Unusual First Presentation of Sjogren's Syndrome - Bilateral Foot Drop

Abstract: We report a case of Sjogren's syndrome in which the sole presenting feature was bilateral foot drop. It is a rare presentation of the disease. Extensive workup for alternate etiologies were negative, this case highlights the neurological feature of Sjogren's syndrome.

Key Words: Sjogren's syndrome, foot drop, neurological feature

Introduction

Sjogren's syndrome (SS) is a chronic inflammatory disorder that usually presents with dry mouth (xerostomia) and dry eyes (xerophtalmia or keratoconjunctivitis sicca) and is frequently associated with arthralgia or a connective tissue disorder. SS can be seen alone (primary) or in association with other autoimmune rheumatic diseases (secondary). Neurological involvement is not uncommon but frequently overlooked. Several forms of peripheral nerve dysfunctions occur, including trigeminal sensory neuropathy, mononeuropathy multiplex, distal sensorimotor polyneuropathy, pure sensory neuropathy and others.

Case Report

A 50-year-old female normotensive, normoglycemic house wife, mother of two children presented in capital hospital Islamabad with burning sensation in the feet, difficulty in walking, arthralgias and fatigue for the last six months. Difficulty in walking and progressive weakness was not associated with pain. Due to unsteady gait she fell frequently. On systemic inquiry she mentioned intermittent dryness of mouth and itching in the eyes. There was no history of muscle pain, difficulty in swallowing, raynaud’s phenomena, mouth ulcers or photosensitivity. Her past medical, surgical, socioeconomic and family history was unremarkable. On physical examination her weight was 65kg and height 138cm, there was no clubbing, lymphadenopathy or pallor. Her pulse rate was 80/min, blood pressure 120/80mmHg, Respiratory rate 14/min, temperature was normal. Peripheral pulses were intact. Neurological examination revealed a normal level of consciousness. Her cranial nerves appeared normal; she had a grade 4/5 muscular weakness in lower limbs. The ankle reflexes were bilaterally absent. Her sensation to pinpricks and temperature was decreased in feet with stocking like distribution. Vibratory and joint position sensations were decreased. There was bilateral foot drops. She stood with a wide stance and used stick while walking. There were no signs of cerebellar ataxia. Gastrointestinal, cardiovascular, respiratory system, eye, ear, throat, dental and fundus examination were normal.

Laboratory study revealed, positive SS-A and, positive SS-B. ESR 51, ALT 67 IU/L, CRP 11.7 (Raised) HEPATITIS B surface antigen Anti HCV was negative. Rheumatoid factors, antinuclear factor, antidouble stranded DNA were negative. The ophthalmologic examination showed a positive Schirmer’s test. The nerve conduction studies showed decreased amplitude of sensory action potentials with normal conduction velocities. The motor conduction velocities were decreased in peroneal nerve. We made a diagnosis of bilateral foot drop due to Sjogren's syndrome. She was given prednisolone 40 mg daily and advised physiotherapy. There was no definite improvement after treatment. The symptoms remained stationary under steroid treatment. She walked with support at the follow-up of 3 months later. She refused treatment of plasma pheresis.

Discussion

The differential diagnosis of a foot drop includes a wide variety of conditions. Sjogren’s syndrome can have multiple systemic manifestations. In our case main manifestation was foot drop due to neuropathy. As
Peripheral nerves control motor and sensory function in the limbs. Their disruption can lead to foot drop or numbness and tingling in an arm or leg. We excluded all other causes of foot drop associated with different diseases.

Our case fulfills the international criteria of diagnosis so we made diagnosis by using Revised Classification Criteria for Sjogren's Syndrome. The presence of anti-Ro/SS-A or anti-La/SS-B is more likely to suggest systemic disease. As our case is not associated with any other connective tissue disorder so we labeled it as primary Sjogren's syndrome. Sjogren's syndrome with gland inflammation is not associated with another connective tissue disease and is referred to as primary Sjogren's syndrome. When it is associated with a connective tissue disease, such as rheumatoid arthritis, systemic lupus erythematosus, or scleroderma, it is referred to as secondary Sjogren's syndrome.

We treated our patient with corticosteroids initially than started immunosuppressive drug but the results were unsuccessful. The recommended treatments includes corticosteroids, azathioprine, cyclophosphamide, cyclosporine, plasmapharesis and intravenous immunoglobulin. It is suggested that treatment in neuropathy associated with Sjogren's syndrome should be individualized and patients with progressive, disabling neuropathy be treated aggressively.

**Conclusion**

The reason to report this case is that neurological features of Sjogren's syndrome are usually missed in many cases and one should always consider this rare possibility in cases of foot drop.

**References**