

Intravenous human immunoglobulin for the treatment of severe longitudinal extensive transverse myelitis associated with systemic lupus erythematosus

Duarte AC¹, Sousa S¹, Nunes T², Cordeiro A¹, Gonçalves P¹

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We report the case of a 39-year-old female, with systemic lupus erythematosus (SLE) diagnosed 13 years ago, with positive antinuclear antibodies (1/1280), anti-double stranded DNA and antiphospholipid antibodies, and known mucocutaneous, articular, lung, gastrointestinal, renal and haematological involvement, under current treatment with hydroxychloroquine and mycophenolate mofetil. She was admitted with a 7-day history of fever, headaches and non-pruritic erythematous rash. Laboratory showed haemolytic anaemia, thrombocytopenia, raised C-reactive protein and erythrocyte sedimentation rate, minimal decreased C3 level, normal anti-dsDNA antibody and worsening proteinuria (2.3g/day) with granular casts. Cerebrospinal fluid (CSF) showed 230 leucocytes/L (mainly polymorphonuclear), total protein of 176mg/dL and glucose of 60mg/dL. Bacterial meningitis was considered and she started ceftriaxone, vancomycin and ampicillin. Fever persisted despite sterile CSF and 8 days later she suddenly developed hyporeflexive paraparesis with total sensory loss below D10. Magnetic resonance imaging (MRI) demonstrated an intradural extramedullary hematoma D9-L3 (after difficult lumbar puncture) responsible for spinal cord compression with spinal cord edema between D9 and conus medullaris. Collection was surgically drained with edema partial resolution. During follow-up surgical scar became infected with *Citrobacter koseri* and *Klebsiella pneumonia* and was treated accordingly. After a month of intensive rehabilitation, neurological deficits persisted and urinary retention developed. A new MRI showed holocord abnormal T2 hyperintense signal, from the conus medullaris to the brainstem, with associated swelling (panel 1, Figure 1). Longitudinal extensive transverse myelitis (LETM) was assumed and complementary exams re-

vealed normal glycaemia, thyroid function, vitamin B12 levels and angiotensin-converting enzyme; infection was ruled out after negative serologies for syphilis, *Borrelia burgdorferi*, human immunodeficiency virus, hepatitis B and C viruses and herpesviruses (herpes simplex, cytomegalovirus and Epstein-Barr virus), sterile CSF (including for *Mycobacterium tuberculosis* and *non-tuberculosis mycobacteria*) and negative CSF protein-chain reaction for neurotropic viruses. Anti-aquaporin-4 antibodies were negative and there were no oligoclonal bands in CSF. An association with the underlying SLE was assumed (SLEDAI-2K 22) and she received

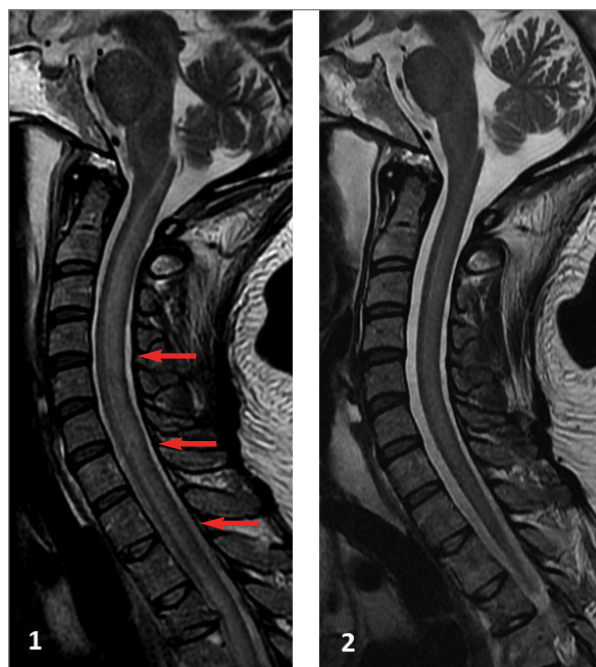


FIGURE 1. Magnetic resonance imaging demonstrating holocord abnormal T2 hyperintense signal at diagnosis (1) and subsequent reduction in extension and intensity after pulsed methylprednisolone and one administration of intravenous human immunoglobulin (2).

1. Rheumatology, Hospital Garcia de Orta, Almada, Portugal
2. Neuroradiology, Hospital Garcia de Orta, Almada, Portugal

Ig methylprednisolone daily for 3 days, followed by oral prednisolone (1mg/Kg/day) and intravenous human immunoglobulin (IVIG;1g/kg/day for 2 days). One week after IVIG administration clinical and imaging improvement was noted, in both intensity and extension of the abnormal spinal cord T2 signal (panel 2, Figure 2). Monthly IVIG was kept for 6 months with resolution of MRI findings after 3 months and progressive neurological recovery, with ability to stand and walk. Four months after IVIG suspension no LETM recurrence occurred and proteinuria improved (0.7g/day), with a SLEDAI-2K 4.

LETM is a rare SLE complication. Although there are reports of patients treated with high-dose steroids, immunosuppressive agents and plasmapheresis, only one-third recovers with minimal deficits^{1,2}. This case illustrates and reinforces the role of IVIG in severe SLE, when infection is present/suspected.

CORRESPONDENCE TO

Ana Catarina Duarte
Serviço de Reumatologia, Hospital Garcia de Horta
Av. Torrado da Silva,
2805-267 Almada
E-mail: catarinaduarte89@gmail.com

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