Acute haemorrhagic encephalomyelitis following dengue infection

Anila Rao Vasireddy, Ami Mehul Mehta, Shubha Seshadri, Sharath P Madhyastha

DESCRIPTION
A woman in her 20s came to our hospital with fever since 12 days and altered sensorium since 4 days for which she was intubated in view of respiratory distress and Glasgow Coma Scale (GCS) of 7. On arrival, the patient was unconscious, pupils were equally reactive to light and vitals stable. Systemic examination revealed right-sided extensor plantar reflex.

Investigations revealed haemoglobin of 81 g/L (normal range: 130 - 160 g/L), total leucocyte count of 7.4 x 10⁹ /L (normal range: 4 - 11 x 10⁹ /L), neutrophil predominance, reactive forms, platelet count of 102 x 10⁹ /L (normal range: 150 - 400 x 10⁹ /L) and elevated liver enzymes. She tested positive for dengue NS1 antigen.

CT revealed two ill-defined intra-axial hypodensities in the left basifrontal region and right parietal region. Differentials include space-occupying lesions, metastasis or high-grade glioma, subacute infarct, haemorrhage, Japanese Encephalitis (JE) and Herpes Simplex Virus (HSV).

MRI was suggestive of multiple intra-axial altered signal intensity lesions involving the white matter of right parietal lobe, left basifrontal lobe and right temporal lobe (figure 1) indicative of acute haemorrhagic encephalomyelitis (AHEM).

The patient was treated supportively with intravenous antibiotics and steroids. In view of worsening sensorium, repeat MRI brain was done which showed increasing size of existing lesions. The patient had received one cycle of plasmapheresis and succumbed on day 5 of admission.

The neurological manifestations of dengue infection may be related to neurotrophic effect of the virus (encephalitis, meningitis, rhabdomyolysis), systemic complications of infection (encephalopathy, haemorrhagic or ischaemic stroke) or postinfectious reaction may be related to neurotrophic effect of the virus.1

ADEM is a neurological manifestation rarely described in association with dengue.2 AHEM is the most severe form of ADEM and is thought to be due to autoimmune cross reaction to the myelin antigens. It is characterised by an acute rapidly progressive fulminant inflammation of the white matter.

It is important to note that MRI is the gold standard for diagnosing this condition. Its features are large tumefactive lesions involving the white matter and sparing the cortex associated with punctate haemorrhages, extensive mass effect and surrounding oedema with possible involvement of ganglia and thalami. In our patient, the location and diffuse nature of the lesions on imaging were paramount in establishing the diagnosis.

Although histopathology is needed for a definitive diagnosis, the presence of focal instead of global lesions on MRI favours a diagnosis of encephalitis over encephalopathy. In cases of dengue, the most commonly affected brain structures include basal ganglia, thalami, brainstem, cerebellum, cortical grey matter, subcortical and deep white matter.3 Furthermore, tumefactive demyelination on biopsy is a rare presentation in ADEM. Since patients with tumefactive ADEM respond well to steroids, it is essential to positively confirm it.4

Since the lesions in AHEM tend to be deep sited, extreme precision is required to obtain a representative tissue sample and increase the chances

Figure 1 MRI brain axial view showing intra-axial altered signal intensity lesions involving the white matter of the right parietal lobe and left frontal lobe with the lesions appearing heterogeneously hyperintense on T2 Fluid-attenuated inversion recovery (FLAIR) (B), hypointense on T1W (A) images showing irregular incomplete ring enhancement on postcontrast images (C) with associated significant adjacent perilesional oedema. Mass effect is noted in the form of effacement of sulci and mild effacement of the lateral ventricles. The walls of the lesions are thick and irregular and there are multiple foci of peripheral areas of blooming on Susceptibility Weighted Imaging (SWI) (D), suggestive of haemorrhage.
Learning points

► Acute haemorrhagic encephalomyelitis is associated with a very poor prognosis. Initially, it has good response to steroids with subsequent new onset of symptoms and a devastating outcome.
► MRI is the gold standard and early diagnosis of this condition is paramount in limiting its progression and thus reducing mortality.
► In our patient, the location and diffuse nature of the lesions on imaging played a vital role in establishing the diagnosis.
► Safely obtaining a positive diagnosis to confirm acute disseminated encephalomyelitis is essential in order to identify and treat these patients.

of positive diagnosis. Thus, stereotactic-guided biopsy can be a potential investigation in the future for the diagnosis of AHEM. The largest neurosurgical series from Mallereau et al comparing different methods of biopsy (frame-based vs robotic vs frame-less neuronavigation) concluded that robotic surgery gives the highest chance of positive histological diagnosis (97.4%, with complication rate of 3.5% and mortality of 0.3%), thus further elucidating the promises robotic surgery hold for the future of medicine.5

Acknowledgements We would like to acknowledge the Department of Radiodiagnosis at Kasturba Medical College, Manipal, for providing the MR images.

REFERENCES