World Journal of Surgery and Surgical Research

Research Article
Published: 17 Jul, 2020

Chiari Malformation-I at Tertiary Center

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Abstract

Introduction: Chiari malformations are one of the most controversial topics in neurosurgery today. It is a congenital disorder characterized by the anatomical defect of the base of skull with caudal displacement of cerebellar tonsils, through the foramen magnum. The objective of the present series was to study clinical outcome following posterior fossa decompression, C1 laminectomy and duraplasty in type 1 Arnold Chiari malformation.

Material and Methods: This study was retrospectively analysis of the Chiari I malformation in the Department of Neurosurgery, NAMS from January 2014 to February 2020. There were 43 admitted patients with Chiari I malformation in our center. OPD follow up or telephonic questionnaires were used to analyze and compare the improvement in neck pain and disability before and after surgery.

Results: The most common age group of presentation was 2nd decade (30.3%) followed by 3rd decade (30.6%). The male to female ratio was 2.5:1. The age ranged from 5 years to 60 years with median age of 32. Out of them 31 (72%) were females and 12 (28%) were male. The most common presenting complaint was sensory disturbances 26 patients (61%) followed by neck pain in 17 patients (39%). The most common sign was limb weakness in 25 patients (58%). All patients were operated with foramen magnum decompression with duraplasty, subarachnoid dissection of CSF flow and C1 laminectomy except three patients who were undergoing foramen magnum decompression only. Two patients were cervical unstable after operation. One patient was managed with occipito-atlanto-axial fixation. We had mortality one which had cervical unstable and chest infection post operatively. There was one patient with surgical site infection requiring debridement and secondary suturing. Patients showed an overall clinical improvement of 90%. There were no motor aggravation occurred in our study. Foramen magnum decompression with duraplasty, subarachnoid dissection of CSF flow and resection of the C1 posterior arch were found to be associated with favorable results on clinical signs and symptoms.

Conclusion: Proper patient selection is critical to prevent unnecessary complication and maximize the outcome. The surgical gold standard consists in three key steps-Foramen magnum decompressions with duraplasty, subarachnoid dissection of CSF flow and C1 laminectomy.

Keywords: Arnold Chiari malformation; Decompression; Duraplasty

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Received Date: *06 Jun 2020*
Accepted Date: *09 Jul 2020*
Published Date: *17 Jul 2020*

Citation:
Shrestha R, Jha R, Bista P. Chiari Malformation-I at Tertiary Center. World J Surg Surgical Res. 2020; 3: 1238.

Introduction

Chiari Malformation (CM) is one of the most controversial topics in neurosurgery today. There is a lack of agreement as to what defines these malformations, their symptoms, their natural history and their management. Over the years there has been a significant improvement in understanding the pathophysiology, diagnostic techniques, and management approaches. Various surgical techniques have been put forth to treat this entity in many series [1,2]. Those techniques may include sub occipital decompression of the posterior fossa with or without duraplasty, arachnoid dissection, shrinking of the cerebellar tonsils or fixation C1 and C2. Symptoms may differ between periods of exacerbation and remission. The diagnosis of CM-I in patients with or without symptoms is recognized with neuroimaging techniques. With the advent of Magnetic Resonance Imaging (MRI), it became the imaging modality of choice in the diagnosis of CM and the associated syrinx. It had been used to enumerate the extent of tonsils below the foramen magnum. The extension of tonsil below the foramen magnum is considered pathologic if it is more than 5 mm [3].

Methods

This study was a retrospective analysis. Patients treated in Department of Neurosurgery, NAMS from January 2014 to February 2020 were included. In the last 5 years, 43 patients with Chiari I malformation have been operated. Their clinical history and OT records were collected from the case records of the patients. The patients were followed up for 6 months and clinical parameters

Table 1: Age distribution.

