Idiopathic Autoimmune Encephalopathy: MR imaging appearance

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Although idiopathic autoimmune encephalopathy (AE) is a rare and poorly understood entity, it is becoming increasingly recognized by the association with autoantibody markers and/or clear response to immuno-modulatory treatment. The clinical manifestations of AE are protean and nonspecific, and make the diagnosis relatively difficult. Since AE is a potentially reversible neurological condition, better understanding its MR appearance is critical to patient's prognosis. In this study, seven cases of AE without specific biological markers were described and their MRI findings were investigated.

Methods: Seven patients (3 females, 4 males), aged 23 to 42 years (mean 34.33 years), with biopsy-proven autoimmune inflammatory meningo-encephalitis were studied retrospectively. The clinical features are listed in Table 1. The laboratory data and MRI findings were analyzed.

Table 1. Clinical features of the patients with AE

patients	Sex/Age (yrs) at onset	Clinical presentations at onset	Interval between onset and referral	Impression before thorough evaluation and biopsy
1	M/23	Headache, low fever, transient aphasia, later seizure	3 months	Demyelination, lymphoma?
2	M/40	Left facial paralysis, weakness of left hand	3 months	Infectious meningo-encephalitis
3	M/42	Seizure, complex partial attack	2 months	Viral encephalitis
4	F/39	Paroxysmal weakness of left lower extremity	40 days	Metastasis,
5	F/33	Dizziness, headache, low fever, naming difficulty	6 months	Viral encephalitis
6	F/27	Right hand clumsiness, speech difficulty	2 years	Inflammatory demyelination
7	M/30	Paroxysmal numbness of extremities, aphasia, seizure	2 years	Venous thrombosis

Results: CSF analysis showed a moderate elevation in protein (33~89mg/dl) in 6 patients, and mild lymphocytic pleocytosis (2~8white cells per field) in 6 patients. Extensive CSF examinations and serological tests ruled out the possibility of infectious causes (bacteria, viruses, syphilis, fungi or cysticercosis), systemic autoimmune diseases, or paraneoplastic reasons. Besides, anti-cardiolipin antibody, anti-neutrophil cytoplasmic antibody, and anti-thyroperoxidase antibody were all negative in this patients group, no autoimmune thyroiditis was found neither. MRI showed multifocal diffuse subcortical and periventricular abnormalities (n=6), basal ganglia involved in 1 patient. Nodular and/or patchy enhancement was disclosed in all 5 patients with Gd-DTPA injection. Follow-up MRI exams were acquired in 6 patients and revealed marked improvement after immunomodulatory treatment, however, relapse was seen in 3 patients. The pathology reports showed prominent T-cell infiltration in brain parenchyma and/or meninges without evidence of organisms. No neutrophilic granulocyte was found. Perivascular lymphocytes infiltration was noted, but mostly not as the classical angiocentric pattern. No microglial nodule was present. Demyelination was secondary to inflammation.

