The prime objective in the surgical treatment of Chiari malformation (CM) and/or syringomyelia (SM) is based on the restoration of the normal cerebrospinal fluid (CSF) dynamics at the craniovertebral junction through the creation of a large artificial cisterna magna. In our current study, a systematic and critical review of the pertinent literature was made for identifying the result of posterior fossa decompression on syrinx in patients of type 1 Arnold Chiari malformation (ACM).

Keywords: Syringomyelia SM, Chiari malformation type I CMI, Foramen magnum decompression FMD.

INTRODUCTION

The pathophysiology of syrinx formation and progression is important for surgeons in their selection of a surgical approach for syringomyelia (SM) secondary to Chiari malformation type I (CMI). There is broad consensus on the role of mechanical blockage of cerebrospinal fluid flow at the craniovertebral junction in the pathogenesis of SM secondary to CMI [1,2]. Posterior fossa decompression (PFD) is currently preferred by most surgeons as the standard surgical option in the treatment of SM secondary to CMI [3-6]. This results in decompression of the cerebellar tonsil restores the normal cerebrospinal fluid flow[7]. In addition, a wide variety of surgical procedures as adjuvants to standard PFD, including syringo subarachnoid shunting, obex plugging and resection of the cerebellar tonsil, have been advocated in previous studies, which potentially induce bias with respect to the evaluation of syrinx resolution. To date, few reports have quantitatively evaluated syrinx resolution after PFD or identified predictive factors for better improvement. In addition to occipital pain and vertigo, dissociative sensory loss is one of the most frequent clinical presentations in children and adolescents with CMI. In this study we have done posterior fossa decompression by midline sub occipital craniectomy with or without subpial cerebellar tonsil resection in a patient of chiary malformation type I with syringomyelia.

MATERIAL AND METHODS

Present study was conducted at Neurosurgery department G.R. Medical College Gwalior from January 2012 to June 2018 in patients which were admitted in this hospital with type 1 ACM with Syrinx and operated for the same by Posterior fossa decompression.

Patients of all age groups and both sexes were included in the study. These cases were analyzed for age, sex incidence, and location of syrinx. Statistical analysis was done by calculating the numbers and percentage for computing the incidence in various age groups, in sexes, location.

Study design: A meta-analysis

Ethical approval

The study was undertaken after consent and clearance by the ethical committee of G.R. Medical College Gwalior

Inclusion criteria

Of all patients admitted with CMI during the period 2012 – 2018 were included.

Exclusion criteria

Patient not willing for operation or managed conservatively
Sample size
Fifty patients

Methodology
Age, Sex, Location, presenting complaints were studied.

End Result
At the end after evaluation of the results of decompression with a minimum follow-up of 6 months, inference made based on the data obtained from study.

OBSERVATION
The present study comprises of 50 patients of CM1 admitted in the department of Neurosurgery, G.R. Medical College & J.A. Group of Hospitals, Gwalior from Feb 2012 to Aug 2018 who were treated by posterior fossa decompression.

Table 1: Age wise distribution

| Age in Years | No. of patients | Percentage |
|--------------|----------------|------------|
| < 20 years   | 1              | 2%         |
| 21-40        | 12             | 24%        |
| 41-60        | 34             | 68%        |
| >60          | 3              | 6%         |

Mean age was 45.7 years

Table 2: Sex wise distribution

| Sex   | Number of patients | Percentage |
|-------|--------------------|------------|
| Male  | 18                 | 36%        |
| Female| 32                 | 64%        |

Table 3: Distribution of syrinx location

| Location of disease | Number of patients | Percentage |
|---------------------|--------------------|------------|
| upper cervical      | 12                 | 24%        |
| cervical and upper thoracic | 15         | 30%        |
| cervical and thoracic | 18              | 36%        |
| cervical thoracic and lumbar | 5          | 10%        |

