ORIGINAL ARTICLE

Posterior Fossa Reconstruction for Chiari Malformation Type I: Experience with 17 Cases

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ABSTRACT

Objective: *To assess the results of posterior fossa reconstruction in patients with Chiari I malformation.*

Material and Methods: *This observational study was conducted at the department of Neurosurgery, medical and teaching institute (MTI), Lady Reading Hospital Peshawar from July 2013 to June 2015. A total of 17 consecutive patients with Chiari I malformation who undergone posterior fossa reconstruction and fulfilled inclusion criteria were included in the study irrespective of their age and gender. The patients were assessed postoperatively for clinical and radiological (syringomyelia) improvement and post operative complications for a period of 6 months.*

Results: *We had total 17 patients who underwent posterior fossa reconstruction for Chiari I malformation. There were 41.2% males and 58.8% females. Age of the patients ranged from 10 – 52 years with the mean age of 31 years. The most common clinical features were headache (64.7%) and sensory impairment (58.8%). We had syringomyelia in 70.1% and scoliosis in 47.1% cases. After surgery headache improved in 81.1% patients, sensations in 70% cases and syringomyelia in 91.1% cases. The overall morbidity in the form of complications was in almost 47% cases. One patient (5.9%) died after surgery.*

Conclusions: *We conclude that posterior fossa reconstruction is a safe and effective procedure for the management of Chiari I malformation. This results in improvement in patients symptoms with acceptable complications. These complications respond to conservative treatment most of time.*

Key Words: *Type I Arnold – Chiari malformation, Posterior cranial fossa, De-compressive craniectomy, congenital abnormality.*

Abbreviations: *CM = Chiari – like malformation, CMI = Type I Chiari malformation, FM = Foramen magnum, FMD = Foramen magnum decompression, MTI = Medical and teaching institute, CTH = Cerebellar tonsil Herniation.*

INTRODUCTION

Chiari – like malformation (CM) is a malformation of the hindbrain and the surrounding caudal cranial fossa. In this condition there is small posterior fossa relative to its contents (cerebellum and brain stem).[1,2]

Chiari malformation was first described in 1895.[3,4] It has four different types. Type I Chiari malformation (CMI) is a complex disorder of the craniospinal system that is radiologically defined by cerebellar tonsillar herniation greater than 3 – 5 mm below the foramen magnum (FM) or McRae line (between basion and opisthion).[5-7] It is measured using T_2 mid sagital images of MRI taken at posterior fossa and cervical spine. In contrast with Chiari II and III malformations, in Chiari type 1 the rest of hindbrain structures remain within the posterior fossa.[4,5]

The clinical features in this condition are due to impaired cerebrospinal fluid dynamics, brain stem compression and spinal cord symptoms due to syrinx formation.[8-10] The most frequent symptoms are headache, ocular disturbances, vertigo, sleep apnea and lower cranial nerve signs, including tongue fasciculation, dysphagia or dysarthria. Motor and sensory symptoms derived from spinal cord disturbances asso-

ciated with syringomyelia or scoliosis.[4,6,11] MRI is the investigation of choice for the diagnosis of Chiari malformation and syringomyelia.[6,10,12]

The aim of treatment for Chiari I malformation is to improve symptoms by maintaining cerebrospinal fluid flow across cranio-vertebral junction, decompression of brain stem and resolution of syringomyelia. Two major surgical procedures, are performed foramen magnum decompression (FMD).

1. With or without reconstruction of posterior fossa.
2. **Shunting** of fluid from syrinx are performed as e.g. syringo-subarachnoid shunt.[13]

As there is limited study on Chiari malformation, this study of posterior fossa reconstruction for Chiari malformation type I will help us to know the effectiveness of the procedure and help to improve health of such patients in our population.

MATERIALS AND METHODS

This observational study was conducted at the department of Neurosurgery, medical and teaching institute (MTI), Lady Reading Hospital Peshawar from July 2013 to June 2015.

