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Case Report

Bickerstaff brainstem encephalitis: A case report☆,☆☆

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ABSTRACT

Bickerstaff brainstem encephalitis (BBE) is a rare inflammatory, demyelinating disease that generally has a good prognosis. It's characterized by an acute dysfunction of brainstem occurring few days after an infection. We report the case of an 11-year-old male child with a history of cold, presented with ataxia in whom a Bickerstaff encephalitis was attested through brain MRI and who has fully recovered after treatment. The main symptoms are ataxia, ophthalmoplegia, and altered consciousness. CSF analysis and serum antiganglioside antibodies are also very suggestive of the diagnosis that can be suspected clinically and confirmed on brain MRI. The interest of this observation lies in its rarity and on the rapid and spectacular clinical improvement under treatment.

Introduction

Bickerstaff brainstem encephalitis (BBE), first described in 1957 by Bickerstaff et al. [1], is a very rare entity that presents several clinical and immunological similarities with Guillain-Barré syndrome (GBS) and Miller-Fisher syndrome (MFS) [2]. This autoimmune encephalitis is characterized by an acute brainstem dysfunction occurring few days after an infection or a vaccination. The aim of this work is to show the contribution of MRI in the diagnosis of this pathology [3].

Case report

We report the case of an 11-year-old male child, born at term by vaginal delivery, from a nonconsanguineous marriage.

He presented to the neurology department of our hospital with a subacute onset of incoordination of movements, gait disorders, loss of balance and muscle weakness.

One week before his admission, the patient had presented with an influenza-like syndrome that resolved after symptomatic treatment.

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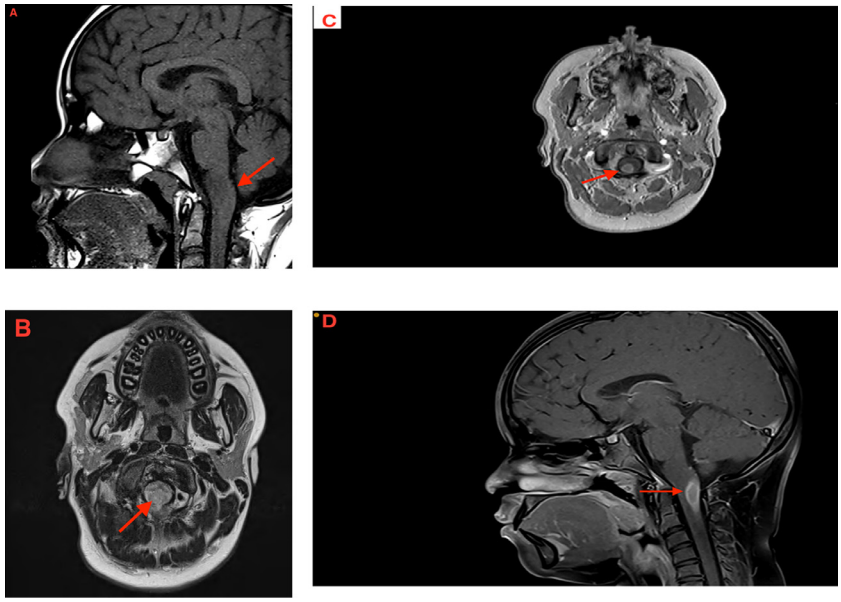

Fig. 1 – Right posterolateral lesion with intermediate T2 signal, T1 asignal, with homogeneous enhancement after gadolinium injection (A) sagittal T1 sequence; (B) axial T2 sequence; (C and D) axial and sagittal T1 injections.

On clinical examination, the child was obese with a weight of 52 kg. Ataxia was observed with impossible standing. The neurological examination showed a posterior cord syndrome with a positive Lhermitte's sign.

A lumbar puncture was performed, revealing lymphocytic pleocytosis, hyperproteinorachia with negative direct examination and cultures.

The child had a brain MRI with T1-weighted, T2-weighted, Flair, diffusion and gadolinium injection sequences, which showed a brainstem lesion with intermediate T2 signal, T1 asignal, with homogeneous enhancement more marked in the periphery after injection (Fig. 1A-D).

