Guillain-Barré Syndrome: Epidemiological, Therapeutic, And Prognostic Characteristics

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Abstract :

Guillain-Barré syndrome (GBS) is the leading cause of acute extensive paralysis since the eradication of acute anterior poliomyelitis (1). A precise and repeated neurological examination often confirms the diagnosis. It is an evolving polyradiculoneuropathy that can lead to death. The diagnosis is not always straightforward, even in its classic form, especially early in the disease when the symptoms may be incomplete. It is not uncommon for patients to make multiple visits to the emergency department before the diagnosis is considered (2).

The objective of our study is to evaluate the epidemiological, clinical, laboratory, therapeutic, and prognostic profile of polyradiculoneuropathies in order to identify patients at risk of developing acute respiratory failure and provide them with prompt and adequate management.

Keywords: GBS, MRC score, electromyography, plasma exchange, immunoglobulin..

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I. Introduction :

Guillain-Barré syndrome (GBS) is a rare but potentially serious neurological disorder. It was first described in 1859 by Jean Baptiste Octave Landry de Théizillat (3), who reported 10 cases of ascending paralysis resulting in death in 2 cases. In 1916, three French neurologists, Georges Guillain, Jean-Alexandre Barré, and André Strohl, described two cases of acute paralysis with areflexia and paresthesia, accompanied by minimal sensory disturbances. The analysis of cerebrospinal fluid (CSF) showed increased protein levels without pleocytosis (4) (5).GBS is an acute autoimmune-mediated polyradiculoneuropathy (1) characterized by muscle weakness with areflexia. It develops rapidly, typically within 4 weeks, and follows a monophasic course. GBS exhibits a wide clinical heterogeneity in terms of severity and prognosis (6).Electroneuromyography (ENMG), along with CSF analysis, are the key diagnostic tests to confirm GBS and rule out differential diagnoses (2).

The objective of this study is to describe the clinical, epidemiological, and prognostic characteristics of GBS..

II. Materials et methods :

This is a retrospective monocentric study conducted over a period of 7 years, from January 2015 to December 2021, involving 111 cases of patients admitted to the Neurology Department at Oran 1 November 1954 University Hospital, Algeria. Data were collected from patients' medical records, including medical history, presenting clinical symptoms upon admission, results of neurological examinations, laboratory test results, and treatment modalities. Information on disease progression and short- and long-term outcomes was gathered from post-hospitalization follow-up records.

All included patients were also examined by at least one of the current co-author neurologists, who confirmed that all necessary investigations to exclude alternative diagnoses had been conducted. Thus, our study patients all met Level 4 of the Brighton criteria.

The data were entered into data extraction sheets and then inputted into the SPSS software.

III. Results:

We identified 111 patients with Guillain-Barré syndrome (GBS). The mean age of the patients was 42 years, with a slight male predominance (58%). The patients were from Oran in 37.8% of cases, while others were evacuated from neighboring provinces such as Mascara, Tiaret, Chlef, Mostaganem, Ain Tmouchent, Sidi Belabbes, Saida, and El-Bayad.

Although 4.5% of patients had a history of GBS upon admission, in 67% of cases, GBS occurred in the absence of any other underlying conditions. The average time between the onset of symptoms and hospitalization was 12.7 days.

The identified triggering factors were flu-like syndrome (20%), SARS-CoV-2 infection (20%), and in 48% of cases, no triggering factor was identified (Figure 01).

The main reason for admission was muscle weakness, with an average score of 32 out of 60 on the MRC scale. This weakness was isolated in 67% of cases or associated with sensory disturbances such as paresthesia (20%), pain (7%), and anesthesia (2.7%). Dysautonomic symptoms, such as tachycardia, were found in 7.2% of cases.

Overall, the prognosis was favorable regardless of the identified triggering factor, except for cases involving SARS-CoV-2 infection and influenza, which had the highest mortality rates.



Figure 01: Clinical progression according to the triggering factor, examples: influenza

Complete recovery is more frequent at 29.8% in cases with an initial MRC score of over 30 points, while the mortality rate is lower compared to an initial score below 30 (Figure 02).



Figure 02: Cross-sectional study on the clinical progression of GBS according to the initial MRC score

Cranial nerve involvement was found in 25% of cases in our series, with a predominant involvement of the bulbar nerves and the facial nerve (unilateral or bilateral).

