ORIGINAL ARTICLE

Posterior Fossa Decompression with Duraplasty in Chiari-1 Malformations

Lal Rehman, Hamid Akbar, Iram Bokhari, Asghar Khan Babar, A. Sattar M. Hashim and Safdar Hussain Arain

Abstract

Objective: To evaluate the symptomatic outcome after PFD (Posterior Fossa Decompression) with duraplasty in Chiari-1 malformations.

Study Design: Case series.

Place and Duration of Study: Department of Neurosurgery, JPMC, Karachi, from July 2008 to September 2012.

Methodology: This included 21 patients of Chiari 1 malformations admitted in department through OPD with clinical features of headache, neck pain, numbness, neurological deficit, and syringomyelia. Diagnosis was confirmed by MRI. PFD followed by C1 laminectomy with duraplasty was done in all cases and symptomatic outcome was assessed in follow-up clinic.

Results: Among 21 patients, 13 were females and 8 were males. Age ranged from 18 to 40 years. All the patients had neck pain and numbness in hands. Only 3 patients had weakness of all four limbs and 12 with weakness of hands. Symptoms evolved over a mean of 12 months. Syringomyelia was present in all cases. All patients underwent posterior fossa decompression with duraplasty with an additional C1 laminectomy and in 2 cases C2 laminectomy was done. Syringo-subarachnoid shunt was placed in one patient and ventriculo-peritoneal shunt was placed in 2 patients. Pain was relieved in all cases. Weakness was improved in all cases and numbness was improved in 19 cases. Syringomyelia was improved in all cases. Postoperative complications included CSF leak in 2 patients and wound infection in one patient. However, there was no mortality.

Conclusion: Posterior fossa decompression with duraplasty is the best treatment option for Chiari-1 malformations because of symptomatic improvement and less chances of complications.

Key Words: *Chiari-1 malformation. Posterior fossa decompression. Duraplasty.*

INTRODUCTION

Arnold-Chiari malformation, or Chiari malformation, is described as a downward displacement of the cerebellar tonsils through the foramen magnum,[1] sometimes causing non-communicating hydrocephalus as a result of obstruction of cerebrospinal fluid outflow. The cerebrospinal fluid outflow is caused by phase difference in outflow and influx of blood in the vasculature of the brain. Despite impressive advances in clinical diagnosis, neuroradiology, and neurosurgery, central questions about the pathogenesis, natural history, and prevalence of this condition remain unanswered.[2] Most patients present in adulthood with slight female predominance.[3]

The clinical presentation of these patients is often highly variable. They often present with a multitude of seemingly unrelated complaints and frequently have normal neurological examinations.[4] Predominant symptoms are usually headaches, fatigue, muscle weakness in the head and face, difficulty swallowing, dizziness, nausea, impaired co-ordination, and paralysis in severe cases. MRI is the best diagnostic choice. The goal of surgery is to restore normal CSF dynamics to the craniocervical junction. However, the extent of surgery required to achieve this has not been elucidated and many different operative techniques have been recommended. Although the necessity for some bony removal is universally accepted, other aspects of Chiari surgery are the subject of debate. The most controversial points include the optimal amount of bony removal, the use of duraplasty (the type of material), the need for subarachnoid dissection, and the need for tonsillar shrinkage.[5] PFD with duraplasty is preferred surgical option in Chiari-1 malformation (CM1), with syrinx. Other options included suboccipital craniectomy, syringostomy, obex plugging, syringo-subarachnoid shunting, 4th ventriculo-subarachnoid shunting and transoral clivus-odontoid resection in cases with ventral brain stem compression. Management of Chiari-1 is controversial, in part because there is no widely used quantitative measurement of decompression.[6]

The objective of this study was to determine the symptomatic outcome and complications after PFD with an additional C1 Laminectomy (CL) followed by duraplasty in these CM1.

Department of Neurosurgery, Jinnah Postgraduate Medical Centre, Karachi.

