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Case Report Open Access

Postpartum Lady with Bleed in the Brain: A Rare Manifestation of Neuroleptospirosis with Acute Haemorrhagic Leucoencephalitis (AHLE)

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Abstract

Acute Haemorrhagic Leucoencephalitis (AHLE) is a rare disorder which was first described by Weston Hurst in 1941. It has been acknowledged as severe form of acute disseminated encephalomyelitis, characterized by acute onset and rapid progression of inflammation of the brain with associated necrosis and haemorrhage. We report an unusual case of a patient in postpartum period presenting with micro bleeds in the brain with cerebral oedema and acute haemorrhagic leucoencephalitis associated with leptospirosis.

Keywords: Acute haemorrhagic leucoencephalitis; Renal tubular acidosis; Leptospirosis

Case Presentation

A 29-year old lady at 40 days postpartum presented with bilateral lower limb weakness for 2 days. There was history of low grade fever but no vomiting, diarrhea, cough, rash or joint pain. Antenatal history uneventful except for a history of herpetic vulvo-vaginitis at 7 weeks gestation for which she was treated with acyclovir. On examination she appeared drowsy, blood pressure was 122/89 mmHg, pulse rate 89 bpm, respiratory rate of 40 and febrile. Neurological examination revealed pinpoint pupils and there was mild neck stiffness. No ptosis or obvious stigmata of connective tissue disease noted. Both lower limbs were hypotonic with motor power grade 1/5 in both ankles, 2/5 in both hips and normal tendon reflexes. She was intubated for airway protection. Investigations revealed hemoglobin of 13.7 g/dL, platelet count 399 × 10^9/L, white blood cell count 22.28 × 10^9/L, urea 6.5 mmol/L, sodium 142 mmol/L, potassium 1.7 mmol/L, chloride 125 mmol/L, creatinine 109.9 µmol/L with normal magnesium and phosphate. Arterial blood gas showed pH 7.147, paO_2 374, $paCO_2$ 40, bicarbonate 13.6 and urine pH of 7.0. Initial diagnosis of distal renal tubular acidosis (RTA) was made based on severe hypokalaemia and hyperchloraemic normal anion gap metabolic acidosis which was presumed triggered by sepsis. Plain and contrasted computed tomography of the brain showed bifrontal bleed with cerebral oedema (Figure 1).

External ventricular drainage (EVD) was inserted and she was kept on cerebral protection. Serology for leptospira (IgM-ELISA) and CMV IgG was positive and other serological tests for dengue, HSV-1 and 2, hepatitis and HIV were negative. Ultrasound of the kidney and urinary system was normal. Magnetic resonance imaging (MRI) of the brain showed foci of haemorrhage at genu of corpus callosum and multiple areas of punctate haemorrhage (Figures 2 and 3).

Cerebrospinal fluid (CSF) examination later showed mildly elevated protein, normal glucose and low cell count. Her acid-base improved shortly before developing rhabdomyolysis with acute kidney injury needing a series of renal replacement therapy in the ICU. Further investigations were negative for biomarkers of connective tissue diseases. Viral screenings for influenza, Para influenza, RSV, adenovirus, coronavirus, bocavirus, and metapneumovirus and rickettsiae serology were also negative. Her neurological recovery remained poor despite appropriate antimicrobial therapy thus methylprednisolone was initiated. However, the treatment was halted after the first dose because of nosocomial sepsis. Paired sera test for leptospira came back positive with microscopic agglutination titer (MAT) 1:400, confirming the etiology of her acute haemorrhagic leukoencephalitis secondary to leptospirosis. She made slow but gradual recovery in the ICU. She was discharged home after 4 months hospitalization.

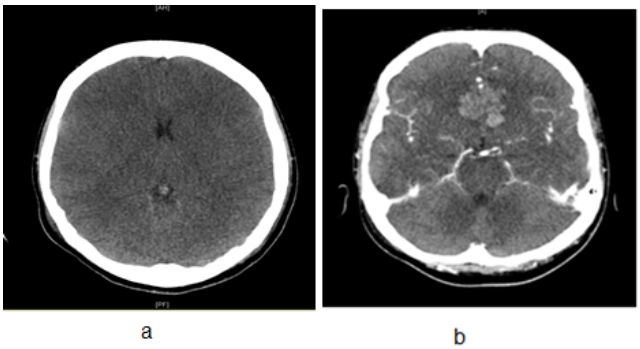

Figure 1: (a) Non-contrasted computed tomography of the brain showing bifrontal hyper densities with cerebral oedema **(b)** Contrasted study showing bifrontal parasagittal hyper densities with subarachnoid haemorrhage configuration and perilesional oedema.

