ORIGINAL ARTICLE

Outcome of Suboccipital Decompression with and without Duraplasty in Adults with Chiari I Malformation

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ABSTRACT

Objective: *To investigate outcome for Arnold Chiari 1 Malformation (ACM1) patients based on intervention methods, i.e., posterior fossa decompression without duraplasty and with duraplasty in terms of symptomatic and functional improvement.*

Materials and Methods: *This study was conducted prospectively over 41 months from January 2013 to May 2016. Patients with confirmed diagnosis of ACM-1 were included in the study. Patients either underwent posterior fossa decompression without duraplasty or with duraplasty, depending upon the severity of tonsillar descent, the presence of syrinx, neurological deficits or hydrocephalous. Data was collected on pre-designed pro forma both pre-operatively and during follow up. Outcome was assessed according to Chicago Chiari Outcome Scale (CCOS). Data analysis was done using SPSS v 22.0.*

Results: *Posterior Fossa Decompression (PFD) without dural opening was performed in 13 patients while in 8 patients duraplasty was performed. Overall mean age was 32.95 ± 5.88 years and mean symptoms duration was 21.62 ± 8.82 months. The most common complaints were headache (76.2%), neck pain (61.9%), hand and arm weakness (47.6%), gait disturbance (47.6%) and cranial nerve dysfunctions (76.2%). The median preop CCOS was 10 ± 1.57 while postoperative CCOS was 13 ± 2.27. There was a significant relief in terms of CCOS improvement in preop and postoperative scores (P<0.0001). Similarly, on Chi-square analysis there was no significant difference between favourability of outcome for the two types of intervention. 50% favourable outcome was obtained for PFD without duraplasty and 50% for PFD with duraplasty. On Chi-square analysis, PFD with duroplasty was significantly associated with the occurrence of postoperative complications (p = 0.006). The commonest complication was CSF leak in 14.3% of patients. There was no mortality. No recurrent cases were noted during the 41-months study period.*

Conclusion: *Posterior fossa decompression forACM-I is simple and effective. Further studies regarding surgical outcome and development of outcome assessment are required with larger patient cohorts.*

Keywords: *Arnold Chiari Malformation, Posterior fossa decompression, Chicago Chiari Outcome Scale, Duraplasty.*

Abbreviations: *CCOS: The Chicago Chiari Outcome Scale. CSF: Cerebrospinal Fluid. CV: Craniovertebral. OPD: Out Patient Department. SD: Standard Deviation. HMC: Hayat Abad Medical Complex. ACM1: Arnold Chain 1 Malformation.*

INTRODUCTION

Arnold-Chiari malformations are closely related abnormalities of the craniovertebral (CV) junction. There are four types out of which the first two are the most prevalent subtypes.[1] The more clinically important disorder, Arnold-Chiari Malformation type 1 (ACM1) is prevalent in young adults and may significantly affect neurological function of an individual.[2] The treatment of ACM1 is frequently surgical in symptommatic cases or when neurological deficits starts appea-

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ring.

The most widely used surgical intervention is posterior fossa decompression also called suboccipital decompression. Various modifications to this basic approach have been described in literature, the main aim of which is to open the shallow posterior fossa in order to allow room for the contents and relieve pressure on the brainstem. These modifications are, dural opening and repair with an expanding graft, C1 laminectomy, tonsillectomy and shunting of associate syrinx.[3]

There is a difference of opinion regarding performing posterior fossa decompression either with or without duraplasty. This difference in opinion originnates from various studies reporting variable outcome for both approaches.[4,5] The most acceptable argument regarding outcome for ACM1 surgery is the paucity of outcome measure instruments. There is no unified instrument which can be used to measure postoperative long and short term outcome. Generally, neurological functional improvement, pain relief, gait improvement and a halt in the progression of the disease are believed to be considered during follow-up evaluation.[6,7]

The Chicago Chiari Outcome Scale (CCOS) was used by several authors to measure the postoperative course of patients. This scale has been subjected to critical external validation and found useful in tracking sensory, motor, functional improvements as well as the occurrence of postoperative complications.[8-10]

We aim to present out experience with the long-term outcome of our patients who were operated for symptomatic ACM1 in terms of improvement in symptomatology, as well as functional improvement and complication occurrence. We hope that this will help understand the postoperative course of the disease and the amount of relief that can be provided with surgical decompression.

