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Case Report

FLAIR-Hyperintense Lesions in Anti-MOG-Associated Encephalitis with Seizures: A Case Report

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Abstract

Introduction: Myelin oligodendrocytic glycoprotein antibody-associated disease (MOGAD) is an immune-mediated demyelinating disorder of the central nervous system. We described, a case of encephalitis associated with anti-MOG antibodies with hyperintense lesions on FLAIR and seizures (FLAMES).

Case Report: We report a case of a 5-year-old girl, with no significant personal or family history, who entered the emergency room with paroxysmal events and anarthria, in a febrile context. On neurological examination she presented right sided facial paresis and pyramidal signs on the right. Lumbar puncture (LP), showed pleocytosis, hyperproteinorrhaquia and the absence of oligoclonal bands. Electroencephalogram revealed encephalopathy and slow paroxysmal left frontal activity. The brain MRI detected hyperintense lesions, however, with an area of left frontal cortico-subcortical hyperintense signal on T2/FLAIR and diffusion restriction. Anti-MOG antibody was detected. These findings pointed to the diagnosis of FLAMES. She completed 7 days of methylprednisolone (30mg/kg/day), with a good response. On the day of discharge, a repeat EEG was conducted, demonstrating significant improvement compared to the initial one, with slightly slow wake and sleep patterns for her age, and no evidence of epileptic activity. She was assessed approximately four months post-hospitalisation and was found to be asymptomatic, with no focal neurological deficits. A further EEG was performed, which showed a structurally age-appropriate wake and sleep pattern with no abnormal graphoelements.

Conclusion: We highlight a rare clinical-radiological entity, which is still poorly described, particularly in the paediatric population, known as FLAMES, with a favourable response to steroids. Early recognition and timely diagnosis of his condition improve clinical and therapeutic management and minimize neurological damage. Immediate initiation of appropriate treatment is important for achieving a favorable outcome.

Keywords: *Seizures; Anti-MOG antibodies; Hyperintense lesions on FLAIR; Immune-mediated demyelinating disorder of the central nervous system.*

Introduction

The disease associated with myelin oligodendrocyte glycoprotein antibody (MOGAD) is a rare, inflammatory demyelinating disease of the central nervous system (CNS)[1,2]. Myelin oligodendrocyte glycoprotein (MOG) is a minor component expressed on the surface of oligodendrocytes, in the outermost part of myelin, and is a highly immunogenic component, making it a potential target for MOG antibodies [2,3]. Although its function is not fully understood, it is believed to play a role as a cell adhesion molecule and stability promoter, regulating and modulating myelin interactions [3]. There are several forms of presentation of this disease, including optic neuritis, myelitis, or acute disseminated encephalomyelitis[1,4]. Recently, a new clinical-radiological subentity was described, the encephalitis associated with anti-MOG antibodies, with hyperintense lesions in FLAIR and seizures (FLAMES). This entity was first described in 2017 by Ogawa et al and is characterized by hyperintense lesions in FLAIR MRI, in patients with encephalitis and seizures [4]. It can present with unilateral or bilateral encephalitis [1,5]. Initially reported as a benign entity, the pathophysiology of FLAMES is unclear and cases of severe relapses have been reported[1,6]. The most common clinical symptoms in addition to seizures are headaches, fever, and focal neurological deficits[3,5,7]. A case of a 5-year-old girl admitted for acute symptomatic seizures, fever, and altered consciousness, assumed to be the most likely diagnosis of FLAMES, is reported. Early recognition and timely treatment are essential for a favorable outcome.

Case Presentation

We report a case of a 5-year-old female child, with no significant personal or family history, admitted to the emergency room due to clonic seizures of the right hemiface, with ipsilateral oculocepahlic deviation and anarthria, accompanied by fever. Upon admission, she was conscious but uncooperative; neurological examination revealed right facial palsy, hyperreflexia (with Achilles clonus), and a Babinski sign in the right lower limb.

Given the initial suspicion of meningoencephalitis, a lumbar puncture was performed, revealing pleocytosis with mononuclear predominance, hyperproteinorrhaquia and the absence of oligoclonal bands. Treatment with acyclovir, ceftriaxone, and ciprofloxacin was initiated. The cerebrospinal fluid culture obtained from this lumbar puncture was negative. Further laboratory tests were performed, including the detection of anti- MOG antibodies, which revealed positive. Cranial MRI showed multiple hyperintense lesions in T2/FLAIR infra and supratentorial regions, suggestive of acute disseminated encephalomyelitis, however, with a hyperintense area in the left frontal cortico-subcortical region in T2/FLAIR and diffusion restriction (Figure 1).

