



An Atypical Case of Primary Sjogren's Syndrome Presenting as Chronic Inflammatory Demyelinating Polyradiculoneuropathy

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Abstract

Sjogren's Syndrome is a chronic autoimmune disorder affecting exocrine glands. During the course of illness, there is multi system involvement. Neurological manifestations may occur in 8.5-70% of diagnosed cases. It is well established that primary Sjogren's Syndrome is associated with peripheral neuropathy to the tune of 10-20% although CNS involvement is much less common. These neuropathies are typically sensory or autonomic. The varieties can range from mononeuritis multiplex, distal sensory neuropathy, painful small fibre neuropathy to sensory neuronopathy. Herein, we describe an uncommon case of Sjogren's syndrome presenting with Chronic Inflammatory Demyelinating Polyradiculoneuropathy. The former denied the hallmark sicca symptoms and was discovered on further immunological studies.

Keywords: Autoimmune disorder, Peripheral neuropathy, Sicca symptoms.

Introduction

Primary Sjogren's Syndrome is a chronic autoimmune disorder with damage to the exocrine glands and multisystem involvement^[1]. The earliest reports of neurological involvement dates back to the 1980s with a prevalence estimate of 8.5-70%^[2,3]. Interestingly, neurological symptoms preceded the diagnosis of pSS by 2 years and in the remaining the symptoms appeared about 6-8 years of diagnosis. Mostly, the peripheral nervous system is involved and it is a negative prognostic factor as it increases the risk of Non-Hodgkin's Lymphoma^[4]. The gamut ranges from

mononeuritis multiplex, distal sensory neuropathy, painful small fibre neuropathy to sensory neuronopathy^[5,6]. Motor predominant neuropathies mimicking Guillain Barre Syndrome and Chronic Inflammatory Demyelinating Polyradiculoneuropathy are now being seen and could be the presenting symptom with improvement on Intravenous Immune Globulin therapy^[7,8,9]. The knowledge regarding CNS involvement is not well structured but the following can be seen: aseptic meningitis, optic neuritis, multiple -sclerosis like manifestations, acute transverse myelitis & cognitive disorders

^[10]. Reports of Amyotrophic Lateral Sclerosis associated with Sjogren's Syndrome have also surfaced.

Case Report

21-year-old post-partum female presented to the Out Patient Department with subacute onset progressive distal followed by proximal weakness of both lower limbs, right followed by left for 3 months. She also complained of difficulty in gripping objects for the past 15 days. Earlier, she was able to walk with support of two persons but after her delivery about 2 weeks back, her weakness deteriorated to the extent that she became bed bound & required help for the activities of daily living. There were also pins and needles sensation below the knees for the same duration. There were no sensory level, cranial nerve, bladder bowel involvement, no preceding history of fever, sore throat, loose stools or rashes and no similar events in the past.

On examination, she was conscious and her vitals were stable. Cranial nerves were intact. The tone was reduced in all four limbs. The power was MRC Grade 1/5 in all muscle groups in both lower limbs and MRC Grade 3/5 in both upper limbs. All the deep tendon reflexes were diminished and plantar was bilateral flexor. Joint position sense was impaired at the great toe & ankle. Vibration sense was impaired at the right up to the tibial shin and the anterior superior iliac spine in the left.

Her labs showed Hb-11.8 g/dl, MCV-94 fl, MCHC-33g/dl, Plt-1,58,000 / cu. mm, TLC-6,500 / cu. mm(N-68.2,L-28.8),Na/K- 136/4.2 meq/l , Urea/Creatinine-15/0.3mg/dl ,Total Bilirubin-0.8mg/dl,AST/ALT/ALP-21/13/75 IU, Total protein-7.1g/dl,Albumin-3.5 g/dl.

NCV was done on day 4 of admission. The motor nerve conduction findings revealed non recordable potentials in bilateral common peroneal and posterior tibial nerves, increased distal latencies and reduced CV in bilateral median and ulnar nerves, reduced CMAP in left ulnar nerve, conduction block in bilateral median

and left ulnar nerves. Sensory nerve conduction findings revealed reduced CV in bilateral median and ulnar nerves, non- recordable potentials in bilateral sural nerves. F waves were prolonged in bilateral median and ulnar nerves. Findings were suggestive of distal symmetrical generalised primary demyelinating with axonal polyneuropathy [Figure 1]. The electrodiagnostic findings fulfilled the criteria of Inflammatory Neuropathy Cause and Treatment (INCAT) for the diagnosis of CIDP.

Lumbar puncture was done and the CSF revealed 10 cells (L-52%, P-48%), protein-525.4mg/ dl, Sugar-76mg/dl, ADA-6 which was suggestive of an albumin-cytological dissociation. A diagnosis of Chronic Inflammatory Demyelinating Polyradiculoneuropathy was made based on the European Federation of Neurological Society Criteria 2010^[11].

On day 6, she was started on IVIg at 2g/kg body weight given over 5 days followed by oral Prednisolone 50 mg at 1mg/kg body weight daily. Patient responded to treatment with gradual improvement in power of the limbs (MRC grade 4-/5) with appearance of the vibration sense over the lower limbs, deep tendon reflexes in the upper limbs after two weeks. The paraesthesia of a lower intensity persisted.

