Therapeutic plasma exchange as a first-choice therapy for axonal Guillain-Barré syndrome: A case-based review of the literature (Review)

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Abstract. Guillain-Barré syndrome is an acute immune-mediated disease that affects the peripheral nervous system, with progressive motor deficit in the limbs, sometimes involvement of the cranial nerves and possible impairment of the autonomic nervous system. Due to the respiratory and autonomic nervous dysfunction, the disease has the potential to be fatal. Although modern methods of treatment have significantly improved patient prognosis, many patients nevertheless experience significant neurological sequelae. The practical applicability of plasmapheresis was illustrated in our case report. We report the case of a 27-year-old man who had a mild viral respiratory tract infection 1 week prior to the onset of disease with gradual development of a motor deficit, urinary retention, slight swallowing difficulties and mild respiratory dysfunction. Nerve conduction studies were performed and the diagnosis of acute motor axonal neuropathy phenotypic variant of Guillain-Barré syndrome was established. Autoimmune and inflammatory diseases, infectious diseases, endocrinopathies, neoplastic diseases, intoxications, metabolic diseases and vitamin deficiencies were ruled out. Our patient attended four sessions of therapeutic plasma exchange performed using peripheral venous approach with two needles with significant recovery of the motor deficit. The patient was discharged 1 week later on maintenance kinetotherapy with further favorable evolution. In conclusion, we report a good evolution as a result of therapeutic plasma exchange in a patient with acute motor axonal neuropathy phenotypic variant of Guillain-Barré syndrome. The procedure is well-tolerated and can be performed safely by peripheral approach not only in the intensive care unit but also in a neurology clinic.

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Abbreviations: GBS, Guillain-Barré syndrome; AIDP, acute inflammatory demyelinating polyradiculoneuropathy; AMSAN, acute motor-sensory axonal neuropathy; AMAN, acute motor axonal neuropathy; TPE, therapeutic plasma exchange; i.v., intravenous; IVIg, intravenous immunoglobulin; MRC, Medical Research Council; HFGS, Hughes functional grading scale; NCS, nerve conduction studies; CMAP, compound muscle action potential; CSF, cerebrospinal fluid; HIV, human immunodeficiency virus; anti-dsDNA, anti-double stranded DNA; ANAs, anti-nuclear antibodies; *C. jejuni*, *Campylobacter jejuni*; *M. pneumoniae*, *Mycoplasma pneumoniae;* MRI, magnetic resonance imaging; FFP, fresh frozen plasma; PV, plasma volume; HHS, hydrocortisone hemisuccinate; CMV, cytomegalovirus; EBV, Epstein-Barr virus; Th1, T helper 1 cell; ASFA, American Society of Aphaeresis; BBB, blood-brain barrier

Key words: Guillain-Barré syndrome, acute motor axonal neuropathy, therapeutic plasma exchange, intravenous immunoglobulins, allergic reaction to plasma

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1. Introduction

The first description of Guillain-Barré syndrome (GBS) dates back to 1859, when Landry published a description of a case with ascending paralysis (1). The clinical and biological picture was later completed in 1916 by French neurologists Georges Guillain, Jean-Alexandre Barré and Andre Strohl (2).

GBS is an acute immune-mediated disease that reaches maximum severity within 2-4 weeks. GBS affects the peripheral nervous system and is characterized by progressive

motor deficit in the limbs with ascending sensory deficits, involvement of muscles innervated by the cranial nerves, reduction or abolition of the deep tendon reflexes, and possible impairment of the autonomic nervous system, sometimes with respiratory failure and albuminocytological dissociation (3). Due to the respiratory and autonomic nervous dysfunction, the disease has the potential to be fatal even when patients are treated at centers that provide optimal care. From the neurophysiological point of view, the following phenotypes of GBS are described: Acute inflammatory demyelinating polyradiculoneuropathy (AIDP), Miller Fisher syndrome and acute autonomic neuropathy (4) and the axonal variants, acute motor sensory neuropathy (AMSAN) and acute motor axonal neuropathy (AMAN). Axonal variants were first recognized in northern China and then reported in other countries (3,5). In North America and Europe the incidence of AMAN is low (6,7).

