

Case Report

Acute Hemorrhagic Leukoencephalitis- A Rare Medical Catastrophe!!!

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Introduction

Acute hemorrhagic leukoencephalitis (AHLE) or Weston-Hurst's syndrome 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, but has some recovery after treatment. It is characterized by a hemorrhagic inflammatory response in the central nervous system primarily affecting the white matter, myelin sheath often sparing the periventricular white matter. Most cases are autoimmune in origin and others have prior history of (h/o) viral illness and post vaccination status. It is a diagnostic challenge at its first attack. Specialised spin-ECHO Magnetic resonance Imaging (MRI) is the diagnostic. AHL usually has a very poor prognosis with high mortality. Treatment is unclear but high dose steroids/immunosuppressants may offer some benefit and promote better recovery although with a residual neurological deficit.

Case Report

Presenting a case of an 8-year-old female child was admitted with sudden onset of slurry speech, headache, urinary retention, vomiting, altered mental status followed by two weeks of febrile illness. She had H/O upper respiratory viral illness six weeks prior. Examination is positive for generalised hypotonia with brisk deep tendon reflexes and with extensor plantar response. Her Glasgow coma scale (GCS) was 8/15 on admission. Pupils were reactive to light and accommodation. Labs showed high inflammatory markers, normocytic normochromic anemia with benign renal, coagulation, hepatic function test panel. The initial impression was acute meningio-encephalitis, so broad spectrum antibiotics and antiviral drugs were commenced and lumbar puncture was performed after some period of stabilization. Viral encephalitis panel was negative along with Malaria, Dengue, Typhoid serologies. Lumbar puncture (LP): CSF protein-14.9, sugar -72mg/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 hemorrhages seen on SW1. Figure 1- 4

SW: SEQUENCE (Susceptibility weighed): Bilateral foci of hemorrhages involving ventrolateral thalamus, lateral mid brain, ventrolateral pons, and bilateral middle cerebellar peduncles.

Patient is diagnosed to have acute hemorrhagic leukoencephalitis based on the clinical history and neuro imaging findings. She received three days of high dose pulse steroids followed by tapering of oral steroids for a total of 8 weeks. The patient showed a gradual improvement in the GCS score and her neurological symptoms resolved. She does have some residual neurological sequelae (spasticity

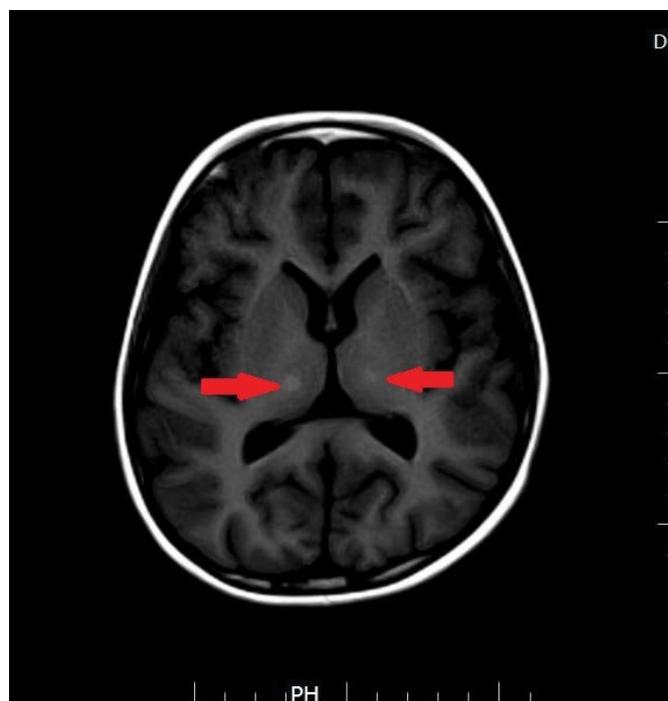


Figure 1: MRI T1 WEIGHTED IMAGES: T1Sequence-With foci of hemorrhages in thalamus.

of her extremities) at present.

Discussion

AHLE is a rare manifestation of ADEM. History is suggestive of 1) acute encephalopathy and neurological symptoms with h/o recent vaccine use or post viral phase and 2) MRI findings of demyelinating punctate hemorrhages in cerebral hemispheres. Differential diagnosis to be considered are acute meningioencephalitis, multiple

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Figure 2: MRI T2 WEIGHTED IMAGE: T2 Sequence-With foci of hemorrhages in thalamus.

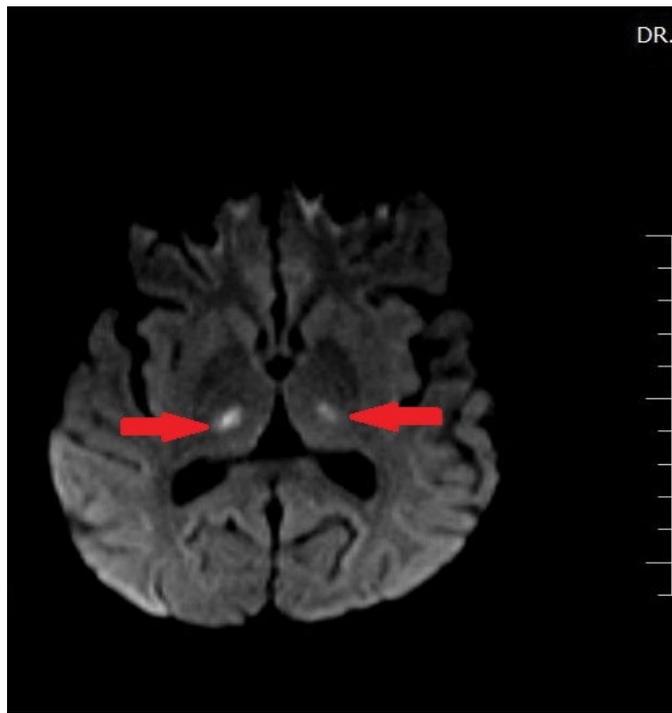


Figure 3: Diffusion restriction: Foci of hemorrhages seen in thalamus.



Figure 4: SW: SEQUENCE (Susceptibility weighed)

sclerosis(MS), transverse myelitis, Devics disease. Demonstration of punctuate hemorrhages in brain magnetic resonance images (MRI) especially in susceptibility weighted images (SWI) is a good distinguishing feature from the classical ADEM. High dose steroids are the first line of treatment once the diagnosis is confirmed. For patients with poor response to steroids, plasma exchange, intravenous immunoglobulins is the next step. Long term follow up from 3 months to at least 5 years with neuro imaging is recommended to confirm the absence of new demyelinating lesions. Prognosis is usually guarded.

Conclusion

At its first attack AHLE is a diagnostic challenge. High index of suspicion in children with multifocal neuro encephalopathic symptoms with h/o post viral and recent vaccination is required for early diagnosis. Our case has thrown light on importance of early detection by MRI and utilizing immune-modulating therapies without delay for a better prognosis. Follow up MRI s are usually recommended.