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Autoimmune encephalitis mimicking sporadic Creutzfeldt–Jakob disease: A retrospective study

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ABSTRACT

Autoimmune encephalitis associated with anti-voltage-gated potassium channel antibodies are most likely to be misdiagnosed as sporadic Creutzfeldt–Jakob disease (sCJD). Our goal was to delineate patients who were initially suspected to have CJD but were later found to have AE. We performed a retrospective clinical review of cases of individuals and made a comparison between groups of patients diagnosed with sCJD and AE. Patients who had rapidly progressing dementia and focal neurological impairment, such as aphasia, gait disturbance, visual disturbance, and depression, at onset were diagnosed with sCJD, whereas epilepsy, hyponatremia and dysautonomia were strong hints for AE. Fluoroscope-positron emission tomography (PET) of patients with AE revealed variable metabolism and normative and long-term immunosuppression were less likely to relapse.

1. Introduction

Autoimmune encephalitis (AE) and sporadic Creutzfeldt–Jakob disease (sCJD) share some clinical features, including rapidly progressing dementia, prominent psychiatric symptoms, memory problems, seizures and occasional ribboning hyperintensity on diffusion-weighted magnetic resonance imaging (DW-MRI) (Mackay et al., 2012). Some types of AE have been confused with sCJD, such as Sjögren's syndrome and anti-voltage-gated potassium channel antibodies (VGKC) autoantibody-associated encephalopathy (Matsuo et al., 2013; Geschwind et al., 2008). Hence, it is necessary to identify potentially treatable causes of dementia and render timely and proper treatment.

The sCJD diagnosis is supported by periodic sharp-wave complexes on electroencephalography (EEG), MRI or increased levels of the 14–3-3 protein; however, diagnostic accuracy depends on the disease phase and genotype. The aim of this retrospective study was to compare the clinical and neuroimaging features, and the prognoses between sCJD and AE with the aim of ameliorating the prognosis and reducing the mortality rate.

2. Materials and methods

2.1. Subjects

We reviewed 89 serial subjects (68 with sCJD and 21 with AE) and the patients with sCJD presented from January 1, 1992 to August 1, 2015 and those with AE presented from June 1, 2014 to August 1, 2015 and both groups were followed up at the Department of Neurology of the General Hospital of the People's Liberation Army. Patients diagnosed with sCJD fulfilled the updated 2009 sCJD clinical diagnostic criteria (Zerr et al., 2009). All twenty-one patients were diagnosed as AE because specific antibodies tested in cerebrospinal fluid (CSF) or serum and administration of immunotherapy resulted in rapid or moderate recovery (Rosenfeld et al., 2012). In group of sCJD, eleven of the sixty-eight enrolled patients were pathologically confirmed. Ten patients were confirmed by brain biopsy and one by autopsy. Eight patients were tested for autoimmune antibodies and the results were all negative. We excluded the remaining 49 (72%) patients for the following reasons: lack of detecting autoimmune antibodies; lack of pathological confirmation. Finally, nineteen of the 68 sCJD patients were included. After reviewing clinical data of all

Abbreviations: sCJD, sporadic Creutzfeldt–Jakob disease; PET, Fluoroscope positron emission tomography; VGKC, voltage-gated potassium channel antibodies; CSF, cerebrospinal fluid; DWI, diffusion-weight imaging; EEG, electroencephalography; FLAIR, fluid attenuated inversion recovery; MRI, magnetic resonance imaging.

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AE patients, eight of the 21 AE patients were previously suspected of having sCJD.

2.2. Methods

After the initial clinical evaluation, CSF and serum samples of the eight AE patients with suspected sCJD and another eight sCJD patients who did not get biopsy were presented at the Peking Union Medical College Hospital Neuroimmunology Laboratory to detect neuronal surface antigens, such as leucine-rich glioma inactivated 1 (LGI1), contactin-associated protein-like2 (CASPR2), and N-methyl-D-aspartate and glycine receptors and intracellular antigens using anti-Hu, anti-Yo, anti-Ri, anti-Ma and anti-amphiphysin antibodies. An indirect immunofluorescence antibody assay was performed to detect the antibodies in CSF and serum. The patients' clinical data were obtained by medical record review and a family telephone interview. The flow of our retrospective study is summarised in Fig. 1. Differences in the symptoms between CJD and AE were compared using the X^2 and Fisher's exact tests.

