

# Neuro-Rehabilitation for Peripheral Neuropathy in Primary Sjogren's syndrome: An OT Perspective

Prabhjot Kaur Gambhir

Max Super Specialty Hospital, Saket, New Delhi, India

---

**Abstract:** Among the constellation of symptomatology associated with Sjogren's syndrome, a patient may present with neurological involvement, which may be central or peripheral. These may present without the primary sicca symptoms. The medical treatment focuses on symptomatic treatment which may still leave the patient dependent for their daily activities along with persisting sensory symptoms making it difficult for the patient to even sustain mobility independently. Neuro rehabilitation for such patients' esp. occupational therapy is beneficial in terms of sensory training, training in daily activities, hand function training and mobility training.

**Keywords:** Primary Sjogren's syndrome, sensory ataxic neuropathy, Occupational Therapy, Sensory, ADL, mobility.

---

## 1. INTRODUCTION

Sjogren's syndrome (SS) is a chronic inflammatory autoimmune disease characterized by exocrinopathy involving mainly salivary and lacrimal glands. The histopathological hallmark is a periductal lymphocytic infiltration of the exocrinal glands, resulting in a loss of their secretory function (1). It could be primary or secondary, if it is associated with connective tissue diseases. It is a multifaceted condition with a broad variety of clinical manifestations. It is commonly seen in middle-aged women. Besides the involvement of exocrine glands entailing the classical sicca syndrome i.e. xerostomia and keratoconjunctivitis, systemic manifestations result from lymphocytic infiltration of organs (2).

Central to the pathophysiology of SS is chronic perpetual stimulation of the autoimmune system. Both B and T cells are implicated in the pathogenesis of SS (2). The most frequent symptoms of SS include the triad of fatigue, polyarthralgia, and sicca symptoms.

Neurological involvement in SS may be manifested in central and/or peripheral nervous system. Such complications occur in about 20% of patients (3, 4, 5, 6). The neurologic symptoms sometimes even precede the diagnosis. The possible pathogenic mechanisms behind it could be vascular, ischemic, or immunologic. The CNS involvement may present with focal disorders like seizures, cerebellar syndrome, optic neuropathies; multifocal disease with cognitive impairment, dementia; spinal cord dysfunction, progressive MS-like symptoms etc.

Peripheral neuropathy is a major neurologic manifestation of SS. It may include axonal polyneuropathies, sensory ganglioneuropathy, motor neuropathy, small-fibre neuropathy, multiple mononeuritis, cranial nerve neuropathy, autonomic neuropathy, or demyelinating polyradiculoneuropathy (7). Diagnosis of neurologic involvement can be confirmed with CSF examination, MRI for CNS involvement, nerve conduction studies, and EMG.

The sensory ataxic form is associated with dorsal root ganglion neurons or mononuclear cell infiltration without vasculitis. SAN is defined as the one with sensory neuropathy predominantly manifesting as impairment of joint position sense leading to sensory ataxia. The initial symptom is usually paresthesias in the digits of the foot or hand. There could be segmental involvement in the progression stage. Pain or painful dysaesthesias may be present (5).

There is no consensus about the specific treatment of neurologic involvement in pSS. Generally corticosteroid therapy is initiated. A higher dose is used with CNS involvement. For peripheral neuropathy, classically symptomatic treatment is recommended. In some patients, immunosuppressive therapy based on corticosteroids, cyclophosphamide, azathioprine, and even plasmaphoresis has shown only mild success (8, 9). IVIg has also been reported as a good therapeutic option in some painful neuropathy cases and in radiculoneuropathy (10).

Much knowledge has been gathered about the outcome of patients with SS. pSS has a slow insidious progression. A steady clinical course is usually observed in patients with peri- epithelial lesions whereas those presenting with extra-epithelial lesions such as polyneuropathy, have increased morbidity and mortality. They require tailoring treatments with higher doses of corticosteroids and immunosuppressive agents (2). It has been that pure sensory neuropathy is the most disabling in the long term.

In order to study the effectiveness of neuro- rehabilitation on neurologic symptoms as well as the impact on daily activities of an individual with pSS, the current case study was undertaken involving Occupational Therapy (OT) to observe the functional improvements in such individuals as a pilot program.

## 2. CASE STUDY

The patient was a 44 years old female with a history of pain in lower back and symptoms of numbness along medial aspect of left foot in Aug'12. Her MRI revealed L5- S1 prolapsed intervertebral disc (PIVD) with compression of left S1 nerve root. She underwent L5-S1 microdissectomy in Oct'12. Physiotherapy was taken post surgery and her functions returned to normal.

