



The Association of Neuromyelitis Optica without Myelitis and Systemic Lupus Erythematosus: A Case Report

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ABSTRACT

Systemic lupus erythematosus is an autoimmune disease. Neuromyelitis optica (Devic's disease) is an inflammatory disorder belonging to the central nervous system. The typical characterizations include severe, immune-mediated demyelination, and axonal damage which mostly involve optic nerves and spinal cord, also the brain and brainstem. Anti-aquaporin-4 antibody has been recently described as a highly specific marker for neuromyelitis optica. Neuromyelitis optica occasionally is linked with systemic autoimmune disorders, including systemic lupus erythematosus. Here, we describe a 26-year-old young woman with systemic lupus erythematosus who had bilateral optic neuritis with no evidence of myelitis or other core clinical criteria. However, aquaporin 4 antibody with high titer was detected. The patient received high-dose prednisolone, cyclophosphamid, and rituximab; but this treatment caused no change in her visual acuity. In patients with systemic lupus erythematosus who complicated by optic neuritis (with or without myelitis), the association of neuromyelitis optica should be considered.

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Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease with adverse effects on various body systems, including central nervous system (CNS) (1).

Neuropsychiatric lupus erythematosus (NPSLE) includes a wide range of neurologic and psychiatric symptoms that can involve any aspect of the central or peripheral nervous system (2). Despite the fact that neurologic

complications of SLE may occur in up to 75% of patients, transverse myelitis is rare, with an occurring rate of only 2% of patients (3).

Neuromyelitis optica (NMO), or Devic's disease, is an inflammatory CNS disorder characterized by concomitant or sequential attacks of optic neuritis and/or transverse myelitis, usually limited to the optic nerves and spinal cord. Anti-aquaporin-4 (anti-AQP4) antibody has been recently described as a highly specific marker for NMO (4). In patients with SLE, NMO is a very rare, but, a very serious neurologic manifestation. The majority of patients with SLE who develop NMO have myelitis with or without optic neuritis. There are very few case reports of patients with SLE who developed NMO without myelitis (5). Here, we report a patient with SLE who had only bilateral optic neuritis with no evidence of myelitis. However, anti-AQP4 antibody with high titer was detected.

Case Report

A 26-year-old Turkmen woman from Iran presented in 30 June 2016 with headache and sudden loss of vision in her eyes (more severe in right eye). She was a known case of SLE. The diagnosis of SLE has been made since 6 years before, based on malar rash, polyarthritis, alopecia, and high levels of antinuclear antibody (ANA) and anti-DNA. She had involved Reynaud's phenomenon. She was on prednisolone 5 mg daily and hydroxychloroquine 400 mg daily. She had a normal childbirth 4 months before, with no history of abortion. On admission, vital signs were normal. Physical examination was normal except for diminished visual acuity in her eyes (more severe in right eye) and a positive relative afferent pupillary defect (RAPD) in right eye. Eye examination by an ophthalmologist showed blurring of disc margins (more severe in right eye). Routine laboratory examinations showed normal results except for a 1st hour erythrocyte sedimentation rate (ESR) of 62 mm. The fluorescent antinuclear antibody (FANA) test

was positive with high titer (1/1000), anti-double stranded DNA antibody (anti-dsDNA) was more than 150 IU/ml (normal range: < 20 IU/ml), and immunoglobulin G (IgG) anticardiolipin was 57.1 GPL U/ml (normal range: < 20 GPL U/ml). Visual evoked response (VER) showed abnormal conduction in optic nerves (severe in right eye and mild in left eye). Brain and cervical magnetic resonance imaging (MRI) were normal. The diagnosis of optic neuritis was made based upon the history, eye examination and, VER findings.

The patient received a course of pulse of methylprednisolone (1000 mg/day for three days) followed by 1 mg/kg of oral prednisolone. She also received intravenous cyclophosphamide (1000 mg/month). After that, only mild transient improvement in her visual acuity occurred and further recovery was not observed. Three months later, despite the fact that she received another two monthly cyclophosphamide pulses, she had severe visual loss in her right eye. She was taking 15 mg prednisolone daily after tapering. The patient was referred to an expert neurologist for reevaluation. He requested blood exam for anti-AQP4 antibody. The anti-AQP4 antibody was highly positive (1/2560, normal range: < 1/10). The patient fulfilled the International Consensus Diagnostic Criteria for NMO spectrum disorders (6) According to this diagnosis, the patient received intravenous rituximab (1000 mg on days 1 and 15, a second course was also administered 6 months later). This treatment caused no change in visual acuity and at present, the patient continues to have severe visual loss in her right eye and to some extent, in her left eye.

Discussion

Here, we described a 26-year-old young woman with SLE who had bilateral optic neuritis with no evidence of myelitis. However anti-AQP4 IgG antibody with high titer was detected. According to presence of optic neuritis and anti-AQP4 IgG antibody, our patient fulfilled the International

Consensus Diagnostic Criteria for NMO spectrum disorders (6). The anti-AQP4 antibody is present in approximately 80% of patients with NMO and has a specificity of 100% for the diagnosis of NMO (7). In SLE-associated forms, anti-AQP4 antibody can help distinguish between SLE-related CNS manifestations and NMO (7). There are some studies regarding to the anti-AQP4 antibody positivity in patients with rheumatic diseases including SLE. In a study, among 170 patients with rheumatic diseases without neurological symptoms, none of them had anti-AQP4 antibodies (8). In a population-based study on patients with SLE, anti-AQP4 IgG was detected in 2 of 208 patients, both of whom had antiphospholipid syndrome and myelitis (9). In another study, of 210 patients with pediatric SLE, 2 patients (0.9%) had NMO. They both had optic neuritis with myelitis (10).

In a study by Maritsi et al., 2 of 89 (2.2%) patients with SLE without CNS involvement were positive for anti-AQP4 autoantibodies. The authors concluded that anti-AQP4 antibodies can be present in patients with SLE and persist for many years, without any radiological or clinical signs (11).

Most of the patients with SLE who develop NMO have myelitis; and patients with SLE who present with optic neuritis (due to MNO) are often complicated by myelitis later. So, NMO without myelitis is very rare in patients with SLE (4, 5, 8, 11).

Costa et al. described a 32-year-old patient with SLE with recurrent optic neuritis. She was seropositive for anti-AQP4 antibody. They treated the patients with low-dose prednisolone. After 5 years, her visual acuity was intact with no relapses and no disability (5). This patient, like our patient, had NMO without myelitis. Unlike this patient, our patient did not have a good prognosis and suffered from severe visual impairment.

All patients with possible NMO should be treated for acute attacks. The treatment of choice here, is pulse methylprednisolone. For severe cases who do not respond to glucocorticoids, therapeutic plasma exchange

is recommended. The best technique to prevent the recurrence of attacks is treatment with systemic immunosuppressants. The best drug regimen and treatment duration have not been determined yet. Azathioprine, rituximab, and mycophenolate mofetil have been listed as the primary treatment option for NMO. The recurrent attacks of NMO cause growing visual, motor, sensory, and bladder deficits (1).

In patients with SLE who complicated by optic neuritis with and without myelitis, the association of NMO should be considered.

Conflict of Interests

Authors have no conflict of interests.

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