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Case report

Limbic encephalitis with phenotypic NMDA receptor antibodies in patients with de novo diagnosis of Systemic Lupus Erythematosus. Case report☆

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ABSTRACT

Malignancy-associated limbic encephalitis was first described in 1968. Since then, cases have been reported in association with the Herpes Simplex virus, Hashimoto's encephalopathy, lupus, Sjögren's syndrome and paraneoplasias. A syndrome with prominent psychiatric symptoms was described in 2005: consisting of memory loss, reduced level of consciousness and central hypoventilation in four young women with ovarian teratoma and antibodies against an antigen highly expressed in the hippocampus.

Shortly afterwards, these patients were found to have autoantibodies against NMDA receptor NR1 (GluN1) subunit. This discovery has been of the greatest importance in clinical practice since it identifies a devastating, life-threatening neurological disorder that is treatable. In 2007, it was recognised as a nosologic entity and today it is the most common cause of autoimmune encephalitis after disseminated subacute encephalomyelitis.

Case report presentation of a patient exhibiting almost all the clinical symptoms described in the syndrome characterised by NMDA receptor-associated encephalitis. Moreover, a neuroradiological correlation was found, with involvement of limbic structures seen on brain magnetic resonance imaging.

Reasonable exclusion was made of the presence of neoplasia and neuroinfection, and clinical and immunological criteria of systemic lupus erythemathosus were found, helping with the categorisation of lupus-associated limbic encephalitis.

Mortality due to limbic encephalitis is 25%, and 75% of patients may have permanent sequelae.

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Given adequate response to immunosuppressive therapy, early and correct recognition are critically important.

Encefalitis límbica con fenotipo de encefalitis por anticuerpos contra receptores NMDA en paciente con diagnóstico de novo de Lupus eritematoso sistémico. Reporte de caso

Palabras clave:
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RESUMEN

La encefalitis límbica asociada con malignidad fue descrita por primera vez en 1968. Posteriormente se han reportado casos en asociación con virus del Herpes Simplex, encefalopatía de Hashimoto, Lupus, Sjögren y paraneoplasias.

En el año 2005 fue descrito un síndrome con prominentes síntomas psiquiátricos: pérdida de memoria, disminución del nivel de conciencia e hipoventilación central en cuatro mujeres jóvenes con teratoma de ovario y anticuerpos contra un antígeno altamente expresado en el hipocampo.

Poco después se determinó que estos pacientes tenían auto anticuerpos dirigidos contra la subunidad NR1 (GluN1) del Receptor NMDA. Este descubrimiento ha sido de gran importancia en la práctica clínica puesto que identifica un trastorno neurológico devastador, potencialmente fatal y tratable.

En el 2007 se reconoció como entidad nosológica y hoy es la causa más común de encefalitis autoinmune después de la encefalomielitis diseminada subaguda.

Se reporta un caso que agrupa casi la totalidad de los síntomas clínicos descritos en el síndrome que caracteriza la encefalitis por receptores NMDA, se encontró además correlación neuroradiológica por compromiso de estructuras del sistema límbico en la resonancia cerebral.

Se excluye de manera razonable la presencia de neoplasia y neuroinfección y se encuentran criterios clínicos e inmunológicos de Lupus eritematoso sistémico que permiten catalogarlo como una encefalitis Limbica asociada a LES.

La encefalitis límbica tiene una mortalidad del 25% y un 75% de pacientes pueden tener secuelas permanentes.

Como hay adecuada respuesta a la terapia inmunosupresora, su temprano y acertado reconocimiento es de suma importancia.

Introduction

Acute encephalitis is a devastating neurological disease that presents in the form of rapidly progressing encephalitis (usually over a period of less than 6 weeks) caused by inflammation. Incidence varies from 5 to 10 cases per 100,000 inhabitants per year.

Malignancy-associated limbic encephalitis was first described in 1968.[1] Since then, cases have been reported in association with the herpes simplex virus, Hashimoto's encephalopathy, lupus, Sjögren's, and as part of the spectrum of paraneoplastic syndromes.[2]

The etiological diagnosis is critical for adequate treatment. There are several types of neoplasms that produce antibodies against different antigens in neuronal synapses.[3] Moreover, other autoimmune diseases that may have an activity against similar terminal neurological structures must be considered.

Given that confirmation tests usually take time and may be difficult to obtain, a sequential diagnostic approach is required, ruling out the main differential diagnoses and achieving a level of evidence of possible, probable or definitive autoimmune encephalitis, in order to initiate immunotherapy in an attempt at reverting the disease and limit sequelae.[4]

Case report

A 27-year-old black woman, IT engineer, with no relevant background history.

The patient visited primary care three times, complaining of flu-like symptoms: rhinorrea, general malaise and subjective fever. Symptomatic treatment was indicated at the first visit. The second time, a complaint of apnea prompted workup consisting of chest X-ray that was normal and IgM for Mycoplasma pneumoniae which was positive.