

Atypical Presentation Syndrome of Guillain-Barré

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Abstract

A case of Guillain-Barre syndrome with autonomic bladder and bowel involvement is described. It's a girl for two years and two months, which debuts with a clinical picture of decay, irritability, vesicular lesions on the lips and fever. In its evolution presents nausea, vomiting, abdominal pain and difficulty in miction. Urological and surgical pathology is discarded. On the third day of clinical evolution develops bladder balloon, absence of miction, constipation, difficulty in ambulation. The neurological examination normal osteotendinous reflexes, muscle strength in the lower limbs 4/5 inconsolable crying to standing, faltering gait. In magnetic resonance imaging of spinal cord a thickening of the nerve roots at the level of the cauda equina and enhancement is identified in sequence T1 with contrast medium. Under suspicion of a syndrome of Guillain-Barre of atypical presentation, gets cerebrospinal fluid which showed 3 cel/mm³, 183 mg/dl protein leukocytes. Immunoglobulin was started a total dose of 2 g/kg given as 400 mg/kg for five days with clinical improvement at 72 hours while walking and static, spontaneous voiding reappears.

Conclusion: Clinical involvement with regional commitment and the correlation with a dissociation albumino - cytological, an enhancement to the gadolinium-based contrast in cauda equina, integrated a diagnosis atypical of Guillain Barre Syndrome.

Keywords: *Guillain-Barre Syndrome; Autonomic Symptoms; Presentation Atypical; Bladder Commitment*

Introduction

Guillain Barré syndrome (GBS) is an acute polyneuropathy mediated immunologically causing motor, sensory and autonomic dysfunction [1]. It is the main cause of acute paralysis in childhood worldwide having an estimated incidence ranged from 0.34/100,000/year to 1.34/100,000/year [2]. It can compromise any myelinated peripheral nerve at any age including the intrauterine or neonatal period, but it seems to dominate in adulthood and its frequency increases with age [3,4].

The GBS has regional variants of presentation: acute inflammatory demyelinating polyradiculopathy (AIDP), acute motor axonal neuropathy (AMAN), axonal motor-sensory neuropathy (AMSAN) and Miller Fisher syndrome.

The pathophysiology of GBS is still under investigation. It is postulated that in the AIDP there is an autoimmune response triggered by a bacterial or viral infection against antigens located in peripheral nerves, which produces degeneration of myelin and a block in nerve conduction [3].

In the AMAN and AMSAN forms, there is no participation of the myelin sheath. Anti-ganglioside antibodies have been detected related to *Campylobacter jejuni* enteritis, which bind to the motor fibers of the axon and block nerve conduction at the level of Ranvier nodules [4]. In the AMAN form, the damage occurs at the level of the peripheral nerves, mainly affecting motor nerves, whereas in AMSAN there is motor and sensory compromise [5].

Miller Fisher syndrome is characterized by ataxia-ophthalmoplegia and areflexia and muscle weakness is absent or scarce [3,4].

Classically, the disease has been divided into three phases: acute or onset phase in which the disease begins with symmetric paresthesia and pain in the feet, occasionally in the hands. Subsequently, symmetric ascending and progressive paresis or flaccid paralysis develops, usually manifested as impaired gait and decreased or absent osteotendinous reflexes. It progress in a sub-acute manner up to a maximum of four weeks, followed by the state phase with a variable duration. The last phase referred to as remission consists of the reversal of the symptoms in the reverse order over a period of weeks or years [1,6].

Guillain Barré syndrome in childhood has atypical variants and the classical presentation is not always present. Karimzadeh P, *et al.* [7] and Mabrouk E., *et al.* [8] found atypical signs and symptoms in 24.2% and 25.8% of children with GBS who required diagnostic confirmation by cerebrospinal fluid study magnetic resonance and electrophysiological studies [7,8].

Because GBS is a disease whose initial symptoms is not specific and may be modified by their atypical variants, there is usually difficulty in detecting them and delaying their diagnosis. Therefore, knowledge about clinical heterogeneity and a high index of suspicion is required for its timely recognition. The following is a case of GBS with atypical presentation and urinary retention.

