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Acute hemorrhagic leukoencephalitis-A medical catastrophe

Spandana Devi Tagaram
Dr D Y Patil Medical College, India

Acute Hemorrhagic Leukoencephalitis (AHLE) is a hyper acute severe variant of acute disseminated encephalomyelitis (ADEM) with an incidence of 0.4 -0.5 per 1,00,000. It manifests with multiple neurologic symptoms, encephalopathy, that may be progressive in nature. Specialised spin-ECHO Magnetic Resonance Imaging (MRI) is the diagnostic. AHLE usually has a very poor prognosis with high mortality. We present a case of 8 year old female child brought to the emergency with sudden onset of slurring of speech, headache, vomiting, retention of urine and altered sensorium followed by 2 weeks of febrile illness. On examination, Glasgow coma scale (GCS) was 8/15, generalised hypotonia with brisk deep tendon reflexes with extensor plantar response was present. Pupils were reactive to light and accomodation. Labs showed high inflammatory markers, normocytic normochromic anemia with benign renal, coagulation, hepatic function test panel. Viral encephalitis panel was negative along with malaria, dengue, typhoid serologies. Lumbar puncture (LP): CSF protein-14.9, sugar -72 mg/dl gross: clear, microscopy: 5 nucleated cells (lymphocytes). MRI: Bilaterally symmetrical areas of altered signal intensity involving ventrolateral thalamus, lateral mid brain, ventrolateral pons, and bilateral middle cerebellar peduncles appearing hyper intense on T2W1. T2W1, FLAIR sequences showing diffusion restriction appearing dark and foci of haemorrhages seen on SW1. Patient is diagnosed to have AHLE based on the clinical history and neuro imaging findings. She received IV immunoglobulin and IV methyl prednisolone followed by oral corticosteroids for 8 weeks with other supportive measures. The patient showed a gradual improvement in the GCS score and her neurological symptoms resolved. She does have some residual neurological sequelae (spasticity of her extremities) at present. Even very rare, an early diagnosis and timely management holds key for a favourable outcome.

spandana.india@gmail.com