The idea of molecular mimicry between pathogens and autologous antigens has been proposed as a possible mechanism of this autoimmune disease. In AIDP, the patient's immune system generates antibodies that cross-react with shared epitopes against myelin or the Schwann cell surface membrane (8).

Characteristic for AMAN is the association with *Campylobacter jejuni* (*C. jejuni*) enteritis; therefore, the antibodies against gangliosides are characteristic (3,8-10). The molecular mimicry and the structural similarity between GM1/GD1a gangliosides and the lipo-oligosaccharides of *C. jejuni* has been proven by numerous bacteriological, immunological and pathological studies (8,11). The immune response is mediated by antibodies against GM1 and GD1a that are situated at the level of Ranvier nodes where the axolemma is exposed and the sodium-channels are clustered (8,12).

Immunohistochemical studies from deceased patients reveal antibody-mediated alteration of the motor axonal membrane, suggesting that the immune response is primarily directed against the motor axolemma (8,13). In AMAN, morphopathological examination have shown deposits of IgG and complement in the axolemma of the motor nerves at the Ranvier nodes, with minimal demyelinating damage and mild lymphocytic infiltration, followed by macrophage infiltration (2,14). The macrophages invade the axon at the Ranvier nodes, where they insert between Schwann cells and the axon without affecting the myelin sheath and produce nerve damage and functional blockage of nerve transmission (15). After the complement activation the development of the complement membrane attack complex occurs and disrupts the sodium channels. The sodium channel dysfunction can explain the changes in the nerve conduction studies, slowing the motor conduction and producing variable degrees of conduction blocks, due to the fact that saltatory conduction is critically altered (8,12).

In advanced stages with ventral root involvement, irreversible changes with severe axonal degeneration may occur (16), if the underlying pathophysiological mechanism is not controlled. Therefore, rapid therapeutic interventions that trigger the neutralization of the autoantibodies, easing the conduction blocks, may lead to a rapid resolution of the symptoms. Contrary, a mediocre recovery is expected if the axonal degeneration occurs at the level of the nerve roots (8,17). The uncertainty is whether which type of intervention may result in a better clinical evolution, depending on the GBS subtype.

Although modern methods of treatment such as therapeutic plasma exchange (TPE) and intravenous immunoglobulins (IVIg) have significantly improved the prognosis, many patients nevertheless experience significant neurological sequelae (5,18).

