FLAMES (FLAIR Hyperintense Lesions in Anti-Myelin Oligodendrocyte Glycoprotein associated Encephalitis With Seizures), a case report.

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INTRODUCTION

Myelin oligodendrocyte glycoprotein (MOG) is a myelin protein expressed at the outermost lamellae of the myelin sheaths in the central nervous system (CNS).

MOG antibody-associated disorder (MOGAD) is an inflammatory disease of the CNS which can present with acute disseminated encephalomyelitis (ADEM), optic neuritis, myelitis or aquaporin-4 (AQP4) seronegative neuromyelitis optica spectrum disorders (NMOSD).

FLAMES, an atypical subtype of the spectrum of MOGAD, is an acronym that stands for FLAIR - hyperintense Lesions in Anti - MOG associated Encephalitis with Seizures which was first described in 2017.

Clinically it presents with encephalitis and at least two of the following symptoms: seizures, headache, fever and cortical symptoms. The MRI scan identified cortical FLAIR-hyperintense lesions mainly unilateral, although bilateral cases have been described.

Lumbar puncture revealed that the patients also presented pleocytosis in the cerebrospinal fluid.

Clinical and radiographic improvement was seen in all the patients after the administration of intravenous methylprednisolone. However, a few had relapses when steroids were rapidly tapered or discontinued.

Keywords: MOG - Encephalitis - MRI - FLAIR - Seizures

OBJECTIVE

Describe a pediatric patient with diagnosis of FLAMES.

CASE REPORT

A 9-year-old male patient, previously healthy, presented with acute gastroenteritis with fever, headache, generalized tonic-clonic seizures, absences and acute sensory impairment. Physical examination did not show motor focus. He received empirical treatment with antibiotics and antivirals. Lumbar puncture was normal.

The brain Magnetic Resonance Imaging showed multiple hyperintense areas in sequences FLAIR/T2 with cortical involvement, of asymmetrical bihemispheric localization, without contrast enhancement. Spinal cord MRI was normal. In addition, an electroencephalogram (EEG) was performed which revealed slight disorganization of the base rhythm with generalized paroxysms of medium amplitude waves.

Antibodies Anti-MOG positive were obtained in the serum. The patient received five pulses of methylprednisolone with posterior tapering and monthly gammaglobulin for five months.

Due to seizure persistence, treatment with Rituximab was started with good response. At present, the patient is under treatment with valproic acid, levetiracetam and clobazam. Images remain stable.

CONCLUSION

FLAMES is a new entity which should be considered in patients presenting encephalitis, seizures, and characteristic lesions on FLAIR sequences, together with serum anti-MOG antibodies. Few cases have been reported in the literature, some with difficult-to-control epilepsy. Current treatment is based on clinical presentation. Further research is needed to establish guidelines ensuring appropriate management of these unusual cases.

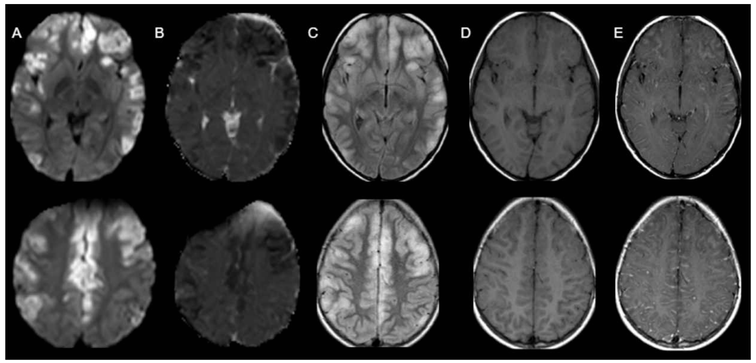

Figure 1. A: DWI. (Diffusion weighted images) Restricted bihemispheric cortical areas (bifrontal, temporo-insular and parietal) **B: ADC** (apparent diffusion coefficient) Hypointensity on ADC map of restrictive focal areas. **C: FLAIR.** Hyperintense focal lesions with exclusive cortical area involvement. Mild cortical thickening in affected areas**. D: T1 without gadolinium**. **E: T1 with gadolinium.** Contrast enhanced images showed leptomeningeal enhancement with left frontal predominance.

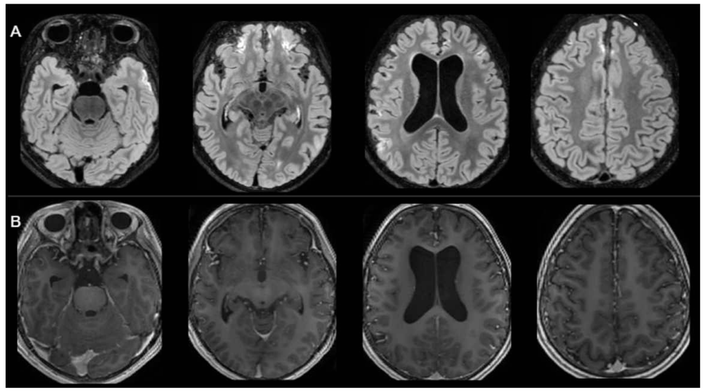

Figure 2. Control MRI. **A:** Vanishing of a few focal lesions, is observed, remaining some thinning and retraction cortical areas which present hyperintense signal in FLAIR in left temporal, frontal and right parietal lobes, possibly linked to gliosis or cortical laminar necrosis. **B:** Disappearance of leptomeningeal enhancement. Of note: ventricular volume increase with predominance of lateral ventricles, probably related to steroid therapy.

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