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Acute necrotizing encephalopathy in a young adult with EBV and COVID19 co-infection: A case report

Dr Sneha s
JSS Medical college

Dr Rudresh Hiremath
JSS Medical college

Dr Gurumurthy M
JSS Medical college

Dr Lakshmi N
JSS Medical college

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Title: Acute necrotizing encephalopathy in a young adult with EBV and COVID-19 co-infection: A case report

Clinical history: A 22yr old female came with complaints of fever and altered sensorium since 2 days. Serology showed positive for Epstein-Barr virus (EBV) and COVID-19 IgG antibodies. Laboratory investigations revealed elevated total counts & inflammatory markers.

Imaging:

MRI brain plain study was done for the patient using Philips Ingenia 3T MRI machine.

The study showed the following Imaging findings –

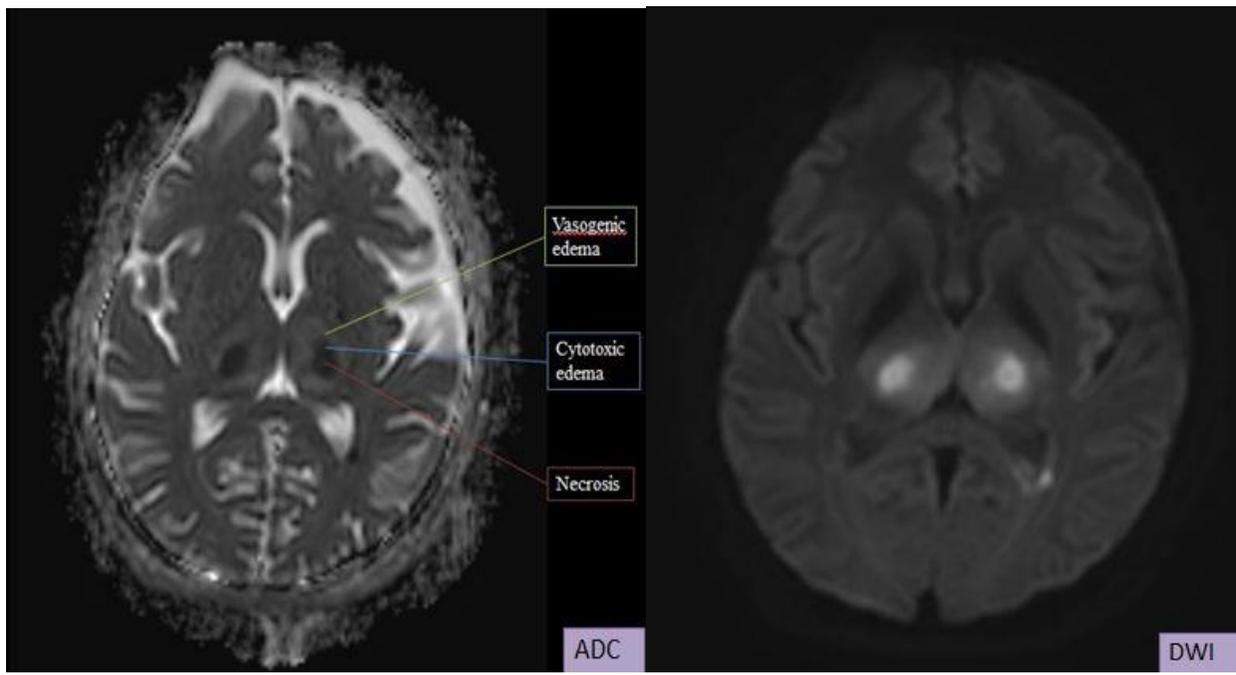


Figure 1 (a and b)

Figure 1(a and b) – DWI and ADC axial images shows higher ADC value in the central region of the bilateral thalami with a surrounding lower ADC ring (cytotoxic brain edema) and outer ring of relatively higher ADC (vasogenic edema) compared to normal brain tissue depicting the typical trilaminar or concentric/laminar structure or tricolor pattern or target-like appearance.

