

Online supplemental material

- Diagnosis, ancillary testing, additional treatment

Diagnosis

Initial diagnoses were viral encephalitis, psychosis, specific epilepsy syndromes, post-infectious encephalitis, and autoimmune encephalitis. In five patients a first onset psychosis was suspected. Therefore, these five patients were initially admitted to a psychiatry department. Six patients were originally sent home after visiting the emergency department or after a short admission, all presenting with a first generalized seizure or status epilepticus.

Ancillary testing

At disease onset, brain MRI was normal in 63% of patients. Abnormal findings were: an increased signal on T2 and/or FLAIR weighted images in mesiotemporal regions (n=4), basal ganglia (n=2), or diffuse in cortical regions (n=2), or post-HSVE abnormalities (n=3)

Except for one patient (4%) with a normal EEG at presentation, all other EEGs were abnormal. Most frequently it showed focal slowing (61%), occasionally combined with typical epileptic patterns (18%). In three patients (11%) the first EEG showed a status epilepticus. Only in patients that required ICU treatment the EEG showed an extreme delta brush pattern at presentation.

CSF analysis was done in all patients. In 61% there was mild pleocytosis. Other abnormalities were increased protein or IgG index, and the presence of oligoclonal bands (OCB).

Using commercial CBA, all available CSF samples were positive (n=27) for NMDAR IgG (NR1) antibodies. In 16/24 serum samples (67%) antibodies were detected. Additionally a specific hippocampal staining pattern was seen in 21/22 serum samples and 27/27 CSF samples.

Additional treatment

Sixty-two percent of patients were treated with anti-epileptic drugs (AEDs). Three of them were treated with > 2 AEDs without seizure freedom during disease course (one with post-HSVE anti-NMDAR encephalitis).