Child Neurology Open
October - December 2015: 1-6

DOI: 10.1177/2329048X15609053
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A New Observation of an Atypical and Severe Variant of the Guillain-Barre Syndrome in a Child: Remaining Challenges for Diagnosis, Nosologic Classification, and Therapeutic Course

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Abstract

Guillain-Barré syndrome is a rare acute polyradiculoneuropathy. Several variants and unusual presentations have been described, particularly in pediatrics. In most cases, making an early diagnosis is challenging due to the treatments that consist in the rapid administration of intravenous immunoglobulin or plasma exchange. The authors present the case of a 7-year-old boy with an atypical and severe axonal Guillain-Barré syndrome, associated with *Mycoplasma pneumonia*. When he was admitted, febrile respiratory failure was the main focus, and then he presented signs of acute polyneuropathy with cranial nerve palsy and brief hyperreflexia. Mechanical ventilation was required for 48 days as well as 2 cycles of intravenous immunoglobulin. The authors describe all the medical challenges that the authors encountered. This case highlights the fact that respiratory distress can be the main clinical symptom in children. This delays the establishment of a correct diagnosis, even more so when neurological manifestations are abundant and unusual.

Keywords

Guillain-Barré syndrome, pediatrics, *Mycoplasma pneumonia*

Received June 19, 2015. Received revised June 19, 2015. Accepted for publication July 6, 2015.

Guillain-Barré syndrome is a rare polyradiculoneuropathy. It is characterized by rapidly progressive symmetric muscle flaccid weakness with areflexia and potentially life-threatening complications (acute respiratory failure, swallowing disorders, and dysautonomia), followed by a stabilization phase and then slow recovery. It was first described in 1916 by French neurologists Guillain, Barré, and Strohl.[1] Similar cases had already been described by Landry in 1859.[2]

The annual incidence ranges between 0.89 and 1.89 cases (median 1.11) per 100 000 person-years, although a 20% increase can be seen with every 10-year rise in age after the first decade of life.[3]

Two-thirds of cases are preceded by symptoms of upper respiratory tract infection or diarrhea. Infectious agents associated with the development of the Guillain-Barré syndrome are *Campylobacter jejuni*, Cytomegalovirus, Epstein-Barr virus, and *Mycoplasma pneumonia*.[4-6] It is considered to be an immune-mediated disorder, with autoantibodies produced by molecular mimicry.[7]

Several variants have been described, such as acute inflammatory demyelinating polyneuropathy, acute motor axonal neuropathy, acute motor-sensory axonal neuropathy, acute

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motor-conduction block neuropathy, Miller-Fisher syndrome (ophthalmoplegia, ataxia and areflexia), Bickerstaff's brain stem encephalitis (alteration in consciousness, paradoxal hyperreflexia, ataxia, and ophthalmoparesis), a pharyngeal–cervical–brachial variant (ptosis, facial, pharyngeal, and neck flexor muscle weakness that spreads to the arms and spares leg strength), and other regional variants.[8-12]

Establishing a diagnosis for Guillain-Barré syndrome can be difficult, particularly in pediatrics, where rare unusual presentations occur, such as acute respiratory distress at the beginning.[13-24] Making an early diagnosis is challenging due to the treatments that consist in the rapid administration of intravenous immunoglobulin or plasma exchange.[25]

The authors present the case of a 7-year-old boy with an atypical and severe Guillain-Barré syndrome, and the authors describe all the challenges that the authors encountered to establish a diagnosis and for the medical management of the patient.

Case Report

Our patient was a previously healthy 7-year-old boy. Vaccines were up to date, except for a booster dose of the combined diphtheria, tetanus, and inactivated poliovirus vaccine. His mother presented a common variable immune deficiency that was treated with weekly subcutaneous immunoglobulin. The patient had not undergone previous screenings.

The symptoms began with a high fever around 39°C, coughing, pain in the legs, and dysesthesia with a waddling gait. His sister had had a flu-like syndrome 1 week before. At first, the treatment was symptomatic.

Aggravations occurred 5 days later with the deterioration of the patient's general state, abdominal pain, food intolerance, and acute respiratory failure.

On arrival at the pediatric emergency department, he presented tachycardia (150 beats/min), normal blood pressure, respiratory retraction, abdominal breathing, diffuse rhonchi, and severe hypoxemia (oxygen saturation of 70%) without hypercapnia. A brief neurological examination was unremarkable. A chest X-ray study showed intense bilateral interstitial pneumonia.

