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Inpatient Physiotherapy Management for Stiff-Person Syndrome: A Case Report

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Abstract

Introduction: Stiff-person syndrome (SPS) is a rare autoimmune neurological disorder presenting with inability to perform daily activities independently. Because SPS is a rare disorder, the efficacy of physiotherapy in the management of SPS is not yet known.

Case Presentation: The patient was a 65-year-old female with SPS diagnosed 1 year before. Assessments were performed, such as range of motion, muscle strength, pain, balance, and functional mobility. She had shoulder pain, with a score of 5/10 on the numerical pain rating scale for both shoulders. Moreover, her muscle strength was impaired. Although the patient could not maintain an upright position or walk, she had sitting balance. The physiotherapy functional mobility profile (PFMP) was scored as 35/63. She received 14 sessions of physiotherapy intervention, which included exercises to improve strength, flexibility, posture, balance, and functional mobility. Walking exercises were performed after the patient gained the ability to maintain an upright stance. Her muscle strength and posture improved, and her pain disappeared. The patient could maintain upright position and began to walk with a walker. At the end of the physiotherapy program, the PFMP was scored as 49/63, and the patient was able to walk 20 m with a walker without need to rest.

Conclusions: Inpatient physiotherapy management for SPS seems effective in improving balance, gait, and functional mobility.

Keywords: Stiff-Person Syndrome, Inpatients, Neurological Physiotherapy, Stiff-Person Syndrome

1. Introduction

Stiff-person syndrome (SPS) is a rare autoimmune neurological disorder and was first reported by Moersch and Woltman in 1956 (1). It is characterized by muscular rigidity and trigger-induced painful muscle spasms predominantly affecting the axial and proximal limb muscles, with emphasis on the paraspinal muscles, leading to gait difficulties and progressive disability (2). SPS affects both females and males equally, with an average age at diagnosis of 40 years (3).

Stiffness is often one of the first symptom of SPS, and this causes pain (4). SPS is typically characterized by an insidious onset, with a slow progression of symptoms over months or years, followed by a long-term stable period (5). Stiffness is also superimposed with forceful episodic spasms that typically last a few seconds as the involved muscles become hardened, resulting in abnormal joint positioning. Spasms typically occur in extension movements in the lower limbs, whereas flexion spasms are more common in the upper limbs (6). Several stimulants trigger the spasms, such as sudden noises and touch. Many patients report fear of walking in open spaces or crossing the street because of the spasms (5). Rigidity and spasms are most frequent in the trunk and proximal lower limbs. The severity of symptoms varies widely, and several factors, such as emotional upset and stress, often cause fluctuations in the symptoms. These symptoms are usually absent during sleep (5).

Postural abnormalities depend on the area of involvement. The affected abdominal muscles cause difficulties in movements, such as bending and twisting. Lumbar hyperlordosis commonly occurs because of the hypertrophy of the paraspinal muscles. Claw toes, plantar-flexion contractures, and excessive supination are also observed in the lower limb involvement (7).

Gait often exhibits a wide step width; it is deliberate and slow as the patient makes an effort to maintain balance. Therefore, falls are frequently reported by patients with SPS (7). Up to 65% of patients with SPS are unable to perform activities of daily living independently, and others need a walker or wheelchair for mobility and become bedridden in the future (8). Some patients deliberately walk slowly or choose not to walk because of fear of falling (9).

Because SPS is a rare disorder, the efficacy of physiotherapy in its management is not yet clearly known, and there are no published standardized physiotherapy pro-

grams. Here, we present a case report on a 65-year-old female with SPS to show the effects of physiotherapy management. This case report was prepared according to the CARE guidelines, which aim for consensus-based, clinical case reporting guideline development (10).

2. Case Presentation

The patient presented in this case report provided written permission for publication without any photographs. The patient was a 65-year-old female with SPS. She was 1.57 m tall and weighed 76.0 kg with a body mass index 30.89 kg/m^2. She was primarily hospitalized for plasmapheresis therapy and stayed in the hospital for 31 days. Plasmapheresis therapy was performed five times for cerebellar symptoms. In addition, the patient received 14 sessions of physiotherapy intervention.

She had received physiotherapy 1 year previously because of weakness and numbness of her right leg. She went to hospital 3 months after physiotherapy because of weakness of bilateral upper and lower extremities and was diagnosed with SPS in November 2012. Her anti-glutamic acid decarboxylase level was determined to be 120 nmol/L. The patient received intravenous immunoglobulin therapy for 5 days after the diagnosis. However, this treatment did not result in any benefits.

Blurred vision, lisping, and being startled with sudden noise or touching symptoms occurred. The patient started to be unable to ambulate independently. These symptoms worsened, and she received plasmapheresis therapy for eight sessions. She regained the ability to walk after the therapy.