Symptom progression was ascending in 88% of cases, and the most common subtype was the demyelinating form (AIDP, 37.8%), followed by the motor axonal form (AMAN, 27%), and the sensory-motor axonal form (AMSAN, 16%). Clinical improvement was significantly more frequent in the demyelinating form (65% of cases) compared to the axonal form (35% of cases). The likelihood of progression to death was less frequent in the AMSAN form compared to the other two forms (Figure 03).

The average time between the onset of symptoms and hospitalization was 12.7 days. Total recovery was more frequent when treatment was initiated in the third or fourth week, while progression to death was more frequent even if treatment was initiated in the first week.



Figure 03: Clinical progression according to the electrical subtype of GBS.



Figure 04: Therapeutic choices for patients with Guillain-Barré syndrome in the Neurology Department between 2015-2021.



Figure 05: Clinical progression according to therapeutic choices.

Regarding therapeutic management, only intravenous immunoglobulins (IVIG) and plasma exchanges (PE) have demonstrated effectiveness in GBS to date. In our cohort, patients received treatment with either PE or IVIG. The clinical outcomes according to the chosen treatment were as follows: good clinical improvement in 83.2% of cases for PE and 75.4% for IVIG. Total recovery in 38% and 22.4% of cases, respectively. The proportion of patients who progressed to death under treatment was 12.9%, with a slight predominance among patients receiving IVIG.

IV. Discussion :

Our results regarding age and sex are similar to those found in studies conducted in Turkey (Kozanoglu et al., 2015) (9) and France (Raphael et al., 1987) (10). The male-to-female ratio is unfavorable for men, with a relative risk of 1.78 for all study series.

The main prodromal agents found in our study were COVID-19 in 20.1% of cases and influenza-like syndrome in 19.8% of cases. In the literature, a recent history of digestive infection (acute gastroenteritis), respiratory infection, or influenza-like syndrome is found in over 60% of cases. The main implicated agents are Campylobacter jejuni, Mycoplasma pneumoniae, Haemophilus influenzae, Cytomegalovirus, Epstein-Barr virus, or human immunodeficiency virus.

GBS is an acute polyradiculoneuropathy, classically characterized by a rapidly progressive, ascending, bilateral, and symmetrical sensory-motor deficit. The diagnosis is primarily clinical. In our study, we observed a correlation between good clinical recovery and an initial deficit evaluated by the MRC score of over 30 points. In the literature, there is no correlation between initial symptoms and disease progression, but there are factors associated with poor clinical prognosis, namely early bulbar involvement, early respiratory failure, and progression in less than 24 hours.

Regarding the electrodiagnostic classification into demyelinating and axonal subtypes, this can be done by nerve conduction studies and electromyography performed within the first three weeks following symptom onset (11). Our results favor a predominance of the demyelinating form compared to the forms found in Europe, North America, and Australia. The most common subtype of GBS is the demyelinating form (up to 90% of cases), while axonal forms constitute 30 to 47% of cases in Asia, South and Central America, and up to 86% in China (11). It should be noted that this classification currently has no therapeutic implications (12).

Specific treatment for GBS is based on plasma exchange or intravenous immunoglobulins (IVIG) (14). In our study, we found a superiority of plasma exchange compared to IVIG in terms of clinical recovery and mortality. In the literature, plasma exchange by centrifugation or filtration has demonstrated efficacy in several aspects: reduced duration of mechanical ventilation, shorter time to walking recovery with and without assistance, and faster recovery from deficits. Long-term effectiveness has been proven, showing a reduction in mortality and sequelae at 1 year (15) (16) (18).

Regarding IVIG, when initiated within 15 days of symptom onset, it is as effective as plasma exchange in terms of mortality and clinical recovery in the short term (4 weeks) and long term (1 year). IVIG is generally better tolerated, with fewer treatment interruptions (8) (11).

V. Conclusion :

GBS is a neurological emergency due to the risks of progression to acute respiratory failure (5). Clinical recovery is related to the severity of initial symptoms, electroclinical subtype, and timeliness of treatment, particularly for plasma exchange (6). New therapeutic agents targeting various components of the complement cascade are at advanced stages of clinical development (4).

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