2. Practical applicability of plasmapheresis: Case illustration

We report the case of a previously healthy 27-year-old man who had a mild viral respiratory tract infection 1 week prior to the onset of disease. Two days before admission to our clinic, the patient experienced paresthesia in the lower limbs with ascending character towards the upper limbs, followed by progressive weakness with the same distribution as the paresthesia. Neurological examination performed at admission did not reveal any changes in the cranial nerves, but detected flaccid tetraparesis of Medical Research Council (MRC) (19) grade 4/5 in the upper limbs and MRC grade 3/5 in the lower limbs, diminished deep tendon reflexes in the upper limbs and abolished deep tendon reflexes in the lower limbs, without pyramidal signs, with no sensitivity disorders. The evaluation performed after the Hughes functional grading scale (HFGS) (20) at admission placed the patient at grade 4. On the following day, the patient's evolution was rapidly progressive, with worsening of the motor deficit to MRC grade 2/5 in the upper and lower limbs with slight swallowing difficulties and mild respiratory dysfunction, requiring oxygen support. He also developed urinary retention, and a Foley catheter was inserted. Nerve conduction studies (NCS) were performed on the same day using conventional procedures, with motor conduction studies on the medial, ulnar, peroneal and tibial nerves bilaterally, and sensory conduction studies on the medial, ulnar and sural nerves bilaterally. A low amplitude of compound muscle action potential (CMAP) was detected in all motor nerves, aspects that are characteristic for marked axonal loss. Examination of the F wave showed a proximal conduction block of 100% at the level of the median and ulnar nerves and a conduction block of 80-90% at the level of the bilateral tibial nerve, suggesting proximal root involvement (Fig. 1). The sensory conduction was within normal limits. Following the NCS examination, the diagnosis of the AMAN phenotypic variant of GBS was established. The peripheral oxygen saturation measured by pulsoximeter was 92%. The acid-base balance revealed a mild respiratory acidosis with pH 7.27, PaO_2 116 mmHg, $PaCO_2$ 65 mmHg at a FiO_2 0.4 with BE-1.2 mmol, bicarbonate 24 mEq/l. Lumbar puncture with cortical spinal fluid (CSF) examination was performed and was normal. Laboratory tests ruled out infectious diseases [human immunodeficiency virus (HIV), syphilis, hepatitis B and C, Lyme disease serology], endocrinopathies and autoimmune diseases [anti-double stranded DNA (anti-dsDNA), anti-nuclear antibodies (ANAs), antiphospholipid antibodies were within normal limits]; serum protein electrophoresis revealed no anomalies. His stool cultures were negative; tests for anti-gangliosides antibody, *Campylobacter jejuni* (*C jejuni*) and *Mycoplasma pneumoniae* (*M. peumoniae*) antibody are not available in our hospital. The patient also underwent a thoraco-abdominopelvic computed tomography
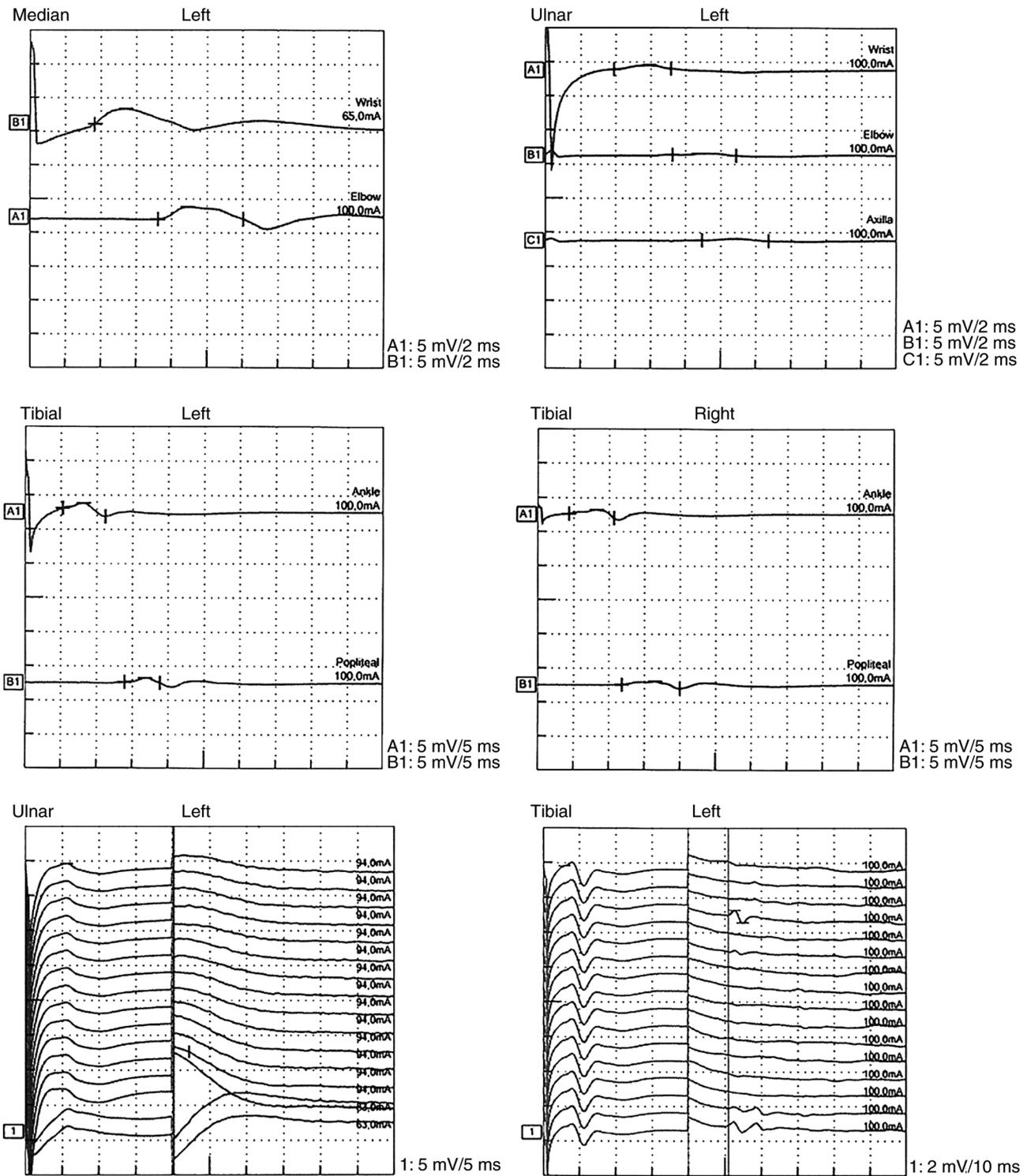
Figure 1. Initial nerve conduction study revealed low amplitude of CMAP, which is relevant for axonal loss. Absence of F-waves is suggestive of proximal conduction blocks. CMAP, compound muscle action potential.

