



Abstracts

Articles appearing in the May 2020 issue

Complement factor I deficiency: A potentially treatable cause of fulminant cerebral inflammation

Objective To raise awareness of complement factor I (CFI) deficiency as a potentially treatable cause of severe cerebral inflammation.

Methods Case report with neuroradiology, neuropathology, and functional data describing the mutation with review of literature.

Results We present a case of acute, fulminant, destructive cerebral edema in a previously well 11-year-old patient, demonstrating massive activation of complement pathways on neuropathology and compound heterozygote status for 2 pathogenic mutations in CFI that result in normal levels but completely abrogate function.

Conclusions Our case adds to a very small number of extant reports of this phenomenon associated with a spectrum of inflammatory histopathologies including hemorrhagic leukoencephalopathy and clinical presentations, resembling severe acute disseminated encephalomyelitis. CFI deficiency can result in uncontrolled activation of the complement pathways in the brain, resulting in devastating cerebral inflammation. The deficit is latent, but the catastrophic dysregulation of the complement system may be the result of a C3 acute phase response. Diagnoses, to date, have been retrospective. Diagnosis requires a high index of suspicion and clinician awareness of the limitations of first-line clinical tests of complement activity and activation. Simple measurement of circulating CFI levels, as here, may fail to diagnose functional deficiency with absent CFI activity. These diagnostic challenges may mean that the CFI deficiency is being systematically under-recognized as a cause of fulminant cerebral inflammation. Complement inhibitory therapies (such as eculizumab) offer new potential treatment, underlining the importance of prompt recognition, and real-time whole exome sequencing may play an important future role.

[NPub.org/N2/9524a](https://pubmed.ncbi.nlm.nih.gov/3249524/)

Age-related visual dynamics in HIV-infected adults with cognitive impairment

Objective To investigate whether aging differentially affects neural activity serving visuospatial processing in a large functional neuroimaging study of HIV-infected participants and to determine whether such aging effects are attributable to differences in the duration of HIV infection.

Methods A total of 170 participants, including 93 uninfected controls and 77 HIV-infected participants, underwent neuropsychological assessment, followed by neuroimaging with magnetoencephalography (MEG). Time-frequency analysis of the MEG data, followed by advanced image reconstruction of neural oscillatory activity and whole-brain statistical analyses were used to examine interactions between age, HIV infection, and cognitive status. Post hoc testing for a mediation effect of HIV infection duration on the relationship between age and neural activity was performed using a quasi-Bayesian approximation for significance testing.

Results Cognitively impaired HIV-infected participants were distinguished from unimpaired HIV-infected and control participants by their unique association between age and gamma oscillations in the parietooccipital cortex. This relationship between age and gamma was fully mediated by the duration of HIV infection in cognitively impaired participants. Impaired HIV-infected participants were also distinguished by their atypical relationship between alpha oscillations and age in the superior parietal cortex.

Conclusions Impaired HIV-infected participants exhibited markedly different relationships between age and neural responses in the parietooccipital cortices relative to their peers. This suggests a differential effect of chronological aging on the neural bases of visuospatial processing in a cognitively impaired subset of HIV-infected adults. Some of these relationships were fully accounted for by differences in HIV infection duration, whereas others were more readily associated with aging.

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