CLINICAL CASE - TEST YOURSELF Pediatric imaging

Bilateral symmetric "owl eye" lesions of thalami in a 7-year-old child

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PART A

A 7-year-old child presented to the emergency department with seizures and altered sensorium. The patient had been suffering from a cold, fever with chills and rigors, headache, and vomiting for three days. Blood and cerebrospinal fluid (CSF) tested negative for dengue and Japanese B encephalitis antibodies. Computed tomography (CT) of the brain and Magnetic resonance imaging (MRI) was performed (Fig. 1,2,3). What are the conclusions from CT and MRI scans?

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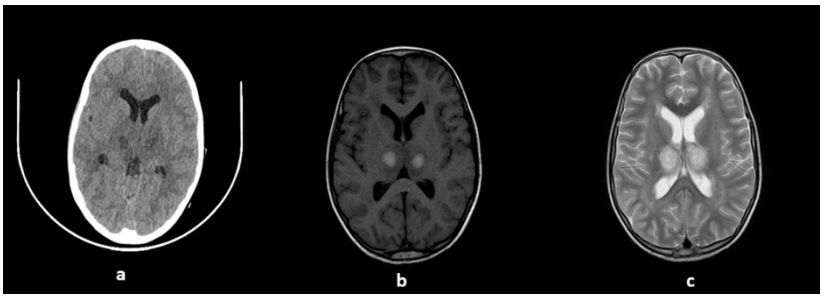

Figure 1: Axial CT (a), T1W MRI (b), T2W MRI (c) images of brain

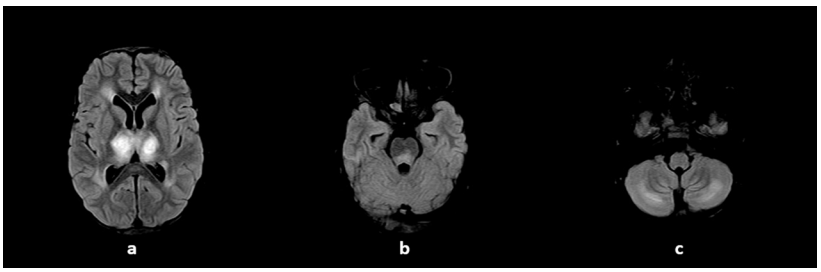

Figure 2: Axial FLAIR (a, b, c) images of the brain

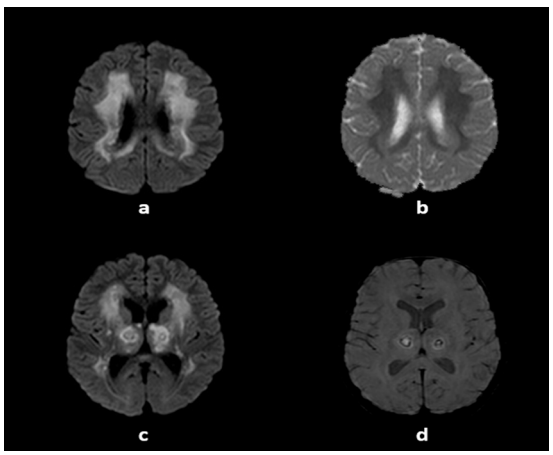

Figure 3: Axial DWI MRI (a, c), ADC (b), and GRE (d) images of the brain

PART B

Diagnosis: Acute necrotizing encephalopathy (ANE)

