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Outcome of Posterior Fossa Decompression for Chiari Type I Malformation with Syringomyelia

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There are various treatment approaches for treating Chiari type I malformation with syringomyelia. Despite various choices, consensus for one particular approach is lacking. The objective of this study is to find out the clinical and radiological outcome of standard posterior fossa decompression incorporating removal of C1 arch with lax duroplasty in such cases.

A retrospective study based on data acquired from a single tertiary center were analyzed. All cases who underwent posterior fossa decompression incorporating removal of C1 arch with lax duroplasty over a period of five years were included and their clinical and radiological progress were recorded during OPD follow up at 6 months.

Out of 21 cases, occipital headache with nape of neck pain was the predominant complaint accounting to 71% followed by sensory symptoms and motor weakness, 61% and 33% respectively. Pain resolved in 93%, weakness in 71% and sensory symptoms in 69% of the cases. Only one patient developed hydrocephalus requiring shunting. Radiological improvement of syringomyelia were documented in 76.1% of the patients. There was no mortality.

Posterior fossa decompression incorporating removal of C1 arch and lax duroplasty is a safe approach with good outcome in patients with Chiari I malformation with syringomyelia.

Keywords: chiari malformation, syringomyelia, posterior fossa decompression, outcomes

Among various types of Chiari malformation, type I malformation (CM-1) is described as caudal displacement of cerebellar tonsils below foramen magnum.[2] Though no exact value is agreed upon as a cut off, more than 5mm is considered pathological.[1] However this diction is clinically irrelevant because it is the symptoms and associated syringomyelia that warrants treatment and the asymptomatic ones found incidentally are better left alone.[10] Various theories are postulated to be the root cause of the problems but common final step is the disruption of the normal CSF flow through foramen magnum.[6]

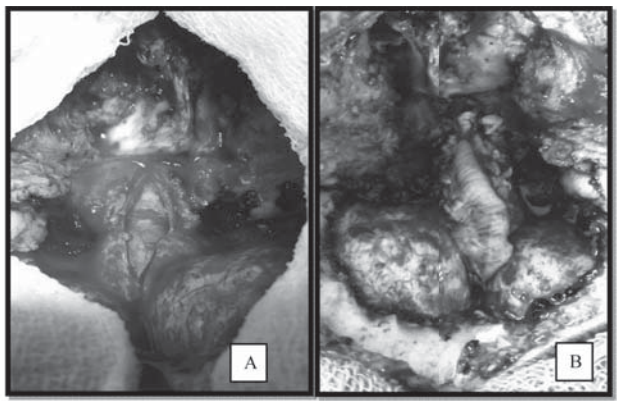

Figure 1. Intra operative view: A. Posterior fossa decompression with C1 arch removal after durotomy with preservation of arachnoid membrane B. Augmentation duroplasty with fascia lata graft.

Two main types of surgical modalities have been advocated for the treatment of CM-I. Posterior fossa decompression with or without duroplasty (PFDD or PFD, respectively) and the other is shunting of syrinx to subarachnoid space in the spine or coelomic cavity.[8] Posterior fossa decompression still remains the primary surgical technique for the treatment of CM-I because the shunt technique produces a risk of iatrogenic spinal cord injury. However, whether duroplasty alone without tonsillar ablation or adhesiolysis is sufficient during posterior fossa decompression remains controversial.[5,8] In this study, we look into the outcome of posterior fossa decompression with C1 arch removal with lax duroplasty for Chiari type I malformation with syringomyelia

Methods

A descriptive retrospective study was conducted after permission from institutional review board. Data were reviewed from the institute's archives. All patients who had undergone Posterior fossa decompression with C1 arch removal with lax duroplasty over the period of five years (January 2013 to December 2017) were included. The demographics of the patients were noted. Their presenting symptoms were noted from the patients file and the follow up of these symptoms at 6 months were done at OPD or over telephone. Reports from the radiologists of the institute were used to confirm the radiological resolution of syrinx.

Posterior fossa decompression of 3 by 3 cm squared with removal of C1 arch, opening the dura above and below the foramen magnum with lax duroplasty was considered standard approach (Figure 1). Operative notes were thoroughly checked to exclude any patient whose procedure were different from the standard.

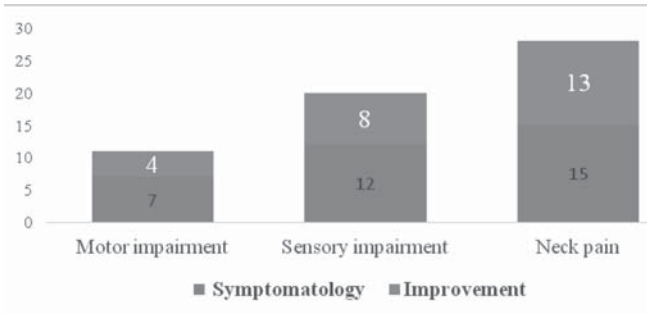

Figure 2. Predominant symptoms and subjective improvement of symptoms

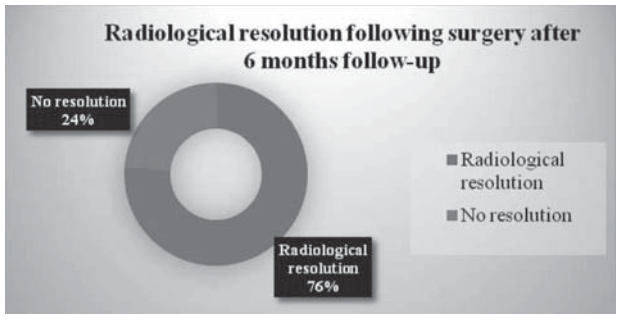

Figure 3. Radiological resolution following surgery after 6 months follow-up

Results

A total of 21 patient underwent the standard approach during study period. The age ranged from 13 to 52 years with median age of 31 (IQR 15). Out of them 11 (52%) were females and 10 (48%) were male.

