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Research Article

Resolution of Syrinx After Surgical Treatment of Chiari Malformation

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Abstract:

Arnold Chiari malformation involves the downward elongation of the cerebellum and brain stem into the cervical portion of the spinal canal, resulting in a variety of symptoms. In 1894, Arnold first described this condition in a newborn with a large myelomeningocele and lower limb deformity, observing herniation of the cerebellum into the spinal canal [1,2]. This syndrome arises from a developmental malformation of the occipital mesodermal somite, leading to the herniation of various structures of the posterior fossa through the foramen magnum. Subtypes of this syndrome are categorized based on the degree and structures of herniation.

Keywords: hydrocephalus; syringomyelia; pathology; surgical treatments

Introduction

Arnold Chiari malformation involves the downward elongation of the cerebellum and brain stem into the cervical portion of the spinal canal, resulting in a variety of symptoms. In 1894, Arnold first described this condition in a newborn with a large myelomeningocele and lower limb deformity, observing herniation of the cerebellum into the spinal canal [1,2]. This syndrome arises from a developmental malformation of the occipital mesodermal somite, leading to the herniation of various structures of the posterior fossa through the foramen magnum. Subtypes of this syndrome are categorized based on the degree and structures of herniation.

The symptoms stem from compression in the posterior fossa, hydrocephalus, and the formation of a syrinx [3]. Syringomyelia, characterized by cystic cavitation of the spinal cord containing fluid akin to cerebrospinal fluid, is predominantly responsible for upper limb symptoms and sensory deficits [4-8]. Approximately 50 to 70 percent of Chiari malformation cases are associated with syrinx formation [9]. Gardner proposed that syrinx formation results from disturbances in cerebrospinal fluid dynamics at the level of the foramen magnum, leading to muscle atrophy, motor weakness, dissociated sensory loss, and exaggerated reflexes. This excess fluid accumulation in the central canal of the spinal cord mimics the pathology seen in central cord syndrome [11]. Another theory, proposed by Williams, suggests that syrinx formation arises from pressure differences between cranial and spinal cavities, exacerbated by tonsillar herniation [12].

This study aims to assess the rate of syrinx resolution following various surgical treatments for Chiari malformation, as well as the persistence or resolution of symptoms post-treatment. This study represents the largest dataset on Chiari and syrinx from Pakistan To date.

Material and Methods:

This was a cross-sectional observational study with prospectively collected data from 46 patients with Chiari Malformation who were presented to the Department of Neurosurgery, Liaquat National Hospital Karachi Pakistan. Liaquat National Hospital is one of the few major hospitals in the city providing Neurosurgical expertise. It has a catchment area of 2 million people also people are been referred to this hospital for neurosurgical care from other hospitals and clinics. These patients were included in the study from the outpatient department of our hospital and were characterized accordingly. The demographic data, radiological findings, and data

on surgical procedures were retrieved from the data bank of the Neurosurgery Department and Health Information and Management Services Department by resident R5. The duration of the study is from 2017 to 2020. The institutional review board approved the research protocols and analyzed them by using SPSS 25 software.

Statistical Analysis:

Patient data was compiled and analyzed through the Statistical Package for Social Sciences (SPSS) Version 25. Qualitative variables are presented as frequencies and percentages. Quantitative variables are presented as mean ± SD. Effect modifiers are controlled through stratification. The chi-square test is used to find association between categorical variables. P ≤0.05 is considered as significant.

Results:

DEMOGRAPHY	No of patients
Male	31
Female	15
Up to 20 years	23
Tonsillar descent on MRI	46
Craniocervial decompression	32
Hydrocephalous	17
CSF diversion	14

In this study we had 46 patients, 31 were males and 15 were female. All of them had tonsillar descent on MRI studies. The majority of the patients were from the first and second decade of their life and the majority were males. Out of 46 patients, 28 patients had syrinx on MRI studies. 5 of them had cervical syrinx, 2 had dorsal and 21 patients had both cervical and dorsal syrinx. 17 out of 46 patients had hydrocephalus on MRI brain.

Out of 46 patients, 32 underwent craniocervical decompression and out of these 32 patients, 14 patients also had some CSF diversion procedure. 14 of our patients who underwent CSF diversion procedure with or without posterior fossa decompression, all of them showed marked resolution in syrinx size and symptoms. Out of these 14 patients, ventriculoperitoneal shunt was placed in 12 patients while in two patients ETV was done and both of these patients showed marked clinical and radiological improvement.

Presence of syrinx	Number of patients
Syringomyelia on MRI	28
Cervical	5
Dorsal	2
Cervico-dorsal	21
SURGICAL PROCEDURE RESULT	**Number of patients**
Resolution of syrinx	30
CSF diversion	14
V-P shunt	12
ETV	2
Unresolved syrinx	2

Of all the patients who underwent any surgical procedure, syrinx was resolved in 30 patients either by cranio-cervical decompression or by CSF diversion procedure. Only two patients did not showed any clinical and radiological improvement in syrinx.

Discussion:

Syringomyelia is a slit-like CSF-filled central canal of the spinal cord [13]. The incidence of syrinx in chiari patients is around 69% in adults and 40% in children [14]. Initially, it was thought that syrinx was in continuity of the fourth ventricle and it is formed by the pressure transmitting from fourth ventricle to the central canal of the spinal cord, this leads to the basic principle of Gardner's procedure that causes plugging of obex at fourth ventricle [15]. However some cases demonstrated absence of communication between fourth ventricle and syrinx that was demonstrated by Milhorat [16]. After this oilfield and colleagues demonstrated that the pathophysiology of syrinx formation is due to the pulsatile movement of CSF from fourth ventricle to central canal of spinal

cord . Some arterial pulsation in subarachnoid space also augments the formation of syrinx [2].

Symptoms form the syrinx depends upon the location of syrinx however it presents with variety of symptoms ranging from mild sensory deficit to motor weakness , spasticity to complete paralysis [6]. Neuropathic pain is also frequently associated with syringomyelia. In addition to addressing the pathology and using sub arachnoid shunts, spinal cord stimulation is also been used for treating syrinx related symptoms. Grower was the first one who described dissociated sensory loss and cape pattern of anesthesia in syringomyelia patient [17].

Regarding the surgical treatment of syrix, many authors have noted that pain related to syringomyelia may not respond to posterior fossa decompressive surgery, but in majority of the patients the size of the syrinx cavity is reduced [18-20]. Persistence of the syrinx cavity after posterior fossa decompression also occurs in a small percentage of patients and the reason could be the untreated arachnoid adhesion [21]. In our study, the data is the largest data of chiari malformation and syrinx formation so far. In our patients majority underwent a surgical procedure including cranio-cervical decompression, CSF diversion or both. Majority of the patients showed significant improvement. Note that the patients who underwent only CSF diversion procedure either ETV or Shunt also showed marked clinical improvement, indicating toward the correction of fluid dynamics by minimal procedure might be a good option in some of the patients.

Conclusion:

In conclusion, the majority of the patients with Chiari malformation have syringomyelia. And the majority of the symptoms are due to syrinx. Surgical treatment for chiari and hydrocephalous will lead to the resolution of symptoms and a decrease in the size of the syrinx in the majority of cases.

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