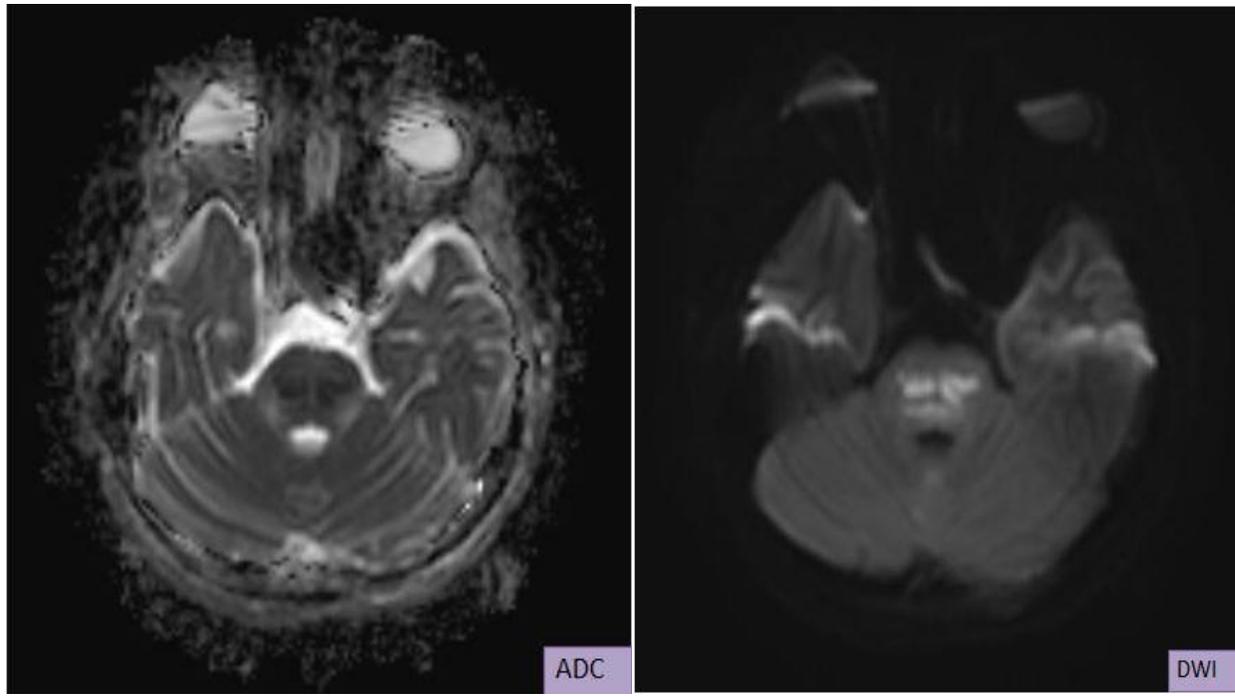
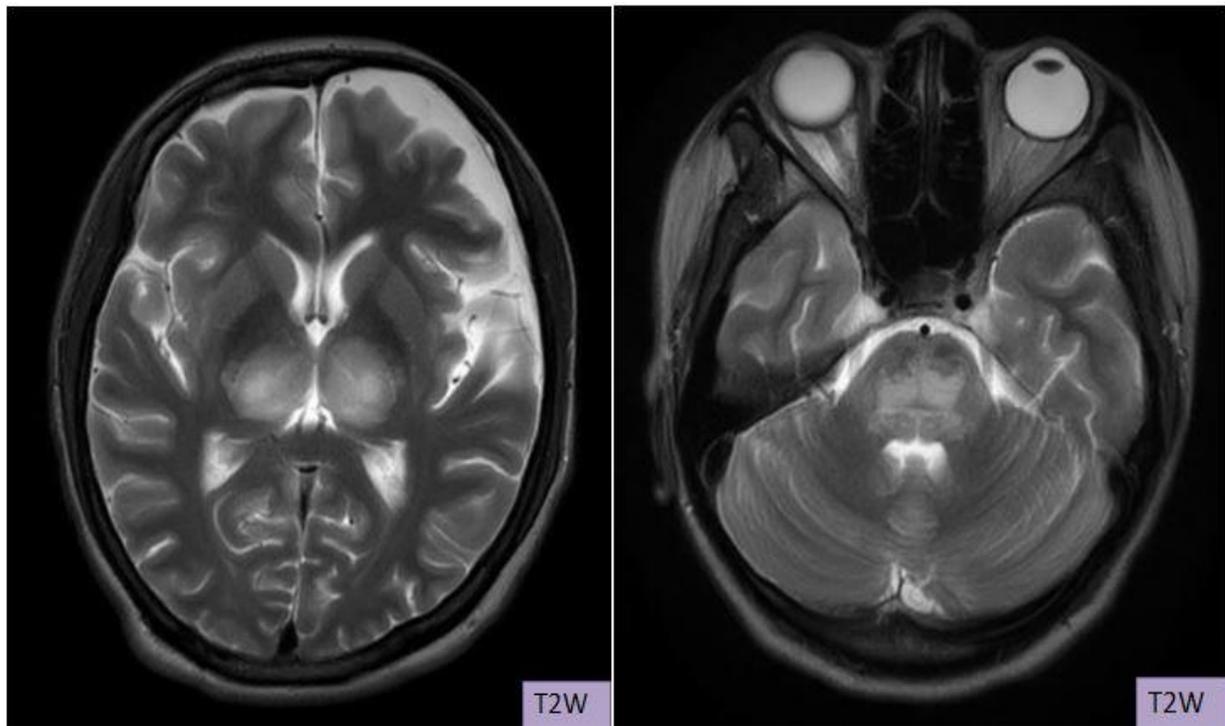


Figure 2 (a and b)

Figure 2 (a and b) - DWI and ADC axial images shows areas of diffusion restriction in the Pons.



