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Occurrence of Pure Sensory Neuropathy in a 45-Year-Old Female with Primary Sjögren's Syndrome: A Case Report Study

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Abstract

Complications of Sjögren's syndrome (SS) and neuropathies may occur in some cases. Almost this complication occurs with peripheral neuropathy. Different forms of sensory neuropathy, multiple mononeuropathy, trigeminal neuropathy, multiple cranial neuropathy, radiculoneuropathy, and autonomic neuropathy can occur in SS. In this novel case presentation, polyneuropathy in a 45-year-old female patient with SS is reported with a complaint of ataxia, vertigo, dysarthria, abnormal gait, foreign body sensation, and deterioration of quadriparesis from six months ago. Fluorescent antinuclear antibody (FANA), anti-RO, anti-RO-52, and RF were strongly detected positive in her rheumatologic examination. Pure sensory polyneuropathy (ganglionopathy) was also reported in electromyography and nerve conduction velocity (EMG-NCV). Although peripheral neuropathy is the major and most common neurological aspect of neuron involvement in patients with SS, here, pure sensory polyneuropathy is reported in a 45-year-old female patient with SS. In conclusion, in this novel case presentation, a novel case of polyneuropathy concurrent with SS is demonstrated in a 45-year-old female patient.

Keywords: Neuropathy, Pure Sensory Neuropathy, Quadriparesis, Sjögren's Syndrome

1. Introduction

Sjögren's syndrome (SS), as an important chronic autoimmune inflammatory disease, can involve exocrine glands primarily (1). It can be categorized into primary SS, when occurs alone, or secondary SS, when occurs in combination with other diseases (almost always connective tissue disease, including systemic lupus erythematosus and/or rheumatoid arthritis) (2). Its prevalence ranges from 0.1% - 4.8% in different reports, also it involves middle-aged women occasionally, and its prevalence in males is 0.11 times its prevalence in females (1, 3). A wide spectrum of clinical signs had been reported, from dry mouth and dry eyes (sicca symptoms) to systemic features (4). Concurrent occurrence of B cell hyperactivity, hypergammaglobulinemia, and serum autoantibodies as a hallmark for the disease (4, 5).

Sjögren's syndrome may become complicated with a wide spectrum of neurological symptoms that peripheral neuropathy is the major and most common neurological aspect of neuron involvement in patients with SS. Different forms of peripheral neuropathies were also reported for SS-related neuropathies such as sensory neuropathy (ataxic form and/or painful sensory form without ataxia), multiple mononeuropathy, trigeminal neuropathy, multiple cranial neuropathy, radiculoneuropathy, and autonomic neuropathy (6). Although progression of neurological signs is slow and they can almost occur in chronic processes in patients, new-onset neuropathies, in the acute and/or subacute forms, are most evident in multiple mononeuropathy and multiple cranial neuropathies (6).

Pure sensory involvement without motor neurons involvement can be predominantly observed in trigeminal, sensory ataxic, painful sensory, and autonomic neuropathies, whereas radiculoneuropathy, multiple cranial neuropathy, and multiple mononeuropathy are characterized by motor neurons involvement (6). Pupils' abnormalities and orthostatic hypotension that are autonomic symptoms are commonly reported in autonomic neuropathies, and multiple mononeuropathy is mostly concomitant with vasculitis (6). However, to the best of our knowledge, there is no report about the occurrence of polyneuropathy in a patient with SS, and we describe a new case of SS with severe poly neuropathic signs in this novel

case report.