The patient was transferred to our pediatric intensive care unit. He required noninvasive ventilation by high-flow oxygen therapy with up to 65% Fio_2. The next day, a chest X-ray control showed right basal consolidation. At that time, his parents reported a dribble on the way to the hospital with probable aspiration.

On day 2, a strikingly rapid neurological deterioration occurred, which consisted in agitation and in bilateral mydriasis but not clonism, and gave rise to suspicions of convulsion, which led to intubation, sedation, and mechanical ventilation.

The neurological manifestations progressed to an unclassical bulbar and peripheral motor deficit, predominant on the upper limb with a brief central involvement. All those manifestations were noticed after respiratory stabilization occurred, and sedation was released on day 4. No seizures were documented.

The patient had normal consciousness. He could not speak because of ventilation but could communicate with eye movements and blinking. There were no spontaneous movements of his arms and legs.

Dysfunctions in the cranial nerves became progressively evident and included facial diplegia, bilateral ptosis and mydriasis without ophthalmoplegia, severe swallowing disorders, and the absence of cough.

The tetraparesia was predominant on the upper limb. It was associated with a brief hyperreflexia without a Babinski reflex for 2 days and then with areflexia, leading to the possibility of the Guillain-Barré syndrome. Dysautonomia was not clearly diagnosed, but the patient needed 4 vascular fillings in the first 3 days to be hemodynamically stabilized. However, he never had hypertension or heart rate lability.

A large-scale screening was performed. *Mycoplasma pneumonia* and *Haemophilus influenza* were respectively identified in nasopharyngeal and tracheal secretions. The blood test showed inflammatory signs with increased C-reactive protein 116 mg/L (N <5 mg/L) and procalcitonin 11 μg/L (N < 0.5 μg/L).

Toxicology studies were negative. Other microbiological tests (Lyme, botulism, respiratory viruses, bacteria and fungi, HIV, pneumocystis, and aspergillum) and immune tests were normal. Two examinations of cerebrospinal fluid performed on days 2 and 5 didn't reveal a cytoalbuminological dissociation (white blood cells maximum 5/mm^3, normal glucose, normal protein and lactate levels, and sterile) or oligoclonal bands.

The analysis for antiganglioside and antigalactocerebroside antibodies detected nonspecific anti-GM4 on day 2 and were negative on day 26. The cerebral tomography scan was normal on day 2. Two cerebromedullar magnetic resonances on days 5 and 11 were normal, without any enhancements of the nerve roots.

An ultrasonography was performed on day 21, when the patient was still completely dependent on invasive ventilation. The ultrasonography showed phrenic paresia. On day 2, an electroencephalogram showed no anomalies for sleep under sedation. Sensitive evoked potentials were normal on day 19.

An electromyogram on day 8 (Figure 1) showed a normal lower limb and the decreased amplitude of motor responses on the upper limb, progressing into an axonal affection on the control electromyogram performed on day 18 (Figures 2 and 3), confirming the diagnosis of an axonal motor polyneuropathy.

The medical management of the patient consisted in administering a first course of intravenous immunoglobulin (1 g/kg/d for 2 consecutive days) on days 5 and 6. It was prescribed because of clinical severity, despite the lack of confirmation on the diagnosis. This treatment didn't result in clinical improvement. Three weeks later, the authors decided to conduct a second course of immunoglobulin because the patient remained tetraparetic and still depended on assisted ventilation. Recovery started quietly and then progressed slowly and regularly.

Nerves	Latency ms	Amplitude 1-2 mV	Velocity m/s	Amplitude S 2-4 %
RIGHT MEDIAN				
Wrist	2,70	1,027		100
Elbow	6,45	0,132	46,7	1,76
RIGHT ULNAR				
Wrist	2,20	0,723		100
Elbow	5,00	0,663	58,9	83,4
Upper elbow	6,00	0,735	75,0	112