Case Presentation

It is a female patient of two years of age, with good growth and development, previously healthy who twelve days before the onset of the picture presented oral ulcerative lesions, asthenia, irritability and thermal rise. Previously his brother of eight years, he had a similar clinical picture. The clinical picture debuts with nausea, vomiting and continuous abdominal pain, difficulty urinating. It is assessed by a pediatrician, integrates a probable urinary tract infection and initiates antibiotic and antiemetic.

Without improvement, on the third day of evolution she is taken to the emergency department, an acute inflammatory abdomen impresses, in her twenty-four hour evolution surgeon rules out appendiceal pathology. Abdominal distension, constipation, bladder balloon are reported, and permanent bladder catheter is placed in the bladder, showing urine output in large volume and relief of abdominal pain. On the sixth day, it is assessed by a urologist who rules out basic pathology. Bladder catheter is removed, but after a few hours bladder balloon reappears and difficulty urinating, bladder catheter is reinstated. On the tenth day of evolution it is valued by neuropediatric, to the re-interrogation directed it is emphasized that from the third day of evolution the girl remains without wanting to walk and in bed. The neurological examination is irritable, easy cry, without alteration of alertness, remains in bed seated, moves four extremities, global osteotendinous reflexes present, without increase of area or diffusion, no pyramidal elements, to the standing appears inconsolable crying, walking hesitant. MRI of the spinal cord is requested, identifying in T1 sequence thickening of the nerve roots at the cauda equina level and enhancement with the contrast medium (Figure 1).

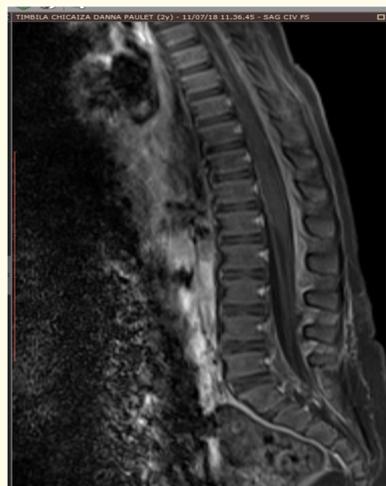


Figure 1: In sequence T1, Gadolinium: thickening of the nerve roots of cauda equina.

With a suspected diagnosis of Guillain Barré syndrome, cerebrospinal fluid is obtained in which the presence of leukocytes of 3 cells/mm³, 183 mg/dl proteins was demonstrated. On the twelfth day of evolution, UTI is established, bladder catheter is removed, spontaneous voiding reappears, it persists with difficulty in walking. Immunoglobulin initiation is considered a total dose of 2 g/kg given as 400 mg/kg for five days, electrical study with electromyography and nerve conduction velocity is requested, and physical rehabilitation is initiated. Clinical improvement is evident at 72 hours in the march and static, spontaneous voiding reappears. The patient remains in clinical follow-up and no clinical relapse is reported.

Discussion

Guillain Barré syndrome is a heterogeneous disorder caused by an autoimmune inflammatory polyradiculopathy. Its clinical manifestation in childhood is heterogeneous, which requires an exhaustive clinical and paraclinical assessment for its diagnostic confirmation.

In the year 2007, Korinthenberg, *et al.* [9] conducted a prospective multicenter study with 95 children to describe the presentation and course of GBS in childhood. The first signs of presentation with greater frequency were instability for gait (45%), neuropathic pain (34%) and inability to walk (24%). At the time of diagnosis, all patients had symmetric weakness and hypo or areflexia, 27% of the children had cranial nerve dysfunction and 33% autonomic dysfunction. After an average of seven days, the maximum point of the disease was reached, highlighting that 60% of patients had inability to walk, 45% had cranial nerve dysfunction and 51% had autonomic dysfunction.