2. Case Presentation

A 45-year-old female patient who had two children was referred to the Razi Hospital of Birjand University of medical sciences and health services to receive Rituximab® for the treatment of SS. She complained of different symptoms, including ataxia, vertigo, dysarthria, abnormal gait, foreign body sensation, and deterioration of quadriparesis from six months ago. Fatigue and weakness were recorded in her examination, adding to xerostomia, white pigmentation in mouth and throat, twitching of fingers, formication sense, tremor of upper limbs. Also, her vital signs were recorded as follows: respiratory rate 16/minute, pulse rate 72/minute, blood pressure 100/60 mmHg, body temperature 37°C, and oxygen saturation of 97%. Excluding dysarthria, macrography, and dysdiadochokinesia, her neurological examinations were normal. Her hematological parameters were normal, and fluorescent antinuclear antibody (FANA), anti-RO, anti-RO-52, and RF were strongly detected positive in her rheumatologic examination. Pure sensory polyneuropathy (ganglionopathy) was reported in electromyography and nerve conduction velocity (EMG-NCV). Abnormal findings in her laboratory analysis had been reported as; decreased neutrophil percentage (29.3%) in her CBC, deceased urine volume (600 ml in 24 hours), elevated RF (Roche-Gen II) approximately 1^{+} (16.2 IU/mL), strongly positive FANA titer: > 1/1200), strongly positive (3^{+}) SS-A native (60 kDa) (SSA) = 102, and strongly positive (3^{+}) Ro-52 recombinant (52) = 132 in ANA profile (Western Blot). Elevated hemoglobin/RBP (0.5 mg/24 h), elevated free light chains (5.9 mg/24 h), Alpha 1-microglobulin (4.8 mg/24 h), elevated free light chain dimers (22.7 mg/24 h), elevated transferrin (1.6 mg/24 h), and elevated IgG/IgA (2.9 mg/24 h) in urine electrophoresis. Interstitial pulmonary fibrosis with a normal heart size was reported in chest X-ray graph (Figure 1).

3. Discussion

Neuropathy is one of the important complications of SS due to severe inflammation of the nervous system (7). Sensory nerves neuropathy with a wide spectrum of patterns in its distribution and sensory modalities is an important involvement of peripheral nerves in SS (8). Although motor nerve involvement related to SS has been demonstrated recently (7), it has not been reported as a prevalent neuron involvement in patients with SS (8, 9). Peripheral neuropathy related to SS is increased with aging (10). In this novel case report, concurrent occurrence of polyneuropathy, including vertigo, dysarthria, abnormal gait, foreign body sensation in pharynx, and deterioration of quadriparesis from 6 months ago, is reported in a 45-year-old patient with SS.

Headache, aseptic meningitis, optic neuritis, transverse myelitis, epileptic seizures, disseminated encephalopathy, cognitive disorders, and lesions in the CNS typical of multiple sclerosis have been reported as important neurological lesions concurrent with primary SS (11). Rapid and progressive cerebellar ataxia, cognitive decline, psychiatric problems, and nystagmus had been reported in a patient with SS with different values of cerebellar atrophy in brain magnetic resonance imaging and glucose hypometabolism of the cerebellum in 18F-fluorodeoxyglucose positron-emission tomography (12). In our novel case presentation, different values of vertigo and abnormal gait were also reported that might be related to cerebellar atrophy and other cerebellar lesions in this patient. Dysarthria, limb tremor, and nystagmus had been also reported as common neurological lesions related to cerebellar damages in patients with SS (13).

Dysarthria, ataxia, dysmetria, intention tremor in the limbs, and dysdiadochokinesia had been routinely reported in patients with SS (13, 14). In our report, dysarthria, macrography, dysdiadochokinesia, abnormal gait, and deterioration of quadriparesis were also reported that might be also related to cerebellar lesions. Pure sensory ganglionopathy, which is named pure sensory ataxic neuronopathy and/or pure painful sensory neuronopathy, is an important characteristic neurological lesion concurrent with primary SS due to severe damages to sensory neurons of Gasserian ganglia and dorsal root (15). In this novel case, pure sensory polyneuropathy (ganglionopathy) was reported in EMG-NCV in both of lower limb and upper limb, which was directly related to damage of the sensory neurons of the dorsal root.

Motor neuropathy; motor, sensory, and/or sensorimotor axonal polyneuropathy; multiplex mononeuropathy and/or mononeuropathy multiplex, autonomic neuropathy, small fiber neuropathy, demyelinating polyradiculoneuropathy, and different cranial neuropathies are other common clinical aspects of SS-related peripheral neuropathies (15). In this case presentation, we introduced a 45-year-old female patient with SS suffering from polyneuropathy in different nerves of lower limb. Also, foreign body sensation in this patient may be related to trigeminal and glossopharyngeal nerves neuropathies.

