Responses to treatment, outcomes of autoimmune cerebellar ataxia

28 September 2015

While autoimmune cerebellar ataxia (a loss of muscle coordination) can lead to severe disability with some patients becoming wheelchair-bound, there are factors that may help to predict better immunotherapy response and neurological outcomes, according to an article published online by *JAMA Neurology*.

Autoimmune cerebellar ataxia in adults, which usually comes on rapidly and progresses quickly, can be divided into disorders that are paraneoplastic (triggered by cancer in the body) or nonparaneoplastic (non-cancer-related autoimmune disorders of the central nervous system). The disabling neurological effects can include speech that is not clearly articulated, gait and balance disorders, and loss of muscle control in the limbs. Little has been published regarding treatment responses and neurologic outcomes among patients with autoimmune cerebellar ataxia. However, at least 17 autoantibodies have been reported as causally linked to autoimmune cerebellar ataxia.

Andrew McKeon, M.D., of the Mayo Clinic, Rochester, Minn., and coauthors reviewed medical records at the Mayo Clinic to examine treatment responses and outcomes in 118 adults with autoimmune cerebellar ataxia who were seropositive for at least one neural autoantibody, had received at least one immunotherapy or cancer therapy, and had neurologist-reported outcomes from 1989 through 2013.

Results indicate the median (midpoint) age at onset of neurologic symptoms was 58, nearly three-quarters of the patients were women, and the median duration from symptom onset to last follow-up was 25 months. Among the patients, 63 had paraneoplastic ataxic disorders and 55 patients had nonparaneoplastic ataxic disorders. Also, 81 patients were seropositive for NNC [neuronal nuclear and/or cytoplasmic] antibodies; 22 patients for neural PMP [plasma membrane protein] receptor or ion channel antibodies; and 15 patients for antibodies from both categories.

Overall, 54 patients (45.8 percent) had physician-reported neurologic improvement with immunotherapy (n=51) or cancer therapy (n=3), according to the study results. Analyses suggest factors that may predict better immunotherapy response and neurologic outcomes include a nonparaneoplastic disorder, the detection of at least one or more PMP antibodies or the detection of GAD 65 [glutamic acid decarboxylase 65-kDa isoform] antibodies.

Regardless of the response to immunotherapy, the final ambulatory outcomes regarding the use of gait aids to get around were 56 patients used a wheelchair, 26 patients had walkers and seven patients used canes. Among the remaining 29 patients, 25 of them required no gait aid but had an abnormal gait and four patients had normal gait, the study reports.

"Although autoimmune ataxia is usually severe, treatment responses can be gratifying, particularly in patients with nonparaneoplastic disorders and in those harboring autoantibodies directed against GAD65 or neural PMPs," the study concludes.

More information: *JAMA Neurol*. Published online September 28, 2015. DOI: 10.1001/jamaneurol.2015.2378