

CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Severe painful lower limbs and refusal of the leg reliance as atypical presentation of Guillain–Barre syndrome

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Introduction Guillain–Barre syndrome (GBS) is the most common cause of acute flaccid paralysis in healthy infants and children. Acute motor axonal neuropathy (AMAN) is a type of GBS characterized by motor syndrome with no sensory symptoms.

Case outline Authors describe a six-and-a-half year old girl with atypical clinical presentation of AMAN with severe painful lower limbs and refusal of the leg reliance with typical findings on nerves conduction studies.

Conclusion Despite the nerve conduction study findings, atypical forms of AMAN and GBS are possible. Pain symptoms must be taken very seriously and treated careful by the clinicians.

Keywords: Guillain–Barre syndrome; acute motor axonal neuropathy; atypical presentation

INTRODUCTION

Guillain-Barre syndrome (GBS) is an acute, immune-mediated, demyelinating, peripheral neuropathy. GBS is the most common cause of acute flaccid paralysis in healthy infants and children [1]. Classic presentation and symptoms of GBS include ascending muscle weakness with sensory symptoms being a relatively minor feature and decreased or absent muscle tendon reflexes. GBS patients were divided into those with acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor and sensory axonal neuropathy and Miller Fisher syndrome [2]. The most frequent subtype of GBS in North America and Europe is AIDP, which accounts for up to 90% of GBS cases [3, 4]. AMAN is characterized clinically by nearly pure motor syndrome without sensory involvement and final diagnosis of AMAN is based on electrophysiological findings such as decreased amplitude of compound muscle action potential (CMAP) without any evidence of demyelination or change in sensory nerve action potential (SNAP) [5].

In this paper, we shall report on an atypical presentation of AMAN with severe pain in the lower limbs and refusal of the legs reliance with typical decreased amplitude of CMAP without any evidence of change in SNAP.

CASE REPORT

A six-and-a-half year old girl was admitted to the emergency department of the University

Children's Hospital. She was somnolent with severe pain and muscle weakness in lower limbs, aphthous ulcers in the mouth and prostration. The symptoms started seven days before admission with a high fever of 39°C and aphthous ulcers in the mouth. The next day, she complained about exhaustion, severe pain, and muscle weakness in lower limbs and she was unable to stand up and walk, so she was admitted to the regional hospital. The following day, her mother found her unconscious in bed, non-responding, with eyes and jaws fixed and livid lips with saliva leaking, so she was immediately transferred to the University Children's Hospital.

On admission to the emergency department of the University Children's Hospital, the girl was somnolent but responsive when called, afebrile, eupneic, acyanotic, and anicteric, pale and cold peripherally. Neurological findings included somnolent, disoriented. Beside dysarthria, other cranial nerves were intact. On upper limbs mild hypotonia, hyporeflexia and mild muscle weakness was presented. On lower limbs, severe muscle weakness and hypotonia were found with areflexia. Babinski sign was negative. Meningeal reactions were positive. Sensations on upper limbs were normal, on lower limbs with severe painful sensations, which were presented until the end of the hospitalization and beside the muscle weakness in the lower limbs, the most dominant clinical symptom. There was no loss of sensory stimuli or pins and-needles sensation. The intestine and bladder sphincters were intact. Autonomic functions were normal.

Personal and family history revealed no specific information. Neurological development

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was compatible with her age and all of immunizations were on schedule with no recent immunizations.

Blood tests revealed elevated C-reactive protein 42.5 and creatine kinase 257 U/L. Later on these parameters decreased. Analysis of the cerebrospinal fluid revealed mild protein content (0.648 g/L), 20 leukocytes, and glucose was 3.7 mmol/L. The serologic tests were negative for the herpes virus, rubella, Epstein–Barr virus, rubeola, toxoplasmosis, cytomegalovirus, enteroviruses, respiratory viruses, Lyme disease, and *Mycoplasma pneumoniae*. There were no microbial proliferations in the culture for *Salmonella*, *Shigella* and *Campylobacter jejuni*. Computed tomography scan of the endocranium was within the normal range. The child underwent nerve conduction studies (NCS). The findings of motor NCS in the lower limbs showed reduced amplitude of CMAPs, whereas distal latencies and motor conduction velocities were normal. The findings in the upper limbs showed also reduced amplitudes, whereas distal latencies and motor conduction velocities were normal. Findings of sensory NCS were interpreted as within normative ranges with normal values for SNAP, latencies and velocities (Table 1).