Age (years)	Numbers	Percentage (%)
0-10	2	4.6
Oct-20	2	4.6
20-30	13	30.3
30-40	14	32.6
40-50	9	20.9
More than 50	3	6.9
Total	43	100

Table 2: Distribution of clinical presentation.

Symptoms	Numbers	Percentage (%)
Headache	15	35
Neck pain	17	39
Sensory disturbance	26	61
Weakness and muscular atrophy	25	58
Limb pain	9	20.9
Spasticity	3	6.9
Cerebellar signs	2	4.6

were evaluated at OPD or *via* telecommunication. Patients included were the ones operated for CM type I and excluded were the ones with Chiari type II, III, and IV. Patients who meet the inclusion criteria were recruited. Recorded data were analyzed with SPSS version 21.

Results

The most common age group of presentation was 2nd decade (30.3%) followed by 3rd decade (32.6%). The age ranged from 5 years to 60 years with median age of 32 (Table 1). Out of them 31 (72%) were females and 12 (28%) were male (Figure 1). The male to female ratio was 2.5:1. The most common presenting complaint was sensory disturbances, reported in 26 patients (61%) followed by neck pain, reported in 17 patients (39%) and Headache, reported in 15 patients (35%). The most common sign was limb weakness in 25 patients (58%) (Table 2). Majority of the patients underwent foramen magnum decompression with duraplasty, subarachnoid dissection of CSF flow and C1 laminectomy (Figures 2-5). However, three patients underwent foramen magnum decompression only. Two patients were cervical instability after operation. One patient was managed with occipitoatlantoaxial fixation (Figure 6). We had mortality one that had cervical instability and chest infection post operatively. One patient had surgical site infection, requiring debridement and secondary suturing. Patients showed an overall clinical improvement of 90% on follow up. There was no reported motor aggravation. Foramen magnum decompression with duraplasty, subarachnoid dissection of CSF flow and resection of the C1 posterior arch were found to be associated with favorable results on clinical signs and symptoms (Table 1,2 and Figures 1-7).

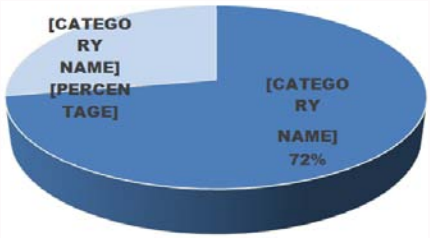

Figure 1: Gender distribution.

Figure 2: Sagittal T2-weighted MR image of the brain showing herniation of cerebellar tonsils.

Figure 3: Intra-operative view: Suboccipital muscle dissection.

Figure 4: Intra-operative view: Suboccipital craniectomy and C1 laminectomy.

Discussion

John Cleland (Professor of anatomy in Glasgow, Scotland) was the first to describe Chiari II or Arnold–Chiari malformation on his report of a child with spina bifida, hydrocephalus, and anatomical alterations of the cerebellum and brainstem in 1883. Since then there have been many theories to explain pathogenesis of Chiari Malformations but main issue is the disruption of the normal CSF flow due to herniation of cerebellar tissue through foramen magnum because of larger cerebellar mass in a smaller posterior cranial fossa. There are various methods of foramen magnum decompression described in the literature [4].

In our study, majority of the patients were in their 2nd and 3rd decade of life, with mean age of 32 years at the time of presentation. This finding was consistent with the findings of study done by Naik et al. [5]. Patients were predominately male in this study whereas, female predominance was seen in study 145 patients conducted by Arnautovic et al. in both adult and pediatric studies [6]. Male

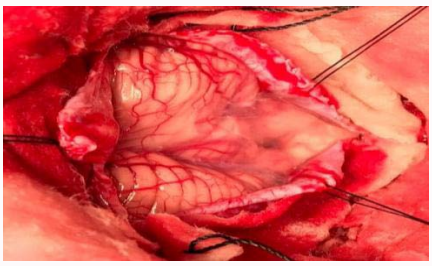

Figure 5: Intra-operative view Y-Shaped durotomy.

