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Case Report

A patient with systemic lupus erythematosus with acute inflammatory demyelinating polyradiculoneuropathy with progression to encephalopathy and status epilepticus: A case report☆

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ABSTRACT

Systemic lupus erythematosus (SLE) is a systemic inflammatory and autoimmune disease which can affect any organ, including the nervous system. We report a case of a woman with SLE with flaccid tetraparesis and facial diplegia, which suggest inflammatory demyelinating polyneuropathy. This syndrome has been described in patients with lupus, however in this case the patient had an unusual evolution, with no response to immunosuppressive treatment and progression to status epilepticus.

Paciente con lupus eritematoso sistémico con polirradiculoneuropatía inflamatoria desmielinizante aguda con evolución a encefalopatía y estatus epiléptico: a propósito de un caso

Palabras clave:
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RESUMEN

El lupus eritematoso sistémico (LES) es una enfermedad inflamatoria sistémica de carácter autoinmune que puede afectar a cualquier órgano y entre ellos el sistema nervioso. Presentamos el caso de una paciente con LES que presenta un cuadro de tetraparesia fláccida y diplejía facial que sugirió polineuropatía inflamatoria desmielinizante. Este síndrome

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está descrito en pacientes con LES, sin embargo, en este caso presentó una evolución poco frecuente, sin respuesta a tratamiento inmunosupresor y desarrollo de estatus epiléptico.

Introduction

Systemic lupus erythematosus (SLE) is a multisystem inflammatory disease that can affect any organ, including the nervous system. There are 19 forms of presentation of neurolupus described according to the criteria of the American College of Rheumatology (ACR), and among them there is the acute demyelinating inflammatory polyneuropathy.[1]

We present a case of severe SLE that develops a picture of acute demyelinating inflammatory polyneuropathy with an unusual evolution to encephalopathy and status epilepticus, with poor response to conventional treatment.

Clinical observation

A 45-year-old woman, born in Colombia, with active hepatitis B virus (VHB) under treatment, SLE diagnosed at 15 years of age with articular, hematological (thrombocytopenia, severe hemolytic anemia with multiple admissions in which she required polytransfusions, corticosteroids, immunoglobulins, rituximab, splenectomy), renal (lupus nephritis treated with cyclophosphamide for years, without reports available), immunological (ANA 1/160, anti-DNA, anti-chromatin, anti-Ro60 and hypocomplementemia) and secondary antiphospholipid syndrome (ischemic ictus, intraventricular thrombus, in treatment with warfarin, but despite this, at the age of 45 years she presented a second ischemic ictus of embolic mechanism secondary to the intracavitary thrombus).

In December 2016, the patient presented an abscessed right nasogenian cellulitis treated with antibiotherapy and surgical excision. Two weeks later, she went to the emergency service due to progressive blurred vision in the left eye, gait instability and alteration in the pitch of the voice. Hypophonia, bilateral ptosis with "frozen gaze" in the horizontal and vertical planes, left dysmetria and ataxic gait stood out in the exploration, with the rest of general, cardiopulmonary and abdominal examination normal. The laboratory analyses showed no cytopenias and the antinuclear antibodies (ANA), antibodies against extractable nuclear antigens (anti-ENA), anti-DNA antibodies and complement were normal. The cranial CT-scan did not show acute alterations. Given the suspicion of myastheniform syndrome vs. neurolupus, the patient was admitted to the hospital and treatment with methylprednisolone 40 mg/day and pyridostigmine 30 mg/12 h was started.

However, the clinical picture evolved into flaccid tetraparesis and facial diplegia, which led us to suspect a Miller-Fisher syndrome (MFS) and 2 g/kg of immunoglobulins divided into 5 days were added to treatment. A lumbar puncture was performed with normal cytobiochemistry, negative cultures and anti-ganglioside antibodies (anti-GQ1b), as well as a brain MRI with hyperintensity in the cortex of the right hemisphere, nonspecific. The patient had a poor evolution with dysphagia and acute respiratory failure, requiring admission to the intensive care unit (ICU) with endotracheal intubation. Five plasmapheresis sessions were carried out, also without response, and she even developed a non-convulsive status epilepticus, with no response to hypnotic and antiepileptic drugs. The lumbar puncture and MRI were repeated without changes. The electromyogram (EMG) showed signs of axonal and demyelinating sensorimotor polyneuropathy, so it was decided treatment with 3 boluses of methylprednisolone 1 g, without response. Given the persistence of the refractory status epilepticus for more than 10 days and the poor prognosis, it was decided to limit the therapeutic effort, and finally the patient died. Autopsy was not performed.

Discussion

The neurological affectation of SLE can have 19 forms of presentation according to the ACR criteria.[1]

Up to 10–80% of patients with SLE may have neuropsychiatric manifestations.[2,3] The affectation may be primary (by neuropsychiatric involvement due to the lupus)[1] or secondary (due to complications of the disease or to the treatments).

One of the neurological manifestations of SLE is the demyelinating inflammatory polyneuropathy. The initial diagnostic suspicion in this patient was Miller Fisher syndrome (MSF), the most frequent variant of Guillain-Barré syndrome (GBS), which typically occurs with ophthalmoplegia, ataxia, and arreflexia, and 25% of patients develop limb weakness. In 85–90% of cases, anti-GQ1b are positive and are associated with involvement of the oculomotor nerves and muscles.[4] The EMG usually shows reduction/absence of sensitive responses with slowing down of the conduction velocity.[5] In most of the cases reviewed, it appears associated with other manifestations of SLE,[6–11] with analytical data of lupus activity and with significant improvement or remission in the majority of cases after the instauration of immunosuppressive medication (high doses of corticosteroids, hydroxychloroquine, intravenous immunoglobulins,[6,7,9–13] plasmapheresis[7,10,11] or intravenous cyclophosphamide.)[6,8–11] Despite the absence of response to treatment and the negativity of anti-GQ1b, this diagnosis was suspected based on the rapid and symmetric progressive areflexic weakness, with involvement of oculomotor pairs and facial diplegia and compatible EMG.

Given the absence of response and the onset of a refractory status epilepticus, other diagnoses were proposed: Bickerstaff encephalitis (it presents the findings of MFS with encephalopathy and hyperreflexia, but usually presents

positive anti-GQ1b and can respond to immunoglobulins and plasmapheresis)[14] or autoimmune or paraneoplastic encephalitis (it is more common that they start with refractory status, but they usually have a subacute onset, with seizures or psychiatric symptoms, pleocytosis in cerebrospinal fluid, MRI and EEG alterations with epileptic activity or slowing down.)[15]

Other less likely options that were proposed were: infectious flaccid encephalomyelitis (unlikely in the absence of data of infection), Wernicke's encephalopathy (unlikely, as it presents nystagmus and the patient received treatment with thiamine without response) or myasthenia gravis (the EMG was not compatible and the patient did not respond to pyridostigmine.)

The final diagnosis accepted was Bickerstaff's encephalitis, given the patient's clinical picture and complementary tests, with autoimmune encephalopathies being the second diagnostic possibility. However, this case has the limitation that the definitive diagnosis could not be reached because the patient died and autopsy was not performed.

Conclusion

We present a rare case of severe SLE with severe neurological involvement, rapidly progressive toward encephalopathy and status epilepticus refractory to conventional treatment.

Conflict of interests

The authors declare that they do not have any conflict of interest.

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