Study on Syringomyelia

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ABSTRACT

Introduction: Chiari malformation encompasses a spectrum of congenital hindbrain herniation syndromes. In 1883 by Cleland initially described the general pathological changes of one type of malformations. So study aimed to see the clinical patterns of the disease, to compare the clinical data with other studies and to study the clinical improvement after surgical treatment.

Material and Methods: The study consisted of 36 patients who presented to this institute between October 2012 and January 2015 in whom a diagnosis of Chiari I malformation with syringomyelia was made and who underwent surgery. Clinical data such as age, sex, chief complaints, presenting complaints, symptoms, related to syringomeylia were recorded.

Results: Average duration of symptoms is 13.5 months of these commonest is Neck pain in 32 patients (89%). Plain X - Rays of the cervical spine were available in 36 patients, The findings were as follows Loss of tunneling / increased canal diameter -15, Magnetic resonance imaging studies were available in 36 patients. Chiari I malformation with syringomeylia was present in all patients, CT Brain - No hydrocephalous, The location of the syrinx was as follows Cervical -14 and Cervico - dorsal – 22. All the patients in this study population underwent small sub occipital craniectomy and foramen magnum decompression. Removal of Posterior arch of CI and Duraplasty with G patch graft. Complications were CSF leak was observed in 02 cases for those cases re-exploration done and duroplasty performed.

Conclusions: Foramen magnum decompression with small sub occipital craniectomy, removal of posterior arch of C1 and duroplasty using G patch graft, with water tight dural closure has given good results.

Keywords: Chiari malformation, Duroplasty, G Patch Graft

INTRODUCTION

First described in 1890 by the Austrian pathologist, Hans Chiari, a range of hindbrain abnormalities found on a study of autopsy material. He described Type I: Caudal herniation of the hindbrain through foramen magnum more than 5mm. Arnold described single patient with type 2 deformity[1,2] but did not focus his attention on the anomalies of the brain and spinal cord junction. No new information or insight was added to the problem to the Arnold description.[3] That's why now it is called as Chiari malformation. The prevalence of Chiari I malformation, defined as tonsillar herniations of 3 to 5 mm or greater, is estimated to be in the range of one per 1000 to one per 5000 individuals. The incidence of symptomatic Chiari is less but unknown worldwide. The prevalence of non post traumatic syringomyelia is 8.4 cases/1,00,000 population between 20-50 yrs. Syringomyelia is associated with chiari type-1 malformation. The word syrinx is derived from the Greek word for "reed or pipe," which in classical mythology is the form that the nymph Syrinx assumed to escape pursuit from the Greek god Pan[11] Caudal displacement of the hindbrain through the foramen magnum more than 5mm (cerebellum, 4th ventricle and brain stem). This malformation is now recognized by caudal displacement of the cerebellar tonsils through the foramen magnum, in to the upper cervical canal. The extent of tonsillar ectopia is variable ranging from few millimeters to a few centimeters. Frequently associated findings include a small posterior fossa, mild caudal displacement of the medulla and / or fourth ventricle, angulation of the cervicomedullary junction, hydrocephalus and various osseous anomalies as well as syringomyelia (dilatation of central canal cavity, not lined by ependymal layer) or hydromyelia (dilatation of central canal cavity lined by ependymal layer.[4] In the days before modern neuro imaging, diagnosis relied on clinical findings, pnuemoencephalography, myelography and intrathecal contrast CT Scanning. However MRI help in better understanding of the pathophysiology of this problem and also correlation between clinical signs and imaging data. Classification was Type 1: caudal displacement of the cerebellar tonsils below the plane of foramen magnum (>5mm).

Type 2: caudal displacement of the cerebellar vermis, 4th ventricle, lower brain stem below the plane of the foramen magnum commonly associated with myelodysplasia.

Type 3: caudal displacement of cerebellum and brain stem into a high cervical meningocele.

Type 4: cerebellar hypoplasia (not a type of cerebellar hernia)

The "Chiari 1.5 malformation" is a condition in which cerebellar ectopia is restricted to the cerebellar tonsils, as in Chiari I malformation, but contrary to Chiari's original description of the Chiari I malformation, the brain stem is also caudally displaced.[5]

MATERIAL AND METHODS

The study consisted of 36 patients who presented to this institute between October 2012 and January 2015 in whom a diagnosis of Chiari1 malformation with syringomyelia was

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made and who underwent surgery.