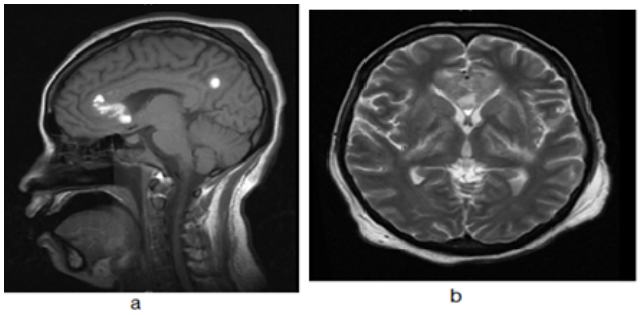

Figure 2: (a) Sagittal view of T1 weighted MRI showing foci of haemorrhage at genu corpus callosum and left parasagittal lobe **(b)** T2/FLAIR no focal white matter hyperintensities apart from perilesional oedema secondary to haemorrhage.

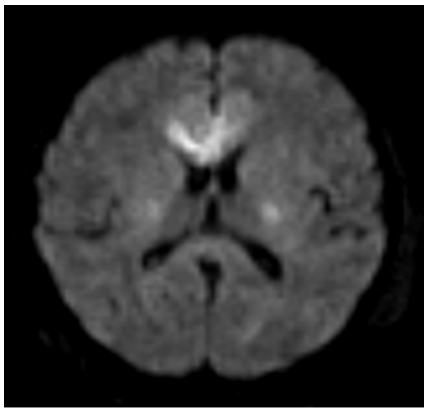

Figure 3: T1 GRE protocol showing corresponding blooming artefact and multiple microbleeds in internal capsule extending down to the crux of midbrain bilaterally.

Discussion

Leptospirosis is a disease caused by spirochetes of the genus Leptospira and is a challenge for clinicians if presented atypically. It carries high morbidity and mortality in selected cases, if untreated [1]. Leptospirosis is transmitted through direct or indirect contact with infected animals or their urine. Typical manifestations include sepsis and septic shock, acute kidney injury, hepatic dysfunction, electrolyte imbalance, pulmonary hemorrhage, cardiovascular instability, thrombocytopenia, pancreatitis, myocarditis, rhabdomyolysis, acalculous cholecystitis, pericarditis, and reactive arthritis [2]. Few uncommon but more severe forms of manifestations may result from immunological phenomena and multi-organ involvement.

Neuroleptospirosis can present as meningoencephalitis, cerebral infarct or haemorrhage, cerebral venous thrombosis, cerebral arteritis, subarachnoid hemorrhage, optic neuritis, cranial nerve palsy, transverse myelitis, Guillain-Barre syndrome (GBS), mononeuritis multiplex, peripheral nerve palsy and psychosis. Acute hemorrhagic leukoencephalitis (AHLE) is a very rare form of neuroleptospirosis characterized by demyelinating process or myelopathy. Some authors also categorized AHLE as a form of acute disseminated encephalomyelitis (ADEM). In certain circumstances the diagnosis remains as a clinical challenge as there is not one single etiological relationship. Exact mechanism is relatively unknown but autoimmune pathophysiology is likely. It has been proposed that presence of immune cross-reactivity between myelin basic protein moieties and various infectious agent antigens causing the demyelination. Two prior case reports by Befort and Francisci [3,4] presented two cases of AHLE after Epstein-Barr virus infection.

The findings in the computed tomography scans or magnetic resonance imaging are used mainly for the diagnosis. The areas involved in the majority of the cases are the parietal lobes, but the lesions can be seen anywhere in the subcortical white matter, mid brain, pons, corpus callosum, basal ganglia, medulla, cerebellum, and even spinal cord. Imaging techniques may also complicate the diagnosis as computed tomography and MRI may show nonspecific findings such as hematoma in the basal ganglia and ventricles in certain patients [5]. Histopathological findings are dominated by features of fibrinoid venous necrosis, leading to acute haemorrhage, with fibrin exudates and neutrophilic debris resembling "ring and ball", white matter ischemic changes adjacent to necrotic post capillary venules haemorrhage which are more prominent than demyelination [6] but some of the times there are no specific immunohistochemical pattern.

Treatment for AHLE is not well-established; some authors describe in recent literature that a combination of immunosuppressant medications and/or therapeutic plasma exchange may be of benefit in the treatment given the underlying cellular-humoral mediation and molecular mimicry [7]. General treatment includes aggressive management of cerebral oedema, surgical intervention in the event of increasing intracranial hypertension and other general measures in caring for the critically ill patients. High index of suspicion is useful in early recognition of the disease. To our knowledge, there is no human case report to tie up association of postpartum tubular renal acidosis with acute haemorrhagic leucoencephalitis in neuroleptospirosis.

Conclusion

Acute haemorrhagic leucoencephalitis is a diagnostic challenge for clinicians, especially when there is no specific or suggesting history of exposure to specific pathogens. This rather unusual and complex case highlighted the urgency of doing things right from the beginning; from making the right diagnosis, implementing the right interventions, and to giving the right treatment timely; for without which can impact on mortality and morbidity.

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