Inclusion Criteria

A total of 17 consecutive patients with Chiari I malformation who undergone posterior fossa reconstruction and fulfilled inclusion criteria were included in the study irrespective of their age and gender. We included patients with Cerebellar tonsil Herniation (CTH) greater than 3 – 5 mm below the Foramen Magnum (Chiari I malformation) and were symptomatic.

Exclusion Criteria

Those Patients were excluded from our study that had secondary causes of CTH, such as hydrocephalus, had undergone previous decompression surgery or had implanted CSF shunts. Those patients who were unfit for surgery or had other types of Chiari malformations were also excluded from the study. After getting approval from the hospital ethical committee to conduct the study and taking informed consent, the medical record of patients who underwent posterior fossa reconstruction for Chiari I malformation was evaluated. The patients were followed for 06 months postoperatively. The patients were assessed postoperatively for improvement of symptomatology, resolution of syrinx (on MRI) and any complication. All information was entered into a proforma especially designed for this purpose. The data was analyzed by statistical program SPSS version 17.

Operative Steps of Posterior Fossa Reconstruction

Under general anesthesia and tracheal intubation, patient was in prone position. Incision is given from occipital protuberance to C_2. Exposure of sub-occipital bone and C_1 arch was made. Sub-occipital craniectomy and C_1 posterior arch excision to make a defect of 3 in to 3 cm. epidural band is released. The dura mater is opened along with arachnoid membrane to facilitate CSF flow and all visible arachnoid adhesions were released. After an adequate intradural decompression the defect was reconstructed (expansile duraplasty) by either large pericranium or facia lata graft to expand the compressed cisterna magna or the creation of a pseudocisterna magna. Then the wound was closed in reverse order.

RESULTS

Gender Distribution

Out of total 17 patients we had 7 male and 10 female patients as given in Table 1.

Table 1:

Gender	No of Patients	Percentage	Ratio
Male	07	41.2%	01
Female	10	58.8%	1.4
Total	17	100%	

Age Distribution

Age of the patients ranged from 10 years to 52 years. The mean age was 31 years.

Clinical Features / MRI

Presenting Clinical features of the patients are given in table 2.

Radiological Investigation

Magnetic resonance imaging (MRI) was done in all the patients.

Table 2:

Clinical Feature	No of Patients (n = 17)	Percentage
Headache	11	64.7%
Dizziness / Vertigo	05	29.4%
Motor weakness	07	41.2%
Sensory features	10	58.8%
Scoliosis	08	47.1%
Syringomyelia	12	70.1%

Improvement of Symptoms

Table 3: *Improvement.*

Clinical Feature	No. of Patients	Improved Patients	Percentage
Headache	11	09	81.1%
Dizziness / vertigo	05	03	60%
Motor weakness	07	02	28.6%
Sensory features	10	07	70%
Scoliosis	08	02	25%
Syringomyelia	12	11	91.7%

Complications

We had wound infection in 3 (17.6%), pseudomeningocele in 2 cases (11.8%), CSF leak in 2 (11.8%) and meningitis in one (5.9%) patient. The overall complication rate in our study is almost 47%.

Mortality

One patient died because of meningitis.

DISCUSSION

Chiari Malformation type I [CMI] is a congenital disorder involving chronic caudal Cerebellar tonsil herniation (CTH) into the cervical canal. It is also known as adult-type Chiari malformation and usually presents after the second or third decade of life.[14]

We had total 17 patients with Chiari malformation who underwent posterior fossa reconstruction surgery. The age of the patients ranged from 10 – 52 years with the mean age of **31 years**. In our study there were 58.8% female patients and 41.2% male, with the **male/female** ratio of 1:1.4. The higher prevalence of **women** with Chiari malformation has been previously documented in the literature and is confirmed here within our study population.[15] Nicholas Shaffer, et al,[13] studied 23 patients with Chiari malformation. Their patients had Mean age of 38years with a range of 17 – 58 years. The gender distribution was 78.3% (18) female and 21.7% (5) male. In another study total patients were 364. There were 275 (75.5%) female and 89 (24.5%) male patients.[6] Williams, et al,[16] studied 34 patients and the mean age was 38.7 years. There were 28 females and 6 males in their study.