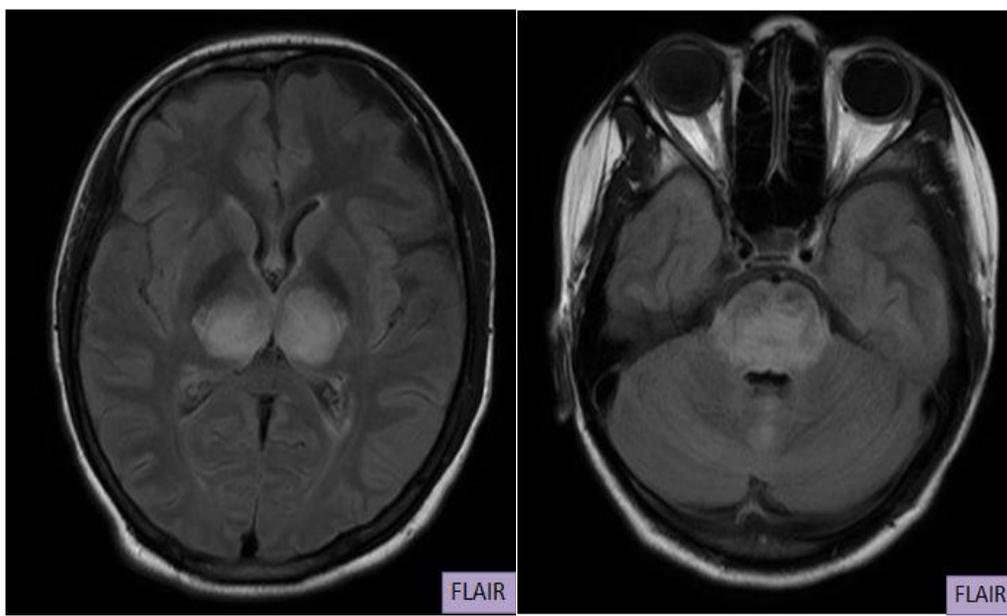


Figure 3 (a, b, c and b)

Figure 3(a, b, c &d) – T2W and FLAIR axial images shows symmetrical hyper intensities and swelling in bilateral thalami and pons. Incidental subdural hygroma noted along the left frontal convexity.

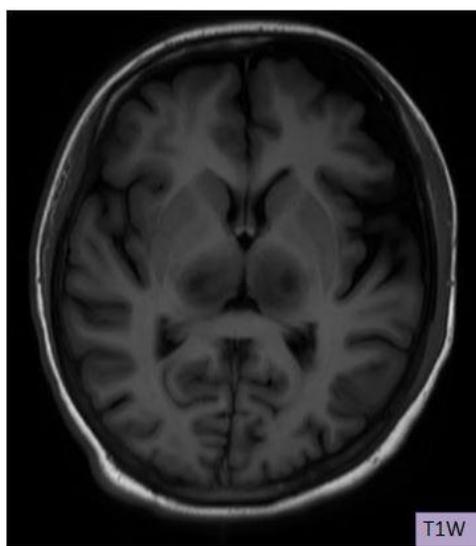


Figure 4

Figure 4– T1W axial image shows corresponding symmetrical hypo intensities in bilateral thalami.

