

Síndrome de Guillain-Barré Recorrente: Caso Clínico

Recurrent Guillain-Barré Syndrome: Case Report

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Resumo

Introdução: A síndrome de Guillain-Barré é uma polineuropatia aguda/subaguda frequente, resultante da resposta imune aberrante dirigida ao sistema nervoso periférico, gerando desmielinização e/ou degeneração axonal. A recorrência é rara e a sua existência não é consensual.

Caso Clínico: Homem de 37 anos, com antecedentes de síndrome Guillain-Barré grave em 2002, classificado como uma forma axonal. Em 2012 iniciou quadro agudo e progressivo de défice sensitivo e motor, com atingimento dos pares cranianos e dificuldade respiratória. Os resultados electromiográficos mostraram-se compatíveis com polirradiculoneuropatia sensitivo-motora desmielinizante com componente axonal. Efetuou terapêutica com imunoglobulinas intravenosas, com recuperação dos défices. Após 3 meses, foi transferido para o nosso Hospital onde cumpriu um programa de reabilitação. Verificou-se melhoria clínica e funcional.

Discussão: Com base na apresentação clínica, nos resultados laboratoriais e electromiográficos consideramos tratar-se de um caso de síndrome Guillain-Barré recorrente.

Palavras-chave: Síndrome de Guillain-Barré/reabilitação.

Abstract

Introduction: Guillain-Barré syndrome is a frequent acute or subacute polyneuropathy, resulting from an aberrant immune response directed to peripheral nerves, characterized by demyelinating and/or axonal neuropathy. Recurrence is rare and its existence is not consensual.

Case presentation: Male, 37 years old, history of severe Guillain-Barré syndrome in 2002, classified as axonal form. In 2012 he presented with acute and progressive motor weakness, involving the cranial nerves and respiratory distress. The electromyography results showed demyelinating motor and sensory polyneuropathy, with axonal component. He started intravenous immunoglobulin treatment with improvement of the neurological disabilities. He was transferred to our Hospital where he completed a broad rehabilitation program. He presented significant functional improvement.

Discussion: based on the clinical presentation, electromyography and laboratory results, we considered it a recurrent case of Guillain-Barré syndrome.

Keywords: Guillain-Barré Syndrome/rehabilitation.

Introduction

Guillain-Barré syndrome (GBS) is an acute or subacute polyneuropathy, resulting from an aberrant immune response directed against peripheral nerves, characterized by flaccid paralysis and other motor, sensory, and autonomic dysfunction.¹⁻⁵ The incidence

of GBS in Europe is 1.2–1.9 cases per 100 000, while worldwide is 0.6–4 cases per 100 000. GBS occurs in all parts of the world, men are more likely to be affected, and its incidence increases with age.³⁻⁶

GBS is considered to be a monophasic disease, although recurrences occur in 2-16% of patients. The existence of recurrent GBS is not consensual. Many authors consider it

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a form of chronic inflammatory demyelinating polyradiculoneuropathy. It is unknown why some patients have a recurrence.⁵⁻⁸ Most are demyelinating forms, in Portugal there was only one case published according to *Índice de Revistas Médicas Portuguesas*, and it was considered a motor axonal form.⁸

Case Presentation

In September 2002, a 28 years Caucasian male without relevant past medical or family history, presented distal lower extremities paresthesias and blurred vision. He reported an episode of diarrhea 7 days before. He was admitted to Hospital Pedro Hispano on the next day, presenting midriasis, anisocoria and symmetrically depressed deep tendon reflexes (DTR). Several exams were performed (Table 1).

Twenty four hours later the patient developed dysphagia and decreased motor function in the lower limbs. Within 48 hours, involvement of respiratory muscles was documented. Intravenous immunoglobulin G (4 mg/kg/day) was started. Within 24 hours developed respiratory failure, left III and bilateral VI cranial nerve palsy, bilateral peripheral facial palsy (PFP), more prominent on the left side, flaccid tetraparesis and absent DTR. Invasive ventilatory support was maintained for 60 days. It was considered a possible axonal form of GBS. The patient developed hypertrophic paraosteopathy in both hips, necrosis of the left femoral head, and a corneal ulcer.

He was transferred to Hospital da Prelada in January 2003, to continue the rehabilitation program. At

admission he presented with:

- Tetraparesis – Medical Research Council (MRC) motor power in the upper limbs graded 4/5, lower right limb 2/5 and lower left limb 1/5, except for dorsiflexion (0/5 bilaterally).
- Bilateral PFP and synkinesis during facial expressions.
- Right lower limb hypoesthesia.
- Severe limitation of the range of motion of both hips.
- Fatigue.

At discharge, 6 months later, the patient presented:

- Diminished fatigue.
- Bilateral PFP, predominant on the left.
- Feet hyperesthesia.
- MRC motor power 4+5 in plantar flexion bilaterally, 0/5 in dorsiflexion and toes extension.
- Independent gait with a bilateral ankle-foot orthosis (AFO), and slight *steppage*.

