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Case Series

Observational outcome in surgery for Chiari malformation patients

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ABSTRACT

Chiari malformation is the commonest anomaly of the craniovertebral junction involving both the skeletal as well as the neural structures. It is congenital anomaly of the hindbrain characterised by downward elongation of the brain stem and cerebellum into the cervical portion of spinal cord. Most common presenting symptoms was pain in the nape of neck with sub-occipital headache and weakness. If not intervened early in these cases they may progress to quadriparesis and respiratory failure. This study includes authors experience of 30 surgical corrections of Chiari malformation performed at civil hospital Ahmedabad from 2017 to 2019. The age and sex of the patient, the presence of syrinx, the type of surgical procedure and the clinical outcome were determined post-operatively and on follow up. Cerebro spinal fluid leak and collection were observed in patient who undergone duroplasty only with no leakage in patient undergone syringo-subarachnoid shunt. Overall, tingling/numbness had best improvement showed improvement in 13 out of 16 patients. Power showed improvement in 20 out of 27 patients and pain showed improvement in 18 patients. Wasting, clawing and cerebellar signs and bony deformity showed no improvement in any of the above procedures. Authors can conclude for Chiari malformation decompression with or without duroplasty with additional procedure with post-operative physiotherapy and analgesia is the suitable treatment.

Keywords: Chiari malformation, Craniovertebral junction, Duroplasty

INTRODUCTION

Arnold-Chiari, or simply Chiari, malformation is the name given to a group of deformities which include characteristic downward herniation of posterior fossa and hindbrain into cervical portion of spinal cord. Chiari malformations include a large spectrum of anomalies of hindbrain formation which appear at different stages of development of the central nervous system. In 1883, John Cleland described a case of hindbrain malformation found during autopsy. Hans Chiari, an Austrian pathologist, described these malformations as congenital anomalies of the hindbrain.[1,2]

Subdivisions of Chiari malformations

- Chiari type 0 (Chiari malformation 0)
- Chiari type I (Chiari malformation I)
- Chiari type II (Chiari malformation II)
- Chiari type III (Chiari malformation III)
- Chiari type IV (Chiari malformation IV)

Chiari malformation type I is the most common cause of syringomyelia. It may not cause any symptoms and often goes unrecognized until adolescence or adulthood. Consequently, this form is sometimes referred to as adult Chiari malformation. Chiari malformation type II is usually more severe than type I and generally symptoms become apparent during childhood. The severity of Chiari malformation type II can vary greatly. The disorder can potentially cause severe, life-threatening complications during infancy or childhood.[3] Chiari malformation type III is associated with an encephalocele, unlike type's I-

III, Chiari malformation type IV is not associated with herniation of the brain but, the brain is underdeveloped. Chiari malformation type IV is the most severe form and is usually fatal during infancy Chiari malformation type 0 have syringomyelia despite the lack of cerebellar tonsil herniation. Chiari malformations appear to be due to a developmental failure associated with genetic factors. Acquired because includes tumours, an arachnoid cyst, prolonged use of a lumboperitoneal shunt, and hematomas. Hydrocephalus and intracranial hypertension have also been linked to Chiari malformations. Chiari malformations affect individuals of every race and ethnicity. Some studies suggest that females are affected more often than males.

Syringomyelia (asymptomatic) associated with a Chiari malformation usually does not require direct treatment. Generally, individuals with no symptoms are not treated, but are regularly monitored to see whether the disorder progresses. If mild symptoms are present such as neck pain or headaches, physicians may recommend conservative treatment such as pain medications and rest. Symptomatic Chiari malformations are most often treated by surgery.[4,5] The most common surgery is known as posterior fossa decompression. With this procedure, by removing small pieces of bone in the back of the skull, thereby enlarging the foramen magnum. This relieves pressure and reduces compression on the brainstem, and may allow the cerebellar tonsils to move back to a more normal position. Also include removal of part of the bony covering of the spinal canal (laminectomy) for cerebrospinal fluid circulation and to remove scar tissue. Individuals with hydrocephalus may require shunt. A myelomeningocele, which is usually associated with Chiari malformation type II, requires surgical repair. Surgery carries risks such as leakage of cerebrospinal fluid or infection. Symptoms related to a Chiari malformation may respond differently from symptoms related to an associated syringomyelia. Although some individuals experience significant improvement, others may have symptoms that persist including residual pain, muscle weakness, and loss of sensation.[6]