Correspondence: Dr. Lal Rehman, Associate Professor, Neurosurgery Department, JPMC, Karachi.
E-mail: drlalrehman@yahoo.com

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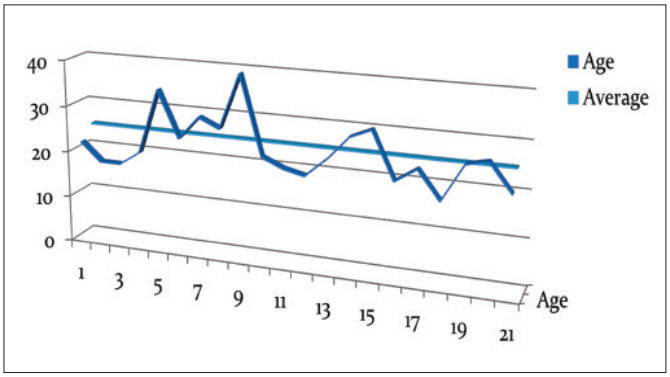

Figure 1: Age of the patients with CM1 (n=21).

Table I: Symptomatic outcome after PFD with duraplasty in Chiari-1 malformation (n=21).

Pre-operative symptoms (n+21)	Patients in whom postoperative improvement was not seen in symptoms			
	Pain	Hand weakness	Numbness	L/L
Pain (n=21)	0	--	--	--
Hand weakness (n=12)	--	03 (25%)	--	--
Numbness (n=21)	--	--	02 (9.52%)	--
L/L* (n=3)	--	--	--	01 (33.33%)

** Involvement of lower limbs.*

Table II: Pre and postoperative complications (n=21).

Complications	Patients
CSF leak	02 (9.52%)
Wound infection	01 (4.76%)
Deterioration	NIL
Vertebral artery injury	O1 (4.76%)
Mortality	NIL

METHODOLOGY

This study included 21 patients of CM1 admitted in department through OPD with clinical features of headache, neck pain, numbness, neurological deficit, and syringomyelia. Those who were previously operated, with other co-morbid or congenital disorders were excluded from this study. Diagnosis was confirmed by MRI. These patients were then admitted in the ward. After taking written consent, PFD followed by an additional CL with duraplasty was done in all cases by a posterior approach. Complications occurring during and after surgery were noted. Symptomatic outcome was assessed in follow-up clinic upto 6 months. In these patients presence of symptom i.e. pain, hand weakness and numbness, and involvement of lower limbs were noted in all patients on admission. After surgery, it was again noted that whether these symptoms were improved or not after surgery in these patients.

The data was collected in a proforma and analyzed on SPSS version 20. Frequencies and percentages were presented for categorical variables while mean and standard deviation were presented for numerical variables.

RESULTS

Among 21 patients, 13 were females and 8 were males. Age ranged from 18 to 40 years (Figure 1). All patients had neck pain and numbness in hands. Only 3 patients had weakness of all the four limbs. Weakness of hand muscles was present in only 12 patients. Symptoms were evolved over a mean of 12 months ranging from 6 months to 3 years. Syringomyelia was present in all

Table III: Synopsis of previously published clinical series with technique and outcome .

