

Poster presentation

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Complement factor B deficiency associated with recurrent aseptic meningitis

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Background

We report complement factor B deficiency in a 16-y-old Caucasian girl, who presented with recurrent episodes of aseptic meningitis. She presented with a 2-week history of headache, vomiting, neck stiffness, facial palsy, equilibrium problems, diplopia, and low grade temperature. 4 months prior to admission she presented with a leucocytoclastic vasculitis. MRI of the brain revealed globally abnormal signal intensity in the cerebral cortex of cerebrum and cerebellum. CSF WCC was 81/ μ L, without rise in glucose and proteins. The patient was successfully treated with high dose glucocorticoids, but tapering of dose was followed by recurrence of the symptoms. Clinical remission was achieved with plasmapheresis, corticosteroids and mofetil mycophenolate.

Methods

A moderately low C3 level (57 mg/dl) together with the clinical picture suggested a deficiency affecting regulation of complement activation

Results

Analysis of haemolytic activity revealed absence of alternative pathway activity with very low level of factor B (1 mg/dl) together with a low level of mannose binding protein (199 ng/ml). Levels of factor H (46 mg/dl) and I (5.5 mg/dl) were normal.

Conclusion

Complement factor B plays an important role in the alternative complement activation pathway by complexing with C3b to create the active C3 convertase. Patients with abnormal levels of mannose-binding protein are known to have recurrent significant infections in the absence of abnormalities in the four major arms of the immune system. To our knowledge this is the first case of combined mannose lectin pathway and factor B deficiency.