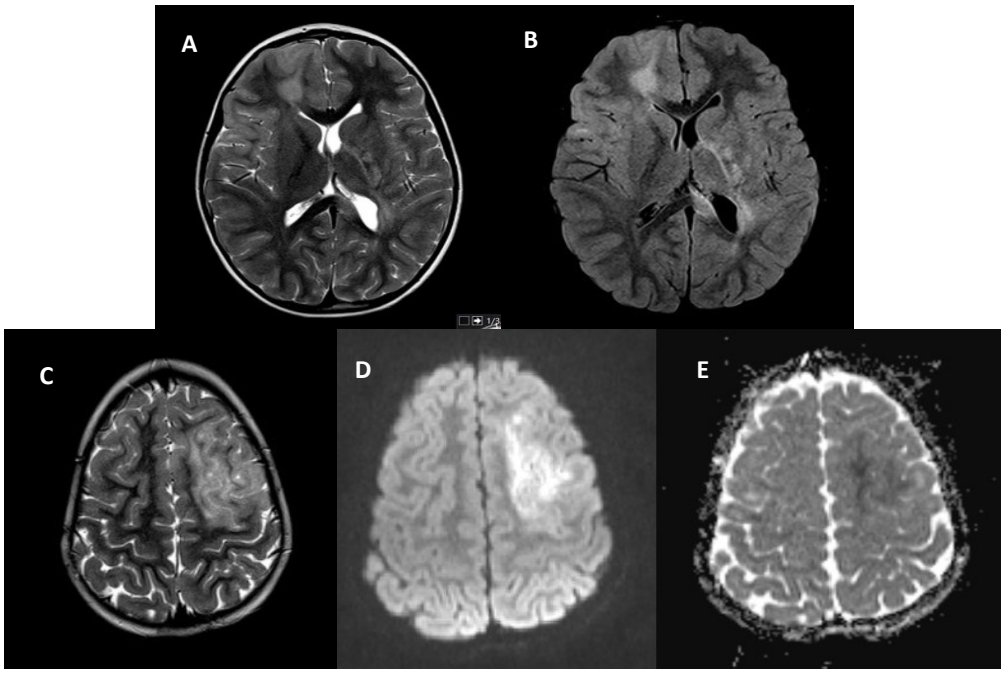

Figure 1. *In the axial T2WI (A) and FLAIR (B) sections, hyperintense lesions are observed at the level of the subcortical and periventricular frontal and frontobasal white matter; in the corpus callosum on the left, and striato-capsular on the left. In the axial T2WI sections (C) we can observe an area of hypersignal with restriction to diffusion which translates into a hypersignal on DWI (D) and hyposignal on ACD (E).*

Spinal MRI showed no abnormalities. The initial electroencephalogram (EEG) during hospitalization revealed encephalopathy and slow and paroxysmal activity in the left frontal region (Figure 2).

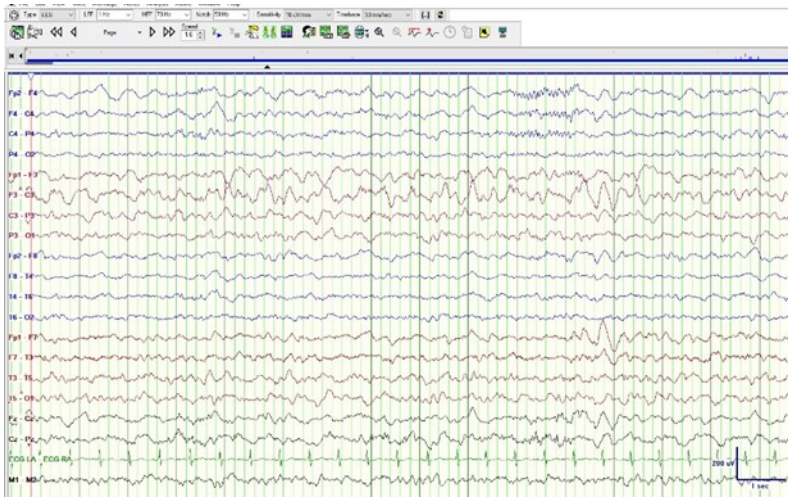

Figure 2. *Interictal sleep EEG in bipolar montage showing slow and paroxysmal left frontal activity*

Therefore, the diagnostic hypothesis of FLAMES was considered the most likely. Treatment was initiated with corticosteroid therapy – methylprednisolone at 30mg/kg/day and valproic acid at 300mg twice a day (27mg/Kg/day).