Authors, year	Patients	Pathology	Surgical approach	Outcome	Morbidity Mortality Complications
Krieger *et al.*[21], 1999	31	CM-1 ± HSM*	Occipital craniectomy, CL + duraplasty	Syrinx: 88% improvement, 3 patients also required a shunt	26% headaches 16% nausea No mortality
Alden *et al.*[16], 2001	21	CM-1 ± HSM	Suboccipital craniectomy + CL ± (i) durotomy + duraplasty:4, (ii) cerebellar tonsillectomy + adhesiolysis + duraplasty: (17)	67% symptom resolution, 29% improvement, 4% no improvement	N/A
Parker *et al.*[21] , 2011	114	CM-1	Occipital craniectomy, CL, duraplasty ± tissue sealant	Re-operations	Graft/ sealant failures, Cumulative complications: 21%
Mottolese *et al.*[17], 2011	82	CM-1 ± HSM,	(i) Occipital craniectomy, CL+ duraplasty (A: 43 patients) (ii) Occipital craniectomy, CL,Gore-Tex (B: 9 patients)	A: 70% improvement B: 89% improvement	Complications; Group a: 18%: Group b: 20,5% No mortality
Sindou *et al.*[13], 2014	44	CM-1, ± HSM	Craniocervical decompression + far lateral foramen magnum opening + duraplasty + arachnoid preservation.	Improvement of syrinx in 60%, stabilization of syrinx in 40%	4.5% CSF leak, 2.3% laryngeal edema, 2.3% pneumonia, 11.4% wound infections. No mortality
Hida *et al.*[18], 1995	70	CM-1, ± HSM	A:33 patients foramen magnum decompression, B:37 patients shunting	A: 94% reduced syrinx, 82% :improved B:100% syrinx reduced, 97%: improved	N/A

** HSM: hydrosyringomyelia*

cases. All patients underwent PFD followed by an additional C1 laminectomy with duraplasty. However, in 2 patients C2 laminectomy was added further. Ventriculo-peritoneal shunt was placed in 2 patients and syringo-subarachnoid shunt was done in one patient. Pain and weakness in upper limbs was relieved in all cases. Numbness was improved in all except 3 patients. Lower limb involvement was not improved after surgery in one patient (Table I). Syringomyelia was improved in all cases except one patient for which syringo-subarachnoid shunting was done. Vertebral artery was injured during surgery in one patient which led to troublesome bleeding. However, it was ligated and the postoperative recovery was uneventful. Postoperative complications included CSF leak in two (9.52%) patients and wound infection in one (4.76%) patients (Table II). In one patient lumbar subarachnoid drain was placed to decrease CSF leak. However, there was no mortality and none of the patients were re-explored due to complications.

DISCUSSION

CM1 is described as a caudal descent of cerebellar tonsils (> 5 mm) below the foramen magnum. Generally there is no descent of the brainstem and not associated with myelomeningocele. It is associated with syrinx in 50 - 70% of the cases. May be associated with hydrocephalus (1 - 10%). Platybasia, basilar invagination, Klippel-Feil syndrome, and scoliosis are also seen along with these malformations. Various theories have been proposed regarding its pathogenesis but none is universally accepted. The most popular hydrodynamic theory of Gardner stated that delayed opening of the 4th ventricle outlets results in transmission of an arterial pulse through a patent obex, producing a water hammer effect that enlarges the central canal.[7] Another theory of craniospinal pressure dissociation by Williams described that different pressures that existed between the cranial and spinal cavities was a causative factor for syringomyelia.[8] Major presenting symptoms are pain, headache, numbness and weakness of the hand and spastic paresis of the lower limbs as were also reflected in this series. Dyste *et al*. showed the same predominant symptoms.[9]

Where syringomyelia exists as in 30 - 70% of cases, treatment is required.[10] While surgical therapies in these malformations focus on PFD, surgical treatment alternatives for syringomyelia vary widely.[11,12] Sindou *et al*. proposed craniocervical decompression, far lateral foramen magnum opening, duraplasty with arachnoid preservation[14] Logue and Edwards reported cases of laminectomy and syringo-subarachnoid shunt after PFD had failed but obtained poor results.[15] Good results were seen when PFD was combined with obex obliteration, 4th ventricle shunting and a dural graft.[13] Suboccipital craniectomy,[11] tonsil resection,[15] ventriculo-subarachnoid shunt,[12] syringo-peritoneal shunt,[16] and occipital craniectomy with CL and Gore-Tex dural patch[17] are the other common options. Another study showed that PFD, CL and duraplasty for the treatment of Chiari-1 malformations may lead to a more reliable reduction in the volume of concomitant hydromyelia, compared with PFD and CL alone.[6] Similar results were seen in this series as almost all cases improved with disappearance of syrinx formation after this approach.

