



Acute Hemorrhagic Leuko-encephalitis - A rare complication of Dengue infection.

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Abstract

Introduction: Acute hemorrhagic leuko-encephalitis or Weston-Hurst's syndrome is a severe variant of acute disseminated encephalomyelitis (ADEM) with an incidence of 0.4-0.5 per 1,00,000. It manifests with multiple neurological symptoms. It is characterized by a hemorrhagic inflammatory response in the central nervous system primarily affecting the white matter and myelin sheath, often sparing the periventricular white matter. Most cases are autoimmune in origin and others have prior history of viral illness and post vaccination status.

Design: Case report.

Patient: A 10 year old female child with history of dengue fever 1 week prior, presented to us with history of vomiting and altered mental status for 1 day, unresponsiveness for few hours. Her Glasgow Coma Scale was 3/15 on admission. The initial impression was acute meningo-encephalitis. Broad spectrum antibiotics and antiviral drugs were started. Dengue

NS1 was positive in serum, IgM was positive in serum and cerebro-spinal fluid. Patient developed dengue shock syndrome with hemiplegia and required inotropic support. MRI Brain revealed multifocal T2 hyper-intense lesions and hemorrhages consistent with acute hemorrhagic leuko-encephalitis.

Conclusion: In our patient, Intravenous Immunoglobulin was started on day 7 of admission. GCS improved to 10/15 on day 9 and child was successfully extubated on day 14. This disease is rare in children and hence early diagnosis and initiation of treatment increases the importance of its recognition from untreatable forms of encephalitis.

Keywords: ADEM, Glasgow Coma, Dengue NS1.

Introduction

Acute hemorrhagic leuko-encephalitis(AHLE) or Weston-Hurst's syndrome is a severe variant of acute disseminated encephalomyelitis (ADEM) with an incidence of 0.4-0.5 per 1,00,000. It manifests with multiple neurological symptoms. It is characterized by

a hemorrhagic inflammatory response in the central nervous system primarily affecting the white matter and myelin sheath, often sparing the periventricular white matter. Most cases are autoimmune in origin and others have prior history of viral illness and post vaccination status.

Case Report

A 10year old female child with dengue fever for 1 week, presented to us with history of vomiting and altered mental status for 1 day, unresponsiveness for few hours. The Glasgow Coma Scale on admission was 3/15 for which the child was intubated immediately. The initial impression was Acute Meningoencephalitis for which broad spectrum antibiotics and antiviral drugs were started.

On day 2, the child worsened requiring multiple inotropic support and had signs of persistently raised intra-cranial tension(ICT). Emergency CT scan was done, which showed no abnormality. The child had persistent high grade fever spikes unresponsive to higher antibiotics.

On day 7, the child developed hemiparesis of right upper limb and lower limb. Dengue NS1 was positive in serum, IgM was positive in serum and cerebro-spinal fluid(CSF). MRI Brain showed thrombosis of left sigmoid sinus and diffuse involvement of brainstem and subcortical region, suggestive of Acute Hemorrhagic Leuko-encephalitis.

On day 8, subcutaneous low molecular weight heparin(LMWH) at 1U/kg and intravenous immunoglobulin was started at 400mg/kg and given for 5 days. Sedation was tapered as child developed spontaneous eye opening, started responding to commands and fever spikes reduced. Ventilator Settings were reduced to minimal.

On day 14, the child was electively extubated and kept on high flow oxygen, Optiflow at 10L/min. Chest physiotherapy and Limb physiotherapy was continued and the child was started on full feeds via Ryles tube which was well tolerated.

On day 30, examination showed improvement in power of right upper limb from 0/5 to 2/5, left upper limb power continued to be 5/5. Child was discharged on high protein oral diet. Repeat MRI Brain was planned on follow up after 2 months to decide about the continuation of LMWH.

Child was brought for follow up after 1 month of discharge and had normal power in all four limbs with normal higher mental functions.

Brain - MRI



Figure 1: Suggestive of blooming in midbrain favoring hemorrhage in midbrain.

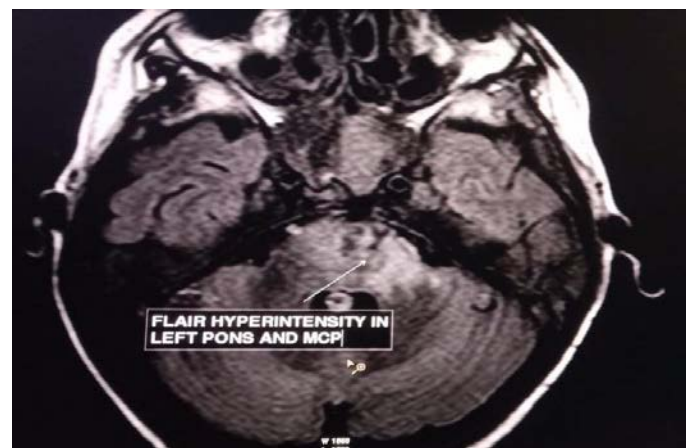


Figure 2: MRI FLAIR: Suggestive of hyper-intensity in left pons and middle cerebellar peduncle (MCP)



Figure 3: Suggestive of left sigmoid sinus thrombosis



Figure 4: Showing the normal basal ganglia and thalamus.

Discussion and Conclusion

In our patient, several findings including acute fulminant course of the disease, a past history of Dengue, CSF profiles, and MRI results, strongly supported the diagnosis of AHLE. Previous studies have shown that treatment with intravenous methyl-prednisolone, intra-venous immunoglobulin (IVIG), acyclovir, and plasmapheresis are helpful and life-saving. Following the same protocol, our patient improved with IVIG and was successfully extubated. This disease is rare in children and hence early diagnosis and initiation of treatment increases the importance of its recognition from untreatable forms of encephalitis.

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