ANE of childhood is a rare disease that causes rapid neurological decline following a respiratory or gastrointestinal infection [1]. Though initially thought to be a disease of childhood, few cases were reported in adults [2]. The disease was initially thought to possess a geographic predilection, with most cases having been reported from Japan and Southeast Asia [3]. However, current evidence suggests that the disease though sporadic, has a global prevalence. Genetic studies have demonstrated positive results for HLA DRB/DQB genes and mutations in Ran Binding Protein 2 (RANBP2) [4]. The exact etiology and pathogenesis of the disease remain unknown. However, immune and cytokine-related injury following influenza, parainfluenza, varicella, mycoplasma, and herpes virus infection has been attributed to the pathogenesis of the disease. The hepatic and renal failure and disseminated intravascular coagulation (DIS) caused by the "cytokine storm" associated with the disease lead to increased mortality in these patients. In addition, tumor necrosis factor (TNF-α), interleukins (IL- 6,10,15), and injury- and brain damage related to the disease are attributed to the disease [5]. There are usually no signs of active inflammation within the brain parenchyma despite the presence of prior viral infection. A common cold, high-grade fever, headache, vomiting, and loose stools are the symptoms that usually precede the rapid neurological decline in patients with ANE. Blood investigations generally test negative for antiviral antibodies and might reveal elevated liver enzymes, while CSF studies can reveal elevated protein levels. The patients suffering from ANE can exhibit diverse clinical features, ranging from mild disease with complete recovery to fulminant disease with high mortality. The prognosis of the disease may range from minimal sequelae due to the disease to high morbidity with irreversible damage. Children suffering from ANE usually present with fever with chills and rigors, headache, gastroenteritis, and vomiting and can proceed to severe neurological symptoms such as altered sensorium and status epilepticus.

In the present case, brain CT examination demonstrated symmetric hypodensities in bilateral thalami (Fig. 1a). MRI revealed symmetrical hyperintensities on T1W, T2W, and FLAIR images (Fig. 1b, Fig. 1c, Fig.2). Diffusion-weighted imaging (DWI) indicated hyperintensities in bilateral thalami and periventricular white matter with reduced apparent diffusion coefficient (ADC) (Fig. 3a, Fig. 3b, Fig. 3c). Hemorrhages were noted in bilateral thalami on Gradient echo T2W (GRE) (Fig. 3d). The signal abnormalities on MRI represented a typical "owl eye" configuration. A diagnosis of ANE was made and the patient was treated with intravenous methylprednisolone and immunoglobulins. However few days later unfortunately the patient succumbed to the disease.

Radiologically, the patients present bilateral symmetrical lesions involving the thalami, putamen, brainstem tegmentum, cerebellar white matter, and periventricular white matter. The lesions in the thalami are usually necrotic with areas of hemorrhages. Wong et al developed a scoring system based on the presence of hemorrhage or necrosis in the thalami, brainstem, and cerebellum with one point each for the presence of hemorrhage or necrosis in the brainstem and cerebral white matter with a minimum score of 0 and maximum of 4 carrying worst prognosis [6]. On T1W images, the signal intensity can vary from hypointensity, resembling areas of necrosis, to hyperintense signals, which indicate hemorrhages. Contrast images can reveal rim enhancement patterns around the necrosis areas. T2W and FLAIR images can reveal hyperintense lesions that are hyperintense with concentric areas of hemorrhage and necrosis, and resemble a "doughnut" or an "owl eye" [7,8]. On diffusion images, lesions appear hyperintense with corresponding areas of reduced ADC suggestive of cytotoxic damage. Lesions demonstrate "susceptibility effects" suggestive of hemorrhages on Gradient and SWI. Lipid/lactate peaks and glutamate/glutamine peaks might be observed on magnetic resonance spectroscopy [9].

The differential diagnoses needed to be considered and subsequently excluded include dengue encephalitis, Japanese B encephalitis, thrombosis of the straight