scan, which was within normal limits, to rule out a paraneoplastic syndrome. He underwent cervical spine magnetic resonance imaging (MRI), which excluded the possibility of transverse myelitis.

On day 2 after admission and on day 4 after the first symptoms, TPE was initiated immediately in hospital using a commercially available system (Spectra Optia Apheresis, TerumoBCT). The exchange fluid was a combination of fresh frozen plasma (FFP) and 5% albumin solution with an Anticoagulant Citrate Dextrose, SolutionA, (ACD-A) anticoagulant solution at 10:1 whole blood:citrate ratio. The flow inlet rate varied between 30 and 70 ml/min according to the type of replacement fluid used and the functioning of the peripheral venous approach. The procedure was performed using the peripheral venous approach with two peripheral catheters 16 Gauge diameter each placed at the antecubital

Table I. Laboratory parameters during the plasma exchange sessions.

	Before session #1	Before session #2	Before session #3	Before session #4
Hematocrit (%)	46.5	48.5	53.2	51.1
Platelet count (/μl)	297,000	346,000	344,000	314,000
Total calcium (mg/dl)	2.14	2.43	2.38	2.3
Magnesium (mg/dl)	0.99	0.9	1.03	0.95
Potassium (mmol/l)	4.27	4.23	4.36	3.81
Sodium (mmol/l)	141	140	141	137
Creatinine (mg/dl)	0.70	0.80	0.69	0.67
Blood urea nitrogen (mg/dl)	24.1	33.1	46.3	52
Glucose (mg/dl)	101	96.9	95.0	88

veins on both sides for the inlet and the outlet circuits. During TPE, the patient received a prophylactic infusion of calcium supplementation consisting of 10% calcium gluconate to prevent a symptomatic fall in plasma ionized calcium due to citrate anticoagulation. After the first TPE session, respiratory symptoms were significantly improved, the peripheral oxygen saturation increased up to 96%, therefore, subsequent arterial blood gases analyses were between normal ranges.

In the first plasma exchange session, 1.1 plasma volume (PV) was exchanged; in the second session, 1.2 PV was exchanged. In the third session, only 0.5 PV was exchanged because the patient developed an allergic reaction to plasma, with generalized rash, pruritus and dyspnoea with hypo-oxygenation and decreased oxygen saturation of 89%. The procedure was stopped immediately, and 50 mg Ranitidine (Medochemie Ltd.) i.v., 200 mg hydrocortisone hemisuccinate (HHS) (Zentiva) i.v. and 10 mg Claritin (Schering Plough Labo NV, Belgium) orally were administered with rapid allergic reaction regression. On the following day, the fourth exchange was performed with 1.2 PV. Each session was separated by 2 days, except for sessions 3 and 4, which were held on two consecutive days, and a total of 4 PVs were exchanged, and a combination of FFP and 5% albumin solution was used as replacement. Following the initial allergic reaction, the patient received 200 mg HHS (Zentiva) i.v. 2 h prior to TPE to prevent other allergic reactions.

