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ORIGINAL ARTICLE

Autoimmune encephalitis: the first observational study from Iran

Masoud Etemadifar[1,2] · Ali Aghababaei[2] · Hosein Nouri[2,3] · Parisa K. Kargaran[4] · Shaghayegh Mohammadi[2] · Mehri Salari[5]

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Abstract

Background Even within the most populous countries in the Middle East, such as Iran, autoimmune encephalitis cases have been rarely reported.

Objective We aimed to describe the demographic, clinical, and paraclinical characteristics of Iranian patients with autoimmune encephalitis positive for anti-neuronal autoantibodies.

Methods This cross-sectional study included all patients diagnosed with autoimmune encephalitis and referred to our hospital, in Isfahan, Iran, from March 2016 to May 2020. Patients' demographic, clinical, laboratory, radiological, and electroencephalographic features were obtained from their medical records.

Results We identified a total of 39 (21 females, 53.8%) patients with autoimmune encephalitis (mean age = 34.9 ± 12.8 years). The most commonly detected antibody was anti-NMDAR ($n = 26$, 66.7%), followed by anti-$GABA_BR$ ($n = 8$, 20.5%), anti-Zic4 ($n = 4$, 10.3%), and anti-GAD65 ($n = 1$, 2.6%) antibodies, in descending order of frequency. Two anti-NMDAR-positive patients had a history of systemic lupus erythematosus (SLE), and four had a prior history of herpes simplex encephalitis. Clinical presentations in patients positive for anti-Zic4 antibodies included cognitive decline ($n = 4$, 100%), seizures ($n = 3$, 75%), parkinsonism ($n = 1$, 25%), and stiff-person syndrome ($n = 1$, 25%).

Conclusion This was the first case series of Iranian patients with autoimmune encephalitis with some interesting observations, including SLE-associated anti-NMDAR encephalitis, as well as an unusual concurrence of anti-Zic4 antibody positivity and cognitive problems, seizures, parkinsonism, and stiff-person syndrome.

Keywords Autoimmune encephalitis · Autoantibodies · Systemic lupus erythematosus · Zic4 · Parkinsonism · Stiff-person syndrome

Introduction

Autoimmune encephalitis (AE) is an inflammatory condition in the central nervous system (CNS) directed by a group of heterogeneous anti-neuronal autoantibodies against cell-surface/synaptic proteins or intracellular components [1]. Patients with AE present with a wide range of clinical signs and symptoms, including cognitive and psychiatric disturbances, seizures, and abnormal movements. Diagnosis is based on clinical, laboratory, magnetic resonance imaging (MRI), and electroencephalogram (EEG) findings. [1]. However, as several other disorders can mimic AE, alternative diagnoses need to be excluded [1].

Our knowledge regarding the epidemiological characteristics of AE is very limited. A recent population-based study in the United States reported the incidence and prevalence rates of AE to be 0.8 and 13.7 per 100,000 person-years,

✉ Ali Aghababaei
aaghababaei78@gmail.com

1 Department of Neurosurgery, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran

2 Al-Zahra Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran

3 Network of Immunity in Infection, Malignancy, and Autoimmunity (NIIMA), Universal Scientific Education and Research Network (USERN), Isfahan, Iran

4 Department of Cardiovascular Medicine, Center for Regenerative Medicine, Mayo Clinic, Rochester, MN, USA

5 Functional Neurosurgery Research Center, Shohada Tajrish Neurosurgical Center of Excellence, Shahid Beheshti University of Medical Sciences, Tehran, Iran