CASE REPORT

A Case Report of Non-Traumatic Spinal Cord Injury Following Large Cervicodorsal Syringohydromyelia

[1]Rajeev Kumar Singh
[2]Prince Rohilla
[*3]Zubina Khan
[4]Nitika Roy
[5]Shazia Mattu

ABSTRACT

Background: Patients having cervicodorsal syrinx are at an increased risk of sustaining a complete spinal cord injury resulting in poor prognosis. Current data on the efficacy of physical therapy for this condition is limited. In this study, rehabilitation aimed to maximize functional independence in ADLs. Subsequently, emphasis was placed on improving general fitness, coordination, and balance.

Case Summary: A female aged 38 years with a history of seizures and tubercular meningitis sustained a non-traumatic spinal cord injury as a result of large cervicodorsal syringohydromyelia. She underwent lumbo-peritoneal shunting surgery. Postoperatively, the patient experienced paralysis and loss of sensation in the lower limbs, along with urinary incontinence. The physiotherapy protocol was planned using a comprehensive neurological rehabilitation strategy so as to relieve her symptoms and improve her QoL. NMES was implemented after 4 months to enhance motor control and facilitate active assisted movement. Sessions were conducted 3-4 times per week.

Outcome measures: ASIA Scale, ASIA impairment Scale, SCIM, Borg rate of perceived exertion, QoL index SCI version-III, modified functional reach test, Spinal Cord Injury Functional Ambulation Index, and Karnofsky performance index were outcomes used to analyse the efficiency of numerous medications and to assess patient prognosis.

Conclusion: The patient's level of functional independence increased, i.e., she was able to get out of bed and move to a chair, stand and walk with a walker, and perform her personal hygiene tasks (eating, dressing, using the bathroom, bathing, etc.). Further research is needed to evaluate potential benefits and optimize treatment protocols.

Keywords: Neurological deficits, Non-traumatic Spinal cord injury, Physical Therapy, Rehabilitation, Syringohydromylia.

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[1]MPT (Neurology), Assistant Professor, School of Physiotherapy and Rehabilitation Science, K R Mangalam University, Gurugram, Haryana, India. E-mail: rajeev.rajeevsingh1085@gmail.com
[2]Physiotherapist, VMMC and Safdarjung Hospital, New Delhi, India. E-mail: princephysio98@outlook.com
[4]PhD Scholar, Delhi Pharmaceutical Science and Research University, Mehrauli-Badarpur Road Pushp Vihar, New Delhi, India. E-mail: nitikaroy95@gmail.com
[5]Associate Professor, School of Physiotherapy and Rehabilitation Sciences, K R Mangalam University, Sohna - Gurgaon Rd, Sohna, Sohna Rural, Gurugram, Haryana 122103, India.

CORRESPONDING AUTHOR

[*3]Zubina Khan

MPT (Neurology), Research Fellow, All India Institute of Medical Sciences, New Delhi, India.
E-mail: zubinak90@gmail.com

