Efficacy of Rituximab Treatment in Rasmussen's Encephalitis

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INTRODUCTION

Rasmussen's encephalitis (RE) is a progressive disorder characterized by drug-resistant focal epilepsy, progressive hemiplegia, cognitive impairment and unilateral cerebral atrophy. T-cell cytotoxicity and microglia-induced degeneration mainly play role on its pathophysiology. Consequently, immunomodulatory treatments (intravenous methylprednisolone, IV immunoglobulin, plasma exchange); T cell-inactivating drugs (tacrolimus, azathioprine, cyclophosphamide) and surgical treatment are traditionally recommended. Immunotherapy is likely to be considered as the way to stabilize prognosis of disease and could protect patients from harmful and destructive hemispherectomy. Recently, it has been reported that rituximab, monoclonal antibody targeted against the pan-B-cell marker CD20, has been used and found to be effective. Previous studies revealed that applying rituximab within the initial year of disease onset may result in better control of seizures and greater stability in disease progression.

METHODS

The effectiveness of rituximab treatment on seizure frequency of a patient with RE was evaluated.

CASE

A 14-year-old female presented with focal right lateralized seizures and diagnosed as RE at the age of 6 years. MRI revealed left hemisphere and nucleus caudatus atrophy. EEG showed diffuse background slowing, more prominent on the left. Seizures were partially controlled with combined therapy of intravenous immunoglobuline (IVIG) and corticosteroids, and varying antiepileptic drugs. During follow-up, the effectiveness of immunologic treatment decreased over the years and seizures became more frequently, up to 3-4 times in a day.

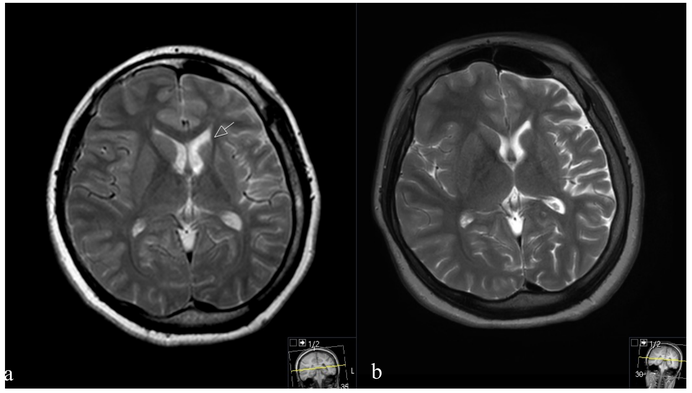

Figure 1: Axial fluid attenuated inversion recovery (FLAIR) images showing left hemisfer and ipsilateral caudat nucleus atrophy (arrow) a: before rituximab treatment, b: after rituximab treatment, no significant progression of atrophy at 2 years follow up.

Rituximab was prescribed instead of IVIG and corticosteroids. Weekly IV infusions of rituximab (375 mg/m^2) applied for 4 weeks. In a succeeding year, rituximab infusion repeated when CD19+ B cells were found to be within the normal range. Meanwhile, antiepileptic treatment (levetiracetam, clonozepam and phenitoin) continued. After administration of rituximab, there was no progression of brain atrophy on MRI (Figure 1 a,b), the frequency of seizures partially decreased, no hemiparesis developed and no cognitive performance deterioration observed.

CONCLUSIONS

Immunologic treatments are frequently used to control disease activity in RE, but the optimal treatment modality is not known. We suggest that effective treatments on B cells, such as rituximab may be an effective choice for immunological therapy.