

5th International Conference on
Brain Disorders and Therapeutics
&
Mental Health and Psychology

November 05-06, 2018 | Edinburgh, Scotland



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Central nervous system inflammatory demyelinating diseases with unusual clinical features - Lessons learned from neuropathology


Central nervous system inflammatory demyelinating disease (CIDD) encompasses a broad spectrum of disorders such as multiple sclerosis (MS), acute disseminated encephalomyelitis (ADEM) and neuromyelitis optica (NMO). Two cases of CIDD with unusual clinical features are presented. In both cases, histopathological examination played pivotal roles in the anatomical diagnosis. Case 1 is a 51 year old female who presented with headache, progressive aphasia and hemiparesis without preceding infection or vaccination. Based on MRI and negative oligoclonal bands in the CSF, a clinical diagnosis of ADEM was made. However, brain biopsy of the affected cerebral white matter revealed both pathological features of ADEM and early stage of active MS including perivenular demyelination, confluent plaque-like demyelination, and subpial demyelination. Case 2 is an autopsied 73 year old female who had been diagnosed as NMO at age 63 based on typical clinical/radiological features and positive serum AQP4

antibody. One year before death, she was treated for an acute myocardial infarction, and one month before death she suffered a massive basal ganglia stroke diagnosed with CT. On autopsy, the corresponding basal ganglia revealed large necrotic lesions associated with several pathological signatures of NMO including inflammatory cell infiltration, perivascular complement deposition, and the presence of numerous corpora amylacea phagocytosed by infiltrating macrophages. These cases illustrate the importance of neuropathological examination which might be of interest in considering the pathogenesis of CIDDs.

Speaker Biography

Shinji Ohara is the Vice Director, Matsumoto Medical Center, Matsumoto Japan. He graduated from Tohoku University School of Medicine, Sendai, Japan, and from postdoctoral course (Neuropathology) at Brain Research Institute, Niigata University, Niigata, Japan in 1986. He then completed Neurology Residency Program at Department of Neurology, Washington University School of Medicine, St. Louis, MO, USA. Since 1995, he has served as Director of Department of Neurology at Matsumoto Medical Center, Matsumoto, Japan

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