MATERIALS AND METHODS

This is a prospective study of 41 – month duration (January 2013 – May 2016) conducted at the department of neurosurgery, Hayatabad Medical Complex Peshawar. Informed consent was obtained from all patients before inclusion in the study. Approval was also taken from the institutional ethical committee before commencement of the study.

All symptomatic patients with a confirmed diagnosis of ACM1 after outpatient and inpatient workup, between the ages of 16 and 65 years irrespective of their gender or ethnicity were included. Patients with significant trauma to the cervical or craniocervical junction, those with significant basilar invagination and those who were previously operated were exclude. Patients who were bed bound for more than 2 years were also excluded.

Patients with confirmed diagnosis of CM-I were admitted to the ward. A detailed preoperative history and examination were performed and the complaints were rated according to the CCOS. The preoperative MRI findings were also recorded about the level of the tonsillar herniation, the presence or absence of syrinx, platybasia, basilar invagination and the presence or absence of hydrocephalous.

The decision for dural opening was based on the presence of higher degree of tonsillar herniation, syringomyelia, severe long tract signs and multiple cranial nerves involvement.

Operative Procedure

After induction of GA, intravenousceftriaxone (1g) were administered. Patient was positioned prone with bony prominences well padded. A vertical midline incision was employed, exposing the occipital bone and downward up to C2 lamina. Suboccipital craniectomy was performed in a standard manner with at least 5×5 cm dimensions or more.C1 ring was removed along with decompression of posterior fossa.

The duraplasty group received a dural opening in a Y-shaped manner, arachnoid adhesiolysis and expansion duraplasty was performed using fascia lata, periosteum or synthetic dural substitute followed by water tight closure.

Follow-up

Patients were called for follow-up at 2 weeks for their initial appointment and monthly for the first 6 months. Thereafter, 3 to 6 monthly follow-up was advised. All patients were assessed in OPD clinic according to the CCOS. Excellent outcome was recorded in those having a score of 16 or more, functional outcome in those with a CCOS score of 12, impaired outcome with CCOS of 8 and incapacitated were those who had 4 or less CCOS.

Data Analysis

Data was entered and analysed using SPSS version 22.0. Mean ± standard deviations were calculated for continuous variables, while frequency and percentages were calculated for categorical variables. A paired

samples t-test was used to ascertain difference between pre- and post-operative improvement or deterioration.

RESULTS

Patient Demographics

21 patients were operated during the 41-month period, including 11 males (52.4%) and 10 females (47.6%) in a ratio of 1.1 to 1. Overall mean age was 32.95 years ± 5.88 SD (range: 20 – 45). The overall mean duration of symptoms was 21.6 months ± 8.82 SD. Overall median CCOS was 10 ± 1.58. The overall post-operative median CCOS was 13 ± 2.27 SD. The overall mean length of stay (LoS) was 6.57 ± 3.06 SD.

Table 1: *Clinical features of patients with ACM-1 and their distribution in the two treatment groups.*