3.1. Conclusions

In this novel case presentation, we described a novel case of polyneuropathy concurrent with SS in a 45-year-old female patient. Also, peripheral neuropathy is the major

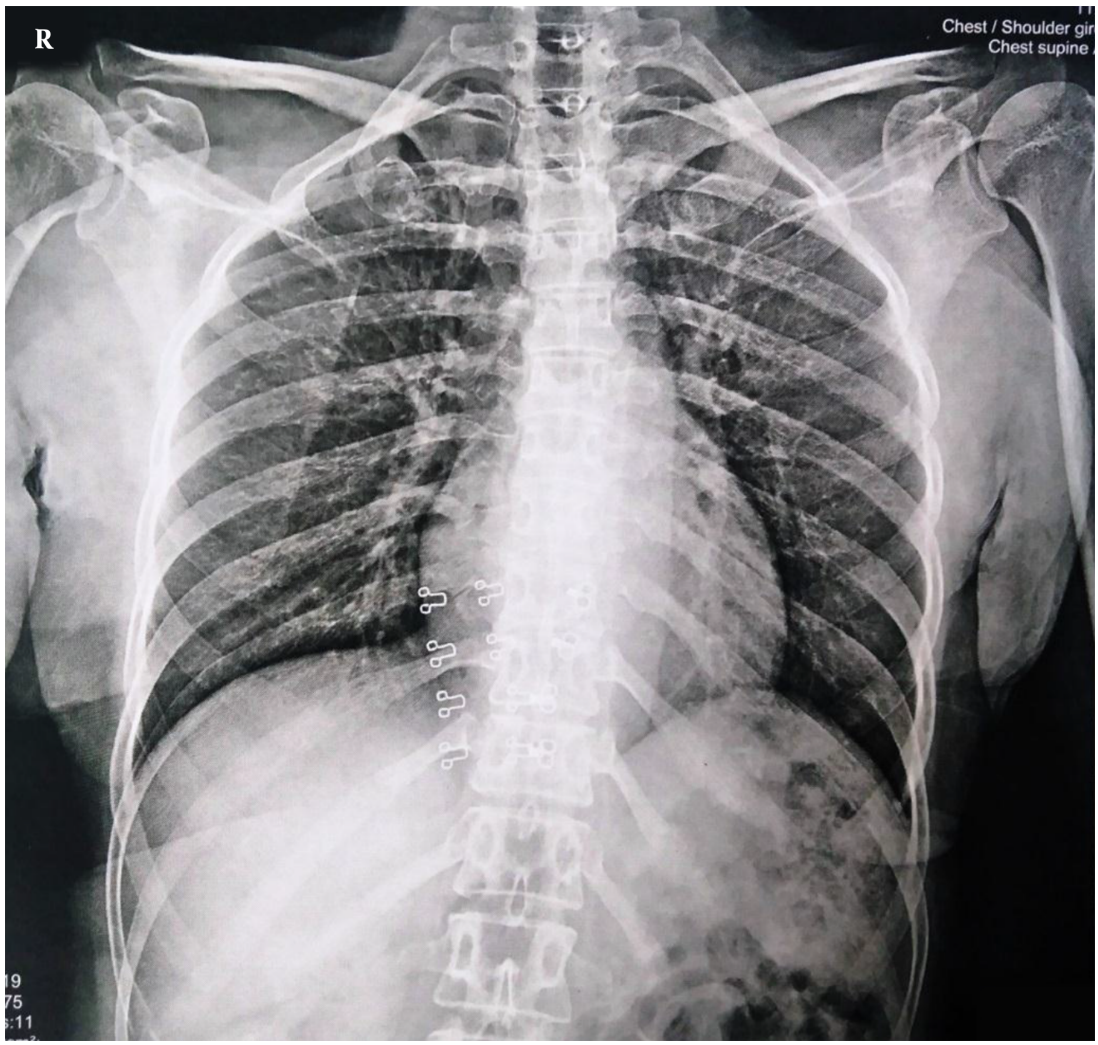

Figure 1. Chest X-ray of the patient. Increase in fibrosis and normal heart size.

and most common neurological aspect of neuron involvement in patients with SS, but in this novel case, pure sensory polyneuropathy is reported in a 45-year-old female patient with SS.

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Footnotes

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