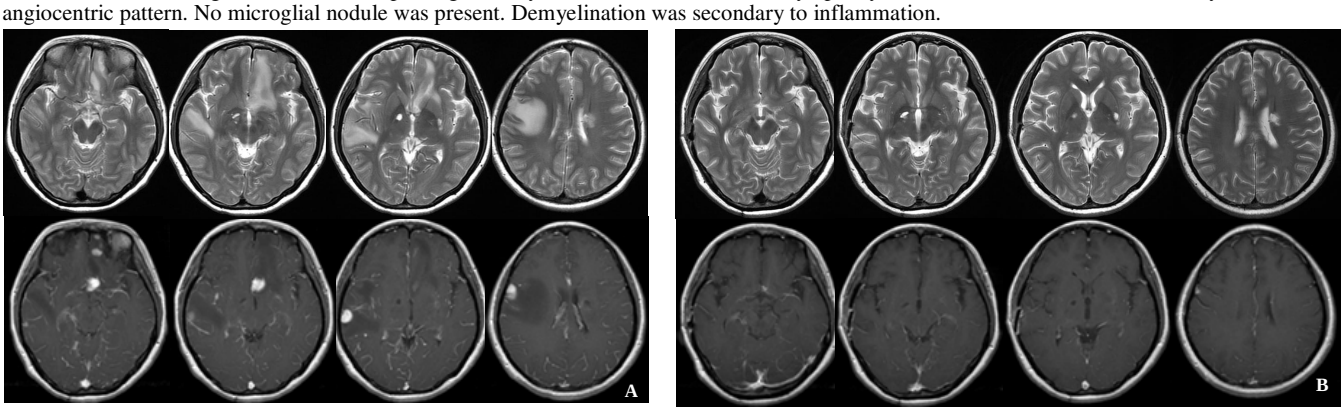


Fig A-B, Patient 4, A: Multiple diffuse T2-prolongation areas (vasogenic edema) in subcortical white matter with multiple enhanced nodules next to meninges were disclosed. B: After steroid+ CTX treatment, 43 days later, the previous enhanced nodules absorbed prominently, and the accompanying T2-prolongation areas also disappeared. Patchy high signals in bilateral basal ganglia showed no changes indicating malacia.

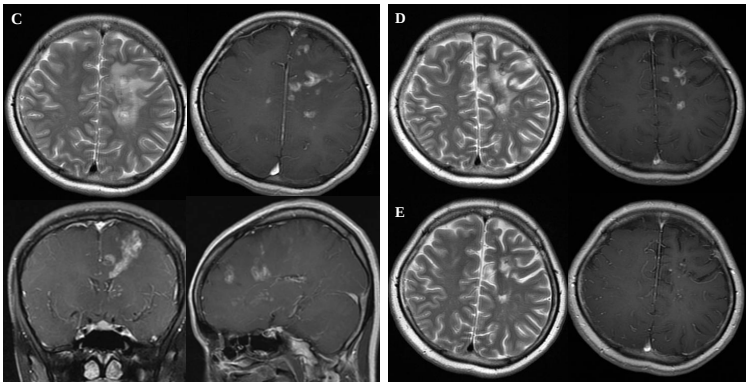
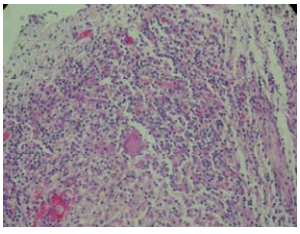


Fig C-E, Patient 6, C: Multifocal T2 prolongation abnormalities in left frontal lobe surrounded with vasogenic edema on T2WI, with multiple nodular and patchy enhancing lesions mainly in white matter area on post Gd-DTPA T1WI were demonstrated. D: After corticosteroid treatment, 50 days later, the lesions decreased in number and size, vasogenic edema also lessened. E: Five months later, the lesions almost disappeared.



prominent T-cell infiltration in cortex, subcortical white matter, and/or meninges. Perivascular lymphocytes infiltration was displayed, but not as the angiocentric pattern

Discussion and Conclusion: The idiopathic autoimmune encephalopathy is a diagnosis of exclusion. Clinically, subacute onset of cognitive impairment with seizures and movement dysfunction is a common scenario. MR imaging characteristics of AE may be nonspecific, as diffuse white matter abnormalities with/without gray matter and/or meninges involved, and enhanced lesions often displayed, the differential diagnosis should include infectious meningoencephalitis, neurosarcoidosis, inflammatory demyelination, toxin exposure and metabolic disturbance. Prompt diagnosis is essential since effective treatment may be available, and permanent impairment can result if the encephalopathy is left untreated. A positive biopsy is required to make a definitive diagnosis for most difficult cases. However, after thorough clinical and laboratory evaluation, in absence of diagnostic serological/CSF findings, MR imaging features may suggest the diagnosis. A rapid improvement both clinically and radiographically shortly after steroid and/or immunotherapy may be the only evidence to validate the diagnosis of AE.

References: 1. Vernino S, et al. Neurologist, 2007;13:140-147. 2. Neeraj J, et al. AJNR 2005;26:642–645. 3. Contreras AS, et al. J Clin Rheumatol 2004;10:339-343.