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Case Report

# Rehabilitation in paraneoplastic stiff-person syndrome – Case Report

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**Abstract**

We aimed to share our rehabilitation experience in a patient diagnosed with paraneoplastic Stiff-person syndrome(SPS). A 45-year-old female patient was admitted to neurology with the complaint of widespread painful contractions. EMG was evaluated in favor of SPS. Amphiphysin-antibody was +++ in CSF. Patients' treatment was arranged and transferred to rehabilitation inpatient-clinic.The patient was included in the rehabilitation program of range of motion,stretching,strength ening, posture&walking exercises, balance&coordination exercises, 5 days/week for 3 months. The patient was screened for breast cancer, diagnosed with invasive breast carcinoma and underwent mastectomy. With the rehabilitation, the patient was mobilized first in the parallel-bar then with tripod-cane in the following months. Significant improvements were found in functional status and quality of life with control of spasticity and mobilization. Although the primary treatment of paraneoplastic SPS is cancer treatment, significant gains have been achieved with rehabilitation. It is necessary to raise awareness of the importance of rehabilitation to physicians who diagnose the disease.

**Keywords:** Case Report, Paraneoplastic, Rehabilitation, Spasticity, Stiff Person Syndrome

## Introduction

Stiff-person syndrome (SPS) is a rare neuroimmunological disease in the axial muscles causing progressive muscle stiffness, rigidity, and spasm, leading to a significant limitation in ambulation[1]. Nowadays the diagnosis of the disease can be made more easily by identifying the autoantibodies causing the disease. The disease is usually associated with higher rates of antibodies to Glutamic acid decarboxylase (Anti-GAD Ab)[2]. But there is also a variant with antibody against amphiphysin, which is mostly associated with paraneoplastic diseases, particularly breast carcinoma. Clinically, these two types have been shown to differ from each other. Amphiphysin-associated SPS is more common in women, more associated with breast ca, which rigidity and spasm are more common, neck or arm involvement is higher, and BDZ response is higher[3].

SPS patients have difficulties in their daily activities and their quality of life deteriorates due to axial muscle spasms that also spread to the appendicular areas in some cases, decreased range of motion and ambulation restrictions. There are many options in the medical treatment of the disease such as benzodiazepine, baclofen, steroid, IVIG, plasmapheresis, rituximab. However, the literature on the rehabilitation of the disease is quite limited. We aimed to share our rehabilitation experience in a patient diagnosed with paraneoplastic SPS.

## Case presentation

A 45-year-old female patient without a known history of chronic disease was admitted to our hospital's neurology clinic with the complaint of widespread painful muscle contractions. The contractions were triggered by emotional stress. Contractions were progressively increased and within 1 month the patient became bed-dependent due to the spasms in lower back and both ankles. In the neurology clinic, EMG performed for etiological research, the presence

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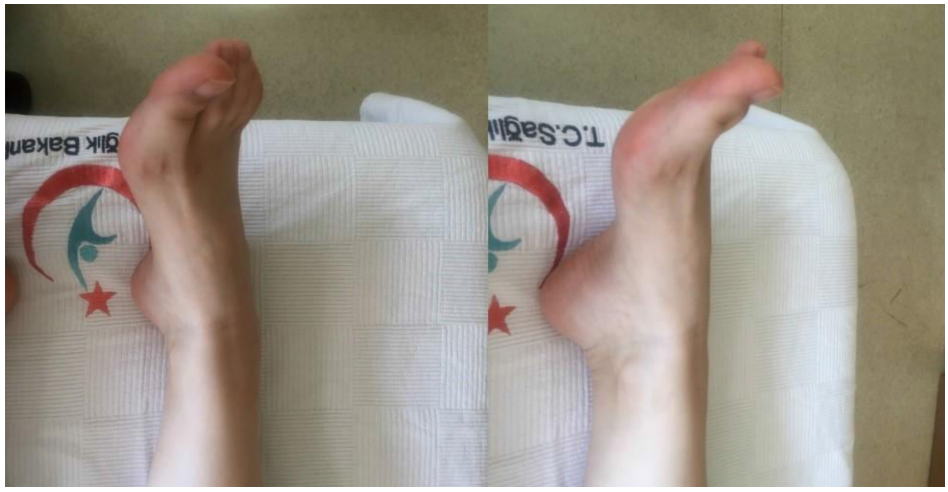

**Figure 1.** Patients ankle plantar flexed and inverted due to spasticity of the gastrocnemius and soleus muscles.

**Figure 2.** Balance and coordination exercises on parallel bar (weight bearing, weight shifting and stepping).

of continuous MUAP activity at rest accompanied by spasm in the right lower extremity was evaluated in favor of stiff-person syndrome. Amphiphysin antibody was detected as +++ in paraneoplastic panel search in CSF. Patients' treatment regime was arranged as 1000mg pulse methylprednisolone for 5 days, followed by methylprednisolone 64 mg/day, IVIG/5 days, diazepam 10 mg/day and baclofen 40 mg/day and patient was transferred to rehabilitation inpatient clinic.

The neurological examination of the patient on admission to rehabilitation inpatient clinic; Patient was conscious, oriented, and cooperative. Muscle strength evaluated via manual muscle test with standard technique (score from 0 to 5 /5)[4]: Left pectoralis major 4/5, biceps brachii 4/5, triceps 5/5, brachialis 5/5, wrist flexors and extensors 5/5, upper

**Figure 3.** Posture of the patient before discharge.

right 5/5 throughout, lower right 3/5, lower left 3/5. Lower extremity selective muscle strength examination could not be performed due to spasticity. Sensory examination: Superficial and deep sense intact. The patient's bilateral hip flexion (R: 100°, L: 95°), bilateral knee flexion (R: 135°, L: 95°), bilateral ankle dorsiflexion (R: 5°, L: 15°), right shoulder abduction (45°) and left shoulder flexion (90°), extension (40°) and abduction (90°) movements were limited. The patient had sitting balance partially and was immobile out of bed due to lumbar extensor spasticity and balance-coordination disorder. In the spasticity evaluation, in addition to the lumbar extensor muscles, there was MAS 2, Tardieu 3 spasticity in the bilateral gastrocnemius and soleus muscles, and MAS 1+ spasticitiy in the left hip flexor muscles (Figure 1).