Later, she presented with gradually progressing tingling sensation or numbness in all 4 limbs along with gait ataxia since June 2014. She started to walk with support. The investigations were conducted in February 2015 which revealed:

**Table I : Investigations conducted to confirm diagnosis**

Nerve Conduction Study	Sensory motor predominantly demyelinating neuropathy of upper and lower limbs
CSF analysis	Albumin cytologic dissociation
Salivary gland biopsy	SS-A positive

She was diagnosed as a case of Sjogren's syndrome, type AMA (anti-mitochondrial antibodies) M2. As part of her medical treatment she underwent a 5-day course of intravenous immunoglobulin (IVIg) in February 2015. She was discharged on Feb'18 2015 and reported on an outpatient basis on Feb 19 2015.

Her chief complaints were numbness in arms and legs (left more than right), slipping of objects from hand when attempting to hold them, inability to balance self in standing and walking and thereby requiring external support assistance. These symptoms made it difficult for her to perform her daily activities which involved self care tasks as well as household work.

A baseline OT assessment was performed which involved:

- Sensory assessment with Semmes- Weinstein monofilaments
- Upper extremity assessment with Upper Extremity Functional Index (UEFI)
- Grip and pinch strength assessment with Jamar dynamometer and pinch gauge
- Balance assessment with Berg Balance Scale (BBS)
- Activities of daily living assessment with Functional Independence Measure (FIM)

### Course of treatment planning

The patient underwent Occupational Therapy treatment for a period of 6 months comprising of:

1. Sensory stimulation using textures, immersion and joint compression
2. Training in activities of daily training for eating, dressing, grooming, and after 3 months for household work
3. Grip and pinch strength training with coordination training.
4. Progression of balance ex from sit to stand, standing balance training, gait training progression from walker to stick to independent walking.

As the patient presented with sensory complaints, based on her initial level as assessed on Semmes- Weinstein Monofilaments, the stimulation was initiated with rough textures progressing towards more smooth textures like cotton in a distal to proximal pattern. Individual joint compressions were added for smaller joints of fingers and toes while proximal joints were approximated through weight bearing exercises. Particle immersion began with coarse items progressing to sand immersion later. This exercise improved weight bearing in the patient as she felt confident in activities of standing with eyes closed as a component of Berg Balance Scale. Also, dropping of objects from hand reduced with better use of upper extremities in her daily activities. The recovery was furthered by addition of grip and pinch strengthening exercises.

The presentation of mobility on the first day of rehabilitation involved standing with support. She was trained for sit to stand followed by walking with the aid of a walker, progressing to stick during which she was also trained for stair climbing and eventually independent walking. She presented with a waddling gait with direct foot flat pattern in the stance phase of gait cycle with each foot. By the end of 6 months, she could walk independently without support with near normal heel strike pattern.

Her training for daily activities included eating, dressing, grooming, and performance of household tasks while a home program was given for bathing and toileting toilet training. She initially had episodes of incontinence due to restriction in mobility. Also, she required assistance with cleaning self, bathing, and was unable to carry out any household tasks. Her therapy program commenced with visual compensation due to diminished sensation with automatic pattern of movement as her sensations recovered.

**Table II : Improvement in various functions as observed after 3 and 6 months**

Assessment	FEB	MAY	AUG
Sensory	4.56	4.31	2.83
Grip strength (lbs)	5	24	38
Pinch strength (lbs)	2	4	5
UEFI	35	54	64
BBG	5	25	38
FIM	64	83	120

### 3. DISCUSSION

The prevalence of Primary Sjogren's Syndrome (pSS) is low in Indian subcontinent which may be attributed to unreported or misdiagnosed cases esp. the ones referred directly to neurologists (11). Also, scant attention is given to problems of dry eyes and dry mouth by the patient- the classical history of sicca syndrome.

Rehabilitation focuses on those factors that may eventually lead to disability. Neuro-rehabilitation aims not only towards biomechanical factors but neurophysiologic domains as well in order to restore the patient to maximum level of independent functioning. The role of exercise in the rehabilitation of patients with pSS was studied by Strombeck et.al (2007). The review concluded that these patients may benefit in terms of aerobic capacity, fatigue, physical function, and depression (12).

The patient studied here presented with neurological symptoms compared to the classical sicca syndrome commonly observed in pSS. The NCV directed the diagnosis towards peripheral neuropathy of a demyelinating nature. She was referred for Occupational therapy as she reported with neurological symptoms of sensory involvement and gait ataxia along with difficulty in performing activities of daily living. The primary line of action including IVIg and corticosteroids added with OT treatment assisted the patient in enhancing her quality of life by improving her mobility as well as participation in daily activities.

