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Atypical Manifestations in Children with Guillain Barré Syndrome

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1. Abstract

Guillain Barré Syndrome (GBS) is an acute single-phase causal disease that occurs after an infection. An analysis ranges from 0.5-1.5/100,000 children, predominates in males. Initiatives with the limbs followed by progressive, symmetrical muscle weakness, with the principle of lower nodes (lower limbs). In children, the predominance is the difficulty of gait and greater involvement of the cranial pairs. Young children are difficult to diagnose because they have atypical complaints and a more challenging neurological examination. In case of suspicion, the patient should be hospitalized in a pediatric ICU, where he/she should remain monitored, with frequent surveillance and treatment should not be delayed, reducing the frequency and severity of complications.

2. Keywords: Guillain Barré Syndrome; Paresthesia; Asymmetry

3. Introduction

Guillain Barré Syndrome (GBS) is classified as an eponym that encompasses acute immune-mediated polyneuropathies. It is considered an acute monophasic paralyzing disease that usually occurs after an infection, being the most common cause of flaccid paralysis in the world [1-3]. The incidence ranges from 0.89-1.89/100,000 people (average 1.1) and 0.5 to 1.5 / 100,000 in children. Although all ages may be affected, the peak incidence ranges from 20 to 40 years and is predominantly male. In addition, there is an increase of approximately 20% every ten years after the first decade of life [1,4-6].

The clinical presentation of GBS begins with paresthesia's in the hands and feet, followed by progressive symmetrical muscle weakness, which begins in the lower limbs, which may last for hours or even a month [1,4,5]. In children, proximal weakness is less common, pain and difficulty in gait predominate and greater involvement of cranial pairs [7,8,9]. In approximately 70% of cases, patients had a previous infection about 1 to 3 weeks before. Generally, this infectious condition was of gastrointestinal or respiratory origin, being the most common etiological agents: Campylobacter jejuni, cytomegalovirus and Epstein-Barr virus, being the first agent associated with the most severe type of this syndrome, axonal [10].

There are four presentations of this syndrome, the most common being acute demyelinating inflammatory polyneuropathy (PAID) and acute motor axonal neuropathy (NAAM), followed by

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Miller-Fischer syndrome (SMF) and acute sensory and motor axonal neuropathy (NAASM). PAID corresponds to 85-95% of cases, while NAAM 10-20% [1].

The diagnosis is primarily clinical. However, complementary exams are necessary to confirm the diagnostic hypothesis and rule out other causes of flaccid paraparesis, such as cerebrospinal fluid (CSF) analysis and electroneuromyography [1,11,12]. After the diagnosis is confirmed, treatment with immunoglobulin or plasmapheresis is initiated, in addition to motor physical therapy [1,13].

In general, the prognosis in children is better when compared to adults, about 85% have an excellent recovery [14]. However, a small amount may be incapacitated or even progress to sepsis, pulmonary embolism and death [1,7].

Thus, we chose to report a case of a female patient of one year and one month of age, with a history of fever for 5 days, with progressive loss of strength, asymmetrical in the lower limbs. She was diagnosed with GBS, but with clinical presentation and atypical exams.

4. Case Presentation

Female patient, one year and one month old, was born at 38 weeks and 4 days, a caesarean section with 3080 g, 47 cm, uneventful during pregnancy, was performed prenatally correctly, without changes. It uses exclusive breastfeeding, walked at 1 year of age.

It was referred due to a history of low fever, accompanied by a flu and crying for five days, progressing to progressive loss in the lower limbs (lower limbs) since the onset of fever and has a concomitant cough. Follow up with a neurologist where macrocephaly has been reported since the age of six months and a magnetic resonance imaging (MRI) of the skull was prescribed, which showed no changes.

A child evolved to regular general state, with oscillations in respiratory rate (80 ipm), associated with tachycardia (185 bpm), slight worsening of strength in the left and right upper limbs, with ascension. In addition, she had generalized hypotonia and she cry the mobilization and cooling in the left lower limb.

It was evaluated by the neurosurgery team where the child had axial and bilateral lower limb hypotonia, positive babinski's sign on the right, patellar and Achilles' flexlexia, slight neck stiffness, lower limb painful hypoesthesia up to the xiphoid appendix. The diagnostic hypothesis of Guillan-Barré was made, as it presented two diagnostic criteria, namely: progressive weakness in more than one appendicular limb and hyporeflexia/vazlexia.

Cerebrospinal fluid (CSF) puncture was requested, where no protein increase, negative cultures, normal glucose and increased cellularity were detected. She was referred for electroneuromyography to confirm the diagnostic hypothesis. The examination detected: reduction of compound muscle action potentials, normal motor conduction velocity, indicating acute motor axonal neuropathy. Evolved with respiratory muscle arrest, requiring ICU space and orotracheal intubation. She was intubated for 10 days using immunoglobulin. There was improvement of the respiratory part, but still intubated, gradual recovery of movements and reflexes. Patient remains hospitalized in the care of neurosurgery and pediatrics.

5. Discussion

GBS was described in 1916 by Guillain, Barré and Strohl, who reported a case of two French soldiers who developed acute paralysis with muscle weakness, CLE and albuminocytological dissociation in the CSF. The combination of these characteristics became known as SGB [15]. Since polio eradication, GBS has become the most frequent cause of acute and subacute flaccid paralysis in the world [16-18].