Functional status of the patients was evaluated with Funtional Independance Measure (FIM). Total score of the patient was 66. Quality of life of the patient was evaluated with SF-36. SF-36 provides a self-reported health status profile consisting of eight dimensions. Results of the patient: Physical functioning was 15%, role limitations due to physical problems was 75%, bodily pain was 22.5%, general health perceptions was 40% , vitality was 5%, social functioning was 25%, role limitations due to emotional problems was 66.7%, and mental health was 28%.

Since the patient was using 64 mg/day methylprednisolone, the patient's bone mineral density measurement with Dual energy x-ray absorptiometry (DEXA) and laboratory tests were performed. Bone mineral density measurement was within normal range, Ca: 9,1 mg/dL, P:2,9 mg/dL, 25-OHVitD: 7,4 ng/mL, PTH: 150,9 pg/mL. 1000 mg/day calcium and 600 IU/day vitamin D supplement was given to the patient.

The patient was included in the rehabilitation program of active range of motion of shoulder, hip, knee and ankle, gastrosoleus and hip stretching (iliopsoas/hamstring), strengthening of abdominal muscles (knee to chest, isometric abdominal exercises, pelvic tilt), posture and gait retraining (symmetrical weight bearing, weight shifting, stepping, single leg standing, push off – calf rise) balance and coordination exercises (heel rise, side stepping, single leg standing, backwards walking) 5 days a week for 2 months (Figure 2).

Meanwhile, the patient was screened for breast cancer with a prediagnosis of paraneoplastic stiff person due to the antibody positivity to amphiphysin. The patient was diagnosed with invasive breast carcinoma by breast ultrasound and subsequent breast biopsy. The patient underwent simple mastectomy, in consultation with general surgery, and was transferred back to our inpatient clinic. The rehabilitation program continued for another month in the same order. With the rehabilitation program, the patient was mobilized in the parallel bar at the end of the first month, and subsequently with a walker and a tripod cane in the following 2 months.

The neurological examination of the patient on discharge from rehabilitation inpatient clinic; The patient was counscious, cooperative and oriented. Muscle strength: upper right 5/5, upper left 5/5, lower right 5/5, lower left 5/5. Sensory examination: Superficial and deep sense intact. The patient had no limitation in range of motion and no spasticity was detected (Figure 3). FIM total score of the patient was 88. SF-36 results of the patient: Physical functioning was 65%, role limitations due to physical problems were 0%, bodily pain was 22%, general health perceptions were 50% , vitality was 55%, social functioning was 37.5%, role limitations due to emotional problems were 0%, and mental health was 84%.

The patient's drug doses were reduced gradually, and she was discharged with baclofen 10 mg/day and methylprednisolone 48 mg/day. Control EMG performed after discharge has been interpreted as; Compared to baseline EMG, there was no pathological spontaneous MUAP activity, and this has been interpreted as noticeably improving condition.

## Discussion

Paraneoplastic SPS accounts for approximately 10% of all SPS patients and is highly associated with breast cancer, lung cancer and Hodgkin lymphoma[5].

The disease usually begins insidiously. As the disease progresses, spasticity develops in the lumbar spinal muscles characteristic of the disease and causes hyperlordosis[6]. Rigidity can spread to the hips and proximal extremities. However, in our case, as the hyperlordotic posture regressed early with oral treatments and rehabilitation, spasticity in the

gastrocnemius and soleus muscles was more resistant and took longer to decrease.

Since SPS is a rare disease, the literature is very limited. Case reports in the literature mostly focused on diagnosis and medical treatment of the disease. Due to rarity of the disease, there is no specific protocol for rehabilitation and physical therapy. In previous cases, massage, electrotherapy, hydrotherapy, relaxation and stretching techniques have been applied at different frequencies and times. In addition, balance and flexibility exercises were performed in some cases, and in some cases casting, walker and orthotic applications were performed[7]. Also in our case, range of motion, stretching, strengthening, posture and walking, balance and coordination exercises were included in the rehabilitation program of the patient, and the program was found to be extremely beneficial. Improvements in rigidity, pain and gait have been demonstrated with Botulinum toxin administration in patients who do not respond to medical treatment[8]. Botulinum toxin application was not deemed necessary as our patient benefited from cancer treatment, oral antispasticity treatment and rehabilitation program.

Spasticity and disturbance of posture and gait caused by SPS affects the functional status and quality of life of the patients. In the literature, the negative effects of spasticity on functional status and quality of life have been shown in studies conducted on many diseases characterized with spasticity[9-11]. Functional status and quality of life in our patient were evaluated with FIM and SF-36 scales respectively, and significant improvement was found in both scales by controlling spasticity and mobilization of the patient.

## Conclusion

Although the primary treatment of paraneoplastic SPS is cancer treatment, significant gains have been achieved with rehabilitation even before cancer treatment. For this reason, it is necessary to raise awareness of the importance of rehabilitation to physicians who diagnose the disease, especially who deals with spasticity patients in clinical practice. The best results will be obtained by conducting the treatment of these patients with a multidisciplinary approach from the moment of diagnosis.

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