In reference to the present case, autonomic dysfunction is common in children with GBS and is present in 50 - 70% of patients during the phase of greatest weakness. It is presented by the involvement of autonomic fibers of peripheral nerves and manifests with arrhythmias, diaphoresis, constipation, urinary dysfunction, paralytic ileus, hypotension or arterial hypertension. The presence of autonomic symptoms have been related to worse prognosis as well as mechanical ventilation requirement [3,6,10].

There are reports in the medical literature of adults with GBS and bladder dysfunction, however reports in children are scarce. The incidence of urinary disorders in children with GBS is variable, is described between 15% and 60% and is present more frequently in motor forms (AMAN) [10,11]. It is postulated that urinary dysfunction in GBS occurs due to an erratic transmission of the lumbosacral autonomic fibers affected by the inflammatory or immune process as well as the alteration of the inhibitory spinal interneurons, which generates an autonomic hyperactivity [12,13].

The most common urinary symptoms are urinary retention, difficulty in emptying and urinary urgency [12,13]. They usually appear in the period of maximum weakness and are rarely reported as initial presentation of GBS and it has been proposed that the presence of bladder involvement in early stages of the disease or persistently excludes GBS and suggests acute myelopathy [11-13].

However, there are cohort studies in children with GBS who report the presence of sphincter involvement in the early stages of the disease [7,8]. In the same way Wu Shih, *et al.* reported a case of a 60-year-old patient with rapidly progressive GBS to respiratory failure whose initial presentation was urinary retention [14]. In our case, there was difficulty in urination and bladder balloon on the third day of clinical evolution.

The presence of osteotendinous reflexes (ROTS) has also been the subject of study in patients with GBS. There is a report of patients with GBS secondary to enteritis due to *Campylobacter jejuni* with normal or exaggerated ROTs which differs from the areflexia described classically. Yuki N., *et al.* [5] conducted a study which describes that 10% of patients with GBS showed normal or hyperexcitable ROTs in the course of the disease. In addition, a non-statistically significant association between GBS with conserved ROTs and a history of diarrhea, pure motor involvement with higher AMAN frequency and presence of immunoglobulin G (IgG) antibodies against gangliosides.

The mechanism of hyperreflexia in AMAN is still unknown but it is suggested that the inflammation in the nerve roots could produce alteration of the blood-brain barrier of the central nervous system in such a way that the anti-ganglioside antibodies would reach the intramedullary collateral branches of the inhibitory interneurons that they cause the disinhibition of the lower motor neurons. The mentioned study suggests considering the presence of AMAN related to *C. jejuni* in patients with motor and normal or hyper reflexia. In addition, it was observed that some patients showed areflexia during the acute phase and hyperreflexia during the recovery phase, which indicates that there is an excitability spectrum of osteotendinous reflexes in the GBS. In the present case, the ROTs were always present.

The diagnosis of GBS is based on clinical assessment supported by nerve conduction studies and CSF analysis. However, neuroconduction findings are usually normal in 20% of patients during the first three weeks of illness. Similarly, albumin cytologic dissociation observed in CSF studies may not be present until the second week of symptoms and is not specific as it is described in infections, demyelination or spinal blockage by cord compression.

For these reasons, spinal MRI has become important in GBS, especially in atypical cases or in situations where neuropediatrician is not available. It has been described that children with GBS present a pathological thickening of the nerve roots after the administration of contrast (gadolinium). MRI does not replace neuroconduction studies as a primary evaluation but its usefulness lies both in the support of clinical suspicion as well as in excluding important differential diagnoses and the lumbar puncture can be performed safely [16].

Conclusion

The previously described can guide us to expand our view of the heterogeneous clinical dimension of Guillain Barré syndrome in childhood. In the present case, an autonomic bladder, subtle intestinal involvement or involvement, accompanied by a minimal paresis to crural predominations, normorreflexia that integrated to a protein/cytological dissociation in cerebrospinal fluid and to a contrast enhancement in cauda equina, supported the atypical presentation of this entity.

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