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Mini Case Study

WEBINO syndrome as a Clinical Presentation of the Fisher - Bickerstaff Overlapped with Guillain Barre syndrome

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Abstract

The Miller Fisher syndrome (MFS) and the Bickerstaff Brainstem Encephalitis (BBE) are clinical variants of the Guillain Barré syndrome (GBS), in which some manifestations correspond to an involvement of the CNS.

We present the case of a 48-year-old male patient with a diarrhea history who was admitted because of horizontal oscillopsia and diplopia. The physical exam revealed a bilateral modification of the horizontal conjugate gaze, compatible with the WEBINO syndrome and hyperreflexia. During progress, presented hyperreflexia and a bilateral involvement of the oculomotor nerves, associated with hyporeflexia and quadriparesis. We diagnosed the BBE-MFS syndrome overlapped with GBS. The imaging, electromyography, GQ1b, and LCR did not show modifications.

Conclusions: The presented case is quite peculiar and unusual since it shows that the WEBINO syndrome can be an atypical central manifestation of the BBE-MFS and GBS spectrum.

***Keywords**: WEBINO syndrome; Clinical; GBS; CNS; Bickerstaff*

Introduction

The Guillain Barré syndrome (GBS) is the most frequent cause of acute flaccid paralysis that affects the peripheral nervous system (PNS). However, the range of clinical variants is very broad, and there can even be an involvement of the central nervous system (CNS) [1-3].

The WEBINO syndrome (Wall-eyed bilateral internuclear ophthalmoplegia) is an ocular motility disorder caused by a lesion of the bilateral medial longitudinal fasciculus (MLF), which clearly involves the CNS. However, it could occur in the context of the GBS [4].

The Miller Fisher syndrome and the Bickerstaff encephalitis are clinical variants of the Guillain Barré syndrome. It is accepted that some manifestations, such as hypersomnolence, hyperreflexia, Babinski, or hemianesthesia are consequences of the CNS involvement; therefore, the possibility of other unusual manifestations should be taken into account [5-8].

We present the case of a patient who started with the WEBINO syndrome and later developed BBE-MFS overlapped with GBS.

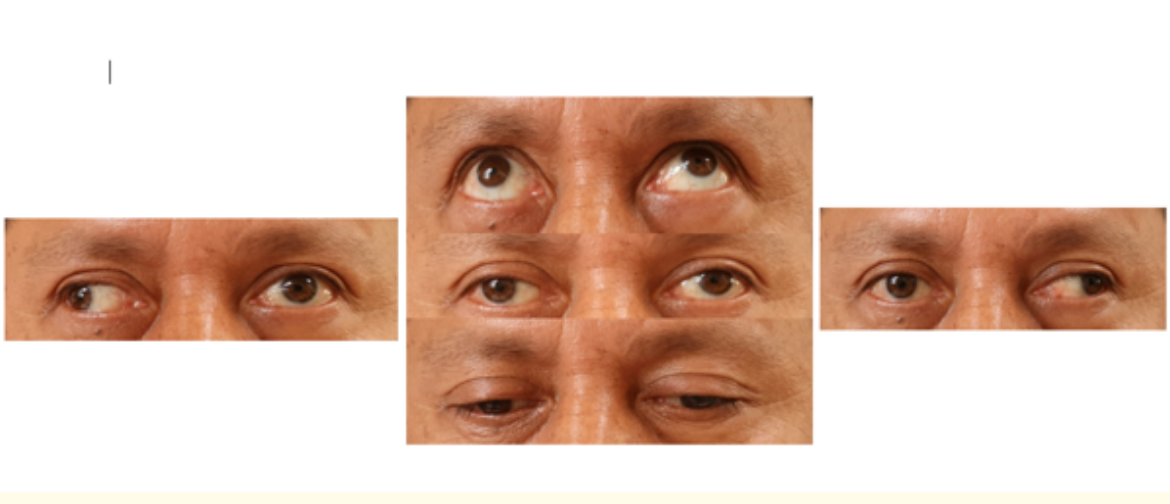
Clinical Case

A 48-year-old man, who can drive and who has no relevant pathological history, started a clinical picture with episodes of temporary oscillopsia and diplopia that lasted for about 10 minutes. Seven days later, it progressed to persistent horizontal diplopia and a decreased muscle strength of the four limbs, which made sanding and gait impossible. That is why he went to the ER.

He alleged having diarrhea for the five previous days.

Emergency physical exam

The patient is awake, lucid, with isochoric pupils which are reactive to light; primary gaze with exotropia on the left eye and alteration of the conjugate gaze with adduction deficit plus nystagmus in abduction of both eyes (Appendix 1). The muscular strength assessment showed weakness in the four limbs with the Medical Research Council (MRC) scale of 5/5 proximal, 4/5 distal in the upper right limb, and 5/5 proximal, 2/5 distal in the upper left limb. In the same way, 4/5 proximal and 3/5 distal in both lower limbs. Preserved sensitivity. Globally increased osteotendinous reflexes and indifferent plantar reflexes. Hughes disability scale: 4.



Appendix 1

Progress

Two days after hospitalization, he developed bilateral ptosis with predominance on the right side, alteration of the ocular movements dependent on the III, IV, and VI bilateral cranial nerve in addition to worsening of the muscular strength of the limbs, and bilateral patellar and Achilles hyporeflexia. That is why we decided to administer IV immunoglobulin 5% (0.4g/kg/day) for 5 days.

On the third day of treatment, the patient showed a partial improvement of the ocular movements and muscular strength and the case was resolved entirely eight days after finishing the treatment with immunoglobulin.