INTRODUCTION

Syringohydromyelia is a term that represents neurological disorder resulting from dilatation of the parenchyma or central canal of the spinal cord due to a cyst filled with cerebrospinal fluid (CSF) [1]. The presence of syrinx in cases of spinal trauma is reported to be up to 28% [2]. This medical condition may exhibit an unclear presentation, having close resemblance to other musculoskeletal conditions of the spine. It may be ignored until it has progressed to a more chronic stage due to its frequent attribution with recent trauma and can only be best validated by an MRI scan [3-5]. Syrinx management entails both surgical and non-surgical approaches. Surgical procedure tries to restore normal CSF flow and effectively treats syringomyelia [3]. However, evidence contradicts the requirement for surgical intervention in cases of severe and progressive neurological impairment [6,7]. Some studies also demonstrate insufficient medical and surgical outcomes, reflecting a poor prognosis [6,8]. The main aim of surgery is to relieve compression over the brainstem as well as correct cranial nerve distortion alongside decreasing any parallelly occurring syrinx cavities, and reestablish normal flow of CSF across the foramen magnum. Surgical approaches vary, but these objectives stay relatively similar for all surgical procedures. Chiari abnormalities are among the most common causes of syrinx cavities. However, any condition that results in a partial restriction of CSF flow in the spinal subarachnoid space, such as trauma, viruses, degeneration, and other conditions, can also result in spinal syringomyelia [9,10]. It can also be caused by the descending movement of the cerebellar tonsils, which partially occlude the subarachnoid space at the level of foramen magnum and act as a piston on the partially confined spinal subarachnoid space [11]. The advancement of the syrinx is caused by elevated cervical subarachnoid pressures, which compress the spinal cord from the outside rather than the inside. This causes the syrinx fluid to move caudally with each pulse. After standard extra-arachnoid decompression surgery, the abnormal shape and position of the tonsils is corrected, proving that Chiari malformation due to cerebellar tonsils' is acquired rather than congenital [12]. Cerebellar tonsils of the vast majority of syringomyelia patients have a Chiari I malformation [13]. Syringomyelia leads to advancing myelopathy and affects the musculature of the back, shoulders, arms, and legs, resulting in stiffness and pain in the muscles of the neck, back, shoulders, arms, and legs. Other symptoms may include migraines, lack of sensitivity to pain and warmth, and atrophy of the muscles. Scoliosis and other postural malformations are frequent and significant in people with syringomyelia [13,14]. This medical abnormality has a significant toll on the life of an individual resulting in many physical, emotional, and social vulnerabilities. All aspects of ill health in such a patient can be treated with the help of physical therapy [15]. Literature search of current data on the efficacy of physical therapy for this condition is limited. Due to a lack of information, experience, and resources, physical therapy is neglected as a form of intervention to restore function. Therefore, this article emphasizes the importance of physical therapy as a rehabilitative modality in improvement of patient symptoms along with enhancing quality of their life, ensuring a more dignified life for the patient.

Patient Information

A right-hand dominant, 38-year-old female who worked in a corporate office was doing well with overall good health and functional status till February 2016. She had an alleged history of severe headaches and fever. The patient consulted a physician regarding her condition and was provisionally diagnosed with a seizure disorder, for which appropriate medication was started. Surprisingly, even the medications did not alleviate the symptoms, with the condition of the patient deteriorating over time.

The patient decided to visit the neurology department of another hospital seeking a second consultation, where she was diagnosed with tubercular meningitis for which relevant medications were prescribed. In 2021, she tripped and fell while walking up the stairs. Her parents noticed that her right limb seemed weak, with inability to develop a good grip of her footwear. On the following day, she went for a neurological evaluation at a hospital in Delhi, where she was diagnosed with large Cervicodorsal Syrinx. She was operated for the same as the doctors advised for emergency surgery (lumbo-peritoneal shunting in July 2021). After the surgery she developed urinary incontinence and had loss of sensation in her lower limb with inability to move her lower extremity. She received physical therapy session after surgery at the hospital till February 2022. In spite of adhering to medication and physical therapy sessions, her symptoms continued to worsen over times with gradual increase of weakness in lower limbs and increased stiffness in right leg.

In June 2022, she visited yet another hospital in Delhi for a third consultation regarding her deteriorating condition. As per MRI findings, she was informed that the Cervicodorsal Syrinx had progressed to syringohydromyelia, extending down from cervicomedullary junction to the terminal end of the cord. Thereafter, she underwent the same surgical procedure of lumbo-peritoneal shunting for the second time. The incision was made at the L4 – L5 region, and lumbar puncture was done with a Tuohy needle. CSF was drained under pressure. The lumbar drain was passed through the Tuohy needle and connected to the abdominal end of the shunt tube.

The two surgeries left her weaker than before. Gradually, as the days passed, she could not sit or stand. Physical therapy sessions were started at home with a frequency of 6 days a week, after discharge from the hospital, and are still being continued.

Diagnostic Assessment

Initial assessment of the patient condition was done at home by a neuro-physiotherapist. The findings of the observation revealed that the patient was cooperative,

conscious, and aware of time, place, and people.

The therapist examined the patient in supine lying, which was the most independently attainable position for the patient.

The patient's build was mesomorphic, with hips externally rotated & adducted, knees slightly flexed, and ankles in plantarflexion.

During a neurological examination, superficial sensation was diminished down to L2 dermatomes on the right limb to pinprick and light touch. In both lower limbs, the deep tendon reflexes were hyperreflexia (Grade +++), tone was spastic grade 1+ and 2 in significant muscles according to the Modified Ashworth Scale (MAS).

Due to bladder involvement, patient underwent foley's catheterization. The neurological examination of the upper extremities was normal. The dynamic sitting balance was poor, while the static sitting balance was fair. It was impossible to stand without substantial support. MRI images of the spine revealed syringohydromyelia extending up to the cervicomedullary junction to the cord terminal, as shown in Figure 1.

