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A Rasmussen Encephalitis in an Adolescent Girl

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Abstract

INTRODUCTION: A 12-year-old female presented to clinic with various features of cognitive decline, including headaches, worsening academic performance, and decline in writing abilities and social skills. On neurological examination, she was found to have intrinsic muscle weakness in the right hand, reduced grip and wrist extension of the right arm, pronator drift of the right arm, and left hemiglossal atrophy, along with lessened spontaneous speech abilities. She had no dysarthria, sensory loss, or reduction of tone. The differential diagnoses considered for this patient were chronic viral encephalitis and autoimmune encephalitis. Initial tests included serology (Full Blood Count and IgG Antibody) and cerebrospinal fluid (CSF) analysis, both of which were unremarkable. Angiography results were also normal. Magnetic Resonance Imaging (MRI) depicted left cerebral hemisphere atrophy and T2 hyperintensity in the frontal lobe. Electrophoresis and flow cytometry were not indicative of oligoclonal bands or NMDA-receptor antibodies. Most interestingly, routine EEG did not demonstrate epileptic activity (the hallmark sign of RE), but right-sided amplitude predominance indicated left hemispheric dysfunction. Surgical intervention involved a biopsy of the anterior middle frontal gyrus. The biopsy depicted CD45+ T-cell lymphocyte infiltration, microglial nodules, cortical atrophy, neuronal loss, and cytomegalic dysplastic neurons, which altogether are indicative of Rasmussen Encephalitis. The patient was started on immunosuppressive therapy, including tacrolimus, methylprednisolone, and Intravenous Immunoglobulin (IVIG). 6-month follow-up demonstrated interval progression of atrophy, and 15-month follow-up demonstrated improvement in right extremity strength. Immunosuppression was continued, and the patient continues to be seizure- free with no further worsening of cognitive abilities.

KEYWORDS: Paediatric Rasmussen encephalitis, immunosuppressive therapy.