Clinical presentation

Occipital headache with nape of the neck pain was the most common symptom seen in 15 patients (71%). This was aggravated while coughing or sneezing (Valsalva maneuver) in 66.66%. The second most frequent symptom was sensory disturbances seen among 12 patients (57%). Symptoms varied from tingling and burning sensation of bilateral upper limb to diminished sensation and numbness. Motor weakness was seen in 7 (33%) patients and was mostly distal parts of the upper limbs (Figure 2).

Resolution of symptoms at 6 months

Among the patients who had pain as the predominant symptom 86.7% had subjective resolution of pain. Improvement in the sensory symptoms were seen in 66.7%. The patients who had motor weakness had improved in 57.1%. Complete radiological resolution of syrinx was seen in 16 patients (76.1%) (Figure 3).

SN	Age	Sex	Pain	Motor impairment	Sensory impairment	Pain improvement	Motor improvement	Sensory improvement	Radiological resolution	Complication
1	35	Male	Yes	None	Yes	Yes	NA	Yes	1	None
2	28	Male	None	Yes	Yes	NA	NA	None	0	None
3	24	Female	Yes	None	Yes	Yes	NA	Yes	1	None
4	32	Female	Yes	Yes	None	Yes	None	NA	1	SSI
5	31	Female	Yes	None	Yes	Yes	NA	None	1	None
6	53	Female	Yes	Yes	Yes	Yes	Yes	Yes	1	None
7	13	Male	None	Yes	None	NA	Yes	NA	1	None
8	52	Male	Yes	None	None	Yes	NA	NA	0	HCP
9	27	Female	None	Yes	Yes	NA	Yes	Yes	1	None
10	18	Female	Yes	None	None	Yes	NA	NA	1	None
11	39	Male	Yes	None	Yes	Yes	NA	Yes	1	None
12	42	Female	Yes	None	Yes	Yes	NA	None	1	None
13	31	Male	Yes	None	None	Yes	NA	NA	1	None
14	25	Male	Yes	None	Yes	Yes	NA	Yes	1	None
15	24	Male	None	None	Yes	NA	NA	Yes	1	None
16	35	Male	None	None	Yes	NA	NA	None	1	None
17	39	Female	Yes	None	None	Yes	NA	NA	1	None
18	28	Female	Yes	Yes	None	Yes	Yes	NA	1	None
19	43	Female	None	Yes	Yes	NA	None	Yes	0	None
20	36	Male	Yes	None	None	None	NA	NA	0	None
21	21	Female	Yes	None	None	None	NA	NA	0	None

Note: SSI: Surgical site infection; NA: Not applicable; HCP: Hydrocephalus requiring shunt.

Table 1. Characteristic of patient: Clinical presentation and resolution of symptoms.

Complications

Surgical site infection requiring debridement and secondary suturing occurred in 1 patient. One patient developed hydrocephalus 2 weeks after the primary surgery requiring hydrocephalus (Table 1).

Discussion

Most patients in our study were young adults with almost equal male and female distribution. Most patients with Chiari Malformation presented with headache with nape of neck pain followed by sensory symptoms are similar to the published literature.[9]

The resolution of symptoms has been reported to be as high as 87% and 97% in independent studies by Munsi I et al and Chen JC et all respectively.[3,11] Our study shows resolution of pain, motor weakness and sensory disturbances were 93%, 71% and 69%. Lower rates were likely because of short follow up of 6 months.

A meta-analysis of 5 retrospective and 2 prospective study done by Durham et al concluded that posterior fossa decompression with duroplasty is associated with a lower risk of reoperation than posterior fossa decompression alone but has a greater risk for cerebrospinal fluid–related complications. There was no significant difference between the 2 operative techniques with respect to clinical improvement or decrease in syringomyelia.[4]

Another meta-analysis of 3481 patient done by Lin W et al recommended that posterior fossa decompression with duroplasty can be an optimal surgical strategy because of its higher clinical improvement and lower recurrence rate in the patients with syringomyelia however in patients without syringomyelia posterior fossa decompression alone can be a preferred choice because of its similar clinical improvement.[7]

Conclusions

Posterior fossa decompression incorporating removal of C1 arch and lax duroplasty is a safe approach with good outcome in patients with Chiari I malformation with syringomyelia. Comparative study with other surgical procedures for this condition is required to find out the best treatment option for these patients.

Ethical Clearance was taken from Institutional review committee (IRC) of Upendra Devkota Memorial National Institute of Neurological and Allied Sciences, Bansbari, Kathmandu, Nepal.

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