3. Results

3.1. Clinical characteristics and signs

Six of the eight AE patients were men, and the median patient age was 54.1 years. Six patients were diagnosed with anti-LGI1 encephalitis, and two patients were diagnosed with CASPR2 encephalitis. The clinical presentations, EEG and MRI findings, antibody types, treatments and responses are summarised in Table e-1. All patients had rapidly progressive cognitive decline, focal neurological signs with or without cortical hyperintensity and suspicious myoclonus. Therefore, sCJD was suspected in these eight patients at disease onset. The correct AE diagnosis was ascertained later during the disease course. AE was diagnosed in eight patients after detecting

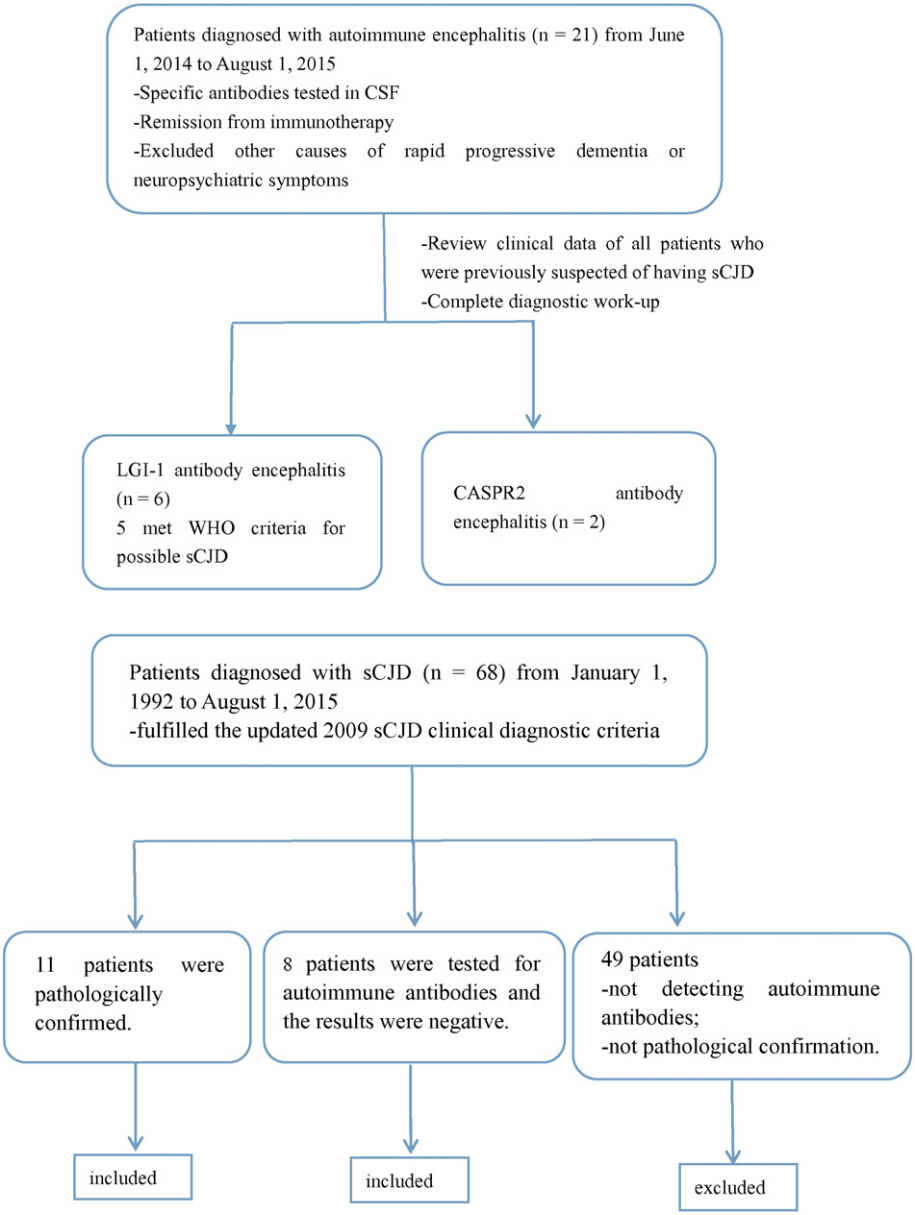

Fig. 1. Study flow chart.