



CENCIA E INNOVACIÓN EN
SALUD

Unusual clinical presentation of Guillain Barre syndrome: a case report

Presentación clínica aguda inusual del síndrome de Guillain Barre: A propósito de un caso

Montaño - Lozada, JM¹, Licona, Erick.¹, Marengo Gómez, Aristides², Espejo - Zapata, LM³, Montoya-Jaramillo Mario, Herrera, Felipe⁴

¹Universidad del Sinú, Cartagena, Colombia,

²Universidad Metropolitana, Barranquilla, Colombia.,

³Unidad Central del Valle del Cauca, Tuluá, Colombia.,

⁴Clínica Cartagena del mar, Cartagena, Colombia

Submitted 13 Jun 2018

Accepted 19 Jun 2018

Published 06 Dec 2018

Editor in chief

Isaac Kuzmar
editor@revcis.com

Academic editor

Vladimir Ospino

Corresponding author

Montaño - Lozada, JM,
Juanma2499@hotmail.com

DOI 10.17081/innosa.60

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ABSTRACT

Background: Guillen barre syndrome is the most frequent cause of flaccid paralysis in the world, it is characterized by an acute demyelinating, autoimmune and multiple etiology polyneuropathy, among which are included infectious agents such as Campylobacter jejuni, Zika virus, of genetic and environmental factors.

Case Report: We present the case of a 56-year-old Colombian male patient with a history of hypertension, who entered the intensive care unit with symptoms of atypical asymmetric motor neurological compromise, which rapidly progressed to ventilatory failure and subsequent confinement syndrome.

Discussion: Guillen barre síndrome establishes a potentially fatal disease, the semiology of pain, paresthesia, symmetric-progressive, distal weakness, instability, hipo/areflexia, constitute a neurological emergency. There are Clinical variants establishing a great.

RESUMEN

Introducción: El síndrome de guillan barré es la causa más frecuente de parálisis flácida en todo el mundo, se caracteriza por ser una polineuropatía aguda desmielinizante, autoinmune y de etiología múltiple, entre los que se incluyen agentes infecciosos como el campilobacter jejuni, virus del zika, además de factores genéticos y ambientales.

Caso clínico Se presenta el caso de un paciente masculino de 56 años, colombiano, con antecedentes de hipertensión arterial, quien ingresa a la unidad de cuidados intensivos con síntomas de compromiso neurológico motor asimétrico atípico, el cual progresó rápidamente a falla ventilatoria y posterior síndrome de enclaustramiento.

Discusión: El síndrome de guillan barré establece una enfermedad potencialmente mortal, la semiología de dolor, parestesia, debilidad distal simétrica-progresiva, inestabilidad e hipo/arreflexia, constituyen una emergencia neurológica. Existen variantes clínicas estableciendo un gran reto diagnóstico, teniendo en cuenta que el tratamiento oportuno podría tener relación directa con el pronóstico de la enfermedad.

Keywords Guillain-Barre Syndrome; Inflammatory Demyelinating Polyradiculoneuropathy,; Acute Inflammatory Polyneuropathies

Palabras clave Síndrome de Guillain-Barré; Polirradiculoneuropatía Desmielinizante Inflamator; Polineuropatía Inflamatoria Aguda

I. BACKGROUND

Guillain Barré syndrome (GBS), described in 1916 by Charles Guillain (*Kusunoki, 2015*), is currently the most frequent cause of flaccid paralysis in the world and constitutes a neurological emergency (*Nayak, 2017*). It is related to autoimmune response characterized by progressive paralysis of the extremities, acute areflexia and cytosolic albuminous dissociation (CAD) in cerebro-spinal fluid (CSF) (*Doctor, Alexander, Radunovic, 2018*), eventually preceded by respiratory tract infections (58%), gastrointestinal (22%) (*De Wals, Deceuninck, Toth, Boulianne, Brunet, Boucher, Landry, De Serres, 2012*) even certain vaccines (*Sejvar, Baughman, Wise, Morgan, 2011*). The GBS incidence is between 0.8-1.9 cases/100,000 inhabitants (*Van den Berg, Bunschoten, van Doorn, Jacobs, 2013*) Mortality can reach up to 5% of cases, despite treatment with immunotherapy or plasmapheresis (*Yuki, H.-P., 2012*) within the forms of expression of GBS are described clinically variants associated with anti-ganglioside antibodies (GM1, GD1a, GT1a, GQ1b) including: Acute inflammatory demyelinating polyneuropathy and its facial variant (diplegia and facial paresthesia), acute motor axonal neuropathy and its extensive form (acute motor sensory axonal neuropathy, multifocal neuropathies due to acute conduction block), pharynx-cervical-brachial variant, Miller Fisher syndrome, its incomplete form (acute ophthalmoparesis without ataxia), acute ataxic neuropathy (without ophthalmoplegia) and an even more rare variant, Bickerstaff's brainstem (*Etzeberria, Lonneville, Rutgers, Gille, 2012*).

II CASE REPORT

A 56-year-old male patient, Colombian, with a history of high blood pressure, who was admitted to the emergency department due to clinical symptoms of 1 hour of evolution characterized by paresthesia's in the left side of the face with deviation of the labial commissure to the right, weakness of the left cerebral brachium of progressive behavior; as related, she reported an episode 15 days prior of liquid diarrhea without signs of dysentery and uncalcified fever, symptoms that had spontaneous resolution without drug treatment. Upon physical examination, there was an increase in blood pressure, dyspnea, afebrile, alertness, oriented, with judgmental and conserved reasoning, fluent, coherent language, adequate executive function, cranial nerves: with clinical evidence of paralysis of the left seventh cranial nerve, Bell sign positive score House Brackmann: IV / VI. Not finding other alterations of the cranial pairs. Dynamic taxi with evidence of a Parética march requiring support, score Daniels for strength in the left upper limb 2/5, ipsilateral lower limb 3/5, right hemibody 5/5, preserved surface and deep sensitivity, normal osteotendinous reflexes (++ / ++++), without pathological reflexes.