Table 4: Sign and symptoms

| sign & symptoms                  | no. of patients | %   |
|----------------------------------|----------------|-----|
| Neck pain                        | 45             | 90% |
| numbness and tingling            | 36             | 72% |
| Dizziness                        | 30             | 60% |
| swallowing difficulty            | 5              | 10% |
| limb weakness                    | 18             | 36% |
| Dissociative sensory loss        | 28             | 56% |
| imbalance                        | 12             | 24% |

Table 5: Result of posterior fossa decompression on resolution of syrinx

| no. of patients | %   |
|-----------------|-----|
| total resolution of syrinx | 18  | 36% |
| partial resolution | 28  | 56% |
| no resolution    | 4   | 8%  |

Table 6: Improvement of symptoms after posterior fossa decompression

| sign & symptoms                  | no. of patients | improvement | %   |
|----------------------------------|----------------|-------------|-----|
| Neck pain                        | 45             | 40          | 88% |
| numbness and tingling            | 36             | 32          | 88% |
| Dizziness                        | 30             | 28          | 93% |
| swallowing difficulty            | 5              | 3           | 60% |
| limb weakness                    | 18             | 10          | 55% |
| Dissociative sensory loss        | 28             | 20          | 71% |
| imbalance                        | 12             | 8           | 66% |

DISCUSSION
This combined retrospective and prospective cohort study describes the long-term clinical and radiological outcomes of consecutive patients with syringomyelia associated with CM-I who underwent surgery between 2012 and 2018. Overall, the results confirm that posterior fossa decompression yields high rates of radiological (92%) and clinical (93.4%) improvement and a low complication rate. Our data are not dissimilar from those reported in previously published studies, [8-13] and the complication rate reflects that seen in eminent past publications [14, 15]. The improvements were stable during long-term surveillance. Equally noteworthy is that the duration of symptoms and the age at surgery were significantly correlated with the postoperative clinical and
radiological courses. As previously described, this simple technique can improve or eliminate both spinal and cranial symptoms. Whether it is necessary to open the arachnoid space is a matter of dispute. Some authors indicate that the inner dural layer and arachnoid should be left intact to avoid CSF leakages [14, 16].

A study in a large series of patients found that the mean age at onset of syringomyelia in CM-I was 25 years, and women accounted for 75% of the patients [17]. The majority of the patients in our series were also women (64%), but the mean age was considerably higher (45.7 years). This is probably due to different referral mechanisms, cultural differences, or the necessary association with CM-I. Since syringomyelia develops late in CM-I, it is possible that the natural evolution of the CM-I process is syrinx development. Moreover, we confirmed statistically that longer duration of symptomatology and older age at surgery negatively influenced outcome. This finding suggests that, while indications of appropriate treatment for CM-I are controversial [18], surgery should be performed early in the presence of syringomyelia.

The present study has several limitations. It was observational, and mixed prospective and retrospective data were collected over a period of 7 years. Consequently, the cohort is highly heterogeneous, and the results must be interpreted with these considerations in mind. Because clinical signs and symptoms are complex and vary from patient to patient, we simplified the clinical features into cranial (due to brain-stem compression or hydrocephalus) and spinal (caused by the syrinx). This simplification can result in a loss of information. However, it is notable the surgical procedure was equivalent in all cases, and study enrolment was limited to CM-I patients presenting with a syrinx.

CONCLUSION

The clinical manifestations of syringomyelia related to CM-I are directly attributable to compression of the brainstem and/or cranial nerves, syrinx, and CSF drainage obstruction or hydrocephalus. Surgical decompression with durotomy, arachnoid opening, tonsillar shrinkage, and re-creation of the cisterna magna is a safe and effective procedure. The prognosis is excellent, with global clinical improvement and radiological improvement in greater than 90% of patients. Definite clinical predictors of poor clinical and radiological prognosis were identified—namely, age at the time of surgery and symptom duration. The results of this study provide additional data that support the effectiveness and safety of treating CSF flow obstruction by performing cranio-cervical decompression and artificial cisterna magna recreation.

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