Patients with co existing other anomalies at the cranio-vertebral junction having these symptoms and signs were specifically excluded. Age, sex, chief complaints, presenting complaints, symptoms, related to syringomeylia were recorded. Imaging including X ~ rays were reviewed and the findings were recorded. MRI scans were reviewed with respect to the presence of Chiari malformation, presence of syrinx, extent of syrinx and presence of other anomalies.

STATISTICAL ANALYSIS

Descriptive statistcs like mean and percentages were used with the help of microsoft office 2007 to interpret the results.

RESULTS

Table 1 shows that the maximum number of patients were in the age range of 35-45 years who constituted to 16(44%). Females were more when compared to males.

Symptom duration ranged from 3 months to 2 years with an average duration of symptoms of 13.5 months. The commonest presenting symptoms were Neck pain in 32 patients (89%), Weakness in 31 patients (86%), Sensory symptoms in 26 patients (72%). Unusual presenting symptoms were Charcots joints in 2 patients.

As atypical presentation was deemed to be present when the overall presentation, of the patient did not fit into the classical pattern of presentation. There were 02 patients in all, who had some atypical feature in their presentation of Charcots joints with sensory loss.

Radiology

Plain X - Rays of the cervical spine were available in 36 patients, The findings were as follows Loss of tunneling / increased canal diameter -15, Magnetic resonance imaging studies were available in 36 patients. Chiari I malformation with syringomeylia was present in all patients, CT Brain - No hydrocephalous, The location of the syrinx was as follows Cervical -14 and Cervico - dorsal – 22. All the patients in this study population underwent small sub occipital craniectomy and foramen magnum decompression. Removal of Posterior arch of CI and Duraplasty with G patch graft.

Complications were CSF leak was observed in 02 cases for those cases re-exploration done and duroplasty performed.

DISCUSSION

The clinical presentation of patients with Chiari malformation with or without syringomyelia is complex and heterogenous. And the number of studies aimed at elucidating the presentation and attempts at classification bear testimony of this fact. Also the advantage of modern neuro imaging has increased not only the detection of the malformation but also our understanding of it. The aim of this study

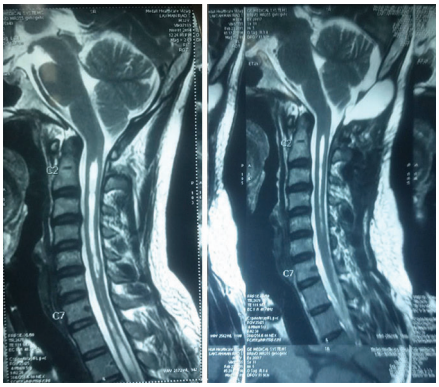

Figure-1: Before surgery; 4 Months after surgery

Age (years)	Number of patients	Percentage
15-25	8	22%
25-35	13	36%
35-45	15	42%
Sex		
Male	16	44%
Female	20	56%

Table-1: Demographic distribution in study.

Symptoms	Number of patients	Percentage
Sub-occipital and High cervical pain	32	89%
Upper limb weakness	31	86%
Lower limb weakness and spasticity	25	69%
Tingling and numbness	26	72%
Small muscle wasting	20	56%
Bladder involvement	11	30.5%
Swaying	15	41.6%
Charcots Joints	2	5.6%

Table-2: Symptoms complained by patients

Signs	Number of patients	Percentage
Short Neck	2	5.5%
Upper limb weakness with diminished reflexes	25	69.4%
Lower limb weakness with Babinski's positive	15	41.6%
Dissociated sensory loss	26	72.2%
Posterior column disturbances	15	41.6%
Incordination	18	50%
Wasting of small muscles hand	16	44.4%
Fasciculations	6	16.6%
Horner's syndrome	4	11.1%
XI nerve involvement	3	8.3%
Nystagmus	15	41.6%
Horizontal	12	33.3%
Down beat	2	5.6%
Charcoats joints	2	5.6%

Table-3: Signs elicited in patients

Symptom/sign	Pre-operative	Improvement
Suboccipital and High Cervical pain	33	29
Weakness	32	26
Sensory symptoms	25	17

Table-4: Clinical improvement after surgical procedure.

