

Poster presentation

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Severe herpes simplex virus encephalitis in a pediatric patient – the role of immunological mechanisms in diagnosis and treatment

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Introduction

Herpes simplex encephalitis (HSE) is the most common sporadic viral encephalitis in children in Western countries. Only early administration of antiviral drugs can improve the mortality rate, which goes up to 70% without treatment. In different pediatric studies on children with HSE treated by acyclovir about 50% of patients recovered fully, whereas about 40% stayed with moderate to severe neurological impairment. Recent studies reported that specific genetic immunodeficiencies lead to increased HSE susceptibility and that secondary immune-mediated processes can modify disease evolution. Some of these immunodeficiencies such as defects in Toll-like receptor signalling can be detected by cheap and fast tests.

Case report

A 2 years old boy with uneventful medical history and psychomotor development presented with prolonged clonic seizures, followed by gradually loss of consciousness. Immediately acyclovir treatment was started in spite of negative HSV PCR in cerebrospinal fluid (CSF) analysis. While the boy further clinically deteriorated and cMRI revealed diffuse hyperintensity in the right hemisphere, a repeat analysis of HSV PCR in CSF was initiated and gave a positive result. Antiviral treatment was continued for a total of 24 days. The boy was discharged on day 25 with still severe impaired consciousness and neurological sequelae. He was readmitted on day 30 with newly

occurred dysphagia. HSV PCR in CSF was negative and cMRI showed no significant disease progression. An immunomodulatory treatment with corticosteroids (methylprednisolone) was given in combination with acyclovir for another 14 days to prevent an early relapse. Standard blood tests and medical history revealed no immunodeficiency. However, specific stimulation of the boy's peripheral blood monocytes showed impaired antiviral interferon-production indicating a specific immunodeficiency, which predisposes him for severe HSE. Therefore, prophylactic treatment with acyclovir orally was started. Molecular studies to identify a possible genetic defect in the Toll-like receptor pathway are ongoing.

Conclusion

We report on a boy with increased immunological susceptibility to life-threatening HSV infections. In the diagnostic and therapeutic work-up of patients with secondary disease progression one should consider HSE relapse as well as postinfectious immune-mediated processes.