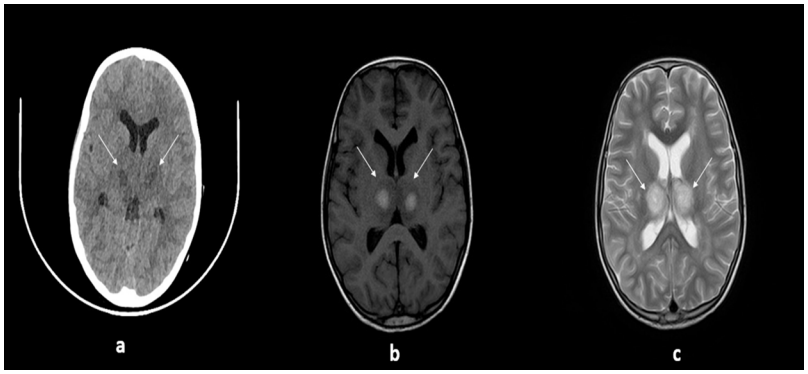

Figure 1 : *Axial CT (a) and T1W MRI (b), T2W MRI (c) images of the brain demonstrating bilateral symmetric hypodensities with areas of hemorrhage (white arrows in image a) and hyperintense signals on T1W, T2W MRI (white arrows in images b,c) suggestive of hemorrhages and surrounding edema in the bilateral thalami.*

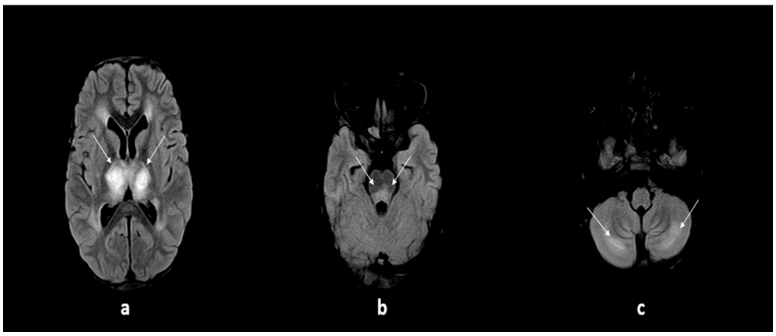

Figure 2 : *Axial FLAIR (a, b, c) images of the brain demonstrating bilateral symmetric hyperintense signals in thalami, pons, and cerebellum (white arrows).*

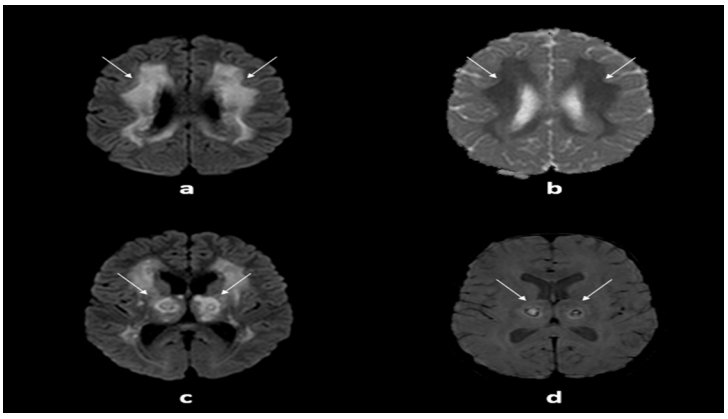

Figure 3: *Axial DWI MRI (a, c), ADC (b), and GRE (d) images of the brain demonstrating hyperintense signals on DWI and corresponding areas of reduced ADC suggestive of cytotoxic edema/damage in bilateral periventricular white matter and thalami (white arrows in images a, b, c). Susceptibility effects noted on GRE images suggestive of hemorrhages (white arrows in image d).*

sinus, artery of Percheron infarcts and bilateral thalamic gliomas, acute disseminated encephalomyelitis, and Leigh's disease. The treatment is mainly supportive care and intravenous steroids, including methyl prednisolone and intravenous immunoglobulins are the main drugs used for the treatment in addition to antibiotics and antiviral drugs. The prognosis of the disease is usually variable with patients with increased liver enzymes and radiologically brainstem lesions having poor prognosis. **R**

KEY WORDS

Computed tomography, Magnetic resonance imaging, Diffusion weighted imaging,Acute necrotising encephalopathy, thalami, GRE

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