Laboratory testing was performed before the first session of TPE, and then performed after each session to check the complete blood count, calcium, magnesium, potassium, sodium, glucose, creatinine and blood urea nitrogen. There were no significant changes in the above parameters before the first TPE and after the last TPE session (Table I).

The neurological evaluation performed after plasma exchange session 4, i.e. on day 8 after admission, revealed significant improvement, with regression of motor deficit in all limbs at MRC grade 4/5, and he was able to pass urine, therefore the Foley catheter was removed. He experienced no additional TPE-related complications during or after the procedure and had no difficulties related to peripheral venous access. We decided to continue kinetotherapy and the supportive treatment with further favorable evolution, and he was discharged 1 week later. The patient was discharged with HFGS grade 2 with an improvement of 2 points compared with that at admission. NCS performed 2 weeks after admission revealed also significant improvement (Fig. 2). One month later, the patient was completely recovered, and the NCS no longer showed pathological changes.

3. Discussion

The proportion of different GBS subtypes varies by region. AMAN is more common in China, but there are also percentage differences between northern and southern China (21-23). In a study on the pediatric population, also conducted in China, 22% of patients with GBS had AMAN (21), while another study conducted in two centers in France (Toulouse and Montpellier) that included children showed that AMAN was present in 17 of the total 110 patients included in the study (24). In North America and Europe, the AIDP subtype is the most common, and only 5% are axonal subtypes of GBS (25), where the incidence of AMAN subtype is 1-3% (6,7).

Clinical and epidemiological observations show that GBS is associated with a preceding illness, and 75% of patients report acute enterocolitis or upper respiratory infection (26). The most frequent pathogens involved are *C. jejuni*, cytomegalovirus (CMV), Epstein-Barr virus (EBV), Zika, post-flu or other events involving immunization with vaccines such as rabies vaccines (27,28), surgery and anesthesia (5). In the case of AMAN, the most frequently involved pathogen is *C. jejuni* (15). Our patient had a respiratory viral infection prior to the onset of the GBS that could be considered the trigger of the immune process but specific serological and immunological tests did not reveal infections without having data concerning *C. jejuni* and *M. pneumoniae*.