Patients with Chiari Malformation I (CMI) can have a variety of neurological symptoms and pain ranging from mild to severe.[6] CMI causes neurological dysfunction by direct compression of the neural tissue at the cranio-vertebral junction or cerebrospinal fluid disturbances that give rise to syringomyelia or hydrocephalus.[4,6,11]

In our study the most common features were headache (64.7% cases) and sensory abnormality (58.8% cases) in the form of paresthesias or anesthesia. Syringomyelia was observed in 70.1% and scoliosis in 47.1% patients. Almost similar results were also published in other studies. In a study of 415 cases, headache was the most prevalent symptom (69.1%), followed by sensory abnormality (paresthesias / sensory loss) in 60.4% cases. Syringomyelia was present in 35.1% of cases.[4] Hwang and colleagues[14] in their study reported that the main symptoms of patients with Chiari malformation I included sensory changes (58.3%), posterior neck pain (50%) and motor weakness (33.3%). Almost same results were published in other studies. In one study of 364 patients the most common associated features with Chiari malformation were syringomyelia (65%) and scoliosis (42%).[6] In literature 50 – 76% of patients, Chiari malformation is associated with hydromyelic cavitation of the spinal cord and medulla oblongata.[17]

MRI is the investigation of choice for the diagnosis of Chiari malformation and syringomyelia.[6,10,12] Because of this reason we did Magnetic resonance imaging (MRI) in all the patients.

In our study period the most common clinical features which improved after surgery were headache (in 81.1% cases) and sensory symptoms (70%). Syringomyelia improved in 91.7% cases and scoliosis in 25% cases. Here again results are identical to the literature. Guyotat and colleagues[18] studied 75 patients with having syringomyelia associated with Chiari I malformation of which 8 patients undergone foramen mag-

num decompression (FMD) and cerebellar tonsils resection. Postsurgical MRI syrinx resolution was observed in all 8 patients either in the early postoperative course or on delayed follow up. It is suggested that tonsils resection might enhance the results of FMD in individuals having Chiari I – related syringomyelia.

Chiari malformation Type I (CM-I) is often associated with scoliosis. Attenello et al studied 21 patients underwent hindbrain decompression for CM – I – associated scoliosis. They concluded that over one third of patients with CM – I – associated scoliosis will improve after cervico-medullary decompression. Euleues and colleagues had[19] patients with Chiari malformation and scoliosis out of which 5 patients improved after foramen magnum decompression. This makes about 26.3% results which is again comparable to our study.

Like other surgical procedures posterior fossa reconstruction can also have post op complications. In our study the most common complications were wound infection 3 (17.6%), pseudomeningocele 2 cases (11.8%), CSF leak 2 (11.8%) and meningitis in one (5.9%) patient. The overall complication rate in our study is almost 47%. Williams, et al,[16] studied 34 patients with posterior fossa reconstruction for Chiari malformation and reported the overall complication rate of 41%. In their study the pattern of complications was different. Meningitis occurred in 9% of the patients, pseudo-meningocele in 26.5% and CSF leak in 5.9% cases. None of their patients had post operative wound infection. The exact reason for the difference in complications is not clear but could be because that we had more cases of wound infection and small sample size as compared to their study.

In our study the post operative complications were treated conservatively and none of the patients needed re-exploration of the wound. Pseudomeningocele cases were picked on MRI and on clinical examination they were not significant. It should be noted that one of the limitations of our is the small sample size.

CONCLUSION

We conclude that posterior fossa reconstruction is a safe and effective procedure for the management of Chiari I malformation. This results in improvement in patients symptoms with acceptable complications. These complications respond to conservative treatment most of time.

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