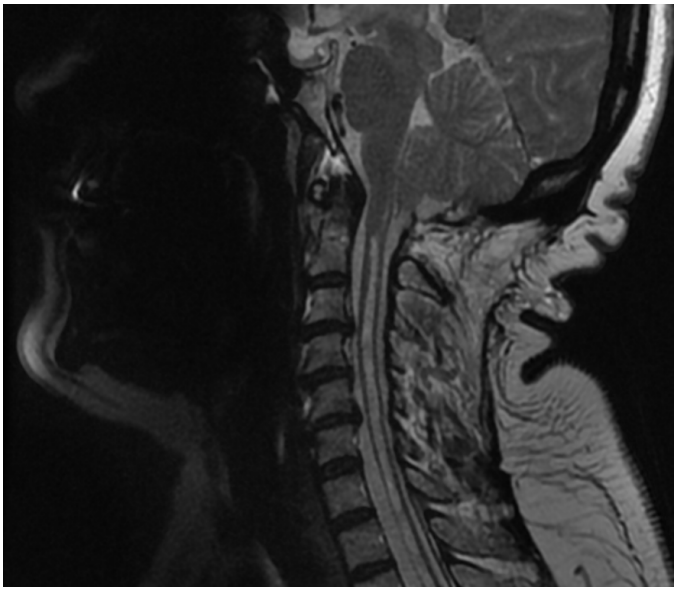

Figure 2. Postoperative Chiari type 1 malformation MRI.

Discussion

Even though its etiology is still controversial, CM1 is the most common congenital anomaly of the craniovertebral junction, characterized by sagging of the ectopic cerebellar tonsils from the foramen magnum [1]. The most prevalent subtype of Chiari malformation is type 1 [3]. Similar to the literature, the most common cases encountered in our clinic were Chiari type 1.

In the study conducted by Elster and Chen, the female:male ratio was reported as 3:2 [23]. In the study conducted by Meadows et al., this ratio was reported as 1:1, which is consistent with our study [24].

In a study in which patients who underwent surgery with a diagnosis of CM1 were prospectively evaluated in the preoperative period, it was reported that 81% of them had accompanying headache [25]. It was reported that suboccipital headache was triggered by the Valsalva maneuver in

Table 1. Distribution of quantitative variables.

	Mean	Standard Deviation	Median	Minimum	Maximum
Age	36.39	16.69	41.50	.50	59.00
Postoperative Intensive Care Unit	2.57	2.68	2.00	1.00	11.00
Stay Postoperative Stay	10.93	7.02	10.00	3.00	28.00
Herniation Amount	13.36	5.38	15.00	5.00	23.00

Table 2. Distribution of qualitative variables.

		n	%
Gender	Female	7	50.0
	Male	7	50.0
Discharged	No	1	7.1
	Yes	13	92.9
Died	No	13	92.9
	Yes	1	7.1
Syrinx	No	3	21.4
	Yes	11	78.6
Postoperative Incisional Discharge (CSF fistulae)	No	12	85.7
	Yes	2	14.3
Hydrocephalus	No	14	85.7
	Yes	2	14.3
Occipital Decompression, C1 Laminectomy	Negative	2	14.3
	Positive	12	85.7
Occipital Decompression, C1 Laminectomy, C2 Laminectomy	Negative	12	85.7
	Positive	2	14.3
Instability	No	14	100.0
The Need for Instrumentation	No	14	100.0

Table 3. Patients' complaints at the time of admission.

Presenting complaint	Yes	No
Headache and neck pain	10 (71.4%)	4 (28.6%)
Paresis-paresthesia	9 (64.2%)	5 (35.8%)
Dizziness	5 (35.7%)	9 (64.3%)

40 (63%) of 63 patients treated for CM1 [26]. They were also the two most frequent presenting complaints in our clinic. About 71.4% of our patients complained primarily of headache and neck pain, consistent with the literature (Table 3). In addition, headache and neck pain were followed by paresis-paresthesia and dizziness, respectively.

In a study by Elster and Chen, 37% of patients with CM1 were found to have syringomyelia [23]. In another study evaluating pediatric and adult Chiari malformation patients, the incidence of syringomyelia was reported as 65% [27]. In our clinic, syringomyelia was noticed in 78.6% of the operated patients, a higher rate compared to the literature.