Figure 1: The T2-weighted Magnetic Resonance Imaging (MRI) scan indicates syringohydromyelia extending up to the cervicomedullary junction to the cord termination.

INTERVENTION

The patient's treatment plan was designed to alleviate her symptoms and enhance her quality of life. Table 1 details the procedure that was followed.

Duration	Treatment Goals	Therapeutic Interventions	Dosage
1st- 4th month	Patient and family education To prevent secondary complications. To reduce spasticity. To improve bed mobility. To improve pulmonary hygiene and function. To maintain and improve the ROM, flexibility, and motor control of affected joints and muscles.	The patient and her parents were informed about the condition, its course, and the advantages of exercise at the start of the physical treatment program. Quick Icing and static stretching. Ankle-toe movement every two hours and regular positioning over bubble mattress were advised to prevent bedsores and DVTs. Rolling in bed as well as back extension exercises. Passive range of motion exercises for bilateral lower limbs. ACBT to prevent accumulation of chest secretions. Use of an incentive spirometer for facilitation of pulmonary capacity. Active exercises of Upper limbs and trunk. Active-assisted exercises for lower limb muscles. Stretching exercises to hip adductors, quadriceps, gastrocnemius, soleus, and hamstring muscles.	10 reps x 3 sets 10 reps x 1 set 5 reps x 2 sets 10 reps x 2 sets 3 reps x 2 sets
4th- 8th month	Maintain upper extremity strength. To improve trunk control and sitting balance. To strengthen the pelvic muscles. To maintain pulmonary hygiene and capacity. To improve strength of affected muscles. To release the tightness of affected muscles. To improve transfers. To alleviate paraesthesia.	Active Exercises for UL using 1 KG weight cuff and body weight exercises like chair push ups, cobra push ups etc. Seated perturbations in all directions and side touches with elbow on bed with elbow to train trunk side flexors. Scooting was added after the 5th month. Kegel's exercises, strengthening for transverse abdominis, hip abductor and adductor roll. Thoracic expansion exercises, Deep breathing exercises and Inspiratory Muscle Trainer (IMT) were used to train respiratory muscle function. Lower limb selective strengthening was done by giving varied progressive resistance manually and by weight cuffs ranging from 0.5- 1.5 kgs. Stretching exercises to gastrocnemius, soleus, quadriceps, hip flexors and hamstrings muscles. Neural mobilization for the femoral and sciatic nerve to release the compressed nerve and relieve paraesthesia. Sensory retraining activities were implemented cotton cloth, silk cloth, feathers, jelly balls, harsh Velcro, sponges, etc., from distal to proximal.	3 reps x 2 sets 10 reps x 2 sets 5 reps x 2 sets 10 reps x 2 sets 3 reps x 2 sets 5 reps x 2 sets 3 reps x 3 sets 10 reps each

8 th - 1 2 th month	To continue the strength protocol and maintain flexibility. To improve trunk control and balance. To increase standing time. To ambulate the patient. To alleviate paraesthesia. To correct postural abnormalities.	Strengthening was done by giving progressive resistance exercises via 1-1.5 kg weight cuff to bilateral muscles of lower extremities. Stretching and neural mobilisation continued. Trunk and Core training by crunches, scooting, quadruped weight shifting forward backwards and side. Supported standing and marching on spot. After the 9th month the patient was ambulating with the support of walker. as seen in picture 2. Posture correction exercises included shoulder retraction exercises to reduce increased kyphosis.	7-8 reps x 3 sets 5 reps x 2 sets 10 reps x 2 sets

Table 1: Detailed Protocol Followed for Rehabilitation.

Neuromuscular Electrical Stimulation (NMES) (Chattanooga Primera) was used (Picture 1) after the 4th month to improve motor control, and active assisted movement was done. The following parameters were used: frequency: 50-80 Hz, pulse width: 250- 300 µs, Intensity: Varied as per muscle response, Ratio: 8:8 secs. The sessions were done 3-4 times /week.

The patient's condition was progressively improving, and this was communicated to the patient's family. A home workout regimen was taught to the patient and her family/ caregivers. It involved strengthening activities at home under the supervision of a family member, stretching the gastrocnemius and soleus muscles, hamstrings, and dynamic quadriceps exercise, as well as posture correction and sensory retraining exercises.

