

# A 36-year-old Woman with Spastic Quadripareisis: A Case Report and Review of Literature

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**Abstract** Neuromyelitis Optica (NMO) is a necrotizing autoimmune disease that affects the spinal cord and optic nerve. It can be associated with systemic autoimmune disorders such as systemic lupus erythematosus (SLE), thyroid diseases, myasthenia gravis and Sjögren's syndrome. The incidence NMO is very rare in lupus and only have been reported as a case report. In this paper, we present a case of SLE who developed NMO in the course of the disease and we discuss that it is essential in any SLE patient with myelitis, Devic syndrome should be considered.

**Keywords** Neuromyelitis Optica, Systemic lupus erythematosus, Myelitis

## 1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can affect any organ [1]. Its clinical manifestations and course are highly variable. Central nervous system (CNS) complications of SLE is different including seizures, cerebrovascular disease, myelopathy, mood disorders, cognitive impairment and aseptic meningitis [2]. Neuromyelitis Optica (NMO) is a necrotizing autoimmune disease that affects the spinal cord and optic nerve. The disease can be idiopathic or associated with other inflammatory autoimmune diseases [3]. Myelitis is a rare complication of lupus that involves 2% of patients. It can occur as the first manifestation or in the course of a disease [4]. Spinal cord involvement is characteristic in NMO. Its lesion is necrotizing and longitudinal extending over three or more vertebral segments [5]. For this reason, the incidence of this myelitis and devic is very rare in lupus and only have been reported as a case report. We report a case of SLE that developed NMO in the course of the disease.

## 2. Case Report

We report a case of a 36-year-old woman with a history of SLE. The patient's lupus was diagnosed 8 years ago when she presented with morning Stiffness and pain in joints of hands and since then, her disease was in remission with prednisone doses ranging from 5 mg to 20 mg once a day and hydroxychloroquine 200 mg daily. The patient presented to

our hospital with 2 weeks history of lower limbs weakness and gait disturbance that progress to upper limbs during 1 day. She also reported paresthesia of the lower extremity over 1 month. Her family history, social history, and review of systems were negative. On examination, she was conscious and had stable vital signs, there was no peripheral lymphadenopathy palpable. He had normal rate and regular rhythm, no murmur. The abdomen was flat, soft and non-tender with normal liver and spleen. Muscle strength in proximal and distal upper limbs were 4/5 and in lower limbs were 3/5. Sensory examination shows sensory level in T5 with abnormal position and vibratory in lower limbs. Deep tendon reflexes were 3+ in upper limbs and 4+ in Achilles and knee and the plantar response was bilateral extensor. Brain MRI showed no abnormality. MRI of spine showed longitudinal hypersignality lesion in whole cervical and upper thoracic. Cerebrospinal fluid (CSF) was normal for cell count, protein and glucose. Oligoclonal bands (OCB) was negative. NMO Ab was positive. Plasma exchange was started with total dose 250 cc/kg that marked improvement observed in the period of hospitalization and 1 month later in follow-up visit.

## 3. Discussion

NMO, also known as Devic's disease, is a rare autoimmune disorder characterized by involvement of optic nerve and spinal cord. The pathogenesis of NMO is different from MS [6]. In this disease the humoral immune system has important role, NMO-IgG (anti-AQP4) plays a direct role in the pathogenesis of NMO. It is against of Aquaporin-4 (AQP4), a water channel protein that is highly concentrated in brain and spinal cord, in addition, NMO mainly causes necrosis of white matter and gray matter of the brain and the