The patient's symptoms recurred and she received plasmapheresis therapy again in July 2013. She had leg pain during the treatment and deep vein thrombosis occurred. Plasmapheresis therapy was completed after four sessions, and the patient was discharged. She began to suffer from nape pain, diplopia and blurred vision 2 months before presenting to our department. She was hospitalized in November 2013 at the department of neurology, Dokuz Eylul university hospital. The timeline of the history is presented in Table 1.

At the first physiotherapy session, initial assessments were performed. The patient was alert and had person, time, and place orientation. She was able to follow comments. There was no speech deficit. She was using medication for her depressive complaints.

Range of motion was assessed with observation while the patient was carrying out active movements in both the sitting and lying positions; it was found to be within normal limits. There was minimal hypertonia in the proximal muscles of the upper and lower extremities. The standard manual muscle test technique was used to assess the muscle strength and scored from 0 to 5 (11). A significant decrease in muscle strength, especially in both proximal upper and lower limbs, was observed.

The patient had shoulder pain during rest, and the pain was assessed using a numerical pain rating scale. This is a simple, frequently used method for the assessment of variations in intensity of pain. The shoulder pain was scored as 5/10 for both shoulders. Superficial and deep sensations were observed to be normal.

She had no balance problems in sitting. She could perform some basic activities of daily living in the sitting position. However, she could not maintain an upright position, and as a result, she could not ambulate. Although the patient could use her wheelchair, her husband drove it for her most of the time. Her sitting posture was characterized by more rounded shoulders and forward head. Cerebellar function was impaired, and there were significant cerebellar signs in both lower extremities; furthermore, trunk ataxia was observed.

The Physiotherapy Functional Mobility Profile (PFMP) was used to assess the functional level of the patient; the patient's score was calculated as 35/63 (12). The detailed information was presented in Table 2.

Plasmapheresis is an immunomodulatory treatment, and one cycle of 5 plasma exchange sessions is usually performed over 1 or 2 weeks (13). This treatment was performed five times to reduce cerebellar symptoms, such as trunk ataxia and the patient's lack of coordination. Her general medication did not change significantly during the physiotherapy program.

The physiotherapy program started 3 days after the onset of plasmapheresis. It was scheduled according to the standard physiotherapy protocol of our inpatient neurorehabilitation department. Daily physiotherapy sessions were performed 14 times except on weekends. A session took approximately 45 min, ending once the patient was perceived to be fatigued. All therapy sessions were performed in the mornings. The program was designed according to the assessment findings and included exercises to improve balance, coordination, posture, strength, flexibility, and functional mobility. The treatment type and progress of physiotherapy protocol are presented in Table 3.

Stretching exercises were mostly performed for the cervical and pectoral girdle muscles. Posture correction exercises were performed to correct the rounded shoulders and forward head posture. Posture correction and stretching exercises were performed as a warm-up and cool-down period in all sessions, and the patient was instructed to perform these exercises two more times throughout the day under the supervision of her husband. Repetitive task training mostly included practicing functional activities, such as reaching while sitting and sit-to-stand from the bed or chair. When she maintained an upright stance with a walker, she started to perform walking exercises. The patient performed hip-knee flexion and hip abduction movements while standing. The walking distance progressed day by day while reducing the manual assistance of the physiotherapist. Stair-climbing activities were practiced on a step board. Towards the end of the program, the patient started to practice climbing activities on stairs while holding the handrails.

The patient's functional mobility improved after the physiotherapy program, reaching a PFMP score of 49/63 (Table 2). This means that she experienced an improvement of about 22% in the functional mobility level, and she could walk 20 m with a walker without needing to rest. Her coordination ability and muscle strength exhibited improvement. Her shoulder pain disappeared, with a numerical pain rating scale score of 0/10. The observed static and dynamic balance improved dramatically while sitting. The patient could also independently maintain an upright position while holding a walker, and she started to walk with the walker in the ninth physiotherapy session. Her walking speed, distance, and independence improved at the end of the physiotherapy program.

Table 1. Timeline of Patient History

Date	History
Early 2012	Onset of the symptoms
August 2012	Physiotherapy intervention for the symptoms
November 2012	Diagnosis of SPS and intravenous immunoglobulin therapy
December 2012	New symptoms and plasmapheresis therapy
July 2013	Repeated symptoms and repeated plasmapheresis therapy
September 2013	Worsening symptoms
November 2013	Hospitalization for plasmapheresis and physiotherapy

Abbreviation: SPS, stiff-person syndrome.

