ACUTE HEMORRHAGIC LEUKOENCEPHALITIS
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Abstract:
Acute hemorrhagic leukoencephalitis (AHL) is a rare disease that is characterized by a rapidly progressive, fulminating, monophasic, inflammatory hemorrhagic demyelination of the white matter. Early diagnosis and management of this rare entity, which may mimic other conditions, is essential as it carries a very high mortality. We present here a lady who developed this rare disease 3 days post partum. Timely initiation of therapy with Intravenous immunoglobulin and pulse steroid led to a good improvement of her neurological status.

Keyword: Acute hemorrhagic leukoencephalitis, postpartum

CLINICAL PRESENTATION
A 35-year-old lady developed vomiting and loose stools 3 days post partum. She was known to have primary infertility for 10 years but had conceived spontaneously. Her pregnancy was complicated by gestational diabetes mellitus. She delivered a live male child by a low forceps delivery and had an uncomplicated immediate postnatal period. She had no history of fever, seizures, bleeding manifestations or recent immunization. 3 days after the onset of above symptoms she was found to be lethargic. On examination, she was drowsy and sluggishly responding to commands. Her vital signs were stable. Neurological examination revealed paucity of movements of left upper and lower limbs with exaggerated reflexes and extensor plantar response bilaterally. Rest of the systemic examination was normal. The clinical differentials considered were Cortical venous thrombosis (CVT), Post partum vasculitis/Antiphospholipid antibody syndrome, Thrombotic thrombocytopenic purpura (TTP), Viral Encephalitis or Acute disseminated encephalomyelitis (ADEM).

On laboratory investigations she was found to have anemia with thrombocytopenia, normal electrolytes, normal creatinine, mild direct hyperbilirubin and normal coagulation tests. An emergency magnetic resonance imaging (MRI) of the brain with magnetic resonance venogram (MRV) done was negative for cortical venous thrombosis. TTP was excluded as she had no fever, schistocytes on blood picture or renal failure. Though complements were below normal she had no other positive serology to suggest vasculitis/Antiphospholipid antibody syndrome. Cerebro-spinal fluid (CSF) analysis showed neutrophilic leucocytosis, elevated protein and low sugars, CSF pressure was not measured.

The MRI done showed multi focal areas of patchy restricted diffusion and micro hemorrhages in the sub-cortical regions in both cerebral hemispheres. There were no focal areas of enhancement. There was no venous or arterial thrombosis. Since patients sensorium worsened further she required intubation and mechanical ventilation and was hence admitted to the intensive care unit (ICU). Her GCS on admission to the ICU was documented to be 2/15. She was treated with intravenous immunoglobulin (0.5 grams per kilogram per day for 4 days) along with high dose steroid pulse (methylprednisolone 1 gram intravenous daily for 5 days). Six days after the initiation of the intravenous immunoglobulin and the pulse steroid, her sensorium improved to 9/15. Her stay in the ICU was complicated by a catheter related blood stream infection with Pseudomonas aeruginosa which was treated with Piperacillin Tazobactem for 2 week as per the sensitivity pattern. She was shifted to the ward and her power was noted to be grade 2 in upper limbs and grade 1 in lower limbs. Over the next 15 days with regular physiotherapy and supportive care, her power improved to grade 4 in the upper limbs and grade 3 in lower limbs. At a follow up visit, 4 months later, she was ambulating well and was independent for activities of daily living.
MRI of the brain: 1a,b FLAIR images showing subcortical areas of hyper intensity in the peri-Rolandic regions. 2a,b Diffusion weighted imaging in the same areas showing restricted diffusion. 3a,b Susceptibility weighted images (SWI) showing associated micro hemorrhages

DISCUSSION:
Among the known white matter diseases only two are monophasic, rapidly progressive, potentially catastrophic, with extensive central nervous system involvement: Acute Disseminated Encephalomyelitis (ADEM) and the other being Acute Hemorrhagic Leukoencephalitis (AHL). (1,2)

Acute hemorrhagic leukoencephalitis (AHL) is a rare entity that is characterized by an acute onset, rapidly progressive, monophasic, fulminant, with inflammatory hemorrhagic demyelination of white matter. (1) It is usually post infectious and can lead to death or severe morbidity within a few days. AHL is thought to be a hyper acute form of the more common disease, acute disseminated encephalomyelitis (ADEM); both diseases result from an autoimmune process which targets the CNS myelin. However AHL has a more fulminant course than ADEM, with rapid progression and is more frequently fatal.

Though our patient clinical profile fit into a diagnosis of cortical venous thrombosis, MRI Brain with MRV was negative for a CVT. Other differentials were also ruled out by appropriate investigations. The possibility of post partum vasculitis , a close differential on MRI was considered unlikely as clinical presentation was not consistent with such illness. Patients with post partum vasculitis present with headache and seizures,there is no rapid worsening of the sensorium or abnormal CSF findings. ( 3) Serological tests for an underlying vasculitis, except for low complements, were also negative

Differentiating ADEM from AHL on the MRI Brain could be difficult. On MRI Brain the lesions of acute hemorrhagic leukoencephalitis are usually larger and are associated with more edema and mass effect as compared to acute disseminated encephalomyelitis and frequently show foci of hemorrhage which are not usually seen in acute disseminated encephalomyelitis. (4 ).Brain biopsy though essential for diagnosis was not done as the patient sensorium showed good improvement with institution of therapy.

Management of AHL has been largely restricted to case reports. (5,6 )Successful treatment with immunoglobulins, plasma exchange and high dose steroids had been reported in literature. Our patient was treated with combination of immunoglobulins with pulse steroids and she showed good improvement in her sensorium and neurological deficits.

Our case report highlights the importance of considering this rare but potentially fatal illness which can mimic the other common differentials in this setting which include CVT, Post partum vasculitis,TTP or a viral/bacterial encephalomyelitis. An MRI imaging of the brain is essential in making the diagnosis.
Initiation of treatment with immunosuppressive therapy at the earli-
est may result in prevention of major morbidity or mortality

REFERENCES: