

Case Report

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Sjogren's Syndrome Involving the Central Nervous System Lesions as the First Performers: A Case Report and Literature Review

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Abstract

Sjogren's syndrome is primarily a chronic systemic autoimmune disease that involves many systems. However, Sjogren's syndrome involving the central nervous system lesions, particularly in the central nervous system symptoms as the first performers is uncommon. In this report, we present a 33-year-old woman having PSs with CNS involvement, whose first symptoms presented progressive weakness on the left leg, 2 years later the left upper limb appeared the same symptoms, being diagnosed with Sjogren's syndrome. To give methylprednisolone pulse therapy, combined with cyclophosphamide and anetholtrithione treatment, her condition improved. After six months' following-up, no recurrence.

Keywords: Sjogren's syndrome; central nervous system lesions; First performers

Introduction

Primary Sjogren's syndrome (PSs) is an autoimmune inflammatory disorder characterized by lymphocytic infiltration of exocrine glands, mainly the lacrimal and salivary glands leading to a chronic sicca syndrome. However, extra glandular organ systems may frequently be involved, including both central and peripheral nervous systems. The reported prevalence ranged from 20% to 60%, among which peripheral nervous system lesions more common, but the incidence of central nervous system diseases is low. Especially, Sjogren's syndrome involving central nervous system lesions as the first manifestation is rarer, which is insidious and difficult to distinguish from central nervous system diseases? [1]. Here, we report a case of a patient with PSs who firstly presented with the central nervous system lesions, and review of the relevant literature. We hope to provide help for the diagnosis of Sjogren's syndrome involving central nervous system lesions, reducing the misdiagnosis.

Case Report

A 42-year-old woman was admitted with a 4-year history of progressive weakness on the left leg, 2 years later the left upper limb appeared the same symptoms. In the last 3 months, the weakness was aggravated, and suffered from pruritus, dry mouth and dry eye the last two months. On admission, her general examination was normal. The neurological examination revealed following positive signs only: Left upper limb muscle strength 4+ level, the left lower limb proximal muscle strength 4- level, bilateral Achilles tendon reflex reduced (+), bilateral Babinski sign (+), bilateral Chaddock sign (+), the left Hoffmann sign (+). Urine of patients had no obvious abnormalities. The patients had no other previous diseases. All of the additional examination results were normal except for brain and spinal cord MRI, which showed that cerebral white matter of the cerebral hemispheres with periventricular appeared multiple lesions (Figure 1A), C5-C6 level of the spinal cord also appeared multiple lesions (Figure 1B and C).

The results of routine laboratory studies were also at normal levels, including complete blood cell count, coagulation, liver function, kidney and thyroid, lipids, glucose, glycosylated haemoglobin, C-reactive protein, and anti-O chain. Serological tests for HBsAg, hepatitis C virus, human immunodeficiency virus, syphilis, and tumour markers were negative. She was positive for anti-SSA antibody (+) and antinuclear antibody positive quantitative ANA 1:80. The CSF examination showed that the cell number was 14, small lymphocytes accounted for 90%, and

general monocytes accounted for 10%, IgG increased as 81.400 mg/L. The histopathological results was per ductal lymphocytic infiltration of the lower lip gland (Figure 2). To give methylprednisolone pulse therapy, combined with cyclophosphamide and anetholtrithione treatment, her condition improved. After six months' following-up, no recurrence.

Discussion and Literature Review

Sjogren's syndrome is primarily a chronic systemic autoimmune disease that affects exocrine organs characterized by focal lymphocytic infiltration and destruction of these glands. However, extra glandular organ systems may frequently be involved, including both central and peripheral nervous systems. However, Sjogren's syndrome involving the central nervous system lesions, particularly in the central nervous system symptoms as the first performers is uncommon [2]. According to the domestic and foreign literature, the incidence rate of Sjogren's syndrome involving in central nervous system disease (CNS-pSS) is about 10% [3]. Various neurological manifestations have been reported in SS affecting the central nervous system (CNS), including brain, spinal cord, and optic features. Central nervous system involvement is composed by multiple sclerosis-like manifestations including acute and chronic myelopathies and by more diffuse manifestations (cognitive dysfunction, subacute aseptic meningitis, encephalopathy, psychiatric symptoms, chorea, seizures...) [4-6]. In addition to brain lesions, spinal cords are more often involved. The clinical manifestations of myelopathy were acute or subacute transverse myelitis, chronic progressive myelopathies or relapsing and remitting cord syndromes. Most commonly, thoracic spinal cord was affected [7]. Currently, the diagnosis of PSs is based on the following: high titters for anti-SS-A/Ro, SS-B/La, and recombinant Ro-52 antibodies and rheumatoid factor; elevated IgG and IgA; accelerated ESR; decreased exocrine glandular function; no trace of other autoimmune or infectious disease; and the revised classification criteria for Sjögren's syndrome proposed by the

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Received May 19, 2015; Accepted June 03, 2015; Published June 06, 2015

Citation: Qiao S, Liu XW, Han T, Li WN, Meng MZ, et al. (2015) Sjogren's Syndrome Involving the Central Nervous System Lesions as the First Performers: A Case Report and Literature Review. J Clin Case Rep 5: 549. doi:10.4172/2165-7920.1000549

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Figure 2: The histopathological results were periductal lymphocytic infiltration of the lower lip gland.

American-European Consensus Group [8]. Our patient with PSs firstly presented with the central nervous system lesions, after suffering from 4-year progressive weakness on the left leg and 2-year weakness on the left upper limb she was affected by dry mouth and dry eye. The brain and spinal cord MRI showed focal involvement. The laboratory tests which was positive for anti-SSA antibody (+) and antinuclear antibody positive quantitative ANA 1:80 also support the diagnosis of Sjogren's syndrome. The histopathological results showed focal lymphocytic infiltration. The clinical data of our patient was consistent with the Sjogren's diagnosis. According to previous literature, corticosteroid therapy was applied with good therapeutic response [9]. In our case, we treated her with a large dose of methylprednisolone treatment, combining with cyclophosphamide and anetholtrithione. As a result, the patient's symptoms were well relieved, and after six months' following-up, no recurrence, which reminds us that treatment of Sjogren's syndrome with high-dose steroid pulse therapy combined with immunosuppressant, may achieve a good clinical efficacy.

Conclusion

Sjogren's syndrome involving the central nervous system lesions is uncommon, and its diagnosis is more difficult. Only have considered the clinical symptoms, pathological examination, neuroimaging and laboratory tests, can we accurately diagnose and ensure the treatment.

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