Variable	Decompression without Duraplasty N = 13	Decompression with Duraplasty N = 8	Significance
Gender			0.1
Male	5 (38.5%)	6 (75%)	
Female	8 (61.5%)	2 (25%)	
Headache	10 (76.9%)	6 (75%)	0.9
Neck Pain	8 (61.5%)	5 (62.5%)	0.9
Gait disturbance	6 (46.2%)	4 (50%)	0.8
Upper limb weakness	7 (53.8%)	3 (37.5%)	0.5
Muscle wasting	6 (46.2%)	5 (62.5%)	0.50
Cranial nerve dysfunction	10 (76.9%)	6 (75%)	0.9
Syrinx	7 (53.8%)	3 (37.5%)	0.5
Complications			0.006
CSF leak	-	3 (37.5%)	0.01
Pseudomeningocele	-	2 (25%)	0.06
Wound infection	-	2 (25%)	0.06
Meningitis	-	1 (12.5%)	0.1
Age (years)	32.69 ± 5.92	33.38 ± 6.19	0.8
Symptoms duration (months)	22.92 ± 7.94	19.50 ± 10.29	0.4
Preop CCOS	9.23 ± 1.3	10.63 ± 1.68	0.05
Postop CCOS	12.15 ± 2.44	12.88 ± 2.03	0.5
Length of Stay (days)	5.0 ± 1.35	9.13 ± 3.39	0.001

Clinical Presentation & Perioperative Findings

Headache was present in 16 (76.2%) patients, neck pain in 13 (61.9%), gait disturbance in 10 (47.6%) and dysesthesias in 13 (61.9%). Upper limb weakness was present in 19 (47.6%), muscle wasting in 11 (52.4%), cranial nerve dysfunction in 16 (76.2%). 10 (47.6%) of patients had underlying syrinx, however, none of them were significant (> 2 mm diameter).

13 (61.9%) patients underwent posterior fossa

Table 2: *Outcome representation for the two treatment types; shaded cells show decompression without duraplasty, clear cells show decompression with duraplasty.*

	1	2	3	4
Pain	-	N = 2 (15.4%)	N = 4 (30.8%)	N = 7 (53.8%)
	-	N = 2 (25%)	N = 6 (75%)	-
Non-pain	N = 4 (30.8%)	N = 2 (15.4%)	-	N = 7 (53.8%)
	-	N = 1 (12.5%)	N = 2 (25%)	N = 5 (62.5%)
Functionality	N = 2 (15.4%)	N = 4 (30.8%)	N = 5 (38.5%)	N = 2 (15.4%)
	N = 1 (12.5%)	N = 1 (12.5%)	N = 5 (62.5%)	N = 1 (12.5%)
Complications	N = 2 (15.4%)	-	N = 2 (15.4%)	N = 9 (69.2%)
	N = 3 (37.5%)	-	-	N = 5 (62.5%)
Total score	4: Incapacitated	8: Impaired	12: Functional	16: Excellent

decompression without duraplasty while 8 (38.1%) of patients were operated by decompression plus duraplasty.

The mean follow-up duration was 18 months ± 5.6 SD. Complications included CSF leak in 3 (14.3%), wound infection in 2 (9.5%), meningitis in 1 (4.8%) and pseudomeningocele in 2 (9.5%) of patients.

Overall the outcome as graded by CCOS was excellent in 6 (28.6%), functional improvement with some residual symptoms in 12 (57.1%) and 3 (14.3%) patients obtained very little improvement in their symptoms.

Table and Table represent the clinical features stratified for the two intervention techniques along with the Chi-square and independent samples t-statistics.

A paired samples t-test was performed to see the mean difference in preoperative and postoperative CCOS score. The correlation between pre- and postoperative score was strong (0.61, p = 0.003). A mean difference of -2.67 ± 1.79 was obtained (95% CI: -3.5 to -1.8), with a t score = -6.8, and p value of < 0.0001.

Outcome at Follow-up

The mean postoperative Chicago Chiari Outcome Score for decompression without duraplasty was 12.1 ± 2.4while for the duraplasty group, it was 12.8 ± 2.03 SD. The table below shows scores for both treatment groups with their frequencies and percentages.