The lab tests performed during the admission showed

Cerebrospinal fluid (CSF): proteins: 41,1 mg%, cells: 0 per field. Anti-GQ1b serum: negative, vitamin B12 165 pg/ml.

Brain Magnetic Resonance Imaging with contrast

Normal.

Electromyography

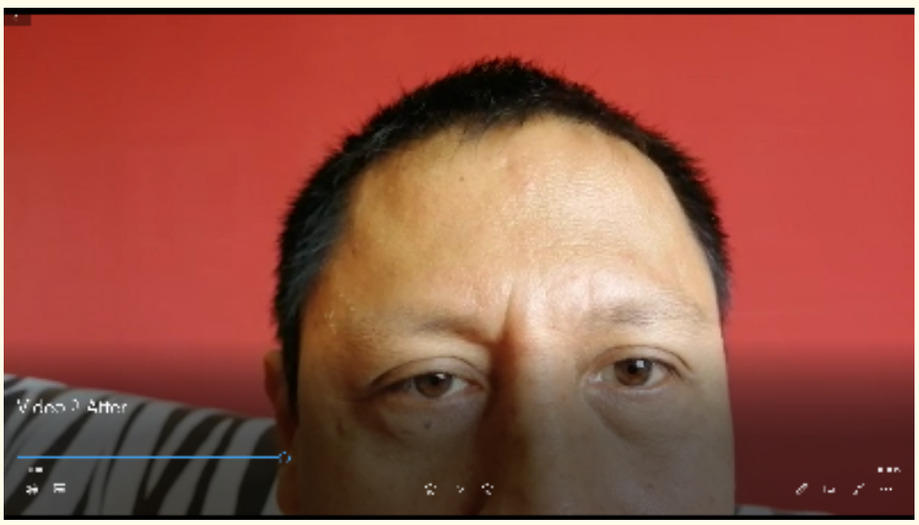
Normal.

Discussion and Conclusion

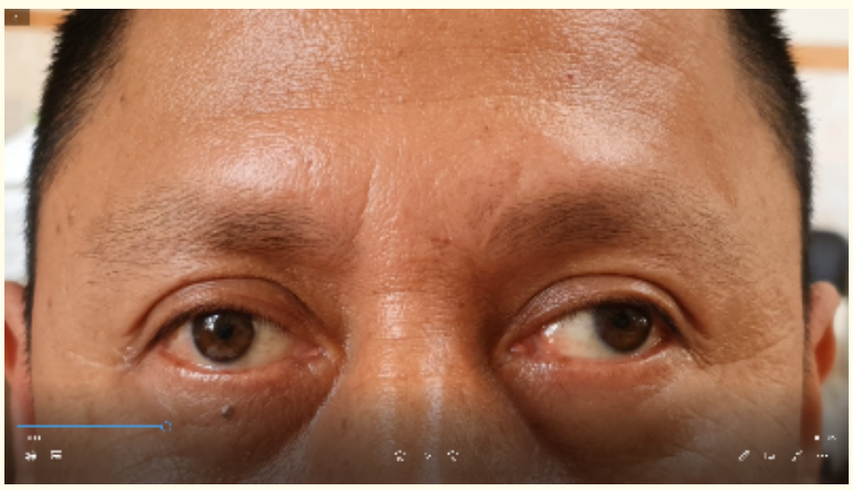
We present the case of a 48-year-old male patient who started with the WEBINO Syndrome (video 1,2) and hyperreflexia. His subsequent progress allowed us to define the diagnosis of BBE-MFS overlapped with GBS.

Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) is a rarely reported syndrome. It clinically occurs along with exotropia in primary position of one of the eyeballs, with a bilateral adduction deficit and horizontal nystagmus on abduction, as in the case of our patient.

Caused by a lesion of the bilateral MLF, the most frequently reported etiologies are autoimmune: multiple sclerosis and neuromyelitis



Video 1



Video 2

optica, spectrum disorder cerebrovascular disease, brain stem tumors, among others. Even though there is no available bibliography that describes the syndrome associated with BBE/MFS, we believe that its association is possible [9].

MFS and BBE are clinical variants of the GBS, and their characteristics suggest an involvement of the CNS. Both can be related to the presence of anti-GQ1b antibodies. However, there are reported cases in which anti-GM1 antibodies were found instead of the latter. Ophthalmoplegia and ataxia are common features in both variants, and the added presence of hyporeflexia or areflexia suggests MFS, while the presence of hyperreflexia and hypersomnolence strongly suggests BBE. Although we can sometimes find incomplete forms and more, we can consider these variants as a spectrum of the Fisher - Bickerstaff syndrome [5,10,11].

The atypical forms and overlaps of MFS, BBE, and GBS, as in our case, can be hard to diagnose and this can imply a challenge, but they should always be considered in the context of an infectious and single-phase evolution history; most of these variants are exceptionally rare. The diagnosis can be supported by the presence of albuminocytologic dissociation in the CSF and alterations in the nerve conduction study; however, its absence does not prevent diagnosis, as in our case. The treatment is based on the correction of the autoimmune disorder, and to do that we use intravenous human immunoglobulin or plasmapheresis, obtaining a favorable response as the one observed in our patient.

It is important to note that our patient started with the WEBINO syndrome and hyperreflexia, which suggested an involvement of the CNS. However, the brain MRI was normal, during progress, there was an involvement of the III, IV, and VI bilateral peripheral oculomotor nerves, a progressive decrease in muscular strength and hyporeflexia. This allowed us to conclude that the manifestations matched a BBE that later evolved to MFS overlapped with GBS, and this is not reported in medical literature. For this reason, we should consider this form of unusual presentation to make an early diagnosis and offer a timely treatment, improving the patient's prognosis.

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