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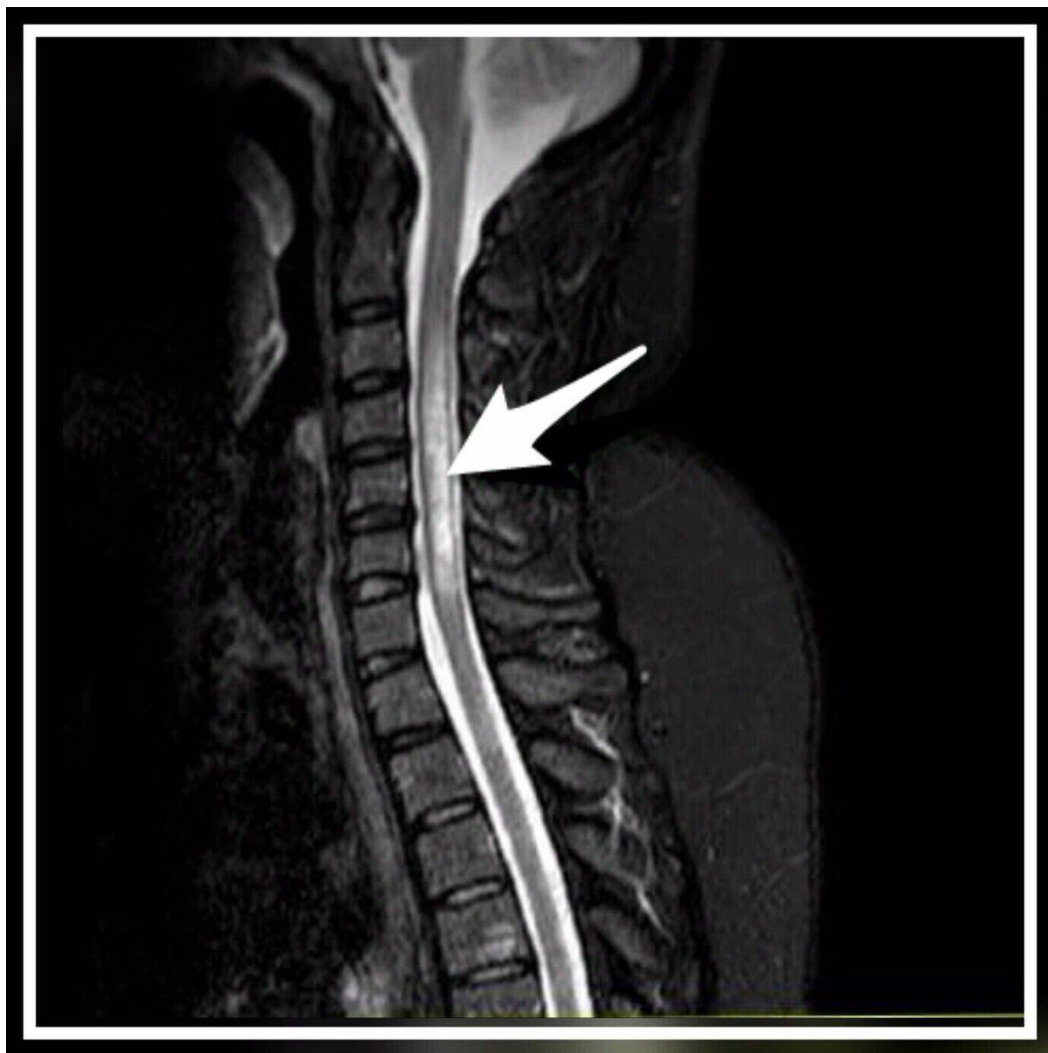
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spinal cord and usually with absence of OCB in cerebrospinal fluid (CSF) [7, 8]. It can be associated with systemic autoimmune disorders such as systemic lupus erythematosus, thyroid diseases, myasthenia gravis and Sjögren's syndrome [9]. SLE has variable effects on the central nervous system (CNS). Neuropsychiatric complications of SLE is different including stroke and transient ischemic attacks, seizures, aseptic meningitis, myelopathy, polyneuropathy, organic psychosis, mood disorders, cognitive impairment and movement disorder such as chorea [10]. The Manifestations of myelitis or optic neuritis in SLE are very unusual that only 1–2% of patients with SLE exhibit myelitis or optic neuritis and only half of the patients affected by myelitis have criteria for the NMO spectrum of disorders [11]. Deodhar et al. reported the first case of SLE with longitudinal myelitis in 1999. He described a 21-year-old woman with SLE that developed extensive involvement of the spinal cord with good response to

cyclophosphamide and high dose corticosteroids [12]. The optic neuritis in SLE is usually bilateral with poor prognosis compared to multiple sclerosis [13]. Treatment in this disease consisted of plasma exchange and IV methylprednisolone for acute attacks and azathioprine, rituximab, mycophenolate mofetil, methotrexate, or mitoxantrone for maintenance therapy [14]. In a study Mok CC and colleague showed use of cyclophosphamide was successful in halting relapses in a patient with systemic lupus erythematosus-associated NMO who was unresponsive to high-dose oral and intravenous corticosteroids, intravenous immunoglobulin, mycophenolate mofetil, tacrolimus, low-dose daily oral cyclophosphamide and rituximab [15]. In conclusion it is necessary, in any patient with SLE that present with myelitis, Devic syndrome should be considered because prompt diagnosis and treatment prevents disability and mortality.



