

Primary Sjogren's Syndrome Presented with Sensory Ataxia Associated with Bilateral Hearing Loss and Dementia

Nastaran Madjdinasab* and Armaghan Moravej Aleali

Department of Neurology, Golestan Hospital, Jondi Shapour University of Medical Sciences, Ahwaz, Iran

Received: 21 Oct. 2007; Received in revised form: 4 Apr. 2008; Accepted: 14 May 2008

Abstract- Primary Sjogren syndrome is one of the commonest autoimmune diseases with characteristic of involvement of lachrymal and salivary glands, but other organ involvements as peripheral and central nervous system are also possible. The reported case is a 23 year old lady presented with progressive sensory ataxia and weakness of four limbs, bilateral sensory hearing loss and cognitive impairment with minimal score equal to 15/30 since one year prior to admission with associated bilateral central corneal opacity, dry mouth and dry eyes. Electro physiologic studies showed sensory motor axonal polyneuropathy. A biopsy of sural nerve and salivary glands of lower lip showed lymphocytic infiltration. Serologic evidence showed positive Anti Ro (SS-B), negative HCV and HIV antibody, thereafter the diagnosis was confirmed and according to this diagnosis she received high dose of intravenous methyl prednisolon then both hearing loss and cognitive impairment improved partially (minimal score 21/30). At last, she underwent plasmapheresis and her sensory ataxia improved greatly.

© 2009 Tehran University of Medical Sciences. All rights reserved.

Acta Medica Iranica 2009; 47(5): 423-426.

Key words: Sjogren, hearing loss, dementia, lip biopsy, poly neuropathy

Introduction

Sjogren syndrome is a slowly progressive inflammatory autoimmune disease, affecting primarily the exocrine glands. It affects mostly females with a female to male ratio of 9/1.

The peak incidence is in the fourth and fifth decades of life but Sjogren syndrome (SS) can occur in any age groups (1-3). Studies looking at the prevalence of SS estimate the rate at approximately 3% (1).

Case Report

The patient is a 23 year old right handed lady, graduated from elementary school, living around the Abadan city, came with chief complaint of inability to walk and weakness of both lower extremities and ataxia especially with closed eyes which has progressed insidiously since five months before admission and then she felt imbalance and was not able to walk alone and had to be helped by others while walking. She developed weakness of distal and proximal parts of both lower extremities. She got completely bedridden due to involvement of her upper extremities since around 4

months prior to her admission. The patient had also a complaint of foreign body sensation in both eyes since one year prior admission and according to this problem, she was referred to an ophthalmologist and received artificial tear drop and then improved partially but her visual acuity decreased due to central corneal scar, in the same time the patient suffered from some degrees of bilateral hearing loss and also feeling of dry mouth and excessive water demanding while speaking and fissures at the angles of her mouth. She also experienced constipation, frequency and dribbling since 3 months prior to her admission. There was no weight loss, fever, night sweating, diplopia, facial pain, numbness, artherlagia or articular swelling. Her drug history contained artificial tear drop, used 3 times a day since one year prior to this current evaluation. No one in her family has had any kinds of neurological problems.

In physical examination, the patient appeared well with stable vital signs and without any orthostatic change. She had bilateral corneal scar (Figure 1) without conjunctiva redness and congestion, also had some fissuring at the angles of her mouth and some Decayed teeth (Figure 2). No parotid enlargement was seen. The lungs, heart, abdomen and extremities were normal.

*Corresponding Author: Nastaran Madjdinasab

Department of Neurology, Golestan Hospital, School of Medicine, Jondi Shapour University of Medical Sciences, Ahwaz, Iran
Tel: +98 611 3743012, Fax: +98 611 3743012, E-mail: n.madjdinasab@gmail.com

Folstein's mini-mental state: for orientation to time (2/5) to place (2/5) registration (3/3) serial 7 (5/5) reading (0/1) recall (1/3) naming (2/2) repeating (1/1) for three stage command (1/3) writing (0/1) copying (0/1); estimated scores: 15/30.

The pupils were round, equal and reaction to light and accommodation and the retinas were normal but the right optic disk was pale and the left one was pink, visual acuity of right eye was equal to 3 meter finger count (FC) & the left eye was 4 meter FC. Extra ocular movements were full with normal saccades and no nystagmus was seen. Facial sensation and facial movements were normal. Finger rubbing test showed bilateral decreased hearing ability. The remaining cranial nerve functions were preserved. The strength of the limbs were as below: left arm: 4/5, right arm: 4/5⁺, left hand: 4/5⁻, right hand: 4/5⁺, proximal lower extremities: 4/5⁺ in both left and right sides, dorsiflexion of both sides: 3/5 and plantar flexion: 4/5.