Picture 1 and 2 (from left to right): NMSE-assisted dynamic quadriceps exercises and therapeutic walking of the patient with the help of walker and bilateral AFO

FOLLOW UP AND OUTCOME

The patient's condition significantly improved after a year of consecutively adhering to the treatment. The ASIA Scale, the ASIA Impairment Scale, the SCIM, the Borg rate of perceived exertion, the QoL index, SCI version-III, modified functional reach test, Spinal Cord Injury Functional Ambulation Index, and Karnofsky Performance Index are various tools used for assessment of functional impairment, evaluation of patient prognosis and comparing the effectiveness of different treatments. Pre-rehabilitation and post-rehabilitation comparisons are provided in Table 2.

Outcome measure	Pre- rehabilitation value	Post- rehabilitation value
ASIA Scale	UEMC- 50/50 LEMC- 1/50 LT Total- 98/112 PP Total- 98/112	UEMC- 50/50 LEMC- 14/50 LT Total- 112/112 PP Total- 112/112
ASIA Impairment scale	B	C
SCIM	25/100	80/100
Borg rate of perceived exertion	At rest: 10/20 At activity: 18/20	At rest: 8/20 At activity: 13/20
QoL index SCI Version -III	Part-A: 49 Part-B: 47	Part-A: 156 Part-B: 158
Modified functional reach test	0 inches	22 inches
Spinal Cord Injury Functional Ambulation Index	Gait parameter- 0 Assistive devices-0 Temporal measures-0	Gait parameter- 12/20 Assistive devices- 6/14 Temporal measures- 2/5
Karnofsky performance index	50/100	80/100

Table 2: Pre and Post Intervention Values of Outcome measures and Differences among them.

DISCUSSION

This report represented a severe case of syringohydromyelia, for which physical therapy management was given. Novelty of this case was in establishing the efficacy of multi-model physical therapy protocol as another way of restoring function after surgical intervention. Arnold Chiari Malformation- I are reported to be leading cause for this neurological disorder [16]. In addition to trauma and post-traumatic or viral adhesive arachnoiditis, spinal cord tumors can also induce syringomyelia. Syringomyelia is more frequently found by accident even when sensory signs like pain and temperature sensitivity are present [17]. Depending on ethnicity and geography, the incidence of syringomyelia ranges from 8.4 per 100,000 to 0.9 per 10,000. A large proportion of people are between the ages of 20 and 50 [18]. The syrinx's strain on the spinal cord frequently results in progressive weakening and stiffness in the arms, shoulders, and back. It can also impair pain perception, make walking difficult, cause problems controlling the bowels and bladder, cause facial pain and numbness, and induce scoliosis deformity [19]. An enlarging cavity in the central spinal canal is

a characteristic of syringomyelia. The neurons of the spinothalamic tract that decussate in the anterior white commissure are compressed by this CSF-filled "syrinx". Conversely, the posterior columns are protected by their distal placement. Segmental dissociated sensory loss occurs when touch and vibratory feelings remain while pain and temperature sensations vanish. The "cape-like" distribution involves a disproportionate amount of the upper limbs. Syringomyelia can be treated surgically using a variety of techniques, including syringe cisternal shunting, removal of extramedullary obstructions at the rostral end of non-communicating syrinxes, posterior fossa decompression, and shunting from the syrinx to the cerebellopontine angle cistern [20]. It has been demonstrated that physical therapy improves syringomyelia patients' overall quality of life and helps them manage their discomfort. Parents and patients are also educated about the ailment and the advantages of physical activity as part of the rehabilitation program. According to certain research, hydrotherapy and stretching have also been demonstrated to be effective in reducing stiffness and pain. Furthermore, it is claimed that the advantages of physical therapy are on par with those of surgery [21].

CONCLUSION

By reducing symptoms and improving the patient's quality of life, we conclude that a thorough neurological rehabilitation approach that comprised both traditional treatment and a well-planned physical therapy program and NMES application significantly improved the patient's condition in a patient with syringohydromyelia. This case study details how a meticulously watched neurorehabilitation program helped the patient become stronger and experience fewer post-operative problems.

This case report has been structured in accordance with the CARE (Case Report) guidelines to ensure clarity, transparency and completeness in reporting.

Abbreviations:

SCI – Spinal cord injury
NMSE - Neuromuscular electrical stimulation
ASIA – American spinal injury association
SCIM – Spinal cord independence measure
QoL – Quality of life
AFO – Ankle foot orthoses

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