It has been revealed that 30% of patients with CM1 have about 60% scoliosis in the presence of syringomyelia [28]. Moreover, during the diagnostic process, hydrocephalus is seen in around 4-18% of cases with CM1 [28]. The development of postoperative hydrocephalus was observed in two patients included in our study. One of them had a ventriculoperitoneal (VP) shunt inserted, the most commonly used treatment method for the diagnosis of hydrocephalus [29]. The other patient developed respiratory arrest secondary to hydrocephalus, an external ventricular drain was inserted, meningitis developed in the subsequent period, and the patient died despite treatments.

CSF fistulae, pseudomeningocele, cerebellar sag, aseptic meningitis, surgical site infection, and even death are possible postoperative complications. Hence, close monitoring is of paramount importance in the treatment and follow-up of patients [3, 18-22]. Proper duraplasty, which stops CSF leakage as the layers are closed, will help prevent complications like CSF fistulae and discharge at the wound site. In 14.3% of our cases, cerebrospinal fluid fistula developed in the operation area in the postoperative period and resolved without the need for surgical intervention.

In a study evaluating 612 pediatric CM1 cases, it was reported that approximately 2.1% of these patients had genetic disorders [16]. In our study, there were no patients with genetic disorders. This may be due to the small size of our patient population.

The aim of surgery is craniocervical decompression. The

common treatment method is suboccipital decompression and C1 total laminectomy (C2 partial laminectomy when necessary). Additional duraplasty or duratomy can be added to this approach [15, 17]. Suboccipital decompression + duraplasty + C1 laminectomy were performed in 85.7% of the patients, whereas suboccipital decompression + duraplasty + C1 total and C2 partial laminectomy were conducted in 14.3% of the patients. Instability may be observed at a higher rate, particularly in patients who underwent additional C2 laminectomy. It is not recommended to receive it as much as possible to minimize the rate of instability [10, 30]. Two of the patients in our study underwent C2 laminectomy. However, no instability was observed during follow-up. The incidence of spinal deformity due to multi-level laminectomy in the pediatric population is 53% [31]. Despite this rate given in the literature, none of our patients developed signs of instability in the postoperative period and no revision surgery was performed.

Limitations

However, our study has some limitations. First of all, it is a single center. Also, the number of patients that can be evaluated is limited due to being a single center. Multicenter studies that evaluate more patients are needed.

Conclusion

The presence of syringomyelia with cerebellar tonsillar herniation in patients with CM1 has stood out as a valuable parameter in the decision to operate. Although headache is the most typical symptom in symptomatic patients, it is significant to recognize that atypical symptoms can also appear. Patients who have had CM1 surgery need to be closely observed for possible complications in the postoperative phase. Clearly, CM1-operated patients in our clinic are primarily compatible with the literature. Nevertheless, it is worthwhile to conduct larger-scale studies.

Ethical approval

Ethical approval was received for this study from Tokat Gaziosmanpaşa University Clinical Research Ethics Committee (24-KAEK-206).

