Autoimmune Encephalitis (AE)

AE is a severe medical condition in which the body’s immune system creates auto-antibodies that mistakenly attacks healthy brain cells. The auto-antibodies attack by targeting distinct neural receptors and binding to the healthy brain cell on the outside of the cell, and as such are considered extracellular. This accidental assault causes health brain cells to become destroyed leading to inflammation or so much damage that the cells no longer function properly resulting in impaired neural function.

**What are the symptoms?**

AE can produce a wide variety of neurologic and psychiatric symptoms. Neurologic symptoms can include impaired memory and cognition, abnormal movements, seizures, or issues with balance, speech, or vision. Psychiatric symptoms may include psychosis, aggression, inappropriate sexual behaviors, panic attacks, compulsive behaviors, euphoria or fear. Neurologic and psychiatric symptoms can vary drastically in frequency, and elongated progression can even result in a patient to lose consciousness or go into a coma.

**Prevalence?**

AE can occur in all age groups, although some types are more prevalent in younger individuals. AE does actually occur at a higher frequency in women than in men, and the condition can develop with or without the presence of a tumor. According to Dubley’s article on *Autoimmune encephalitis epidemiology and a comparison to infectious encephalitis*, it is estimated that approximately 1 million people worldwide had autoimmune encephalitis in their lifetime. Likewise, it was estimated that, currently, about 90,000 people around the world develop autoimmune encephalitis each year.

**Diagnosis?**

Many with AE are not immediately diagnosed, as AE is considered such a rare disease. For an individual to be diagnosed with AE they must have subacuate onset of working memory deficits/altered mental status/psychiatric symptoms, new focal CNS findings/unexplained seizures/CSF pleocytosis/MRI features suggesting encephalitis, and reasonable exclusion of alternative causes.

There is a lot of difficulty in diagnosing AE. According to Graus and his colleagues, “existing criteria for autoimmune encephalitis are too reliant on antibody testing and response to immunotherapy, which might delay the diagnosis. Likewise, some of the difficulty in accurately diagnosing AE is that many of these symptoms may appear at different times and appear at different degrees. Often this disease is mistaken for similar common disorders.

**Treatment and Management?**

Luckily, AE is treatable. The most positive outcomes for treating AE often come from early and aggressive treatment techniques. According to Eric Lancaster, “most forms of autoimmune encephalitis respond to immune therapies, although powerful immune suppression for weeks or months may be needed in difficult cases.” In some instances, these types of treatment have even assisted in preventing the further progression of the condition.

There are two lines of treatment options available for combating AE: “first line” and “second line”.  A “first line” treatment option is often prescribed to a patient immediately following diagnosis. Some of the commonly utilized “first line” treatments are removal of a teratoma, prescription of anti-inflammatories, prescription of plasmapheresis, and intravenous immunoglobulin treatments. When “first line” treatment options fail to work “second line” options are used. “Second line” treatments include immune system suppressants such as Rituximab, CellCept, and Cytoxan.

**References:**

1. Dubey et al., (2018). Autoimmune encephalitis epidemiology and a comparison to infectious encephalitis. *Annals of Neurology*, 83(1), 166-177. http://doi.org/10.1002/ana.25131
2. Lancaster, E., (2016). The Diagnosis and Treatment of Autoimmune Encephalitis. *Journal of Clinical Neurology (Seoul, Korea)*, *12*(1), 1–13. [http://doi.org/10.3988/jcn.2016.12.1.1Links to an external site.](http://doi.org/10.3988/jcn.2016.12.1.1)

<https://synapse.koreamed.org/Synapse/Data/PDFData/0145JCN/jcn-12-1.pdfLinks to an external site.>

[Links to an external site.](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4712273/pdf/jcn-12-1.pdf)

3. Graus et al., (2016). A clinical approach to diagnosis of autoimmune encephalitis. Lancet Neurol, 15(4), 391-404. [http://doi.org/10.1016/S1474-4422(15)00401-9Links to an external site.](http://doi.org/10.1016/S1474-4422(15)00401-9)

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