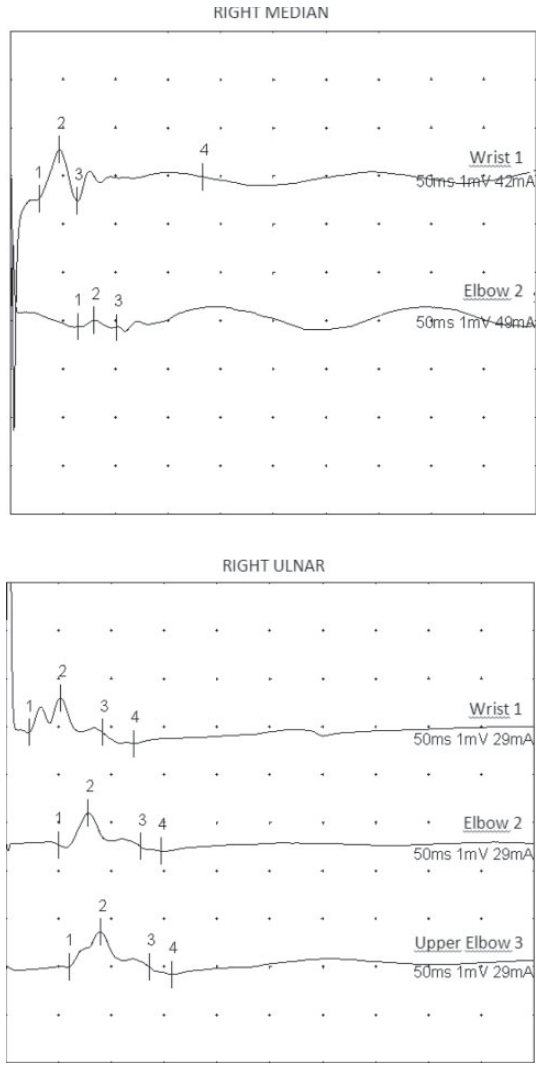

Figure 1. Electromyogram recordings of motor nerves on day 8. Decreased amplitude of motor responses on the upper limb. Normal conduction velocity.

Nerves	Latency ms	Amplitude 1-2 mV	Velocity m/s	Amplitude S 2-4 %
RIGHT MEDIAN				
Wrist	3,55	0,498		100
Elbow	7,25	0,247	47,3	59,8
RIGHT ULNAR				
Wrist	2,55	1,533		100
Elbow	5,45	1,327	56,9	89
Upper elbow	6,65	1,222	58,3	104

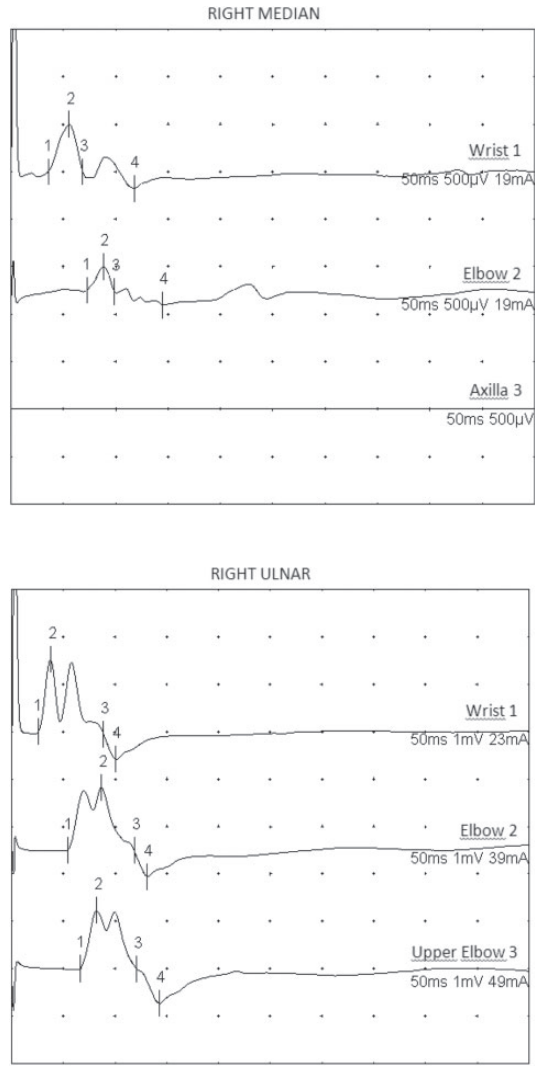

Figure 2. Electromyogram recordings of motor nerves on day 18. Decreased amplitude of motor responses on the upper limb. Normal conduction velocity.