Approximately 70% of cases appear about one to two weeks after an infection, either respiratory, gastrointestinal or that induces an aberrant autoimmune response. Most often it is benign and can

be minimized or forgotten by the patient [17-20]. Campylobacter jejuni is the most widely reported agent, other infections associated with this condition are those due to cytomegalovirus, Epstein-Barr virus, measles, influenza virus, Mycoplasma pneumonie, as well as enterovirus D68 and Zika virus [21-29].

One of the hallmarks of this syndrome is progressive, bilateral, ascending weakness that usually begins in the lower distal extremities but may begin more proximally in the lower and upper limbs, which may give the false impression of a pyramidal lesion [30-33]. Weakness peaks within 2 to 4 weeks after the onset of symptoms. A small number of patients have paraparesis during the disease period [33]. In addition to weakness, patients may initially have sensory signs, ataxia and characteristic autonomic dysfunction. About one third of patients have muscle or root pain before they even develop weakness [34]. Most patients have, or develop, reduced tendon reflexes in the affected limbs and may initially be normal [35]. This syndrome may progress for up to six weeks after its onset. About 20 to 30% of patients develop complications and may require mechanical ventilation [30].

Regarding the pathogenesis of this disease, it was considered for many years as a disorder whose severity was related to the extent of axonal lesion due to demyelination. However, it is currently known that there are several phenomena among them acute inflammatory demyelinating polyneuropathy, where the injury is effective in the immune system affects a myelin sheath, in addition to acute axonal motor neuropathy, where axonal membranes are the main [17]. It allows, occurs in healthy people and is not associated with autoimmune disorder or other systemic disorder. It is primarily a humorous mediator rather than T-cell mediated disorders [36].

It can be difficult to diagnose in young children because their responses are typically atypical and have a more challenging neurological examination [37].

Table 1: Diagnostic criteria of Guillain-Barré syndrome.

Clinical features
Progressive weakness over a period of up to 6 weeks in the legs and arms (sometimes only in the arms).
Hypo or Flexlexia (sometimes normal or even hyperreflexia).
Relative symmetry.
Mild sensory symptoms or signs.
Ache.
Autonomic Dysfunction.
Complications, such as respiratory failure, requiring ventilation, aspiration pneumonia, sepsis, cardiac arrhythmia, hyper or hypotension, and urinary retention.
Lumbar puncture
Cytoalbuminological dissociation (ie normal cell count with increased protein levels) in cerebrospinal fluid.
Nerve conduction study
Evident after 2 weeks, showing motor impairment and / or sensory amplitudes.
In the case of demyelinating polyneuropathy, prolonged distal motor latency, reduced nerve conduction velocity, prolonged F-wave latency, increased temporal dispersion, and conduction blocks.
Magnetic resonance
Post-gadolinium enhancement of peripheral nerve roots and equine tail.
Serum
Anti-ganglioside antibodies (in about 50% of patients).

Table 1 summarizes GBS diagnostic requirements that can be used in clinical trials that can be used for further research [38]. The use of two main tests, CSF evaluation and electroneuromyography (ENMG) is allowed. In the first, leukocytes separate within normal limits and should increase the value of albumin, thus having what they call albuminocytological dissociation. In the ENMG, it is

neuronal lesion, demyelinating or axonal [1,11]. Although not specific, post-gadolinium enhancement of peripheral nerve roots and the equine tail can be seen on MRI in up to 95% of patients [39,40].

The neurological examination helps in the diagnosis, as there is symmetrical weakness of the lower limbs with decreased or absent osteotendinous reflexes and neuropathic pain.

The Hughes Scale (Table 2) is used to assess the patient's motor impairment [1].

Table 2: Hughes scale.

0 Totally healthy.
1 Minimal signs and symptoms, walkable.
2 Able to walk more than five minutes without assistance but unable to run.
3 Able to walk more than five minutes with astonishment.
4 Bed or Wheelchair.
5 Requires ventilatory assistance for at least part of the day.
6 Death.

In general, it is a life-threatening disease, with a mortality rate of 3-7% [41,42]. There is a higher prevalence of death due to ventilatory insufficiency, complications or autonomic dysfunction [43]. Patients show improvement mainly during one year from the beginning of GBS. Worst outcomes are usually associated with older age (> 40 years), diarrhea or C. jejuni infection in the four weeks preceding the disease.

Treatment requires a multidisciplinary approach consisting of general medical care in addition to immunological treatment (Table 3).

Table 3: Therapeutic approach to Guillain-Barré syndrome.

General Health Care
Respiratory, cardiac and hemodynamic monitoring.
Deep venous thrombosis prophylaxis.
Management of possible bladder and bowel dysfunctions.
Early physical therapy and rehabilitation.
Psychosocial support.
Treatment of pain using non-steroidal opioids or anti-inflammatory drugs.
Immune treatment with documented efficacy
Intravenous immunoglobulin therapy.
Plasmapheresis.
New controls under evaluation
Interferon-beta.
Cyclophosphamide.
Rituximab.
Eculizumab.

Respiratory, cardiac and hemodynamic function should be monitored, in addition to preventing or treating complications [17]. For pain control, the use of non-steroidal opioids or anti-inflammatories is recommended [44]. Although there is no specific drug, various drugs have been used to target the components of the immune response. Immunomodulatory treatments with immunoglobulin or plasmapheresis have been shown to be effective in accelerating recovery in addition to improving outcome [1,45].

6. Conclusion

It is very important to emphasize that if you are facing a suspicion of GBS, even if it is atypical, you should hospitalize the patient in a pediatric intensive care unit, where he should remain monitored and frequently monitored, providing life support and reducing frequency and severity of complications. In addition, treatment should not be delayed and the individual should be adequately monitored for hemodynamic and ventilatory aspects in order to detect autonomic changes early.

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