Although PFD remains first line treatment in patients with Chiari-1 malformations complicated by syringomyelia in many cases. Hida *et al*. proposed that clinical symptoms and radiological findings improved much more quickly in the patients in whom syringo-subarachnoid shunting was done.[19] In one patient in this study, syrinx which was already large in size failed to resolve after surgery with persistence of symptoms in this particular patient. So syringo-subarachnoid shunting was done after which syrinx collapsed along with improvement in symptoms in this patient. Thus, indicated placement of shunt in the presence of a sufficiently large syrinx appears to be beneficial. The question of when to place a shunt, however, requires further, preferably prospective investigation and further studies.

Symptomatic improvement was seen in majority of patients included in this study after surgery. However, in few patients hand weakness, numbness and involvement of lower limbs was not improved after PFD. The probable explanation for these may be that probably these patients presented late. Thereby, permanent changes may have occurred in the cord. Thus, there was no improvement seen in such patients. However, it may be expected that further deterioration may be delayed in such patients. It has been observed, in patients with Chiari associated syringomyelia, that motor symptoms are more likely to improve with hindbrain decompression than paraesthesias or anaesthesia symptoms. However, Alzate *et al*. concluded from a series of 66 patients that pain was most likely to resolve than sensory and motor deficit after decompressive surgery in such patients.[20]

Surgical intervention in such malformations has been associated with a wide spectrum of intraoperative and postoperative complications.[20] The complication rate in this series was 14.2% which is much lower than when by using other methods as seen in Table III. Meticulous knowledge of all potential complications is of paramount importance for their complete prevention or their minimalization and also for their early and prompt management when they occur. Thorough knowledge of the posterior fossa anatomy is mandatory for avoiding any vascular injuries to a remnant circular venous sinus, the extracranial vertebral arteries, or the posterior inferior cerebellar arteries. Brain herniation can lead to

variations in the vascular anatomy thus making patient susceptible to vascular injuries. This complication occurred in one of the patients included in this series. The probable explanation could be failure to recognize the vessel due to variable course due to distortion of anatomy due to cerebellar descent. It was, therefore, ligated. However, the postoperative recovery was uneventful.

Another recognized postoperative common complication includes CSF leak,[21] after PFD with duraplasty. Thus, meticulous water-tight dural closure is important for avoiding any postoperative leaks. The optimal suture material for sewing in dural grafts is 7 - 0 Gortex suture utilizing interrupted sutures, as running stitches may loosen or pull out. Furthermore, the advantage of this suture is that the needle is smaller than the suture itself; thus the hole created by the needle is filled with the suture itself limiting the hole as a potential source of leakage. Absence of CSF leak should further be confirmed by Valsalva's maneuver before closure of the wound. However, the failure to prevent a CSF leak following duraplasty closure results in the most common complication; pseudomeningocele formation.[22] This results in incisional CSF leaks, chemical/aseptic, and bacterial meningitis, the development of hydrocephalus, and the potential for wound dehiscence. Despite taking meticulous care during closure of duramater and, thereafter, checking for leak by Valsalva's maneuver, CSF leakages were observed in two of the patients included in this study. Both these patients were managed non- operatively. In one of the patients the CSF leak stopped after few days and was thus thereafter discharged. However, it was managed by placing a lumbar drain and with high dose antibiotics to prevent meningitis in the other patient as the leak was large. Although it took longer to settle but, however, it stopped after a drain and patient was discharged on 14th postoperative day.

Several deaths have been reported after posterior fossa decompression for Chiari malformations, with a reported mortality rate of 3% and 1%.[23-25] However, in this series, there was no mortality and none of the patients had to undergo re-exploration thus indicating PFD with duraplasty as a safe and effective technique in terms of complications and symptomatic relief.

CONCLUSION

PFD with duraplasty is best treatment option for Chiari-1 malformations because of symptomatic improvement and less chances of complications as well as with zero mortality rate.

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