She was investigated further to rule out any primary cause of CIDP and her serology was non-reactive, ANA was positive (3+ on IF) with a homogenous pattern, ENA profile revealed anti SS-A antibodies being strongly positive, positive RO-52 antibodies and a borderline positive U1 SM/RNP antibodies. Schirmer's test revealed >5mm wetting in 5 minutes. Lip biopsy was performed and showed irregular mucosal & mild peri appendageal chronic inflammation & unremarkable minor salivary glands. The finding though not pathognomonic of Sjogrens but the subdued inflammatory finding was probably due to the effect of steroids as consulted with the pathologist.[Figure 2]. Other laboratory data including hepatitis B surface antigen, anti-hepatitis C virus, HIV I & II,

Immunoelectrophoresis and Rheumatoid factor were all negative. This pointed to a diagnosis of Primary Sjogren’s and the CIDP was a presenting

feature of the former. Patient was discharged on maintenance dose oral steroids and kept in regular follow up.

Figure 1: Motor nerve conduction study

NERVE	LATENCY(ms) D	LATENCY (ms) P	AMPLITUDE (Mv) D	AMPLITUDE (Mv) P	%dec	DURATI ON D	p	% incr.	CV (m/s)	F-Min(m/s)
RT.MEDIAN	13	25.75	9.53	3.33	65.06	25.50	24.50	210	16.47	49.75
RT.ULNAR	7.37	16.12	5.77	3.31	42.63	31	31.12	210	26.29	46
LT.MEDIAN	9.62	15.75	10.52	4.34	58.75	26.67	29.50	210	34.26	52.87
LT. ULNAR	7.25	15.75	4.19	1.84	56.09	25.62	33.0	210	27.06	50

Sensory nerve conduction study

NERVE	LATENCY(ms)	AMPLITUDE (Uv)	DISTANCE (mm)	CV(m/s)
RT.SURAL				
LT.SURAL				
RT.MEDIAN	3.45	15.14	120	34.78
LT.MEDIAN	4.16	10.29	120	28.85
RT.ULNAR	3.11	11.01	90	28.94
LT.ULNAR	3.23	14.10	90	27.86

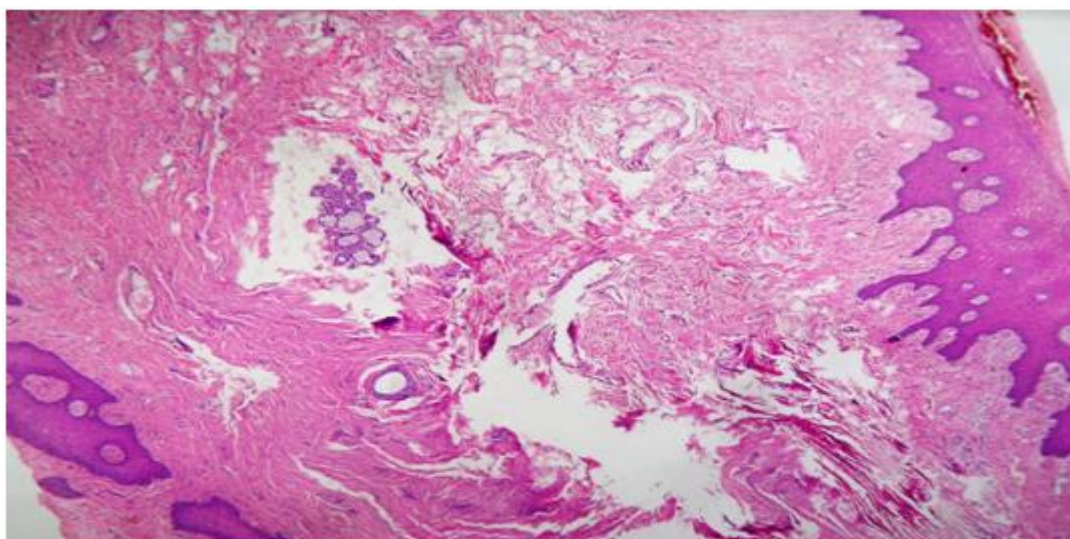


Figure 2: biopsy from lower lip showing irregular mucosal & mild peri appendageal chronic inflammation & unremarkable minor salivary glands

Discussion

To our knowledge, this is the first case of Primary Sjogren's syndrome presenting with CIDP without any sicca symptoms clinically. What is remarkable is the course of evolution of symptoms in the disease process of Sjogren's Syndrome with extra glandular involvement antedating glandular involvement. Furthermore, there was worsening of symptoms in the postpartum period. The patient improved with IVIG and showed signs of motor recovery within two weeks. Hence, the patient could be educated about the symptoms which could ensue and about early reporting of the same.

CIDP can be thought of as window to an underlying disorder. There is limited data on CIDP in Sjogren's Syndrome^[12,13]. Both are thought as separate processes and not one as a manifestation of the other. CIDP & Sjogren's Syndrome may share the same underlying pathogenesis including vasculitis and humoral-and/or cellular-mediated immune responses. The important take home message is that any CIDP should be investigated thoroughly for underlying autoimmune disorders & early treatment can cause marked improvement of symptoms as in our case^[14,15]. Neurological manifestations in absence of sicca symptoms often result in delayed diagnosis of Primary Sjogren's. A positive immunological test leads to an early diagnosis of Primary Sjogren's syndrome several years before the onset of typical sicca symptoms and timely institution of treatment would help prevent the systemic complications like chronic organ damage, lymphoma^[16].

Conflict of Interests

The authors declare no conflict of interests.

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