Early treatment improves outcomes in rare, often undiagnosed form of encephalitis

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A mysterious, difficult-to-diagnose, and potentially deadly disease that was only recently discovered can be controlled most effectively if treatment is started within the first month that symptoms occur, according to a new report by researchers from the Perelman School of Medicine at the University of Pennsylvania.

The researchers analyzed 565 cases of this recently discovered paraneoplastic condition, called Anti-NMDA Receptor Encephalitis, and determined that if initial treatments fail, second-line therapy significantly improves outcomes compared with repeating treatments or no additional treatments (76 percent versus 55 percent). The research is being presented at the American Academy of Neurology's 64th Annual Meeting in New Orleans.

The condition occurs most frequently in women (81 percent of cases), and predominately in younger people (36 percent of cases occurring in people under 18 years of age, the average age is 19). Symptoms range from psychiatric symptoms, memory issues, speech disorders, seizures, involuntary movements, to decreased levels of consciousness and breathing. Within the first month, movement disorders were more frequent in children, while memory problems and decreased breathing predominated in adults.

"Our study establishes the first treatment guidelines for NMDA-receptor encephalitis, based on data from a large group of patients, experience using different types of treatment, and extensive long-term follow-up,"

said lead author Maarten Titulaer, MD, PhD, clinical research fellow in Neuro-oncology and Immunology in the Perelman School of Medicine at the University of Pennsylvania. "In addition, the study provides an important update on the spectrum of symptoms, frequency of tumor association, and the need of prolonged rehabilitation in which multidisciplinary teams including neurologists, pediatricians, psychiatrists, behavioral rehabilitation, and others, should be involved."

The disease was first characterized by Penn's Josep Dalmau, MD, PhD, adjunct professor of Neurology, and David R. Lynch, MD, PhD, associate professor of Neurology and Pediatrics, in Annals of Neurology in 2007. One year later, the same investigators in collaboration with Rita Balice-Gordon, PhD, professor of Neuroscience, characterized the main syndrome and provided preliminary evidence that the antibodies have a pathogenic effect on the NR1 subunit of the NMDA receptor in the Lancet Neurology in December 2008. The disease can be diagnosed using a test developed at the University of Pennsylvania and currently available worldwide. With appropriate treatment, almost 80 percent of patients improve well and, with a recovery process that may take many months and years, can fully recover.

In earlier reports, 59 percent of patients had tumors, most commonly ovarian teratoma, but in the latest update, 54 percent of women over 12 years had tumors, and only six percent of girls under 12 years old had ovarian teratomas. In addition, relapses were noted in 13 percent of patients, 78 percent of the relapses occurred in patients without teratomas.

As Anti-NMDA Receptor Encephalitis, the most common and best characterized antibody-mediated encephalitis, becomes better understood, quicker diagnosis and early treatment can improve outcomes for this severe disease.

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