CASE SERIES

This study consists of 30 case of Arnold chiari malformation that has undergone surgical treatment in neurosurgery department civil hospital Ahmedabad from period September 2017 to august 2019. Minimum age was 6 years and maximum was 58 years. Commonest in 3rd-4th decade. Duration of symptoms was in range of 3 months to 6 years. Majority of patients in our series were female. Most common presenting symptoms was pain in the nape of neck in which sub-occipital headache constitute and limb pain. Weakness was commonly observed involving upper limb mainly. Second most common sign was altered sensation. Cerebellar signs positive was observed in few.

The study included all cases of Chiari malformation that had undergone surgical treatment. All patients with incidental findings and asymptomatic were excluded and associated anomalies as atlanto-axial dislocation and basilar invagination, also those treated conservatively.

Table 1: Comparison of improvement with different procedure.

Symptoms/ signs	Improved on discharge	Percentage	Total improved follow-up		Total	Percentage
			PFD+LAM+D	PFD+D+SS shunt		
Pain	20/25	80.00	0	0	20/25	80.00
Power	19/24	79.16	2	2	23/24	95.83
Sensation	04/20	20.00	5	9	18/20	90.00
Tingling/numb	10/14	71.42	1	0	11/14	78.57
Wasting	00/14	0.00	0	4	04/14	28.57
Clawing	00/13	0.00	0	8	08/13	61.53
JPS	03/08	37.5	1	1	05/08	62.50
CN	00/10	0.00	1	1	02/10	20.00
Cerebellar sign	00/08	0.00	2	2	04/08	50.00
Gait	03/08	37.5	2	2	07/08	87.50

PFD: Posterior fossa decompression, LAM: Laminectomy, D: Duraplasty, SS: Syringosubarachnoid.

After detailed neurological evaluation of all patient admitted from OPD, appropriate radiological investigation like haematological investigation X-ray, MRI and CT CV junction with screening of all spine and brain to rule out associated anomaly, cardiac work up as and when required for general anaesthesia during surgery. All patients were operated in prone position. Operative findings were noticed with immediate post-operative outcomes and complications assessed. Post-operatively antibiotics were given as per institution protocol and physiotherapy of chest and limb was given in all patients. Neurological evaluation was done on discharge of patients. Suture was removed on the 10th day post-operatively and advised to follow-up after three months of surgery with neurological examinations and MRI of CV junction. There is variety of conditions include multiple sclerosis, chronic fatigue syndrome, and fibromyalgia and spinal cord tumors that mimic Chiari malformation.

Surgery done in our institute was posterior fossa foramen magnum Decompression with posterior arch of C1 removal and fascia lata graft duroplasty (PFD+FMPR+ C1PA+FLGD). Second common surgery done was foramen magnum removal with posterior arch of C1 removal with syringo-subarchnoid shunt and fascia lata graph duroplasty (PFD+FMPR+C1PA+FLGD+SSS). Few patients were operated for posterior fossa foramen magnum decompression with posterior arch of C1 removal with G-patch duroplasty (PFD+FMPR+ C1PA+GPDL).

The overall improvement in symptoms/signs of the procedures then pain, power, tingling/numbness, gaits have best improvement. Sensation, clawing has intermediate improvement. While wasting, JPS, cerebellar signs, CN have the worst improvement or in other words those symptoms or signs have poor outcome and very late improvement in follow-up.[8-10]

Table 2: Complications of surgery.

Type of complication	PFD+FMPR+ C1PA+FLGD	PFD+FMPR+C1PA +FLGD+SS shunt	PFD+FMPR+ C1PA+GPDL	No. of patients	Percentage
CSF leak	01	00	01	02	6
CSF collection	01	00	01	02	6
Neurological detoriation	02	0	0	02	6
Wound infection	01	00	00	01	3
Meningitis	00	00	00	00	0

Table 3: Syrinx size versus type of surgery complications.