Table 2. Physiotherapy Functional Mobility Profile Scores[a]

Item	Pre-Intervention	Post-Intervention
Bed mobility	7/7	7/7
Lie to sit	7/7	7/7
Sitting balance	7/7	7/7
Sit to stand	4/7	6/7
Standing balance	1/7	5/7
Transfers	3/7	4/7
Wheelchair	4/7	5/7
Ambulation	1/7	5/7
Stairs	1/7	3/7
Total	35/63	49/63

[a]Scores are ranged from 1 to 7. 1, total assistance; 7, independent.

Table 3. Treatment Type and Progress of the Physiotherapy Protocol

Session	Treatment Type
1	Initial assessments; posture correction and stretching exercises were taught
2	Repetitive task training (e.g., bed mobility, lie-to-sit activities)
3	Repetitive task training (e.g., sitting balance, functional reach during sitting)
5	Repetitive task training (e.g., sit-to-stand from bed or chair)
7	Repetitive task training (e.g., standing balance, functional reach during standing)
9	Ambulation with walker with one assistant
11	Repetitive task training (e.g., stair practice on a step board)
13	Repetitive task training (e.g., stairs practice)
14	Last assessments and discharge plan

3. Discussion

The aim of this case report was to show the effects of inpatient physiotherapy intervention for a patient with SPS. The patient showed significant improvements, especially in balance, gait, and functional mobility. Since SPS is a rare autoimmune neurological disorder, the efficacy of physiotherapy in the management is not yet known, and there is no described guideline. Conducting clinical trials in patients with SPS is difficult because of the rarity of the disease. Therefore, more case reports on this issue should be published to better understand the effects of physiotherapy interventions.

Two previous case reports were published on physiotherapy intervention for SPS (7, 14). The patients in these case reports showed improvements after physiotherapy intervention. However, the major problem for the patients was their limited range of motion. Our patient had a range of motion within normal limits; her major problem was her inability to maintain an upright stance or ambulate. In Potter's case report, medication changes occurred at the same time as the physiotherapy program (14); this makes it difficult to claim that the improvement was only caused by physiotherapy. Although in Hegyi's case, the medication did not change, the patient received physiotherapy in an outpatient setting for 15 weeks (7). Thus, the program setting and duration differed completely from those of our case, as our patient completed a 14-session physiotherapy program in an inpatient setting. Plasmapheresis therapy was performed five times for our patient, although her medication did not change significantly during the physiotherapy program. It is considered that the improvements were likely caused by this treatment. However, the patient had two additional plasmapheresis therapies without physiotherapy intervention before the treatment, with poor results, and the main reason for use plasmapheresis therapy was do address the cerebellar symptoms. In addition, plasmapheresis therapy was found to have conflicting results in patients with SPS (13). Therefore, it seems that the observed improvements in the balance, gait, and functional mobility occurred due to physiotherapy intervention. Our findings showed that short-term physiotherapy in an inpatient setting will be effective in SPS patients.

Spasms and stiffness are the characteristics of SPS, and our patient had minimal hypertonia with the use of a muscle relaxant medication. Although our patient had a normal range of motion, stretching exercises were performed to maintain this. The stretching program was focused on neck and pectoral girdle muscles due to her abnormal posture. The patient showed some improvements in her posture after combining the stretching and posture correction exercises. Strengthening exercises for both the upper and lower extremities were applied with manual resistance while performing range of motion activities.

The greatest improvements were observed in upright stance, gait, and functional mobility. The patient was not able to maintain an upright position, although she had sufficient muscle strength and range of motion. In our opinion, the major cause of this was the spasmodic episodes and fear of falling, as reported in the literature (9). The progress in the physiotherapy program resulted in increased self-confidence, and the patient was able to maintain her upright stance. Subsequent to gaining balance, she was able to walk with a walker in the ninth session. The walking speed and distance were progressively increased, and manual assistance was decreased in the other sessions. The patient could walk 20 m with a walker at discharge, without rest or manual assistance. She could also climb a few stairs.

Plasmapheresis was performed for the cerebellar symptoms, and the patient had two more plasmapheresis therapy session without physiotherapy intervention before this treatment, with poor results. Therefore, it seems that the observed improvements in the balance, gait, and functional mobility were caused by the physiotherapy intervention. Inpatient physiotherapy management for SPS seems effective in improving functional mobility and impairments associated with SPS.

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Footnote

Authors' Contribution: Study concept and design: Turhan Kahraman, Birgul Balci, and Ihsan Sukru Sengun; acquisition of data: Turhan Kahraman; analysis and interpretation of data: Turhan Kahraman and Birgul Balci; drafting of the manuscript: Turhan Kahraman; critical revision of the manuscript for important intellectual content: Turhan Kahraman, Birgul Balci, and Ihsan Sukru Sengun; administrative, technical, and material support: Ihsan Sukru Sengun.

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