

A successful case of lupus myelitis treated with intravenous pulse methylprednisolone and pulse cyclophosphamide therapy

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SUMMARY Lupus myelitis is a rare but serious condition characterized by myelopathy in patients with systemic lupus erythematosus (SLE). Its presentation is usually acute or subacute, and it is often refractory to treatment. We reported a rare presentation of lupus myelitis in a 38-year-old Japanese woman with a 20-year history of SLE. She developed paraparesis and bladder/bowel dysfunction 6 months prior to presentation. Magnetic resonance imaging revealed atrophy of the entire thoracic spinal cord with high intensity on T1-weighted sequence. She was initially treated with intravenous pulse steroid therapy, and prednisolone (20 mg/day) was continued; mizoribine was changed to azathioprine (100 mg/day). In addition, she underwent a rehabilitation program to improve lower-extremity muscle weakness. Moreover, because of the refractory clinical condition, intravenous cyclophosphamide pulse therapy was added. Within 1 month, she could walk with a cane and had a desire to urinate and defecate. In conclusion, early and aggressive treatment improves the permanent damage of lupus myelitis.

Keywords systemic lupus erythematosus, central nervous system lupus, myelitis, methylprednisolone, cyclophosphamide

Lupus myelitis is a rare and serious complication in patients with systemic lupus erythematosus (SLE). Partial remission and complete recovery occur in 50.0-62.5% and 7.1-27.8% of these patients, respectively (1). Here, we report the improvement of a rare case of lupus myelitis using intravenous pulse methylprednisolone and pulse cyclophosphamide therapy.

A 38-year-old woman with SLE received oral prednisolone (20 mg/day) and mizoribine (100 mg/day) for 20 years at a nearby hospital. She was admitted to our hospital because she developed paraparesis and bladder/bowel dysfunction 6 months prior to presentation. She was bedridden because of the exacerbation of these symptoms. On admission, she presented with discrete maculopapular erythema on her face (Figure 1A), ulcers on the buttock and oral mucosal ulcers. Laboratory findings were as follows: antinuclear antibody at a titer of 1:1,280 in a speckled pattern, anti-double-stranded DNA antibody level > 200 IU/mL (< 12 IU/mL); total functional hemolytic complement (CH50) < 10 U (31.6-57.6 U/mL); C3, 22.8 mg/dL (73-138 mg/dL); C4, 1.3 mg/dL (11-31 mg/dL); anti-cardiolipin β -2-glycoprotein I, 1.3 U/mL (< 3.5 U/mL); anti-cardiolipin immunoglobulin G < 8 U/mL (< 10 U/mL); an anti-aquaporin 4 antibody < 1.3 U/

mL (< 5.0 U/mL). Magnetic resonance imaging (MRI) revealed atrophy of the entire thoracic spinal cord with high intensity on T1-weighted sequence (Figure 1B). Cerebrospinal fluid (CSF) examination showed elevated protein levels of 47.6 mg/dL (8-43 mg/dL), and the immunoglobulin G index was 1.18. Oligoclonal bands in CSF were also positive. Pretherapeutic SLE disease activity index (SLEDAI) score was 16 (vasculitis, new rash, mucosal ulcers, low complement, increased DNA binding). Based on these results, she was diagnosed with myelitis associated with SLE (lupus myelitis). She was initially treated with intravenous pulse steroid therapy, and prednisolone (20 mg/day) was continued; mizoribine was changed to azathioprine (100 mg/day). In addition, she underwent a rehabilitation program to improve lower-extremity muscle weakness. One week after the initiation of this therapy, the SLEDAI score improved up to 4 (low complement, increased DNA binding). Because of refractory hypocomplementemia, high titer of anti-double-stranded DNA antibody and leukopenia, intravenous cyclophosphamide pulse therapy was added. Within 1 month, she could walk with a cane and had a desire to urinate and defecate.

Overall, 1-2% of patients with SLE have lupus myelitis (2). It occurs most commonly at the thoracic

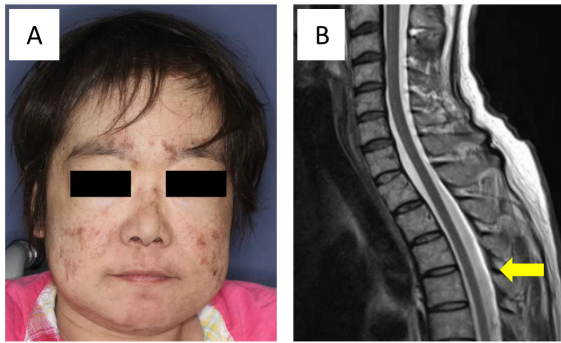


Figure 1. (A) Discrete maculopapular erythema on her face. (B) Magnetic resonance imaging (MRI) showed an atrophy of whole thoracic spinal cord.

levels, especially at the levels of T5-T8 (2). The factors associated with neurological outcomes of lupus myelitis include: extensive spinal cord lesions, severity of the symptoms at onset, antiphospholipid antibodies, and delay of treatment (3). Although the therapeutic strategy for lupus myelitis is based on SLE, initial treatment often involves intravenous pulse methylprednisolone and pulse cyclophosphamide (1). Rituximab, intravenous immunoglobulin, and plasma exchange therapy are sometimes added as combination therapies (4). Anticoagulation therapy has also been shown to be effective in patients who are antiphospholipid-positive (1,5). In conclusion, early and aggressive treatment improves the permanent damage of lupus myelitis.

References

1. Li XY, Xiao HB, Pai P. Myelitis in systemic lupus erythematosus. *J Clin Neurosci.* 2017; 44:18-22.
2. 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; 59:120-124.
3. Bertias GK, Ioannidis JPA, Aringer M, *et al.* 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.* 2010; 69:2074-2082.
4. Ye Y, Qian J, Gu Y, Chen X, Ye S. Rituximab in the treatment of severe lupus myelopathy. *Clin Rheumatol.* 2011; 30:981-986.
5. D'Cruz DP, Mellor-Pita S, Joven B, Sanna G, Allanson J, Taylor J, Khamashta MA, Hughes GR. Transverse myelitis as the first manifestation of systemic lupus erythematosus or lupus-like disease: good functional outcome and relevance of antiphospholipid antibodies. *J Rheumatol.* 2004; 31:280-285.

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