Autoimmune Encephalitis: Not Rare and Increasing

Silky Pahlajani . Neurology Alert ; Atlanta Vol. 37, Iss. 8, (Apr 2018).

ABSTRACT (ENGLISH)

SYNOPSIS: Detection of autoimmune encephalitis is increasing over time. According to the results from this population-based study, its prevalence and incidence are comparable to infectious encephalitis.

FULL TEXT

Dr. Pahlajani reports no financial relationships relevant to this field of study.

SOURCE: Dubey D, Pittock SJ, Kelly CR, et al. Autoimmune encephalitis epidemiology and comparison to infectious encephalitis. *Ann Neurol* 2018;83:166-177.

The traditional diagnostic approach to encephalitis, a broad term for brain inflammation, has focused primarily on infectious causes. Recent discovery of various neuronal antibodies and associated forms of autoimmune encephalitis has led to a paradigm shift. Despite increasing recognition and diagnosis of autoimmune encephalitis, studies pertaining to epidemiology are scant.

The authors of this population-based, comparative study compared the prevalence and incidence of autoimmune encephalitis and infectious encephalitis in residents of Olmsted County, MN. The analysis includes patients of all ages, both sexes, and ethnic minorities. Search of medical records from Jan. 1, 1995, to Dec. 31, 2015, selected all patients with encephalitis and other potentially relevant diagnostic codes. Of the 570 patients identified, 57 met inclusion criteria, 28 for autoimmune encephalitis (definite or probable) and 29 for infectious encephalitis. Diagnosis of definite or probable autoimmune encephalitis was determined by the 2016 diagnostic criteria published by Graus et al.[1] The comparison group with infectious encephalitis included meningoencephalitis and progressive multifocal leukoencephalopathy (PML) but required confirmation of an infectious pathogen (bacterial, viral, fungal, or parasitic). Patients with encephalitis were excluded if the final diagnosis was possible autoimmune encephalitis not meeting criteria for definite or probable, presumed infectious encephalitis without a confirmed pathogen, a prion disorder, and encephalitis of unknown etiology or immune-related disorder.

Results of statistical analysis and age- and sex-adjusted calculations revealed a prevalence of 13.7 per 100,000 and incidence of 0.8 for autoimmune encephalitis, comparable to that of infectious encephalitis with a prevalence of 11.6 per 100,000 and incidence of 1.0. Additionally, the authors included two tables that compared rates of occurrence for each subtype. The prevalence of autoimmune encephalitis was almost three times higher in African-Americans than Caucasians (38.3 per 100,000 and 13.7 per 100,000, respectively), and incidence was four times higher in African-Americans than Caucasians (2.8 per 100,000 person-years vs. 0.7 per 100,000 person-years, respectively). No differences were found in ethnic proportionality pertaining to prevalence or incidence of infectious encephalitis. Over two decades, the incidence of autoimmune encephalitis tripled from 0.4 between 1995-2005 to 1.2 between 2006-2015, mainly because of increased detection of neural-specific IgG-associated encephalitis. In comparison, incidence of infectious encephalitis remained unchanged (1.0) during both time

intervals. In addition to increased detection of autoimmune encephalitis, there is also a greater tendency to relapse, therefore increasing disease burden.

COMMENTARY

This is the first population-based study to analyze the prevalence and incidence of autoimmune encephalitis, two of the most fundamental measures in epidemiology. Many clinicians still consider autoimmune encephalitis to be a “diagnosis of exclusion” or “rare” when compared to infectious encephalitis. Results of this study contradict that presumption and serve as an eye-opener. Knowing the epidemiology of autoimmune encephalitis is crucial; it provides context for diagnostic decision-making and allows for appropriate allocation of resources and healthcare planning. A practical example of this is demonstrated by the fact that immunoglobulin (IVIG), commonly used for acute and maintenance treatment of autoimmune encephalitis, is not covered by most insurance companies and is considered experimental. The cost of IVIG is in tens of thousands of dollars. Patients who are unable to afford treatment and therefore are left untreated perpetuate the existing severe disease burden from encephalitis. A few limitations of this study include small sample size, lack of a population-based study for direct comparison, and variations in diagnostic criteria for autoimmune vs. infectious encephalitis. Nonetheless, this study is a great starting point and demonstrates the importance of epidemiological data as well as the need for more studies to assess frequency and occurrence rate of autoimmune encephalitis in other populations.

References

1. Graus F, Titulaer MJ, Balu R, et al. A clinical approach to diagnosis of autoimmune encephalitis. Lancet Neurol 2016;15:391-404.

DETAILS

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