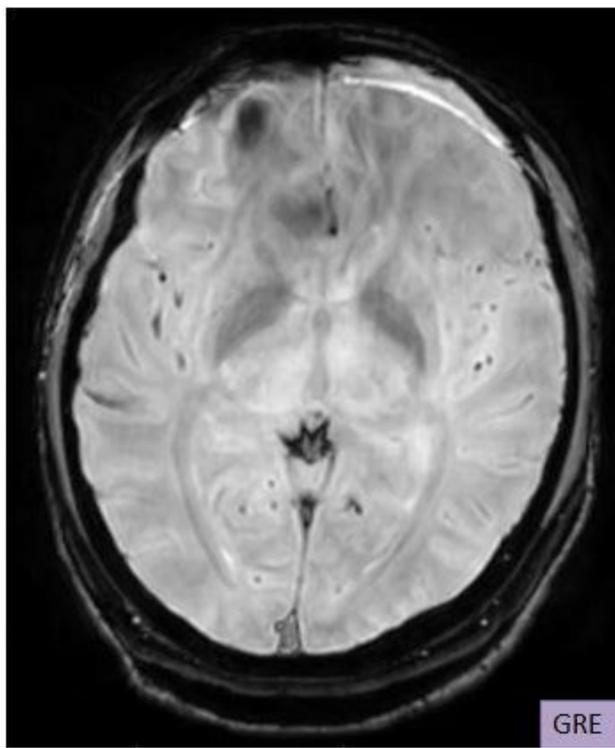


Figure 5

Figure 5 – GRE axial image shows no evidence of blooming suggestive of no hemorrhage in the bilateral thalami.

Differential diagnosis:

- Reye's syndrome.
- Metabolic disorders - Leigh encephalopathy, glutaricacidemia, methyl malonic aciduria, Wernicke encephalopathy.
- Acute disseminated encephalomyelitis.
- Acute hemorrhagic leukoencephalitis.

Final diagnosis: Acute necrotizing encephalopathy. Secondary to EBV and COVID- 19 viral co-infection.

Discussion:

Etiopathogenesis –

Currently the etio-pathogenesis of acute necrotizing encephalopathy (ANE) remains obscure. Prodromal viral infections have been reported as common causative agents. (1) The influenza virus and Human Herpes Virus – 6 (HHV-6) are the most common viral infections predisposing to ANE. Other viral infections include EBV, parainfluenza, varicella, enterovirus, rotavirus, herpes simplex virus, rubella, coxsackie A9, COVID-19 and measles.[1,2]

Though the pathogenesis is unclear, the most proclaimed hypothesis is the hypercytokinemia/cytokine storm. Cytokine storm results in brain injury through alteration of vessel wall permeability without vessel wall disruption. They also cause proteolytic destruction of the blood-brain barrier (BBB) which subsequently increases vascular permeability and causes brain edema, petechial hemorrhage, and necrosis. [1]

Apart from the above viral infections, diphtheria, tetanus toxoid, and whole-cell pertussis (DPTw) vaccination can also result in ANE. [1]

Imaging features–

- General features
 - Imaging hallmark of ANE - Multifocal, symmetric brain lesions involving both the gray matter and the white matter.
 - Characteristic Location – Bilateral thalami, brain stem, cerebral white matter, and cerebellum.
- CT findings – hypo densities in the above mentioned sites. Petechial hemorrhage appear as irregular hyperdense spots at the center
- MRI findings –
 - T1 – low signal in the above mentioned sites
 - T2 and FLAIR- high signal intensity in the above mentioned sites. When hemorrhage and necrosis are present, the lesions have mixed signal intensity.
 - DWI– Trilaminar or concentric/laminar structure or tricolor pattern or target-like appearance in bilateral thalami on ADC [as shown in the figure 1(a)] is characterized by:
 - Center of thalamic lesions characterized by hemorrhage and necrosis.
 - Periphery of the central thalamic lesions characterized by cytotoxic edema.

- Outer portions of the thalamic lesions suggesting vasogenic edema
- Gradient sequence- blooming represents hemorrhage
- Post contrast images - Occasionally faint ring enhancement may be noted around necrosis.[3]
- MR prognostic scoring system (0–4) - One point awarded for each of the following: presence of hemorrhage, cavitation, and location of lesions, including the brain stem and the white matter (cerebral, cerebellar, or both). As the thalami were involved in all patients, their involvement was not scored in terms of location of lesions. Based on the above scoring system, as the MR score increased the outcome of the patients worsened. The patients having the highest MR scores 3 or 4 were in the poor outcome category; whereas the patients with lower MR score 1 or 2 had better outcomes.[3, 4]

Therapeutic options-

There is no standardized protocol of management and treatment of ANE thus far. Medical management mainly remains supportive with some studies showing benefits with the use of antivirals, immunomodulatory agents and steroids. [1, 5] Therapeutic hypothermia and anticytokine therapy, has also been proved pivotal in the management. [1]

Teaching points –

- ❖ Imaging hallmark of ANE includes Multifocal, symmetric brain lesions involving bilateral thalami, brain stem, cerebral white matter, and cerebellum.
- ❖ Classical imaging sign - concentric/laminar structure” or “tricolor pattern” or target-like appearance in bilateral thalami on ADC.
- ❖ MRI helps in diagnosis and MR scoring system helps in predicting the prognosis of the disease.

References:

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