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C0259 - DEEP BRAIN STIMULATION IN A CASE OF RASMUSSEN'S ENCEPHALITIS-RELATED DYSTONIA

J. Rumià Arboix¹, L. Reyes², P. Roldán², F. Valldeoriola³, M.J. Martí³ and M. Carreño⁴

¹Servicio de Neurocirugía, Universidad de Barcelona; ²Servicio de Neurocirugía; ³Unidad de Parkinson y Trastornos del Movimiento; ⁴Unidad de Epilepsia, Hospital Clínic de Barcelona, Barcelona, Spain.

Resumen

Objectives: Rasmussen's encephalitis (RE) is a rare neuroinflammatory disease, characterized by frequent and severe seizures, loss of hemispheric functions, and cognitive deterioration. RE affects a single cerebral hemisphere and generally occurs in children under the age of 15. Immunomodulatory treatments may improve temporarily seizures and prevent further tissue loss. However if seizures persist, surgery (hemispherotomy with disconnection of the involved hemisphere) is often indicated. Here we report an unusual case of adult RE in which the seizures have been relatively well controlled with medication, but later in the disease the patient developed an extremely disabling leg and foot dystonia which has been resistant to multiple medical treatments.

Methods: The patient is a 39 y.o. lady without relevant risk factors for epilepsy. Focal seizures in her left hemibody started at the age of 22. She was treated with different antiepileptic drugs without achieving seizure control. Initial Cranial MR was normal. At age 33 consulted for pain and abnormal posturing (flexion and inversion of the left leg with knee flexion) of the left leg, very evident during deambulation. Sensation was normal and there was no motor deficit. All diagnostic workup for metabolic, genetic and autoimmune movement disorders were negative. A second MRI showed atrophy, and a PET scan showed hypometabolism in the right hemisphere. A tentative diagnosis of adult Rasmussen encephalitis was done, confirmed by brain biopsy.

Results: During the following years seizures were mostly controlled by medical treatment. However, the dystonic posturing of the left leg impaired seriously impairing patient's gait, and did not respond to medical treatments, except for mild and transient improvement with botulin toxin. Therefore, surgical treatment by deep brain stimulation of the right globus pallidus internus (GPi-DBS) controlled the dystonia in few hours to date, allowing normal gait function.

Conclusions: Movement disorders related to RE may benefit from DBS as an effective symptomatic treatment, and therefore should be considered.