When the patient was admitted, he was suspected of being immunodeficient and was therefore treated with a large probabilistic association of medication, including acyclovir, clarithromycin, ceftriaxone, cotrimoxazole, and oseltamivir. Of all the medication, clarithromycin was the only drug that was administered for 21 days. Amoxicillin–clavulanate was associated with it for 8 days in order to treat ventilator-assisted pneumonia.

Mechanical ventilation was necessary for a total of 48 days. However, the oxygen requirement rapidly decreased, but muscle weakness and swallowing troubles compromised autonomous breathing for a long time. Since the neurological prognosis was guarded, a tracheostomy was performed on day

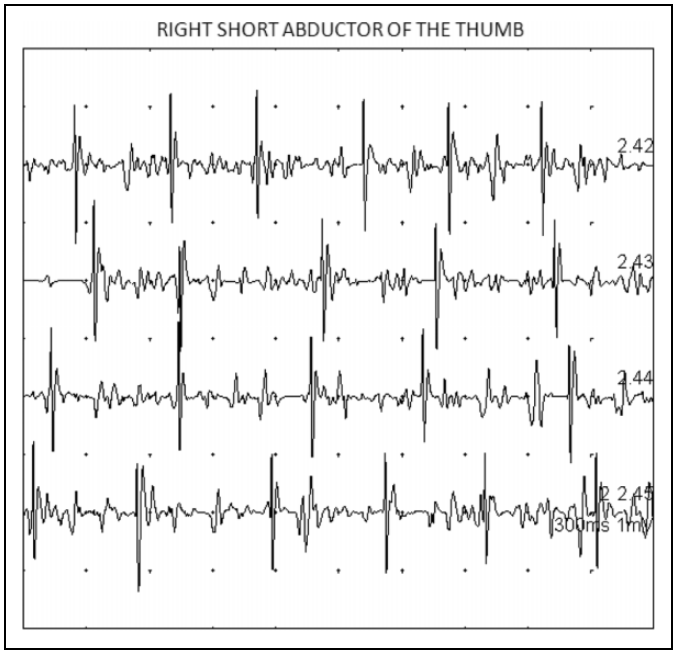

Figure 3. Electromyogram of the right short abductor of the thumb on day 18. Neurogenic muscle activity.

29, improving comfort. The patient could then be progressively taken off ventilation support.

Enteral feeding for 18 weeks (first with a gastric tube and then through gastrostomy from day 55) and salt supplementation, because of intense salt loss when swallowing, were required. Then speech and oral intake improved. Once life-threatening complications were excluded, the patient started intensive functional and psychotherapeutic rehabilitation.

The patient's progress was slowly favorable, and he left the hospital after 11 weeks. An examination after 20 weeks only showed a proximal weakness and decreased upper limb deep tendon reflexes. Walking was fluid. No breathing or swallowing troubles persisted. An electromyogram was conducted as a control after 18 weeks. It showed definite improvements in the upper limb.

The tracheostomy and the gastrostomy could be removed after 5 months without any complications. During the last examination, 1 year after the patient's admission to the hospital, the authors still noticed discrete hypotonia in the upper limb with fine motor skill difficulties, fatigability, and intermittent hand tremors. Deep tendon reflexes were normal. The electromyogram was still improving and showed only a discrete neurogenic involvement.

Discussion

The authors present the case of an atypical and severe axonal Guillain-Barré syndrome, associated with a *M pneumonia* lung infection, in a 7-year-old boy. Difficulties were encountered to establish a diagnosis. The elaboration of a specific treatment was therefore delayed.

The authors initially focused on respiratory distress, due to the fact that our patient first presented febrile acute respiratory failure linked to an interstitial pneumonia, in a context of pain and probable weakness of the lower limb. Swallowing disorders and perhaps aspiration were retrospectively reported. Lower limb weakness seemed to be the first sign, but it was associated with pain and dysesthesia, which mimicked flu-like diffuse myalgia. Neurological disorders were only noticed when vital respiratory functions had been managed through mechanical ventilation and through the release of sedation.