The patient underwent left hip arthrolysis in 2004, with significant improvement of the range of motion. He maintained follow-up in our Physical Medicine and Rehabilitation consultation, with no relevant functional alterations.

In April 2012, at the age of 37, the patient attended to Hospital Pedro Hispano due to discoordination, dysphagia, vomits (the first episode occurred 24 hours before) paresthesia, tetraparesia, predominant in the upper limbs, aggravation of the bilateral PFP and fatigue. Several exams were performed (Table 2).

Table 1 - Workup and Results in 2002.

Study	Result
Brain computed tomography scan Brain and medulla magnetic resonance imaging (MRI)	No alterations
Lumbar Puncture	Albumino-cytologic dissociation
Electromyography (EMG)	No muscular activity at rest Somatosensory evoked potentials were present but motor evoked potentials were not

Table 2 - Workup and Results in 2012

Study	Result
Lumbar Puncture	Albumino-cytologic dissociation
EMG	Demyelinating motor and sensory polyneuropathy, with axonal component
Brain MRI	Radicular contrast accumulation and in the left jugular foramen
Serologies and Immunologic Tests	Negative

Later, he presented dysphonia, dysarthria, absent DTR and tetraparesia aggravation. It was considered a case of recurrent GBS and intravenous immunoglobulin G (32gr/day) was administered for 5 days. He developed respiratory failure, requiring mechanical ventilation support for 15 days. Afterwards methylprednisolone was administered for 3 days. Although the motor deficits improved, the patient presented signs of dysautonomia (arterial hypertension and tachycardia), controlled with clonidine 150 mg/day. The patient completed a 5 day cycle of intravenous immunoglobulin G, 27.5 gr/day, with deficit improvement. Due to depressive symptoms, Psychiatric care was required. The pain complains were controlled with gabapentin and tramadol.

He was transferred to Hospital da Prelada in June 2013, at the admission he presented:

- Tetraparesis - global motor power 4/5, except for dorsiflexion of the feet (0/5) and plantar flexion (2/5).
- Decreased pinprick sensation and absent joint position sense on the right foot.
- Bilateral PFP.
- Depressed DTR.
- Indifferent cutaneous plantar reflex bilaterally.
- Dysarthria and mandibular tremor during speech.
- Intention tremor of the hands.
- Dysmetria.
- Poor orthostatic balance.
- Dependent on a third person to perform most activities of daily living, grading 31/100 in Barthel Modifide Scale and 63/126 in Functional Independence Measure scale (FIM) – motor score of 28/91.

At discharge, 3 months later, the patient presented with the same symptoms and disability remaining after the first episode. He was functionally independent, scoring 122/126 in FIM (87/91 in motor score).

After discharge, the EMG revealed motor and sensory polyneuropathy, predominantly demyelinating, with an axonal component and significant improvement compared with the previous exam.

Discussion

GBS presentation is heterogeneous and different subtypes have been described including acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor and sensory axonal neuropathy (AMSAN), Bickerstaff’s brain stem encephalitis, Miller Fisher syndrome, facial weakness and pharyngeal cephalobrachial variant.⁹⁻¹¹ In

North America and Europe AIDP accounts for 90% of cases and 5-10% are axonal subtypes.⁵

Approximately two-thirds of patients report a preceding infection, such as diarrhea or upper-respiratory-tract infection, within 6 weeks prior to symptom onset.^{5,12,13} Respiratory failure occurs in 30% of patients.^{14,15}

A typical case presents rapidly ascending symmetrical weakness. The acute or progressive phase lasts up to 4 weeks, during the *plateau* phase the symptoms remain stable for 2-4 weeks and the recovery phase lasts for 1-2 years.^{14,15}

Functional recovery takes place mainly during the first 6-12 months and no further recovery is expected after 2-3 years. In 10-35% residual symptoms remain and 4-15% die in the first year. Many continue to refer fatigue. The functional residual deficits may affect the work situation and require emotional support and substantial changes in social activities in more than one third of the patients.^{3,7}

There appears to be no significant difference between monophasic and recurrent GBS episodes considering clinical symptoms and triggering events. The nerve conduction studies showed similar findings. A weaker respiratory burst was the only immunological predisposing factor identified for recurrence. Patients may have similar or different presentations at recurrence, and many have rapid recovery following therapy although multiple recurrences tend to have slower recovery and residual neurologic deficits. The recurrence cases have been reported in every age group including pediatric age.^{10,16}

Recurrent GBS is rare and not consensual entity. Based on clinical, laboratorial and neurophysiological findings, we consider our case a probable recurrent AMSAN. Despite presenting many adverse prognostic factors (Table 3), the patient completed an intensive rehabilitation program with favorable functional evolution.

Table 3 - GBS Adverse Prognostic Factors⁵

- Older than 50 years at onset
- Rapid onset
- Severe disease at nadir
- Campylobacter jejuni or Cytomegalovirus infection
- Signs of axonal loss in neurophysiological tests

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