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Case Report

Acute Disseminated Encephalomyelitis

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Abstract

Acute Disseminated Encephalomyelitis (ADEM) is an inflammatory disorder of the CNS characterized by generalized demyelination, primarily of the white matter of the brain and spinal cord. The condition is usually triggered by a viral infection or vaccination. We report a case of ADEM following a cutaneous varicella infection. Children are preferentially affected.

Keywords: *Acute Disseminated; Encephalomyelitis; Children; MRI; Neuroimaging; Varicella-Zoster Virus*

Abbreviations

MRI: Magnetic Resonance Imaging; ADEM: Acute Disseminated Encephalomyelitis; CNS: Central Nervous System; MS: Multiple Sclerosis

Introduction

Acute Disseminated Encephalomyelitis (ADEM) is an inflammatory disorder of the CNS characterized by generalized demyelination, primarily of the white matter of the brain and spinal cord.

Case Report

A 7 years old girl, with no previous history, presents 1 month ago a skin rash and then status epilepticus with fever.

Discussion

It is the cause of various and variable neurological signs. It is generally monophasic but can be multiphasic or recurrent. In the absence of biological markers, the diagnosis is clinical and radiological. MRI is the best means of imaging, the T2 and flair sequences are the most sensitive. They show hyper-signal lesions in general globular asymmetric, unlimited. No segment of the CNS is spared [1]. The lesions generally affect the subcortical and central white matter, the cortical grey matter, the subcortical grey matter and the cortical grey-white junction of the two cerebral hemispheres, the cerebellum and the brainstem [2]. The gray matter of the thalami and the basal ganglia is frequently affected, generally asymmetrically, the corpus callosum is less affected. The large lesions can cross the contralateral hemisphere. There are multiple forms [3]:

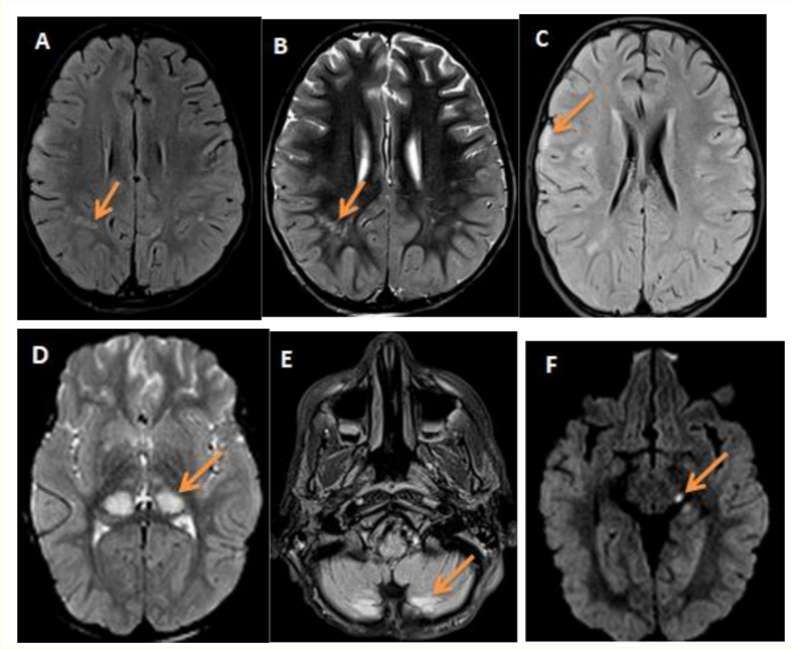

Figure: *Axial MRI sequences showing bilateral and asymmetric supratentorial and subtentorial nodular and patchy signal abnormalities, T2 and Flair hypersignal, of the subcortical white matter (FLAIR/ T2 sequence (A/B)) cortical (FLAIR sequence (C)), temporal, thalamic (diffusion sequence (D)) putaminal, cerebellar (FLAIR sequence (E)) and left mesencephalic (diffusion sequence (F)). These lesions have a diffusion restriction (diffusion sequence (D/F)). They are not enhanced after injection of Gadolinium.*

- ADEM with small lesions less than 5 mm.
- ADEM with large pseudotumor lesions with peri-lesional edema.
- ADEM with bi-thalamic involvement
- Acute hemorrhagic encephalomyelitis (AHEM)
- Isolated monofocal involvement of the brainstem.

The enhancement is very variable; it can be complete or incomplete (annular, nodular, gyral or spots). Meningeal enhancement is rare. Spinal cord involvement may be present, often in the thoracic region, with a large and swollen appearance. Spectroscopy may show an increase in lactate levels, a decrease in N-acetylaspartate (NAA), and some authors recommend at least two follow-up MRI scans to monitor the evolution. It should be noted that the SPECT scan allows a better follow-up of the evolution of the lesions than the MRI. Acute hemorrhagic leukoencephalitis. Acute hemorrhagic leukoencephalitis (AHL), AHEM, and acute necrotizing hemorrhagic leukoencephalitis (ANHLE) of Weston Hurst are variants of an acute, rapidly progressive, and frequently fulminant inflammatory hemorrhagic demyelination [4].

Differential diagnosis:

- Pediatric MS first especially in multiphasic forms [5].
- In ADEM with large lesions, it is done with brain tumors, Shilder's disease, Marburg variant of MS and brain abscesses.
- In ADEM with bi-thalamic involvement, it is done with deep cerebral venous thrombosis, hypernatremia, and extrapontic myelinolysis as well as in post-Japanese B vaccination encephalitis.
- In the basal ganglia; it is done with organic aciduria, poststreptococcal ADEM, or infantile bilateral striatal necrosis.

Treatment: No randomized treatment studies, the protocols are diverse and combine corticosteroids, immunoglobulins, cyclophosphamide and plasma exchange [3].

Conclusion

Epidemiologic evidence from this study suggests an infectious cause for ADEM. The agent is most likely a varicella-zoster virus. Magnetic resonance imaging was the neuroimaging study of choice for establishing the diagnosis and for following the course of the disease. Prognosis for survival and outcome was excellent.

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Conflict of Interest

The authors declare no conflict of interest.

Bibliography

1. Russell C Dale., *et al*. "Pediatric central nervous system inflammatory demyelination: acute disseminated encephalomyelitis, clinically isolated syndromes, neuromyelitis optica, and multiple sclerosis". *Current Opinion in Neurology* 22.3 (2009): 233-240.
2. Schwarz S., *et al*. "Acute disseminated encephalomyelitis: a follow-up study of 40 adult patients". *Neurology* 56 (2001): 1313-1318.
3. Silvia Tenembaum MD., *et al*. "Acute disseminated encephalomyelitis". *Neurology* (2007).
4. SN Krishna Murthy., *et al*. "Acute Disseminated Encephalomyelitis in Children". *Pediatrics* 110.2 (2002): e21.
5. Matilde Inglese., *et al*. "Magnetization Transfer and Diffusion Tensor MR Imaging of Acute Disseminated Encephalomyelitis". *American Journal of Neuroradiology* 23.2 (2002): 267-272.

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