Case report

Ataxic autonomic sensory neuropathy with dorsal root ganglionitis: a rare association of primary Sjögren's syndrome

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Introduction

Sjögren's syndrome first described by Swedish ophthalmologist Henrik Sjögrens in 1933 is an autoimmune epithelitis with lymphocytic infiltration of the exocrine glands, resulting in dry eyes (keratoconjunctivitis sicca) and dry mouth (xerostomia). It can occur alone or in association with other connective tissue diseases. One third of the patients presents with extra glandular neurological, rheumatological, gastrointestinal or pulmonary manifestations. Acute autonomic sensory neuropathy (AASN) with dorsal root ganglionitis is a rare association of primary Sjögren's syndrome (PSS). We report a case of a 29-year-old female who presented with sensory ataxia with autonomic involvement who was found to have PSS.

Case report

A 29-year-old female presented with numbness in all four limbs and face with difficulty in walking and unsteadiness for one week. She had a history of upper respiratory tract infection three weeks back. She had dry eyes and mouth with dry cough and early satiety.

On examination, she had resting heart rate of 120 bpm and supine blood pressure of 160/100 mmHg with blood pressure dropping to 130/90 mmHg on standing. Neurological examination revealed sensory ataxia, generalized absence of joint position sense, vibration sense and global areflexia with abnormal autonomic function tests. Muscle power, pain and temperature sensation and rest of the system examination were unremarkable.

Her ESR was 50 mm/1st hr. Antinuclear antibody was negative. Nerve conduction studies revealed absent sensory nerve action potentials in both upper and lower limbs resembling generalized axonal degeneration. Her motor conduction studies were normal. MRI scan of the brain and cervical spine and cerebrospinal fluid analysis was normal. Her sural nerve biopsy did not reveal any evidence of significant inflammation or granuloma formation.

Schirmer's test and anti SSA antibody were positive (24.3 U/ml) with an equivocal result of anti SSB (3.62 U/ml) antibody. Parotid sialogram was negative. But minor salivary gland biopsy revealed chronic sialadenitis with lymphocytic infiltration.

According to the 2002 American-European classification criteria, the diagnosis of primary Sjögren's syndrome was made with an acute autonomic sensory neuropathy with dorsal root ganglionitis.

She was treated with intravenous immunoglobulin 200 mg/kg/d for five days and oral prednisolone 1 mg/kg/d and thereafter with supportive rehabilitation. Her autonomic functions returned to normal but the dryness and the ataxic sensory neuropathy persisted.

Discussion

AASN is a rare disorder, usually with an antecedent event of either upper respiratory infection or gastrointestinal infection[1]. In the pathogenesis, immune mediated mechanism may be associated with a predisposed risk of lack of blood nerve barrier in autonomic and sensory ganglion. It is associated with connective tissue diseases (Sjögren's syndrome, systemic sclerosis, rheumatoid arthritis, systemic lupus erythematosis, mixed connective tissue disorders) paraneoplastic conditions, coeliac disease, HIV, vitamin B6 toxicity, use of cisplatin and other drugs[2].

Primary Sjögren's syndrome is diagnosed according to the 2002 American-European classification criteria for PSS, which require at least four out of six (two subjective measures of ocular and oral dryness and four objective measures of ocular or salivary gland involvement or elevated anti-Ro or anti-La auto antibodies) or at least three out of the four objective criteria[2,5]. She had fulfilled the criteria needed for the diagnosis of primary Sjögren's syndrome without any association with other autoimmune diseases.

PSS is more common in females with a ratio of 2:1. Neurological manifestations of Sjögrens could be

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peripheral nervous system or central nervous system involvement or myopathies[2,3]. The dorsal root ganglionitis and peripheral nerve vasculitis have been observed in histology. Predominantly sensory and pure sensory neuropathies are most common in PSS. Sensory ataxia, and painful trigeminal neuropathy are due to ganglionitis while multiple mononeuropathy and multiple cranial neuropathy are due to vasculitis[2]. Studies have shown that antibodies against dorsal root ganglion play a role in pathogenesis of sensory neuropathy[4]. In our patient the sural nerve biopsy was normal and with the evidence of autonomic and sensory involvement, the possible site of pathology is at the sensory ganglion.

IV Immunoglobulin is used to treat sensory ataxic neuropathy and small fibre involvement. For multiple mononeuropathies and cranial neuropathies, corticosteroids are used as the first line treatment[5]. Plasmapheresis, D-penicillamine, infliximab, cyclophosphamide, rituximab and Interferon α have been used[2,5].

Considering the age, gender, dry mucous membranes and finding of sensory and autonomic neuropathy with laboratory finding of generalized sensory axonal neuropathy, positive Schirmer's and anti SSA antibody, the most likely diagnosis is PSS with a rare manifestation of ganglionitis[3].

References

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