

A Rare Presentation of Primary Sjogrens Syndrome

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Rec date: Dec 09, 2015, Acc date: Dec 16, 2015, Pub date: Dec 24, 2015

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Abstract

Neurological manifestations are common in primary Sjogrens syndrome (SS). Neurological involvement in Sjogrens Syndrome may be manifested in the central nervous system (CNS) and/or peripheral nervous system (PNS). Motor Neuron disease (MND) is very rare in Sjogrens syndrome. Treatment is steroids.

Introduction

Motor Neuron Disease is a disabling neurological disorder characterized by death of upper and/or lower motor neurons. Although neurological manifestations occur commonly in Sjogrens syndrome and may precede the syndrome, MND is rare. We here present a lady who presented to us with clinical features suggestive of Motor Neuron Disease that ultimately proved out to be secondary to Sjogrens Syndrome.

Case Report

A 47 year old female was admitted to Neurology Department of our hospital with complaints of weakness of all four limbs and progressive dysphagia of three months duration. Three months earlier patient started with difficulty in walking that was gradual in onset and progressive and associated with feeling of stiffness of muscles. Patient noted difficulty buttoning/unbuttoning her shirt and climbing stairs and progressively ambulation became difficult. Patient also developed progressive dysphagia to solids as well as liquids associated with nasal regurgitation. There were no sensory disturbances or bowel and bladder symptoms associated with this weakness. On examination patient was conscious and oriented with normal higher motor functions with no signs suggestive of cerebellar disease or meningitis. Motor examination revealed fasciculations, atrophy of tongue and distal musculature of all four limbs associated with hyperreflexia, hypertonia and Grade III (MRCS Scale) power of all four limbs. Bilateral plantar responses were flexor. There was no sensory involvement. Baseline investigations and CSF were normal. NCV was normal while as EMG showed fasciculation potentials in right Flexor Digitorum Longus and long duration of Motor Unit action potential. MRI cervical spine was normal. A diagnosis of Motor Neuron Disease was made and patient was put on riluzole 50 mg bid and discharged home. Four months later patient presented with progressive bluish discoloration of digits of both hands and feet. Peripheral pulses were felt normally and there were gangrenous changes in digits of hand and feet (Figure 1).



Figure 1: Peripheral pulses.

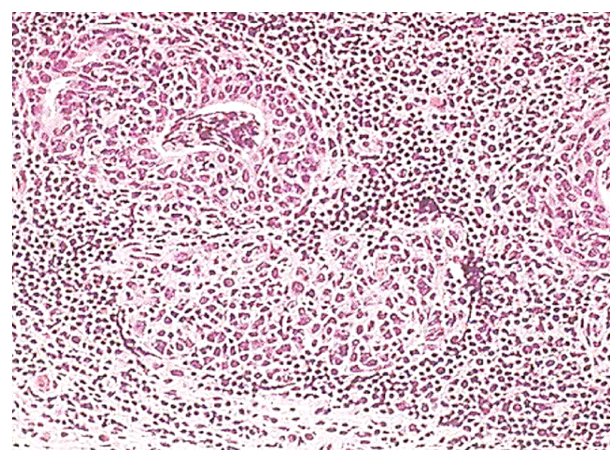


Figure 2: Lip biopsy revealed moderate lymphocytic infiltration in the intervening stroma favouring nodular aggregates.

Raynaud phenomenon was present. The previous weakness was still present, although dysphagia had slightly improved. Patient also gave history of dry mouth and foreign body sensation in her eyes. On evaluation patient had positive Schirmers test. Unstimulated Salivary Secretion was <1.5 ml/15 min. Lip biopsy revealed moderate lymphocytic infiltration in the intervening stroma favouring nodular aggregates at some places (Figure 2).

Features were consistent with Sjogrens Syndrome. ANA, anti dsDNA, pANCA/ cANCA, SCL-70/ACA, anti U1RNP antibody, anti Ro/La antibody, Serum electrophoresis was negative. Sural nerve biopsy revealed thickened nerve with mild to moderate inflammation in perineurium and epineurium and vessels showing severe chronic inflammatory infiltrate, features suggestive of vasculitis. Muscle biopsy revealed foci of degeneration with areas showing vessels with perivascular inflammation and neovascularisation, features suggestive of vasculitis. Our complete diagnosis was primary Sjogrens Syndrome with Motor Neuron Disease and Vasculitis. Patient was managed with corticosteroids (prednisolone 1 mg/ kg) and cyclophosphamide pulse therapy. Patient has received two pulses of cyclophosphamide and has shown marked improvement in her symptoms (Figure 3).



Figure 3: Patient has received two pulses of cyclophosphamide and has shown marked improvement in her symptoms.

Discussion

Sjogrens syndrome is a multisystem disorder that predominantly presents with dry eyes and dry mouth. Although exact etiology of primary Sjogrens syndrome is unknown, role of various viruses have been proposed as causative agents. Epstein-Barr virus (EBV), HTLV-1, human herpes virus 6 (HHV-6), HIV, hepatitis C virus (HCV), and cytomegalovirus (CMV) may have a role. Sjogrens-like syndromes are seen in patients infected with HIV, HTLV-1, and hepatitis C [1]. Neurological complications occur in as many as 20% patients of Sjogrens syndrome [2,3]. In as many as 25-92% of this group, neurological symptoms may precede dry eyes and mouth. [4-7]. The most common neurological manifestations of Sjogrens syndrome include sensory ganglionopathy, painful small fibre neuropathy, and transverse myelitis.

Motor Neuron Disease is a rare presentation of Sjogrens syndrome, described in only a few case reports. The pathogenic mechanisms responsible for most forms of neurological involvement in primary SS are largely unknown [8-10]. Several hypotheses have been put forward including direct infiltration of neurons by mononuclear cells, vascular involvement and ischemia due to small vessel vasculitis [11]. Upper motor neuron signs are more predominant than lower motor neuron

signs and occur early in Sjogrens syndrome patients with MND. Two mechanisms possibly underlie the upper motor neuron involvement in patients with Sjogrens syndrome [12]. One possible mechanism could be ascribed to a primarily autoimmune origin of upper motor neuron involvement as in such patients the presence of several anti-neuronal antibodies is reported in the literature. Another possibility is that subclinical Sjogrens syndrome plays an additive role in the progression of upper motor neuron dysfunction.

There are no consensus guidelines regarding the treatment of neurological manifestations of Sjogrens Syndrome. Steroids, cyclophofamide and IVIG are commonly used, either singly or in combination [13,14]. Some of the manifestations like dementia have an excellent response to steroids while as others like peripheral neuropathy respond poorly. Steroids are usually given in high doses e.g. Prednisolone 1mg/kg upto a total of 80 mg. Azathioprine, Rituximab and Methotrexate are other agents used in steroid unresponsive or steroid dependant cases.

Conclusion

Neurological symptoms occur commonly in Sjogrens syndrome, although MND is rare. Immunosuppressants should be tried in such cases

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