TPE using the Spectra Optia system involves separating plasma from hematocrit by centrifugation, and removing and replacing the patient's plasma with an equal volume of fluid replacement consisting of FFP and 5% albumin solution to maintain appropriate oncotic pressure while the remaining cells are re-infused back to the patient (29). In GBS, this procedure alters the T helper 1 cell (Th1) to Th2 ratio, alters B cell and T cell number and activation, and helps to reduce the concentration of pathogenic plasma components such as circulating autoantibodies, immune complexes, complements, cytokines or other immunologically active substances (29).
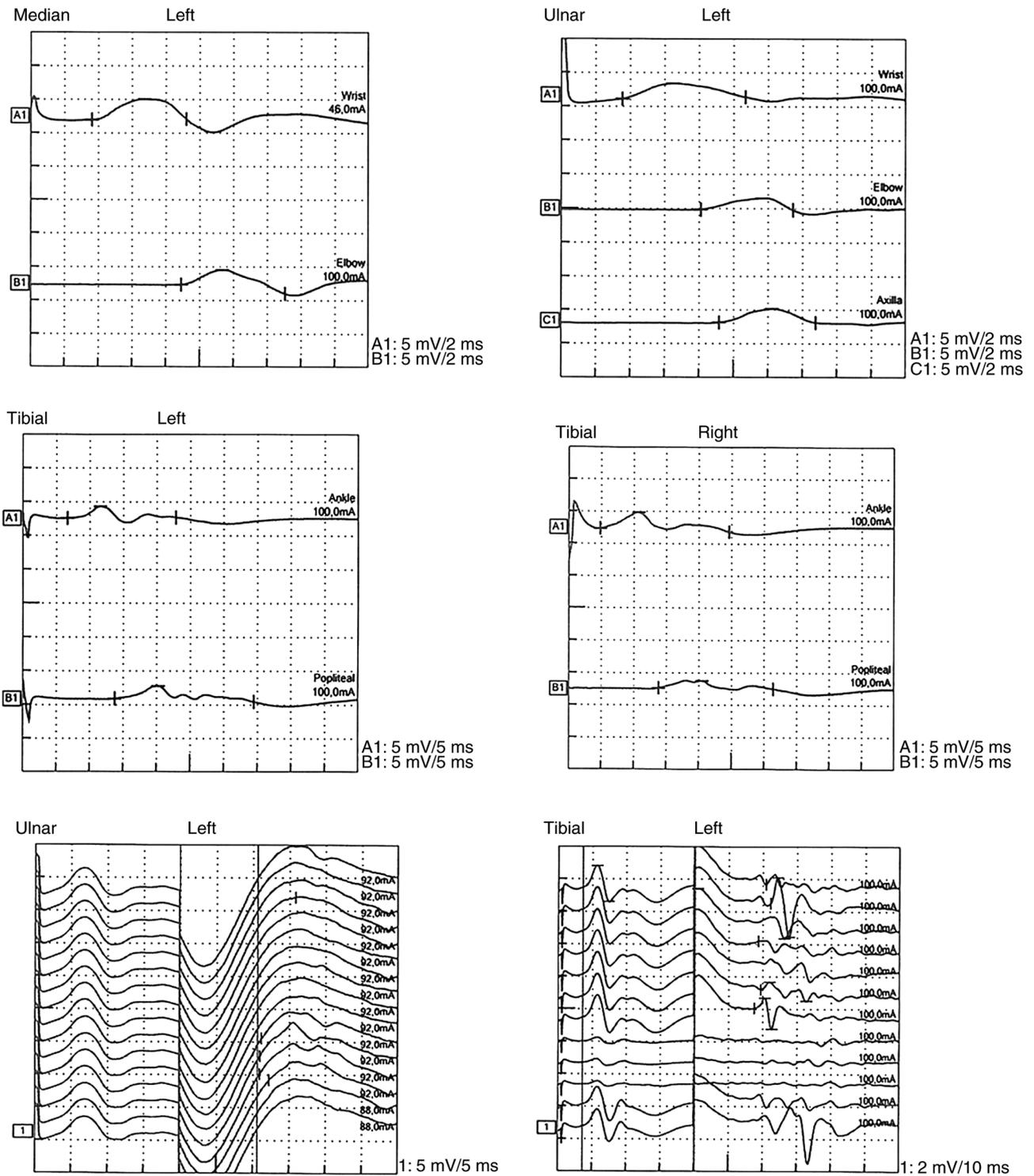
Figure 2. Follow-up nerve conduction study 2 weeks after admission and following plasmapheresis revealed an increase in CMAP amplitude and the appearance of F-waves. CMAP, compound muscle action potential.

The benefit of TPE is increased if the treatment is initiated early, i.e. within 7 days from the disease onset, according to a recent Cochrane review (30,31), but still has beneficial effect in the first 4 weeks from disease onset (16,32).

According to the 2019 American Society of Aphaeresis (ASFA) guidelines the primary use of TPE in GBS is an ASFA category I indication (recommendation grade. 1A) (4). It is recommended to exchange 1-1.5 PV per session, 5-6 times over 10-14 days in an every-other-day regimen (4). A TPE procedure usually removes 60-70% of the substances from the intravascular compartment (4). The interval between procedures is required to allow rebalancing of the intravascular space and to reduce the risk of bleeding caused by the depletion of the anticoagulation factors, of which the main factor is fibrinogen (30-33). In moderate cases, there was no difference between patients who underwent four PV exchanges and those

who underwent six exchanges (16,34). Our patient underwent four exchanges with a total of four PV exchanges. In moderate to severe groups, four sessions are beneficial (34,35), while severe cases require 5-6 TPE procedures (4,29).

The most common adverse effects associated with TPE cited in the medical literature are allergic reactions to plasma (chills, fever, rash, hives, dyspnea and stridor), chest pain, dizziness, headache, abdominal pain, anxiety, hypotension, nausea and vomiting; the incidence is approximately 11% compared to those receiving 5% albumin solution as fluid replacement; another adverse effect is symptoms of hypocalcaemia when citrate is used as an anticoagulant as a result of calcium ion binding in the blood. Albumin is especially associated with adverse effects such as hypotension, nausea and vomiting due to the hypo-oncotic effect (36).