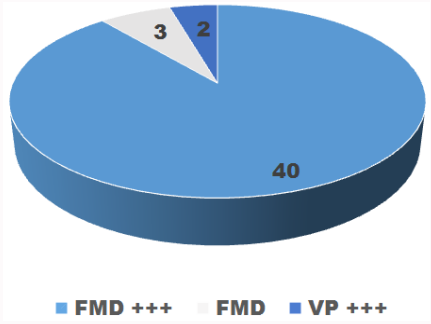

Figure 6: Operation Pattern.

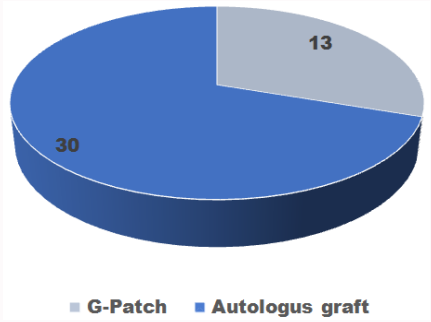

Figure 7: Choice of grafts in patients underwent duraplasty.

predominance was also seen in the study done by Ramnarayan et al. [7].

The most common presenting symptom was sensory disturbances in 26 patients (61%) followed by neck pain in 17 patients (39%) and Headache in 15 patients (35%) in our series. Our finding was consistent with that of study conducted by Ramnarayan et al. [7] and Bharath Singh et al., who reported 62% and 68% of patients respectively with sensory disturbances as the presenting symptom [5]. Other symptoms present at the time of diagnosis included headache, limb pain, spasticity and cranial nerve dysfunction. The most common sign was limb weakness seen in 25 patients (58%). In very severe cases in which compression of the spinal cord or medulla oblongata occurs, symptoms of involvement of the motor or sensory pathways, or lower cranial nerves were seen.

Various approaches were implemented for the treatment of symptomatic patients during our study period. 40 cases underwent suboccipital cranioectomy with duraplasty, C1 laminectomy, subarachnoid dissection of CSF flow, and tonsillar coagulation. Only suboccipital cranioectomy was performed for 3 cases that had congenital anomalies in cervico vertebral junction and morbidly obese patients. Two patients underwent VP shunt prior to suboccipital cranioectomy as they had hydrocephalus (Figure 6). Posterior suboccipital cranioectomy with duraplasty had higher clinical improvement and lower recurrence rate in the patients with syringomyelia. This finding was consistent with that of Batzdorf et al. [8] and Lin et al. [9] meta-analysis. No recurrence was documented in our patients during one year follow up. However, Krishna et al. [10] mentioned high recurrence rates where decompression without dural opening was performed.

Complications were observed in two patients with cervical instability after surgery. X-ray of cervical region (flexion-extension view) before surgery was done based on hospital protocol. One patient was managed with occipitoatlantoaxial fixation. Mortality of one patient with cervical instability and chest infection post-op was documented. Duraplasty with autologous graft was done in 13 cases and G-patch in 30 cases (Figure 7). Others complications noted in our series were CSF leak seen in 3 patients in early days. One patient had surgical site infection requiring debridement and secondary suturing. But this was not significant with G-patch. Study conducted by Krishna et al. [10], stated that the rate of complication is low but recurrence rates are high.

There are many different surgical techniques to treat Chiari type I malformation. The goal of all of these surgeries is to restore adequate cerebrospinal fluid CSF flow at the level of the foramen magnum and reconstruction of the Cisterna magna. In our series, patients showed an overall clinical improvement of 90%. This was based on the findings our study over the period of 5 years in our institution. Nevertheless, we recommend for future randomized studies with larger study group, to provide stronger evidence for surgical decision making of CM-I.

Conclusion

Proper patient selection is critical to prevent unnecessary complications and maximize the outcome. The surgical gold standard consists in three key steps-Foramen magnum decompressions with duraplasty, subarachnoid dissection of CSF flow and C1 laminectomy.

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