All deep tendon reflexes were absent and plantar reflexes were downward.



Figure 1. Central corneal scar



Figure 2. Teeth decades

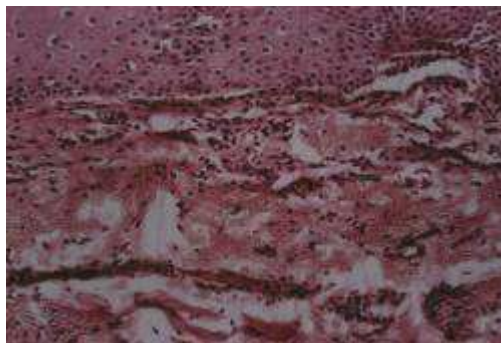


Figure 3. Histologically, the photomicrograph revealed lymphocytic infiltration of mucosal Lower lip biopsy

In sensory examination vibratory sensation was markedly reduced in toes and ankles, and also was reduced in both elbows. The sense of position was impaired in all extremities but pinprick and temperature sensations were normal. The patient was not able to stand or to walk.

The laboratory test consist of C.B.C, liver function test, thyroid function test, biochemistry and E.S.R was normal. Serum protein electrophoresis showed a normal pattern with increase in the gamaglobulin level. Tests of ANA and rheumatoid factor were negative. Pure tone audiometry showed mild to moderate bilateral sensory neural hearing loss.

An electro physiologic study was performed and revealed chronic axonal sensory motor polyneuropathy in lower extremities and sub-acute axonal sensory motor polyneuropathy in upper extremities with sensory predominance.

Sural nerve biopsy showed lymphocytic infiltration and results of serologic tests against hepatitis C and HIV were negative.

Biopsy of abdominal fat pad was negative for amyloidosis and mucosal biopsy of lower lip showed lymphocytic infiltration compatible with focus source equal one (Figure 3). The result of anti SS-A (Anti - Ro) was positive but Anti SS-B (Anti-la) was negative. According to this, the diagnosis of primary Sjorgen syndrome was confirmed for the patient.

At first, the patient was treated with intravenous methylprednisolone, 500mg/day for five days and then prednisolone tablet 60mg/day. One and half a month later her hearing and cognition were improved partially & MMSE showed the score 21/30. The patient was not able to walk but she could stand by aid and in spite of having good muscle force, she suffered from sever sensory ataxia so the patient underwent plasma

pheresis, 250 ml/kg but even after ending the course of plasmapheresis, she could only walk by help of a cane.

Discussion

The syndrome of keratoconjunctivitis sicca, xerophthalmia, xerostomia and chronic arthritis is called Gougerot-Sjogren's syndrome (3). The syndrome can be primary when occurred as an isolated entity or can be considered secondary when associated with any of a wide range of other autoimmune diseases, such as rheumatoid arthritis, systemic lupus erythematosus, chronic active hepatitis, Hashimoto thyroiditis and myasthenia gravis (2, 3).

Sjogren's patients frequently exhibit neurologic manifestations with involvement of the entire neural axis.

Identifying nervous system complications in SS¹ is of growing importance, because SS is more common than what many clinicians suspect and its neurological manifestations are often unrecognized or at least under appreciated. The need for early recognition of its neurologic complications becomes more evident. An earlier diagnosis is essential so that treatment can be initiated earlier in the course of disease (6).

Peripheral nerve involvement occurs in 10–32% of patients with primary SS (4) neuropathic symptoms often precede and overshadow the sicca symptoms and may be the major presenting complaint (4,5).

A distal symmetrical sensory neuropathy is the most common presentation (4,5). Less common patterns of nerve involvements include sensory motor neuropathy, polyradiculoneuropathy, painful dorsal root ganglionopathy and cranial neuropathy (trigeminal sensory neuropathy is the commonest) multiple mononeuropathy (4-6).

Sural nerve biopsy frequently shows nonspecific perivascular lymphocyte (T-cell) infiltration together with diffused decrease in myelinated fiber (4,5).

Laboratory abnormalities include elevated sedimentation rate, positive rheumatoid factor, and hypergammaglobulinemia (4,5).

SS-A antibody is considered to be the most specific serological test for SS but it is less common in patients with neuropathy (4).

