Acute necrotizing encephalopathy secondary to sepsis

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A 46-year-old man was admitted for a sudden onset of generalized tonic-clonic seizure and convulsion as well as deteriorating unconsciousness for 14 hours. He was treated with antibiotics during the preceding 2 weeks for high fever and upper respiratory tract infection. Upon admission, his body temperature was 40.3°C, blood pressure was 86/50 mm Hg, and the Glasgow Coma Scale score was 5/15. Nuchal rigidity was noted. Kernig sign and Babinski sign were positive bilaterally. Other systemic physical examinations were unremarkable. The blood routine test showed increased white blood cells (17.47×10^9/L; neutrophils 86%) and decreased platelet (37×10^9/L), and coagulant dysfunction (fibrinogen degradation product 122.9 μg/mL, D-dimer 6283 μg/L, prothrombin time 25 s, activated partial thromboplastin time 44.1 s) was observed. Acidosis and elevated serum liver enzymes (aspartate aminotransferase 7222 IU/L, alanine aminotransferase 5126 IU/L), creatinine (142 μmol/L), and amylase (705 IU/L) were found. Blood samples were collected for blood cultivation, whereas no pathogen was identified. Serum viral studies (hepatitis B virus, hepatitis C virus, human immunodeficiency virus) and syphilis serology tests were all negative. The cerebrospinal fluid (CSF) test revealed slightly elevated protein level (0.87 g/L), elevated immunoglobulin G (IgG) level (107 mg/L), and normal glucose level without pleiocytosis. CSF IgG and IgM for herpes simplex virus, cytomegalovirus, Epstein-Barr virus, rubella virus, rubeola virus, and human herpesvirus were all negative. CSF rapid plasma reagin for syphilis was negative. The electroencephalography examination demonstrated diffuse generalized and slow background activity. The pulmonary computed tomographic (CT) scan showed mild pneumonia, and the abdominal CT scan showed abnormal attenuation in the right lobe of liver. The brain CT scan showed symmetric low attenuation on the thalamus, and the brain magnetic resonance imaging (MRI) showed symmetric concentric thalamic lesions (**Figure 1**). Sepsis and acute necrotizing encephalopathy (ANE) were diagnosed by ruling out viral encephalitis, acute disseminated encephalomyelities, cerebral vasculitis, and metabolic encephalopathy, according to laboratory tests and imaging features. Intensive antiseptic and supportive treatment was initiated, and the patient turned conscious 2 days later. The follow-up MRI-fluid-attenuated inversion recovery (FLAIR) 2 months later showed a low signal intensity in the core of the lesion in Panel E, which was indicative of iron deposition (**Figure 1**).

ANE, which usually occurs subsequent to viral infections especially the influenza virus, is a rare disease leading to high mortality and mobility.[1-3] The characteristic pathological findings of ANE are perivascular hemorrhage and necrosis of lesions, and vascular congestion with extravasations and the lesion edge. The imaging feature of bilateral and symmetrical lesions in the deep white matter and basal ganglia is typical of ANE (**Figure 1**).[1,2,4-6] It has been reported that prompt treatment with anti-cytokine agents such as corticosteroid brings favorable outcomes in patients with ANE.[6-8] Our patient had a febrile history prior to the neurological deterioration presenting as coma and seizures and was given an insufficient antiseptic treatment. Although no pathogen was identified in this patient, possibly due to the prior use of antibiotics, the sudden withdrawal of the antibiotics might result in a rebound of immune responses. According to our knowledge, ANE secondary to sepsis has not been reported thus far. The prompt recognition of the disease and the earlier introduction of intensive treatment resulted in a good outcome for our patient.[8]

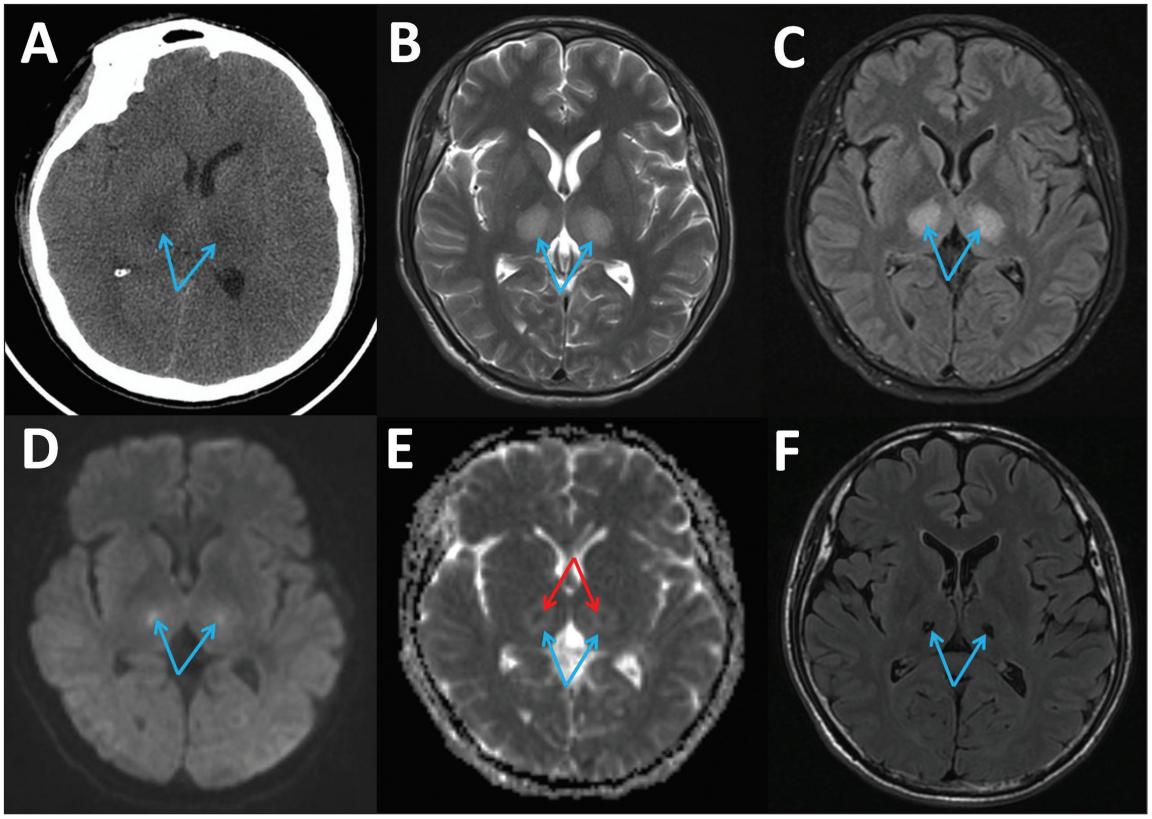

Figure 1. CT (A) and MRI (B: T2WI; C: FLAIR; D: DWI; E: ADC) showed symmetric lesions on the bilateral thalamus. DWI (D) revealed high signal intensity (blue arrow). ADC (E) revealed symmetric concentric thalamic lesions: reduced signal intensity in the core (red arrow), corresponding to diffusion restriction, and increased in the outer layer (blue arrow), corresponding to vasogenic edema and diffusion enhancement. The follow-up MRI-FLAIR showed low signal intensity in the core of the lesion in Panel E (blue arrow), which is indicative of iron deposition (F). ADC: Apparent diffusion coefficient. DWI: diffusion-weighted imaging; FLAIR: fluid-attenuated inversion recovery; MRI: magnetic resonance imaging.

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