Syrinx size	PFD+LAM+D	PFD+D+SSS	Total
Improved	07	09	16
Same	03	04	07
Detoriated	0	0	0
Total	10/13	13/16	23/29
Percentage	76.90	81.25	79.31

PFD: Posterior fossa decompression, LAM: Laminectomy, D: Duraplasty, SS: Syringo subarachnoid.

Table 4: Extent of tonsillar herniation versus outcome.

Extent of tonsillar herniation (in mm)	No. of patients			Percentage
	Very good	Good	Same	
<5	1	2	1	75
6-10	8	15	1	95
11-15	2	4	0	100
16-20	1	1	0	100
21-25	0	1	0	100

CSF leak or collection was not seen in patients operated with syringosubarachnoid shunt group. All of them had improvement in compressive symptoms but persistent symptoms related to syringomyelia. Table 4 shows that extent of tonsillar herniation has no relation with outcome.

DISCUSSION

This study consists of 30 patients of Chiari malformations, all have been operated between the periods September 2017 to August 2019.

Minimum age was 6 years and maximum were 58 years occurs commonly in 3rd-4th decade. Duration of symptoms was in range of 3 months to 6 years.

Majority of patients in this series were female (60%). Most common presenting symptoms was pain in the nape of neck (60 %) in which sub-occipital headache constitute 40 % and limb pain (18%). weakness in limbs (45%) unilateral limb weakness was most common type constituting (34%) and bilateral (12%) of it. Third most common symptoms were tingling, numbness (30%).

Among signs weakness was commonly observed (40%) involving upper limb mainly. Second most common sign was altered sensation (22%). Third one was spinal deformity (18%) of which scoliosis constitute (14%) and kyphosis (4%), hyperreflexia (16%) mainly involved in lower limb. Other signs observed were wasting (15%), clawing of upper limb altered joint position sense and neuropathic joint. Ataxic gait was observed in 3 patients

with positive cerebella signs. Wasting of limb (16%) involving only upper limb with unilateral (10%) of it. Difficulty in walking with tendency to fall was complained by (10%) of patients. Limb deformity involving only upper limb was complained by (6%) of patient. One patient had neurological deterioration in right upper limb weakness and other had whole of left side weakness. Change in voice present in (6%) of patients with difficulty in swallowing in (3%) of patients.[11.12]

Cerebellar signs positive was observed in 4 patients. No patients had FNA/DDK positive. Ninth/tenth cranial nerve complex involved in 3 patients with isolated 11th nerve in one patient. Commonly observed radiological finding in MRI was scoliosis (30%). Others was occipitalised C1 (15%), hydrocephalus (3%). In this series extend of tonsillar herniation most commonly seen between 7 mm to 11 mm. This study shows almost similar results with other study in literature.[13,14]

Table 5: Currentstudy with different studies in terms of clinical parameters.

Symptoms or signs	This studies		Other studies			
			Alzate et al[15]		Hida et al[16]	
	Total	Improved	Total	Improved	Total	Improved
Pain (suboccipital/arm)	25	20	28	27	18	16
Power	24	23	8	7	37	32
Sensory deficit	20	18	20	19	51	46
Cranial nerve	10	02	-	-	8	2

CONCLUSION

Chiari malformations is a central nervous system anomaly which progress with age mostly shows symptoms after 1st decade of life.it presents with subtle neurological findings which when detected early can lead to arrest of the disease process at young age. Incidentally discovered and asymptomatic are managed on watchful observation. whereas, surgery is the definitive treatment for restoration of normal CSF pathway between cerebellum and spinal cord in symptomatic patients. surgery does prevent the subsequent development and/or progression of syringomyelia. Syringo-subarachanoid shunt should be done in patients with large syrinx and having central cord syndrome. Associated bony anomalies have poor prognosis. Compressive symptoms like pain, tingling/numbness have best prognosis after posterior fossa decompression. Weakness and sensation have good recovery after syringo-subarachnoid shunt . extent of tonsillar herniation has no relation with outcome. Authors can conclude from the study that at present, posterior fossa decompression with or without duroplasty with additional procedure (syringo-subarachanoid shunt) according to the individual case with post-operative physiotherapy and analgesia is the best treatment.

Long term follows-up and large meta-analysis of study is required in patients operated for PFD, to correctly evaluate the efficacy of different surgeries and to see for the neurological deteriorations.

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