Bickerstaff Brainstem Encephalitis with extensive medullary involvement. An unusual stroke mimic.

Cebrián. J, Muro. I, Trillo. S, Díaz Pérez. C, Ramos. C, de la Fuente. E, Nombela. F, Vivancos. J.

Stroke Unit. Neurology Department. Instituto de investigación Sanitaria Princesa. Hospital Universitario de la Princesa. Madrid.

BACKGROUND AND AIMS:

Bickerstaff brainstem encephalitis (BBE) Is a rare disease included in the clinical and immunological spectrum of the Miller-Fisher syndrome. It has an autoimmune etiology usually related to anti-GQ1b antibodies

METHOD:.

We report a case of BBE mimicking acute stroke that received early immunosuppressant treatment.

CASE REPORT

<u>24-year-old male</u> patient with relevant medical history of sporadic ketamine consumption.

SYMPTOMPS:

- Wake-up symptoms: lower <u>facial paralysis</u>, <u>dysarthria and impaired</u> <u>consciousness</u>
- Gastrointestinal infection one week before.

EXAMINATION:

- Horizontal diplopia + VI CN palsy and multidirectional nistagmus.
- Supranuclear left facial palsy
- Absent gag réflex. Left palatal palsy.
- Bilateral upper dysmetria.
- Preserved strenght. Myotatic reflexes were globally increased with Bilateral clonus



URGENT CT

Multiparametric-CT: normal.

Acute reperfusion treatment is witheld as the time of symptoms-onset is not known and in absence of any artery oclussion.

Lumbar Puncture is performed (Suspected Inflamatory/infectious rhombencephalitis) → **CSF** had normal values.



ADMISSION AT THE STROKE UNIT

- I. V Aciclovir and I.V Ampicilina are started
- **Esteroid bolus is administered** (soon after normal urgent CSF normal results -leucocyte recount, biochemistry values and Gram stain-).



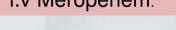
ICU ADMISSION

First day of admission: Clinical worsening

- Urgent MRI: Extensive medullary involvement.
- ICU admission



- Poorly secretion management and compromised airway.
- Orotracheal intubation was performed
- **Urgent** Orotracheal intubation → Left lung atelectasis + pneum
- 5-day immunoglobulin treatment is indicated



ICU complications:

- Tracheostomy and nasogastric nutrition.
- <u>Aspirative Pneuomonia</u> (*S. Pyogenes*) treated with i.v Meropenem.



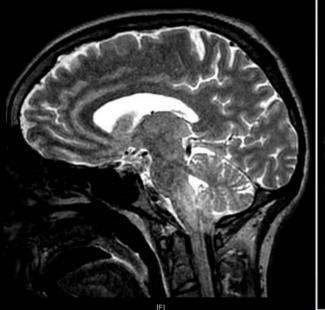
NEUROLOGY HOSPITALIZATION

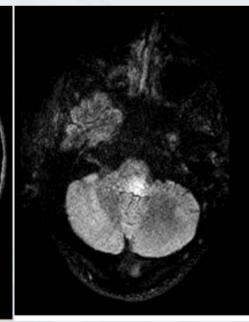
29-day admission:

- Left upper limb mild dysmetria
- Independent wide-base gait
- Positive CSF antiGM2-IgM antibodies.

COMPLEMENTARY EXAMS

- Multiparametric-CT: normal.
- **Lumbar Puncture**: Cell recount, Biochemistry normal. Negative viral and bacterial determinations.
- **Electroneurography**/**electromyography** did not show any alteration.
- Urgent brain MRI: Extensive medullary involvement on T2-weigth images without contrast enhancemet.
- Blood analysis and serologies: Negative.
- Positive antiGM2-IgM antibodies





CONCLUSION:

- This case report highlights the importance of an <u>early</u> <u>diagnosis</u> of stroke mimics, which can lead to a fast clinical worsening.
- <u>Immunosuppressant treatment and compromised</u>
 <u>airway awareness is essentia</u>l if BBE is suspected. Our patient had a good clinical outcome in spite of showing extensive medullary involvement
- The current evidence of BBE mediated by <u>antiGM2-antibody</u> is scarce and dysimmune neuropathies have been more frequently reported.

BIBLIOGRAPHY:

- Y. Fukami et al. Anti-GQ1b antibody syndrome: anti-ganglioside complex reactivity determines clinical spectrun European J Neurol. 2016, 23:320-326.

- Odaka M, et al. Bickerstaff's brainstem encephalitis: clinical features of 62 cases and a subgroup associated with

Guillain-Barré syndrome. *Brain*. 2003;126(Pt 10):2279-90.

- B.Bisel et al. GM1 and GM2 gangliosides: recent developments. *BioMol Concepts*. 2014;5(1):87-93