DISCUSSION

ACM1 is a rare disorder of CV junction with a degenerative and congenital component. There are various theories regarding its development. True incidence is yet unknown, Meadows J et al[11] in MRIs of asymptomatic cases estimated it to be approximately 1%. Out of these, nearly 20% are estimated to become symptomatic over the course of few years.[6,11,12]

MRI of the CV junction with posterior fossa and cervical spine cuts is the study of choice along with help obtained from multiplanar CT for evaluation of bony structures.[13,14] Cine MRI is still used as a research tool in studying the pathophysiology around CV junction.[15] Posterior fossa volume can be estimated using MRI images and is helpful in deciding appropriate surgical modality.[16]

Decompression of the posterior fossa and upper C-spine without opening the dura is one of the simplest surgical intervention. However, the biomechanics and pathophysiological findings around CV junction in ACM1 are not usually that simple and in some cases complex interventions such as dural opening, adhesiolysis, manual tonsillar reduction and expansion duraplasty are necessary. In the present study, we operated 13 patients without opening the dura and simple decompression of the posterior fossa with C1 laminectomy while 8 patients were operated with dural opening, duraplasty and adhesiolysis. However, no cases required manual tonsillar reduction or amputation of the tonsils. The symptomatology improved in more than 75% of our patients while more than 60% achieved good functional outcome. Kennedy BC et al[6] has reported good to excellent outcome in 91% of their patients during the 3-year follow-up period while Chotai S et al[17] has shown favourable outcome in 90% patients. Gurbuz et al[18,19] have purely favoured PFD with duroplasty, however, accepting that PFD with duroplasty have higher complications rates in the form of CSF leaks, pseudomeningocele and subsequent risk of wound infection and meningitis.

Statistically no significant beneficial effect of simple decompression or decompression with duraplasty was observed. Moreover, both procedures showed a favourable final outcome in more than 50% of patients (favourable outcome: 50% for without duraplasty and 50% for decompression with duraplasty). As is shown in a systematic review by Hankinson et al,[20] there are currently no level I evidence which supports the notion that either procedure is significantly superior to the other. However there are multiple reports which favour the application of PFD without dural opening because of the simplicity of the procedure, technically less demanding, low risk of serious complications and equivalent outcome as compared to duraplasty group.[6,21,22,23] Individual case series such as one by Chotai S el al[24]has shown the favourability of PFD without duraplasty especially taking into account the higher rate of CSF related complications in the duraplasty group, which are more worrisome to the neurosurgeon. CSF related complications lead to need for further procedures and consequently longer morbidity and prolonged longer hospital stay. Gurbuz MS et al[18,19] and also some other studies[25] have recommended PFD with duraplasty due to direct manual approach to arachnoid adhesions and removal of the pathophysiologic barriers around the CV junction.

Majority of patients in our series were young adults (mean age: 32.95 ± 5.88 years) with slight male predominance (1.1:1). The mean symptoms duration was 21.62 ± 8.82 months. Clinical features included pain (headache, neck pain) in more than 75%, sensory

abnormalities in over 50% and motor problems such as hand weakness, gait disturbance, sphincters disturbance was present in more than 45% of patients. Cranial nerve involvement was noted in 76% patients. Chotai S et al[17] has reported a mean age of 36.6 years with slight female predominance. They noted syringomyelia in 23% of their patients which in our series was present in 47.6% patients.

PFD with and without duraplasty has also been compared in a series by Lee A et al[23] who has used the CCOS as their outcome assessment instrument. They have not found significance of difference for the two treatment groups with a mean CCOS of 14.6 ± 0.70 SD. Complications in our series occurred in 14% of patients while in Lee et al series it was noted in 19.4% patients while meningitis was found in 8% cases. It is notable that almost all CSF related complications in Lee et al series which is similar to our series. They have concluded that PFD without duraplasty is associated with significantly lower rates of postoperative complications ($p = 0.014$), although they reported a higher re-operation rate ($p = 0.58$).[23]

CONCLUSION

Posterior fossa decompression with or without dural opening are the most favoured procedures with equivalent results in attaining symptoms relief and functional improvement. The most important disadvantage reported for dural opening in posterior fossa decompression is the high incidence of CSF related complications such as CSF leak, pseudomeningocele and meningitis.

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