Autoimmune encephalitis;Updates

Hamidreza Ghalyanchi Langroudi
Rasht;Ghaem International Hospital

The knowledge of immune dysregulation and autoimmunity in neurological disorders has expanded considerably in recent times. Over the past 10 years, the continual discovery of novel forms of encephalitis associated with antibodies to cell surface or synaptic proteins has changed the paradigms for diagnosing and treating disorders that were previously unknown or mischaracterized.

Autoimmune encephalitis is increasingly recognized as the cause of a variety of neuropsychiatric syndromes that can be severe and prolonged. In contrast to the classic paraneoplastic disorders that are poorly responsive to tumor treatment and immunotherapy, autoimmune encephalitis often responds to these treatments, and patients can have full or marked recoveries. As early treatment speeds recovery, reduces disability, and decreases relapses that can occur in about 20% of cases, it is important that the immune pathogenesis of these disorders is recognized.

We describe the differences between limbic encephalitis associated with antibodies targeting intracellular antigens, and neuronal surface antibody syndromes (NSAS) where the antigens are primarily receptors or synaptic proteins located on the neuronal cell surface. We chronologically highlight important developments in NSAS by focusing on voltage gated potassium channel complex-associated antibody mediated encephalitis, anti-N-methyl-D-aspartate receptor (anti-NMDAR) encephalitis, and anti-dopamine 2 receptor antibody-associated basal ganglia encephalitis and DPPX and IgLON5. The therapeutic challenges that clinicians face such as the timing of therapy and the role of second-line therapy will be discussed.

Keywords : Autoimmune encephalitis; Limbic encephalitis.