Fatigue is prevalent and severe in patients with primary Sjögren's syndrome and physical activity is low compared with people from the general population (13). The effect of cognitive behavioural therapy group combined with exercise training on fatigue in pSS patients was studied by van Leeuwen N as part of a RCT.

In contrast to the body of evidence of impaired occupational performance in self-care, productivity and leisure, no papers relating to occupational therapy in pSS were found in the literature. Furthermore, as far as is known, occupational therapy

**International Journal of Novel Research in Healthcare and Nursing**

 Vol. 2, Issue 3, pp: (130-133), Month: September-December 2015, Available at: [www.noveltyjournals.com](http://www.noveltyjournals.com)

is not routinely offered to people with pSS. Impairments in physical functioning have been shown to affect well being and occupational participation in chronic fatigue syndrome (Taylor et al 2010), which has many symptoms in common with pSS including pain, fatigue and cognitive difficulties. Occupational therapy has been shown to be effective in improving functional ability and, for this reason, may have a unique role to play in the management of people with pSS (14).

People with pSS are considerably impaired in occupational performance. Occupational therapy may improve both occupational performance and wellbeing for people with this condition.

**REFERENCES**

- [1] Moutsopoulos H M., "Sjogren's syndrome: autoimmune epithelitis. Clinical immunology and immunopathology", Vol.72, issue 2, pp 162-165, 1994
- [2] Soyfoo M S, Cogan E, "Diagnostic and prognostic features of Sjogren's syndrome" Insights and Perspectives in Rheumatology, Intech Publications, 2012
- [3] Alexander E L, Provost T T, Stevens M B, Alexander G E, "Neurologic complications of primary Sjogren's syndrome", Medicine, Vol 61, Issue 4, pp 247-257,1992
- [4] Barendregt P J, Van der Bent M J, Van der Arssen V J, et.al, "Involvement of the peripheral nervous system in primary Sjogren's syndrome", Annals of the Rheumatic Diseases, Vol 60 Issue 9, pp 876-881, 2001
- [5] Mori K, Iijima M, Koike H, Hattori N, et.al, "The wide spectrum of clinical manifestations in Sjogren's syndrome associated neuropathy", Brain, Vol 128, Issue 11, pp 2518-2534, 2005
- [6] Goransson L G, Herigstad A, Tjensvoll A B, Harbas E, et.al, "Peripheral neuropathy in primary Sjogren's syndrome: a population-based study", Archives of Neurology, Vol 63, Issue 11, pp 1612-1615, 2006
- [7] Tobon G J, Pers J O, Devanchelle-Pensec V, Youinou P, "Neurological disorders in primary Sjogren's syndrome" Autoimmune Diseases, Hindawi Publishing Corp., Doi.10.1155/2012/645967, 2012
- [8] Kastrup O, Maschke M, Diener H C, "Pulse cyclophosphamide in the treatment of ataxic sensory and cranial nerve neuropathy associated with Sjogren's syndrome", Clinical Neurology and Neurosurgery, Vol 107, Issue 5, pp 440-441, 2005
- [9] Chen W H, Yeh J H, Chiu H C, "Plasmaphoresis in the treatment of ataxic sensory neuropathy associated with Sjogren's syndrome", European Neurology, Vol 45, Issue 4, pp 270-274, 2001
- [10] Kizaesa M, Mori K, Iijima M, Hattori N, et.al, "Intravenous immunoglobulin treatment in painful sensory neuropathy without sensory ataxia associated with Sjogren's syndrome", Journal of Neurology, Neurosurgery, and Psychiatry, Vol 77, Issue 8, pp 967-969, 2006
- [11] Mishra R, Hissaria P, Tandon V, Aggarwal A, et.al, "Primary Sjogren's syndrome: rarity in India", Journal Assoc Physicians India, Vol 51, pp 859-862, 2003.
- [12] Strombeck B, Jacobsson L T H, "The role of exercise in the rehabilitation of patients with systemic lupus erythematosus and patients with primary Sjogren's syndrome", Current Opinion in Rheumatology, Vol 19, Issue 2, pp 197-203, 2007.
- [13] Wouters E J M, van Leeuwen N, Bossema E R, Kruize AA, et.al, "Physical activity and physical activity cognitions are potential factors in maintaining fatigue in patients with primary Sjogren's syndrome", Annals of the Rheumatic diseases, Vol 71, pp 668-673, 2012.
- [14] Hackett K, Newton J, Ng W F, "Occupational therapy: a potentially valuable intervention for people with primary Sjogren's syndrome", British Journal of Occupational Therapy, Vol 75, Issue 5, pp 247-249, 2012.