Our patient had peripheral venous access and no access-related complications. He was closely monitored, and the onset of plasma allergy was detected immediately, with immediate cessation of plasmapheresis, administration of Claritin (loratadine, 10 mg tablet), which is a second-generation histamine 1 (H1) receptor antagonist used to treat allergies, and 50 mg of Ranitidine i.v., a H2 receptor inhibitor that relieves the symptoms of acute allergic reactions. It has been observed that the combination of blocking both the H1 and H2 receptors may provide better relief (37).

Geographical variation, clinical course and disease severity probably differ due to gene polymorphism in populations with different susceptibility to developing different types of GBS. In a study conducted by Estrade *et al*, the mean follow-up period was 300 days; 77% of the patients recovered with progressive regression of symptoms, and the mean recovery time was estimated at 280 days. The presence of sequelae was correlated with the short duration between symptom onset and hospitalization and with the axonal form of GBS (AMSAN or AMAN), with 29% of children with the axonal form having sequelae compared to 5% of those with AIDP. We should mention that, in our French colleagues' study, most children received IVIg, unlike our patient who underwent four courses of TPE (24). Other studies report that the axonal forms (AMAN and AMSAN) are correlated with a higher risk of long-term sequelae (38,39), and other authors consider that although the axonal form of GBS has a longer recovery period compared to the demyelinating forms, there are no long-term differences between them (40,41).

In a study published in 2015 in a Chinese population, HFGS at nadir (grade 0, healthy; grade 1, minor signs and symptoms, able to run; grade 2, able to walk independently at least 5 meters; grade 3, able to walk with assistance; grade 4, bedbound; grade 5, requires assisted ventilation; grade 6, deceased) was significantly higher in patients in the AMAN subgroup of GBS, and the necessity for mechanical ventilation was higher than in the AIDP group (21,42). Reports of GBS in the pediatric population also show that patients with axonal involvement have more severe disease progression and a higher rate of morbidity and mortality than patients with AIDP (43). Our patient, diagnosed with AMAN, had an HFGS score of 4 at nadir, which is quite high, and he was close to being intubated, similar to the data reported in the medical literature. In the above study, the patients with AMAN had worse outcome at the 3- and 6-month follow-up than the patients with

AIDP (P=0.001, P=0.000 respectively) (21), unlike our patient, who presented a favorable outcome after TPE.

Another UK study suggests that the pattern of axonal injury is associated with poorer prognosis of recovery at 3 and 6 months (44). According to Zhang *et al*, an HFGS score at onset of ≥3 and the presence of dysautonomia are predictors of poor outcome at 6 months (21).

Generally, patients with AMAN are thought to have poor prognosis compared with patients with AIDP (5), but some patients with AMAN often show rapid recovery, and Hiraga *et al* described these patterns of evolution in a study that included patients with GBS who received immune treatments (3). Out of a total of 35 patients with the AMAN phenotype of GBS who were analyzed for neurological evolution, 19 had a 1 point improvement on the HFGS at 4 weeks; in the other patients, the 1-point improvement was delayed by >1 month (3). Low CMAP amplitudes or absent motor responses on NCS are predictive criteria of poor outcome in adults (43,45), and our patient presented these criteria.

The explanation for patients with AMAN who reach their nadir quickly and recover as quickly as patients with AIDP is that the pathological process does not destroy the axon, but produces a conduction block that is reversible without axonal degeneration in the case of rapid elimination of the autoantibodies directed against the sodium channel, or the degeneration that occurs is located very distally (12,16).

A meta-analysis published in 2012 that included 649 patients enrolled in six trials showed that TPE decreased the need for ventilation support compared with controls (RR: 0.53) and reduced the time needed to regain the ability to walk (30 vs. 44 days) (29,46).

The combination between the HFGS and MRC scores assessed at 1 and 2 weeks are good predictors and are correlated with the outcome at 6 months (29,47). Our patient had very good evolution, with improvement of 2 points on the HFGS within 15 days, significantly higher compared to previously reported data.