The clinical improvement was spectacular under corticotherapy with resumption of normal activity after a few days.

In view of this picture of acute dysfunction of the brainstem, especially after an infectious flu syndrome, the MRI appearance and the favorable evolution under corticosteroid therapy, the diagnosis of Bickerstaff encephalitis was retained.

Discussion

Bickerstaff's encephalitis is an acute demyelinating pathology, affecting the brainstem, and occurring few days after an infectious episode. Although the exact pathogenesis remains unclear, it is presumably related to an immune reaction, triggered by a previous infection from pathogens like *Campylobacter jejuni, Mycoplasma pneumonia*, or *Haemophilus influenzae* [4]. In 2020, a case of Bickerstaff encephalitis was reported occurring after an infection with SARS-COV2 in a 72-year-old woman [5].

Guillain-Barré syndrome (GBS), Miller-Fisher syndrome, and BBE have some similarities, including the presence of antiganglioside antibodies. Together with GBS and Miller-Fisher syndrome, these 3 syndromes form a spectrum of postinfectious demyelinating diseases [4]. Odaka et al. in 2003 suggested that the 3 syndromes are different phenotypes of the same immunological condition. Hamaguchi et al. [6] evoked an anti-GQ1b syndrome by an exemplary clinical case of a patient who over a period of nine years presents an episode of MFS, followed by ophthalmoplegia interna, and finally an episode of EB.

There is no specific biological marker for the BBE and the diagnosis is based on a combination of anamnestic, clinical, and radiological features [7,8]. The clinical symptoms typically associate ophthalmoplegia, ataxia, and confusion occurring after an infectious episode. In 2003, Odaka et al. established the characteristics of this pathology through the study of 62 patients. During their evolution, they all presented an external ophthalmoplegia with ataxia, 74% presented consciousness disorders, 60% a weakness of the belts, 58% a decrease or even an abolition of the ROT, 40% had a positive Babinski, and 38% had diffused ROT [9].

Lumbar puncture is systematically performed to discard infectious meningitis requiring specific antibiotic treatment. CSF analysis shows lymphocytic pleocytosis in 40% of cases with hyperproteinoarrachy in 59% of cases and albuminocytological dissociation in 19% of cases [8,10].

Serum antiganglioside antibodies (anti-GQ1b) are found in two-thirds of patients. However, the absence of these antibodies does not rule out the diagnosis [7,8,11].

The clinical and biological features suggest the diagnosis, which will be verified by imaging. In 30% of cases, brain MRI will show areas of T2 and FLAIR hypersignal in the brainstem, cerebellum or thalamus. Contrast enhancement is inconstant. MRI with injection is performed at the acute stage and during recovery. In the meantime, FLAIR sequences can be performed for the follow-up [7,10]. Interestingly, 66% of patients that underwent a spine MRI showed abnormal findings, such

as hyperintensity and/or contrast enhancement of the cauda equina, the nerve roots and the conus medullarismi [1]. In 2007, Debruxelle et al. [12] reported for the first time a case of Bickerstaff encephalitis revealed by a ventriculitis aspect on MRI in a 74-year-old man.

Further investigations of these patients can include electrophysiological studies. EEG studies may show slow wave activity in the h to d range. Electromyography may reveal decreased motor nerve conduction studies, prolonged distal latency, reduced compound muscle action potential and absent or prolonged F-waves [13].

Most patients can be managed by immunotherapy, using plasmapheresis or intravenous immunoglobulin either singly or in combination. Those patients who fulfill the diagnostic criteria of BBE usually have a good prognosis. Optimal effective treatment has not been established. Intravenous immunoglobulin has proved as effective as plasma exchange, while combined treatment with methylprednisolone and intravenous immunoglobulin is controversial [14,15].

Conclusion

Bickerstaff encephalitis is a disposal diagnosis that must be considered in front of an acute dysfunction of the brain stem, especially after an infectious syndrome with no other evident etiologies. Brain MRI is a key element in the diagnostic strategy. The clinical improvement is sometimes spectacular after treatment, making it important not to ignore this diagnosis.

Patient consent

Informed consent for patient information to be published in this article was obtained.

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