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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 convulsions, as well as for deteriorating unconsciousness for 14 hours. He was treated with antibiotics during the preceding 2 weeks for a high fever and upper respiratory tract infection. Upon admission, his body temperature was 40.3°C, his 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. A routine blood test showed increased white blood cell (WBC) (17.47×10^9/L; neutrophils 86%) and decreased platelet (37×10^9/L) counts; coagulant dysfunction (fibrin degradation products [FDP], 122.9 μg/mL; D-dimer, 6283 μg/L; prothrombin time [PT], 25 seconds; activated partial thromboplastin time [APTT], 44.1 seconds) was also observed. Acidosis and elevated serum liver enzymes (aspartate transaminase [AST], 7222 IU/L; alanine transaminase [ALT], 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 [HBV], hepatitis C virus [HCV], human immunodeficiency virus [HIV]) and syphilis serology tests were all negative. A cerebrospinal fluid (CSF) test revealed a slightly elevated protein level (0.87 g/L), an elevated immunoglobulin (Ig)G level (107 mg/L), and a normal glucose level without pleiocytosis. CSF IgG and IgM for *herpes simplex virus*, *cytomegalovirus*, *Epstein–Barr virus*, *Rubella virus*, rubeola, and *human herpesvirus* were all negative. The CSF rapid plasma regain test for syphilis was negative. Electroencephalography examination demonstrated a diffuse generalized and slow background activity. Pulmonary computed tomography (CT) showed mild pneumonia, and abdominal CT showed abnormal attenuation in the right lobe of the liver. Brain CT showed symmetric low attenuation of the thalamus, and 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 encephalomyelitis, cerebral vasculitis, and metabolic encephalopathy, according to laboratory tests and imaging features. Intensive antiseptic, as well as supportive, treatment was initiated and the patient regained consciousness 2 days later. Follow-up MRI–fluid-attenuated inversion recovery (FLAIR) 2 months later showed low signal intensity in the core of the lesion, indicative of iron deposition (**Figure 1E**).

ANE, which usually occurs subsequent to viral infections, especially following the influenza virus, is a rare disease leading to high mortality and morbidity.[1-3] The characteristic pathological findings of ANE are perivascular hemorrhage and necrosis of the lesions, as well as vascular congestion with extravasations and the lesion edge. The imaging features of the bilateral and symmetric lesions in the deep white matter and basal ganglia are typical of ANE (**Figure 1**).[1,2,4-6] It has been reported that prompt treatment with anticytokine agents, such as corticosteroids, would yield favorable outcomes in patients with ANE.[6-8] Our patient had a febrile history prior to neurological deterioration, which presented as coma and seizures, and he 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 have resulted in a rebound of immune responses. 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]

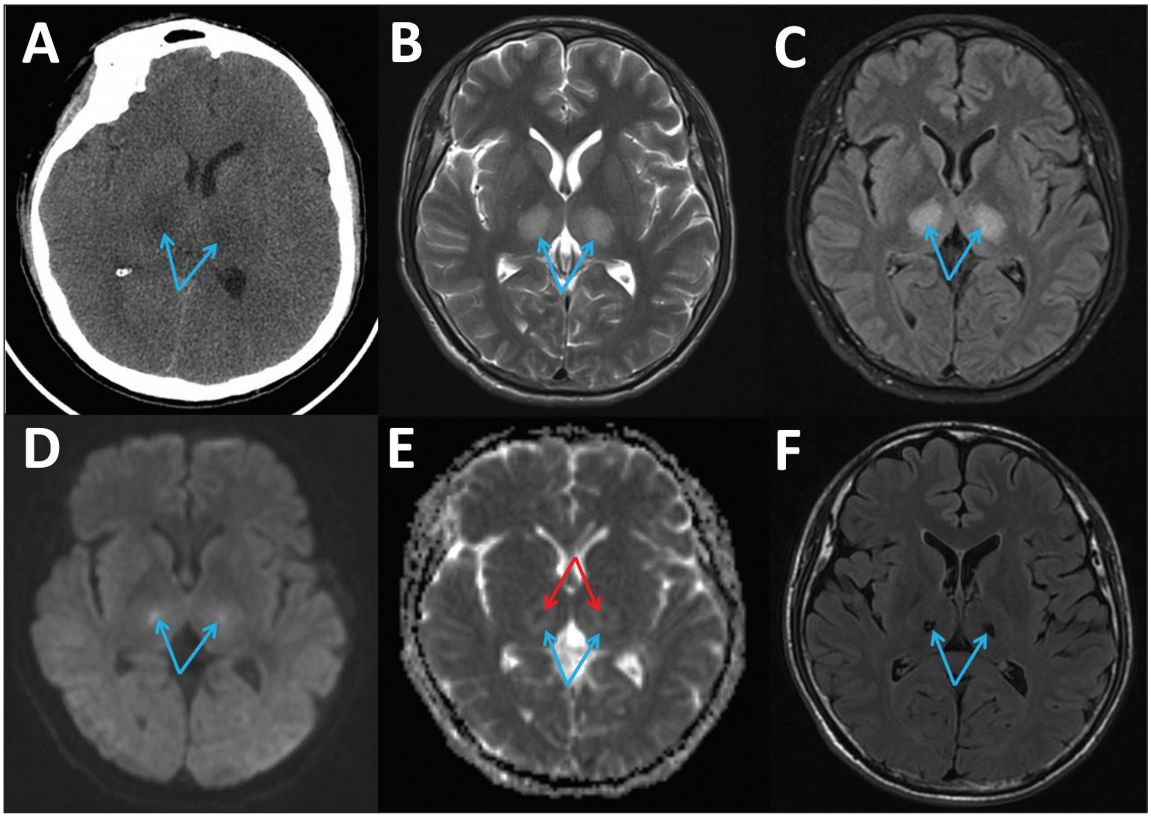

Figure 1. (A) CT and (B–E) MRI (B, T2WI; C, FLAIR; D, DWI; E, ADC) showed symmetric lesions on the bilateral thalamus. (D) DWI revealed high signal intensity (blue arrow). (E) ADC revealed symmetrical concentric thalamic lesions: reduced signal intensity in the core (red arrow), corresponding to diffusion restriction; and increased signal in the outer layer (blue arrow), corresponding to vasogenic edema and diffusion enhancement. (E) Follow-up MRI-FLAIR showed low signal intensity in the core of the lesion (blue arrow), indicative of (F) iron deposition.

Abbreviations: CT: Computed tomography; MRI: magnetic resonance imaging; T2WI: T2-weighted imaging; FLAIR: fluid-attenuated inversion recovery; DWI: diffusion-weighted imaging; ADC: apparent diffusion coefficient.

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