Normal cerebral tomography (CT), laboratory tests, normal chest radiograph, electrocardiogram without alterations. The diagnostic possibility of cerebrovascular event was raised, however, after 5 hours of evolution, it presents dysphagia for fluids, areflexia, generalized weakness and acute respiratory distress, it is transferred to the intensive care unit (ICU) where it presents a ventilatory failure requiring orotracheal intubation.

On day 2 in the ICU, nuclear magnetic resonance (NMR) was performed, cervical, dorsal and lumbar spine without abnormal findings. A lumbar puncture (LP) showed cerebrospinal fluid of normal characteristics. Given the clinical context, GBS was suspected, neurophysiology studies were carried out, confirming the diagnosis. On the 3rd day of stay in ICU plasmapheresis begins. On the 4th day the progression continues in the neurological deterioration conditioning an enclosure syndrome maintaining communication through the ocular movements managing to manifest conservation of sensitive levels, without changes at the motor level; On the 5th day, signs of an acute systemic inflammatory response begin, a broad-spectrum antibiotic is initiated due to aspiration / nosocomial pneumonia. On the 6th day of stay in the ICU after his third plasmapheresis session, he had a torpid evolution, multiorgan failure, cardiorespiratory arrest and death. Necropsy was not authorized by relatives

III. DISCUSSION

GBS is an acute polyneuropathy of autoimmune response with heterogeneous clinical manifestations, which is the most frequent flaccid paralysis in the world ([Webb, Brain, Wood, Rinaldi, Turner, 2015](#)). Most of the studies that estimate GBS incidence rates were conducted in Europe and the USA, showing a range of 0.8-1.9 (median: 1) cases / 100,000 persons per year. The annual incidence of GBS increases with age (0.6 / 100,000 children year and 2.7 / 100,000 people over 80 years), slightly more frequent in men than women ([Yuki, Kokubun, Kuwabara, Sekiguchi, Ito, Odaka, Hirata, Notturmo, Uncini, 2012](#)) The case presented is consistent with a small percentage of patients described in the world literature, in the case of a 56-year-old male patient with acute atypical symptoms, asymmetric weakness with osteotendinous reflexes present. Studies have succeeded in demonstrating GBS variability with normal or hyper-excitability osteotendinous reflexes during the clinical course of the disease in approximately 10% of patients ([Fokke, van den Berg, Drenthen, Walgaard, van Doorn, Jacobs, 2014](#)) Scientific evidence associates respiratory or gastrointestinal tract symptoms up to 4 weeks before the start of GBS ([Musso, Cao-Lormeau, Gubler, 2015](#)) and infectious processes of microorganisms isolated in the laboratory such as *Campylobacter jejuni*, cytomegalovirus, Epstein Bar virus, Influenza A, *Mycoplasma pneumoniae*, HIV, Zika and Chikungunya, among others ([Wong, Umaphathi, Nishimoto, Wang, Chan & Yuki, 2015](#); [Chaverra, & Ayala, 2017](#)) cases of post-vaccine SGB has been described in rabies virus, Influenza A, H1N1.

In the documented case, it is mentioned that 15 days prior to the start of GBS, there were gastrointestinal manifestations that followed a self-limiting course. Regarding the symptomatic progression, the evidence shows an average of 1-2 weeks after the immune trigger and advances to a clinical maximum of 2-4 weeks (Fokkin, Selman, Dortland, Durmus, Kuitwaard, Huizinga et al, 2014). This point is interesting in the presentation of the case, where the symptomatic progression is established in hours up to approximately 4 days managing to severely compromise the patient, conditioning it to the confinement syndrome.

The clinical diagnosis of GBS classically involves acute ascending, symmetric progressive areflexia paralysis with albuminous cytological dissociation; an atypical presentation constitutes a diagnostic challenge for the medical specialist given the symptomatic heterogeneity and diverse diagnostic possibilities. In the case described, his clinical presentation was atypical and manifested by left brachiorural weakness, which later became generalized and initially associated with normal tendon reflexes, in addition to a normal CSF. According to the literature, there are studies that show the approximate time of albumin-cytological dissociation in CSF from the week following the start of GBS, not constituting a confirmatory or excluding test of GBS. 15% of patients with the disease have an increase in the CSF cell count (5-50 cells μ L) (Hughes, Swan, Van Doorn, 2012)

Finally, the treatment of GBS shows efficacy of both immunoglobulin and plasmapheresis, directly related to early onset. In the case presented once the diagnosis was confirmed, plasmapheresis was started, however, the rapid progression favored conditions causing death.

IV. CONCLUSION

It constitutes a diagnostic challenge for the neurologist the atypical presentations of GBS, especially those presenting with rapid progressive evolution, are a diagnostic challenge for the neurologist. It is important to highlight the importance of suspecting this pathology in the emergency department for timely diagnosis. Based on this case, it can be seen that the form of presentation can hinder diagnosis and treatment in the early stages of the disease and thereby delay the patient's chances of effective recovery. However, there is a small percentage of patients who still have diagnosis and timely treatment, his disease continues its natural course progressing to death

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