The respiratory emergency was the main focus when the patient was admitted with severe hypoxemia. As it has previously been reported, this delayed the neurologic diagnosis.[26] It is well known that in emergency departments, where clinical presentations can be elusive at first, misdiagnoses or delayed diagnoses are not uncommon.[27]

Furthermore, the neurological manifestations were unusual, thus making the diagnosis more challenging. First, the neurological evolution rapidly worsened to nadir. It was concomitant with the *M pneumonia* respiratory infection. The authors didn't observe the typical delay that usually occurs between the infectious trigger and the acute polyneuropathy. The authors recall the fact that *M pneumonia* can be the trigger for large peripheral and central neurological manifestations, through several sorts of pathomechanisms, making it more difficult to identify the correct diagnosis.[28-31]

Second, neurological symptoms were misleading because of the association between central and peripheral involvement disorders: involvement of cranial nerves, tetraparesia that was predominant on the upper limb,[32] and deep tendon reflexes first noted as normal, then brisk,[33-35] and finally depressed.

Those brief central manifestations were reminiscent of Bickerstaff's brain stem encephalitis. However, there wasn't a disturbance of consciousness, nor was there ophthalmoplegia, or ataxia. Abnormal T2 hyperintensities weren't found on the 2 cerebromedullar magnetic resonances.[36,37]

Third, the whole paraclinical screening was also misleading. The 2 lumbar punctures didn't serve as support, as they did not document any albuminological dissociations. The examination of antiganglioside and antigalactocerebroside antibodies didn't help either and were both negative.

The repeated use of the electromyogram was the only tool that led to the certain diagnosis of an axonal, bulbar, and predominant on the upper limb Guillain-Barré syndrome.

This case report highlights the fact that the Guillain-Barré syndrome is a large nosologic framework that encompasses several clinical variants and overlaps with Bickerstaff's brain stem encephalitis. This forms a continuous spectrum in which the diagnosis of especially atypical cases can be particularly difficult to establish.

However, establishing an early diagnosis and thus initiating a specific immunotherapy treatment is a real challenge. In severe cases, the Cochrane Database recommends intravenous immunoglobulin.[25] In this case, the first course of immunoglobulin was only administrated on days 5 and 6, although it started before the electromyogram confirmation, which points

to the difficulties met to find the diagnosis. The intravenous immunoglobulin treatment did not contribute to make significant improvements. Because of the severity of such atypical forms, and due to the guarded neurological prognosis in such cases, the authors decided to administer a second course 3 weeks later. This strategy is rare but recommended for severe Guillain-Barré syndrome cases.[38,39] Determining the nosologic framework was also important since the question of adjuvant treatments—such as intravenous corticoids—in the event of Bickerstaff's encephalitis had been discussed.[36]

Finally, reanimation supporting care was essential for our patient who needed 48 days of invasive ventilation assistance and was therefore in the most severe range of ventilation duration. Of the patients with Guillain-Barré syndrome, 17% to 30% require mechanical ventilation with an increased risk of death. Nearly 5% of patients with Guillain-Barré syndrome die from medical complications such as sepsis, pulmonary emboli, or unexplained cardiac arrest. The mean duration of ventilation ranges between 15 and 43 days, suggesting that a proportion of patients can receive a tracheostomy for better comfort and airway safety.[40]

Usually, poor prognostic factors are related to age, clinical stages, and electrophysiological subtypes (axonal form).[41,42] This confirms the severity of our case and the urgent need to provide a correct diagnosis and immunotherapy to fight against a poor outcome.[43-45]

Conclusion

Guillain-Barré syndrome is a life-threatening neurological emergency in pediatrics. This case report highlights the fact that respiratory distress can be the main clinical symptom in children, thus delaying the correct diagnosis of Guillain-Barré syndrome, even more so when rare and unusual neurological manifestations are combined, although they have already been reported separately within the paediatric population. This case also highlights the importance of recognizing the constellation of Guillain-Barré symptoms across a spectrum, especially in children, in order to ensure the rapid and efficient medical management of patients, particularly in severe forms.

Acknowledgments

The authors wish to thank the patient and his family for their kind cooperation, all our colleagues who provided their help for this article and the Bibliothèque scientifique de l'Internat de Lyon and the Hospices Civils de Lyon for their grant support.

Author Contribution

All authors cared for the patient. VM analyzed the electromyograms data. LP wrote the first draft of the manuscript, and all other authors contributed to the final form of manuscript.

Informed Consent

Informed written consent for publication of this case was obtained from the patient and his parents (mother and father).

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

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