was to attempt to understand the clinical presentation of the disease with respect to morphology on MRI. Atypical presentations were also studied which were presented in the form of Charcots joints with sensory loss. Following are the comparisons between the present study and other studies found in literature: Levy et al[6] studied in 127 cases of adult Chairi malformation, of which there were 72 females and 55 males, yielding a sex ratio of 1.3:1, with an age range of 12 to 73 years and an average age of presentation of 41 years. The average duration of symptoms in this group was 3.1 years with a range of 1 month to 20 years. Dyste al (1989)[7] have reported on a group of 50 symptomatic patients comprising 16 patients below the age of 20 years, Bindal et al (1995)[8] have reported on a series of 29 patients with Chiari 1 malformation diagnosed by MRI of which 13 were males and 16 were females ranging in age from 18 to 66 years (mean 45 years). Heiss JD,[9] have reported on 252 patients with Chiari malformation with and without syringomyelia, of which there were 130 males and 122 females. Duration of symptoms in this study ranged from 1-5 years. The average age of presentation in this study was 29 years with a range of 1-57 years. Among the pediatric patients the average age was 11 years (range 1-19 years). The average age in the adult group was 34 years (range 20-57 years). The average duration of symptoms was 50 months with a range of 1 day to 30 years. The pediatric group had a shorter duration of symptoms averaging 20 months (range 1 day to 36 months) compared to 65 months in the adult group (range 1 day to 30 years).

Pillay et al (1991)[10] reported on 35 patients with Chiari malformation in adulthood, of which 19 were males and 16 were females, ranging in age from 18 to 57 years with an average age of 38.5 years. Mean duration of symptoms was 18 months. Versari et al (1993)[11] have analysed a group of patients with "foraminal syringomyelia" consisting of 40 patients including 22 females and 18 males In a retrospective study comprising 141 patients with Chiari I malformation, Stevens et al (1993)[12] analyzed the clinical presentations and related them to outcome. Saez et a[13] reported a retrospective study of 60 patients with syringomyelia. Of these, there were 22 males and 38 females, a sex ratio of 1.7:1 with an age range of 13-68 years and a mean age of 38 years, the mean duration of symptoms was 4-5 years with a range from 6 weeks to 30 years. In present study, the duration of symptoms was in the range of 3 months- 2 years with an average of 13.5 months, Pillay et al[10] the duration of symptoms with average of 18 months, Heiss JDet l[9] had a range of 1-5 years. Sex incidence in the present study i.e. males/females was 16/20 with incidecnce of 0.8:1, Pillay et al[10]; had male/females ratio of 19/6 with incidence of 1:1.8; Verasri et al[11] 18/22 with incidence of 1:1.8, Bindal et al[8] 13/16 with incidence of 1:1.23; Heiss JD et al[9] 130/122 with incidence of 1.06:1. Banerji and Millar[14] studied clinical presentations in a group of 20 patients consisting of 8 males and 12 females and analyzed the clinical presentations. Symptoms were pain of 73%, sensory complaints were 56% in pillay et al.[9] Verasriet et al[10], symptoms due to segmental and compression was 85%, cranial nerve impairment was 63%.

In most studies, the predominant complaints were related to foramen magnum compression syndrome, such as pain, which is probably reflective of the difference in the patient populations. Most studies have included patients with Chiari malformation without syringomyelia whereas in the present study the population is a fairly homogenous group of patients with Chiari malformation and syringomyelia.

CONCLUSION

Clinical presentations of patients with Chiari malformation with or without syringomyelia can be extremely diverse and heterogenous. The understanding of the disease and its pathogenesis is still evolving. Today, we understand the Chiari malformations to involve an abnormality at the craniocervical junction resulting in impaired neural function and CSF hydrodynamics. However, the pathophysiology of each malformation is likely very different, and the management is tailored to each individual. The Magnetic Resonance Imaging is mainly useful for the better understanding of disease pathogenesis and may help to elucidate the different presentations of the disease when compared to other investigations. This would help to elucidate the nature and pathogenesis of atypical presentations. There is no effective nonsurgical alternative to operative decompression for patients with symptomatic Chiari malformation I. All patients with a syrinx regardless of the size, location, or other associated symptoms are offered surgical intervention. Early diagnosis and surgical treatment of syringomyelia is essential to arrest progressive myelopathy and prevent further loss of neurologic function. Craniocervical decompression resolving appropriate symptoms is quite high with minimal operative risk

Restoration of normal CSF dynamic flow from the fourth ventricle to the subarachnoid space and relief of direct brainstem compression are the goals of surgery. Foramen magnum decompression with small sub occipital craniectomy, removal of posterior arch of C1 and duroplasty using G patch graft, with water tight dural closure has given good results in our study.

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