Long-term management of anti-n-methyl-d-aspartate receptor (NMDAR) encephalitis in young children—still a matter of debate

Sandra Moreira¹, P. Pires², and C. Garrido²

¹Neurology, CHEDV, Portugal
²Neuropeditrics, Hospital de Santo António, Portugal

Abstract

Anti-NMDAR encephalitis is the best-characterized and most common antibody-mediated encephalitis. With early aggressive immunosuppression, prognosis is usually good, although recurrences have been reported in up to 20-25% of patients, mostly in patients without teratoma. Guidelines for the best medical management are still lacking, especially concerning its duration, the comparative efficacy of individual treatments and the role of corticoid-sparing agents. It is also unclear if tumors should be sought after an initial negative screening in males and females younger than 18.

We report the case of a 30-month boy with previous speech delay, who presented with insidious onset of irritability, asymmetric dystonia and chorea, sleep disturbance and consciousness fluctuations. Infections and metabolic disturbances were excluded. NMDAR antibodies were identified in serum and CSF. MRI showed right insular and frontal cortex T2-hyperintensity. Tumor screening was negative. He was initially treated with metilprednisolone pulses and IVIG and then kept on monthly IVIG and prednisolone 1mg/Kg/day, followed by slowly tapering after 2 months of sustained clinical improvement. Follow-up MRI disclosed some brain atrophy and the patient remains with a significant speech delay after 5 months. Despite the good response to first-line treatments, as in this case, corticoid side effects in children may be severe and irreversible. On the other hand, quick withdrawal may compromise recovery and increase relapse probability, especially in cases without associated tumor. This case is illustrative of the difficulties faced by clinicians in the long-term management of NMDAR-encephalitis, namely in respect to the need of corticoid-sparing agents and tumor screening repetition.