During the hospitalization, there was good progress with clinical improvement, completing a total of 7 days of treatment with methylprednisolone, later switching to prednisolone 1.3mg/kg/day, which was maintained at the time of discharge.

On the day of discharge, an EEG was repeated showing significant improvement compared to the initial one, with slightly slow wake and sleep patterns for age, without any record of epileptic activity (Figure 3).

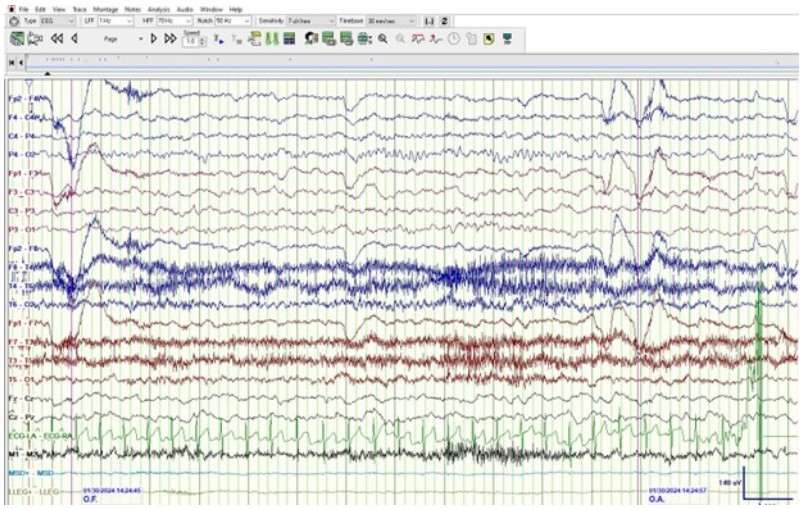

Figure 3. *Interictal awake EEG in bipolar montage showing dominant basal rhythm in alfa range.*

She was assessed approximately 4 months post-hospitalisation and was found to be asymptomatic with no focal neurological deficits. She repeated the EEG, which showed a structurally age-appropriate wake and sleep pattern with no abnormal graphoelements. She will begin tapering off corticosteroids and will also undergo a repeat MRI while continuing with regular follow-up appointments.

Discussion

We report a case of FLAMES in a 5-year-old child, an entity still poorly described particularly in the paediatric population[1]. Some authors report that this may be a disease with an indolent clinical course, with one of its initial manifestations being a decline in school performance in children[3,8]. However, in our case, this was not observed, given the more abrupt course of the condition. The main symptoms we found, consistent with those described by other authors, were seizures, headaches, fever, and cortical symptoms related to the location of the lesions[1,3,5,6,7]. CSF pleocytosis has also been reported in many studies, with a predominance of lymphocytes[1,4] . The detection of anti-MOG antibodies in the serum is important for establishing this diagnosis. The fact that its concentration in the cerebrospinal fluid is lower than in the serum makes the serum the best medium for its detection. The timing of the test is extremely important, as with the onset of remission, its titer usually decreases, and may even become undetectable [3,10]. In some studies, the presence of subcortical hyperintensities in FLAIR on MRI was considered an exclusion factor for this diagnosis. However, Maturu et al. suggest that this finding may be explained by the presence of post-ictal edema, as was the case in our situation[3]. As with the majority of cases of diseases within the MOGAD spectrum, there was a favorable outcome in this case, under treatment with methylprednisolone at a dose of 20-30mg/kg/day for 5-7 days [3,4,8,9]. To reduce the likelihood of relapse, described in various articles, it is important to gradually reduce this therapeutic regimen[3,4,8,9]. Recurrences, as in other disorders within the anti-MOG spectrum, although not common, can be associated with long-term sequelae, highlighting the importance of regular follow-up for these children[3,6].

Conclusions

We describe a case of scientific interest as it is a recently described clinical-radiological entity. As this is a new entity still poorly described in the literature, there are no evidence-based guidelines regarding its diagnosis and treatment. As described in the literature, this child had a favorable outcome, was discharged symptom-free, and so far, without apparent immediate sequelae. The low prevalence, varied clinical characteristics, and differences in presentation make recognizing this entity a challenge for clinicians. The method and timing of anti-MOG antibody detection are crucial.

Early recognition and timely diagnosis of these entities improve clinical and therapeutic management and minimize neurological damage. Immediate initiation of appropriate treatment is important for a favorable outcome.

Conflicts of Interest

The authors declare no conflict of interest.

Acknowledgement

None

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