References

1. Barkovich A, Wippold F, Sherman J, Citrin C. Significance of cerebellar tonsillar position on MR. American journal of neuroradiology. 1986;7(5):795-9.
2. Chiari H. Uber Veranderungen des Kleinhiens, des pons und der medulla oblongata. Folge von congenitaler hydrocephalie des grossherns. Deskschr Akad Wiss Wien. 1895;63:71-116.
3. Nyland H, Krogness K. Size of posterior fossa in Chiari type 1 malformation in adults. Acta neurochirurgica. 1978;40:233-42.
4. Aboulezz AO, Sartor K, Geyer CA, Gado MH. Position of cerebellar tonsils in the normal population and in patients with Chiari malformation: a quantitative approach with MR imaging. Journal of computer assisted tomography. 1985;9(6):1033-6.
5. Batzdorf U. Chiari I malformation with syringomyelia: Evaluation of surgical therapy by magnetic resonance imaging. Journal of neurosurgery. 1988;68(5):726-30.
6. Aydoseli A, Sencer A. Chiari Tip 1 Malformasyonunda Klinik Tablo. Türk Nöroşir Derg. 2015;25(2):243-7.
7. Bindal AK, Dunsker SB, Tew Jr JM. Chiari I malformation: classification and management. Neurosurgery. 1995;37(6):1069-74.
8. Godzik J, Kelly MP, Radmanesh A, Kim D, Holekamp TF, Smyth MD, et al. Relationship of syrinx size and tonsillar descent to spinal deformity in Chiari malformation Type I with associated syringomyelia. Journal of Neurosurgery: Pediatrics. 2014;13(4):368-74.
9. Kumar R, Kalra SK, Vaid VK, Mahapatra A. Chiari I malformation: surgical experience over a decade of management. British Journal of Neurosurgery. 2008;22(3):409-14.
10. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery. 1999;44(5):1005-17.
11. Milhorat TH, Johnson WD, Miller JI, Bergland RM, Hollenberg-Sher J. Surgical treatment of syringomyelia based on magnetic resonance imaging criteria. Neurosurgery. 1992;31(2):231-45.
12. Papanastassiou AM, Schwartz RB, Friedlander RM. Chiari I malformation as a cause of trigeminal neuralgia: case report. Neurosurgery. 2008;63(3):E614-E5.
13. Bogdanov E. Epidemiology/Flint G., Rusbridge C.(eds.). Syringomyelia. Springer-Verlag Berlin Heidelberg; 2014.
14. Krieger MD, Falkinstein Y, Bowen IE, Tolo VT, McComb JG. Scoliosis and Chiari malformation Type I in children. Journal of Neurosurgery: Pediatrics. 2011;7(1):25-9.
15. Meadows J, Guarnieri M, Miller K, Haroun R, Kraut M, Carson BS. Type I Chiari malformation: a review of the literature. Neurosurgery Quarterly. 2001;11(3):220-9.
16. Sadler B, Kuensting T, Strahle J, Park TS, Smyth M, Limbrick DD, et al. Prevalence and impact of underlying diagnosis and comorbidities on Chiari 1 malformation. Pediatric neurology. 2020;106:32-7.
17. Aslan A, Rakip U, Boyacı MG, Yildizhan S, Kormaz S, Atay E, et al. Posterior Fossa Decompression and superficial durotomy rather than complete durotomy and duraplasty in the management of Chiari 1. Neurological Research. 2021;43(6):440-6.
18. Tubbs RS, Beckman J, Naftel RP, Chern JJ, Wellons JC, Rozzelle CJ, et al. Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I. Journal of Neurosurgery: Pediatrics. 2011;7(3):248-56.
19. Dubey A, Sung W-S, Shaya M, Patwardhan R, Willis B, Smith D, et al. Complications of posterior cranial fossa surgery—an institutional experience of 500 patients. Surgical neurology. 2009;72(4):369-75.
20. Durham SR, Fjeld-Olenec K. Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients: a meta-analysis. Journal of Neurosurgery: Pediatrics. 2008;2(1):42-9.
21. Vanaclocha V, Saiz-Sapena N. Duraplasty with freeze-dried cadaveric dura versus occipital pericranium for Chiari type I malformation: comparative study. Acta neurochirurgica. 1997;139:112-9.
22. Alden TD, Ojemann JG, Park T. Surgical treatment of Chiari I malformation: indications and approaches. Neurosurgical focus. 2001;11(1):1-5.
23. Elster AD, Chen M. Chiari I malformations: clinical and radiologic reappraisal. Radiology. 1992;183(2):347-53.
24. Meadows J, Kraut M, Guarnieri M, Haroun RI, Carson BS. Asymptomatic Chiari Type I malformations identified on magnetic resonance imaging. Journal of neurosurgery. 2000;92(6):920-6.
25. Sperling NM, Franco Jr RA, Milhorat TH. Otologic manifestations of Chiari I malformation. Otology & neurotology. 2001;22(5):678-81.
26. Alperin N, Loftus JR, Oliu CJ, Bagci AM, Lee SH, Ertl-Wagner B, et al. Imaging-based features of headaches in Chiari malformation type I. Neurosurgery. 2015;77(1):96-103.
27. Arnautovic A, Splavski B, Boop FA, Arnautovic KI. Pediatric and adult Chiari malformation Type I surgical series 1965–2013: a review of demographics, operative treatment, and outcomes. Journal of Neurosurgery: Pediatrics. 2015;15(2):161-77.
28. Loukas M, Shayota BJ, Oelhafen K, Miller JH, Chern JJ, Tubbs RS, et al. Associated disorders of Chiari Type I malformations: a review. Neurosurgical focus. 2011;31(3):E3.
29. Kiyak V, Deniz FE, Oksuz E, Demir O, Demir O. Epidemiological and complication assessments of patients with ventriculoperitoneal shunts. Medicine. 2022;11(3):1223-6.
30. McGirt MJ, Attenello FJ, Datoo G, Gathinji M, Atiba A, Weingart JD, et al. Intraoperative ultrasonography as a guide to patient selection for duraplasty after suboccipital decompression in children with Chiari malformation Type I. Journal of Neurosurgery: Pediatrics. 2008;2(1):52-7.

31. Bell DF, Walker JL, O'Connor G, Tibshirani R. Spinal deformity after multiple-level cervical laminectomy in children. Spine. 1994;19(4):406-11.