Autonomic impairment is common in GBS, especially in cases where respiratory dysfunction is present, as happened with our patient (2). Urinary dysfunction is one manifestation and has been reported in 50% of patients with axonal GBS, more frequently than in classic GBS (21%), but the underlying mechanism is not well known and is believed to be either bladder areflexia or a non-relaxing urethral sphincter (43,48,49).

The CSF protein level is generally elevated, with albuminocytological dissociation as a result of increased blood-brain barrier (BBB) permeability if the protein level is measured 2 or 3 weeks after onset, but in the first week of the illness, it may be normal, as it was in our patient (43). Anti-ganglioside antibodies are identified in many patients with GBS, but tests are expensive and not always available (2).

We believe that our case presented rapid evolution towards recovery with lack of neurological deficits 1 month after discharge despite the presence of factors correlated with negative evolution (short interval between symptom onset and admission, presence of axonal form, low CMAP amplitude) due to the application as the first-line treatment of plasmapheresis with the elimination of autoantibodies, which prevented axonal degeneration, and to the very short time elapsed between symptom onset and the first session of plasmapheresis.

The existence of good recovery potential, evidenced by our patient's spectacular evolution with improvement, suggests that axonal degeneration is not always the basis of the changes found by NCS. It is possible that there is antibody-induced blockade at the level of the Ranvier nodes in the motor fibers, and in the case of rapid removal by plasmapheresis, axonal degeneration does not recur, and this may explain our patient's favorable evolution.

IVIg and TPE appear to have approximately equal efficacy for treating GBS, but most studies have predominantly included the AIDP subtype (15,50,51). The therapeutic response to IVIg is good in the case of AIDP, but is unsatisfactory in patients with the axonal forms (16,52); Buzzigoli *et al* suggested that patients with GBS with axonal involvement could have a better response to TPE, which should be considered early (53), and Dada and Kaplan also suggest that TPE may be superior to IVIg for treating patients with severe forms of GBS and axonal involvement (54). TPE is a therapeutic option in the case of the AMAN phenotypic variant of GBS, with even higher efficacy, when it is initiated earlier. Due to the increased percentage of patients that become disabled, larger prospective trials that include patients with the axonal form of GBS treated with TPE and IVIg are needed.

Other favorable data for the use of TPE are represented by a study conducted in Southern India and published in 2014, which compared the two treatments and did not show a significant difference between the two groups in terms of improvement rate, but found that IVIg costs more (USD 4250-5300) compared to plasmapheresis (USD 2600-4100) (55). These data are similar to that reported by Western European countries (56-58); therefore, plasmapheresis could be the preferred method for treating GBS in low-socioeconomic countries.

The main aim of the present article is to report the efficacy of TPE in a severe and rapidly progressive AMAN subtype of GBS. Another recently published case report showed the beneficial effect of TPE after IVIg failed to improve the neurological condition of a child diagnosed with the AMAN subtype of GBS (59). Cases such as this may be included in retrospective studies on the efficacy of different treatments for the axonal forms of GBS.

The particularity of the presented case consists in a good clinical resolution of the symptoms using TPE in the presence of a severe disease with rapid onset and evolutionary potential with severe nerve conduction impairment and respiratory dysfunction, in a young patient with no associated comorbidities.

4. Conclusions

The good evolution of our patient suggests that TPE could be considered first choice treatment for the AMAN subtype of GBS.

Close monitoring makes it easy to detect possible complications, and prompt intervention makes it easy to treat them, proving that, in these conditions, TPE is a relatively safe and well-tolerated procedure.

The possibility of performing TPE in the neurology clinic by the peripheral venous approach makes it possible to be performed quickly from the moment of therapeutic indication, saving precious time and relieving colleagues in intensive care units of additional work.

Availability of data and materials

Data and materials are available from the corresponding author on reasonable request.

Authors' contributions

AS and GŞ performed the literature research and drafted the manuscript. ZB, SA and OM analyzed and interpreted the patient data regarding the neurological disease and treatment. AB provided critical review for all aspects of the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Not applicable. Ethical approval was not required as the patient was not included in a clinical study and did not receive experimental treatment.

Patient consent for publication

Informed consent forms were signed by the patient who agreed to the use of his medical records for publication.

Competing interests

The authors state that they have no competing interests.

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