**Figure 1.** Sagittal STIR MRI reveals the large longitudinal lesion in cervical and upper thoracic spinal cord

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## REFERENCES

- [1] ACR Ad Hoc Committee on Neuropsychiatric Lupus Nomenclature (1999) The American College of Rheumatology nomenclature and case definitions for neuropsychiatric lupus syndromes. *Arthritis Rheum* 42: 599-608.
- [2] Kampylafka EI, Alexopoulos H, Kosmidis ML, Panagiotakos DB, Vlachoyiannopoulos PG, et al. (2013) Incidence and prevalence of major central nervous system involvement in systemic lupus erythematosus: a 3-year prospective study of 370 patients. *PLoS One* 8: e55843.
- [3] Kolfenbach JR, Horner BJ, Ferucci ED, West SG (2011) Neuromyelitis optica spectrum disorder in patients with connective tissue disease and myelitis. *Arthritis Care Res (Hoboken)* 63: 1203-1208.
- [4] Carmona-Fernandes D, Santos MJ, Canhão H, Fonseca JE (2013) Antiribosomal P protein IgG autoantibodies in patients with systemic lupus erythematosus: diagnostic performance and clinical profile. *BMC Med* 11: 98.
- [5] Fujihara K, Leite MI: Seronegative NMO: a sensitive AQP4 antibody test clarifies clinical features and next challenges. *Neurology* 2013; 80:2176–2177.
- [6] Akman-Demir G, Tüzün E, Waters P, İçöz S, Kürtüncü M, Jarius S, et al: Prognostic implications of aquaporin4 antibody status in neuromyelitis optica patients. *J Neurol* 2011; 258: 464–470.
- [7] Takahashi T, Fujihara K, Nakashima I, Misu T, Miyazawa I, et al. (2007) Anti-aquaporin-4 antibody is involved in the pathogenesis of NMO: a study on antibody titre. *Brain* 130: 1235-1243.
- [8] Trebst C, Jarius S, Berthele A, et al. Neuromyelitis Optica Study Group (NEMOS). Update on the diagnosis and treatment of neuromyelitis optica: recommendations of the neuromyelitis optica Study group (NEMOS). *J Neurol.* 2014; 261: 1–16.
- [9] Weinshenker BG, de Seze J, Cedex L, Vermersch P, Pittock S, Lennon V. The relationship between neuromyelitis optica and systemic autoimmune disease. American Academy of Neurology 58th Annual Meeting. April 2006. San Diego, California.
- [10] Popescu A, Kao AH (2011) Neuropsychiatric systemic lupus erythematosus. *Curr Neuroparmacol* 9: 449-457.
- [11] Kovacs B, Lafferty TL, Brent LH, DeHoratius RJ. Transverse myelopathy in systemic lupus erythematosus: an analysis of 14 cases and review of the literature. *Ann Rheum Dis* 2000.
- [12] A.A. Deodhar, T. Hochnedel, R.M. Bennet Longitudinal involvement of the spinal cord in a patient with lupus related transverse myelitis *J Rheumatol*, 26 (1999), pp. 446-449.
- [13] Bertsias GK, Ioannidis JP, Aringer M, Bollen E, Bombardieri S, et al. (2010) EULAR recommendations for the management of systemic lupus erythematosus with neuropsychiatric manifestations: report of a task force of the EULAR standing committee for clinical affairs. *Ann Rheum Dis* 69: 2074-2082.
- [14] Polgár A, Rózsa C, Müller V, Matolcsi J, Poór G, Kiss EV. Devic's syndrome and SLE: challenges in diagnosis and therapeutic possibilities based on two overlapping cases. *Autoimmun Rev.* 2011; 10:171–4.
- [15] Mok C.C., Ying K.Y., Tang S., Leung C.Y., Lee K.W., Ng W.L., et al. (2004) Predictors and outcome of renal flares after successful cyclophosphamide treatment for diffuse proliferative lupus glomerulonephritis. *Arthritis Rheum* 50: 2559–2568.