Table 1. Nerve conduction studies values for tested nerves

Type of nerve	Name of nerve	R/L ms (Normal values) [6]	R/L mV (Normal values) [6]	R/L m/s (Normal values) [6]
		Distal latency	CMAP	NCS
Motor	Median	2.85/2.8 (≤ 4.4)	2.8/3 (≥ 4)	55/58 (≥ 49)
	Ulnar	2/2.1 (≤ 3.3)	4.5/4.7 (≥ 6)	62/60 (≥ 49)
	Peroneal	2/1.8 (≤ 6.5)	1.1/1 (≥ 2)	55.1/55.7 (≥ 44)
	Tibial	3.15/3.2 (≤ 5.8)	1.2/1 (≥ 4)	54.3/50.3 (≥ 41)
		Latency	SNAP	NCS
Sensory	Sural	1.7/1.7 (≤ 4.4)	12.9/14 (≥ 6)	50.1/47.7 (≥ 40)

CMAP – compound muscle action potential; NCS – nerve conduction studies; SNAP – sensory nerve action potential

Based on clinical features with motor involvement, electrophysiological investigation, as well as the normal laboratory findings, this patient was diagnosed with AMAN subtype of GBS. Findings of sensory NCSs were normal in spite of the severe pain in the lower limbs.

The patient was treated with intravenous immunoglobulin and general supportive therapy. She did not require respiratory support. Rehabilitation treatment started early, right after the stabilization of the general condition. Muscle strength of the lower and upper limbs started to improve immediately after medication, but pain did not decrease with the introduction of Carbamazepine, so she required opioids for pain management in the beginning. After five weeks, muscle strength on the upper limbs was grade five and muscle strength on lower limbs had improved to grade four, so she could stand independently, but severe pain in the lower limbs persisted and the child refused to stand up and walk. Eight weeks after admission, muscle strength on lower limbs improved to grade five and painful sensations decreased so she could stand up and walk independently.

Three months later, at the outpatients' follow-up, she had fully recovered.

DISCUSSION

Guillain-Barre syndrome (GBS) is an acute, immune-mediated, demyelinating, peripheral neuropathy. The disease is thought to be autoimmune and triggered by a preceding infection in two thirds of cases, most frequently respiratory or gastrointestinal infections [7, 8].

Campylobacter jejuni infection has been associated with GBS. GBS that occurs after *Campylobacter jejuni* infection is usually more severe related with extensive axonal injury [9]. The following infections have also been associated with GBS: cytomegalovirus, influenza, *mycoplasma pneumoniae*, Epstein–Barr virus infection, or mononucleosis, HIV or AIDS. The incidence of GBS after immunization was not different from the background incidence of GBS, thereby precluding any firm conclusions about the significance of these findings. However, because of the close temporal association of GBS with selected vaccines, the risks and benefits of immunization merit individual review by the clinician and patient [3]. The incidence of GBS is between 1.1/100,000/year and 1.8/100,000/year with lower rates reported in children (< 16 years) of around 0.6/100,000/year. The age of onset of GBS in our patient was in accordance with previous reports [4]. The review reported mostly on studies from Europe and North America [8]. As it was previously stated that the frequency of AIDP subtype of GBS is high in US and Western Europe, it should be stated as well that the AMAN is frequent (up to 65%) in the study conducted in China [4]. NCS and cerebrospinal fluid analysis is important investigations that help confirm the diagnosis of GBS [5]. Although GBS is a relatively uncommon condition, the consequences of a missed or delayed diagnosis and delayed treatment can lead to progression of muscle weakness and a worse outcome [10]. Our findings on NCS correlated with classical patterns for AMAN [11]. Treatment of GBS patients requires a multidisciplinary approach [3, 12]. Based on clinical features with motor involvement, electrophysiological investigation, as well as the normal laboratory findings, this patient was diagnosed with AMAN subtype of GBS with severe painful legs as atypical presentation of this condition. Lee and Han [13] as well as Neocleous et al. [14] reported the first case of AMAN confirmed by electrophysiological studies that was accompanied by severe pain of the entire body. In our patient, at the beginning, severe pain sensations were treated with opioids and Carbamazepine. Later on, opioids were excluded and painful sensations were successfully treated only by Carbamazepine. Muscle strength was the first to return, but pain persisted after full muscle recovery. Three months later, at the outpatients' follow-up, she had fully recovered.

Despite the NCS findings, atypical forms of AMAN and GBS are possible. Painful symptoms must be taken very seriously and treated carefully by the clinicians.

Conflict of interest: None declared.

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Јак бол у доњим екстремитетима и одбијање вертикализације као атипична презентација Гилен–Бареовог синдрома

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САЖЕТАК

Увод Гилен–Бареов синдром (ГБС) представља најчешћи узрок акутне флацидне парализе код здраве одојчади и деце. Акутна моторна аксонална неуропатија (АМАН) представља тип ГБС-а и карактерише се моторним синдромом без сензорних симптома.

Приказ болесника Аутори приказују шестоипогодишњу девојчицу са атипичном клиничком презентацијом АМАН-а.

У налазу доминира јак бол у доњим екстремитетима и одбијање вертикализације са типичним електронеурографским налазом.

Закључак Иако електронеурографски налаз показује типичне знаке ГБС-а, клинички су могуће појаве атипичних форми АМАН-а у склопу ГБС-а. Болни симптоми се морају правовремено препознати и адекватно клинички третиранти.

Кључне речи: Гилен–Бареов синдром; акутна моторна аксонална неуропатија; атипична презентација