The diagnosis depends on specific inquiry about sicca symptoms and ophthalmological tests for keratoconjunctivitis sicca (Rose Bengal staining of the cornea or Schirmer Test < 5mm wetting a paper strip at 5 minutes), a minor salivary gland biopsy of the lower lip showing chronic lymphocyte infiltration is also helpful for confirmation (4,5).

Treatment recommendations for polyneuropathy include corticosteroids either alone or in combination with immunosuppressant, neutralizing tumor necrosis factor- α antibodies (4) intravenous immunoglobulin (6,10) and plasma pheresis (3,9). Selecting a suitable treatment regime depends on type of neuropathy (8,10), for example intravenous or plasmapheresis for sensory ataxia (8,9) and methyl prednisolone pulse therapy for vasculitis neuropathy.

Involvement of brain can cause focal or diffused neurological abnormalities (4,7) Focal deficits include motor or sensory hemi paresis, aphasia, dysarthria, absence or complex partial seizure, extra pyramidal disorders or brain stem and cerebellar involvements (4,3).

Diffused manifestations include sub-acute or acute encephalopathy, recurrent aseptic meningitis, cognitive dysfunction and dementia (4,7).

Affective disturbance especially anxiety and panic attacks are the most common psychiatric abnormalities (11). Spinal cord involvement can be manifested as transverse myelitis, chronic progressive myelitis, Brown-squared syndrome, neurogenic bladder (7). In conclusion, in our patient, dry eyes and central corneal scar of both eyes with dry mouth were associated with dementia (MMSE³ equal to 15/30), bilateral sensory neural hearing loss, right optic disk pallor and sensory ataxia since one year prior to admission. Diagnosis of primary SS was confirmed by results of serological tests & lower lip biopsy; therefore accurate history taking about sicca complex at first & then serological interventions and even lip biopsy are very important in practice of any patient coming with multiple neurological complaints.

References

1. Koopman WJ, Boulware DW, Heudebert GR, eds. Clinical Primer of Rheumatology. Philadelphia: Lippincott Williams & Wilkins; 2003.
2. Klippel JH, Dieppe PA, editors. Rheumatology. 2nd ed Barcelona: Mosby; 1998.
3. Lafitte C. Neurologic manifestations of primary Gougerot-Sjögren syndrome. Rev Neurol (Paris) 1998;154(10):658-73.
4. Bosch PE, Smith EB. Chapter 82: Disorder of peripheral nerves. In: Bradley WG, Daroff RB, Fenichel GM, Jankovic J, editors. Neurology in Clinical Practice. 4th ed. Philadelphia: Elsevier; 2004. p. 2374.
5. Victor M, Roper AH. Adams and Victor's Principles of Neurology. 7th ed. New York: McGraw-Hill; 2001.

6. Yanagihara C, Nakaji K, Nishimura Y. A patient with primary Sjögren's syndrome featuring polyneuropathy and oculomotor paralysis and associated with asymptomatic left middle cerebral artery stenosis. *Rinsho Shinkeigaku* 2002;42(3):216-20.
7. Tumati B, Casoli P, Parmeggiani A. Hearing loss in the Sjögren syndrome. *Ann Intern Med* 1997;126(6):450-3.
8. Noguchi Y, Tsuchiyama T, Matsumoto T, Fujigasaki H, Inaba A, Yokota T, et al. Two distinct types of neuropathy associated with Sjögren's syndrome developed in one patient. The importance of the selection of an appropriate therapeutic regimen. *Rinsho Shinkeigaku* 2003;43(9):539-43.
9. Chen WH, Yeh JH, Chiu HC. Plasmapheresis in the treatment of ataxic sensory neuropathy associated with Sjögren's syndrome. *Eur Neurol* 2001;45(4):270-4.
10. Takahashi Y, Takata T, Hoshino M, Sakurai M, Kanazawa I. Benefit of IVIG for long-standing ataxic sensory neuropathy with Sjögren's syndrome. IV immunoglobulin. *Neurology* 2003;60(3):503-5.
11. Soliotis FC, Mavragani CP, Moutsopoulos HM. Central nervous system involvement in Sjogren's syndrome. *Ann Rheum Dis* 2004;63(6):616-20.
12. Aminoff MJ. Chapter 55: Neurologica complications of systemic disorder. In: Bradley WG, Daroff RB, Fenichel GM, Jankovic J, editors. *Neurology in Clinical Practice*. 4th ed. Philadelphia: Elsevier; 2004. p. 1083.

Archive of SID