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CASE REPORT

Paraparetic Variant of Guillain-Barré Syndrome in First 24 Hours of Postpartum Period

A case report

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طور متعلق بالخزل السفلي في مُتَلاَزِمَةُ غيَّان-باريه في غضون الأربعة وعشرين ساعة الأولى ما بعد الولادة

تقرير حالة

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ABSTRACT: Guillain-Barré syndrome (GBS) is a heterogeneous disorder with a diverse clinical presentation ranging from weakness of certain body regions to tetraparesis with autonomic dysfunction and respiratory failure. Paraparetic GBS is a variant of GBS which is characterised by weakness limited to the lower limbs only. It is crucial to identify such topographical presentations, as a delay in diagnosis can lead to delayed initiation of specific treatment, which can negatively impact the outcome. We report a 29-year-old female patient who presented to the King Fahd Hospital of the University, Al Khobar, Saudi Arabia, in 2017 with rapid onset asymmetrical weakness of lower extremities associated with bladder dysfunction during the immediate postpartum period. The weakness spared cranial nerves and arms and imaging studies of the spine was unremarkable. Cerebrospinal fluid investigations showed cyto-albuminologic dissociation and nerve conduction studies showed features of demyelination. The patient was diagnosed with a paraparetic variant of GBS and treated with intravenous immunoglobulin. She had almost recovered completely at the two–month follow-up.

Keywords: Paraparesis; Guillain-Barré Syndrome; Demyelination; Postpartum Period; Case Report; Saudi Arabia.

الملخص: مُتَلاَزِمَةُ غيَّان-باريه هي اضطراب متغاير المنشأ، ويجيء بصور سريرية متنوعة، تتراوح بين ضعف في أجزاء معينة من الجسم إلى خزل رباعي يترافق مع خلل وظيفي اتونومي (مستقل) وفشل تنفسي. ويُعد الخزل السفلي طورا من أطوار مُتَلاَزِمَة غيَّان-باريه يتصف بضعف يقتصر على الطرفين السفليين فقط. ومن المهم الاستعراف على مظاهره الطبوغرافية، إذ أن التأخير في التشخيص قد يفضي إلى تأخير في بدء العلاج المحدد، وهذا من شأنه أن يؤثر على النتيجة. وفي هذا التقرير نقدم حالة مريضة عمرها 29 عاما جاءت لمستشفى الملك فهد بجامعة الخبر في المملكة العربية السعودية في عام 2017م وهي تشكو من ضعف عضلي لامتناظر في طرفيها السفليين حدث سريعا مترافقا مع اختلال في وظيفة المثانة في غضون الفترة التالية مباشرة للوضع. ولم يحدث ذلك الضعف في الأعصاب القحفية ولا في اليدين، ولا في العمود الفقري (حيث أظهرت التصوير الاشعاعي سلامته). وأظهر فحص السائل الدماغي النخاعي تفارقا خلويا البيوميني المنشأ. وأثبتت دراسات التوصيل العصبي بعض مظاهر زوال الميالين. وتم تشخيص حالة المريضة على أنها طور متعلق بالخزل السفلي في مُتَلاَزِمَة غيَّان– باريه، وعولجت بالحق الوريدي بغلوبيولين مناعي. وشفيت المريضة بصورة شبه كاملة عند فحصها بعد شهرين من العلاج.

الكلمات المفتاحية: الخزل السفلي؛ مُتَلاَزِمَة غيَّان-باريه؛ زوال الميالين؛ الفترة التالية للوضع؛ تقرير حالة؛ المملكة العربية السعودية.

Guillain-Barré syndrome (GBS) is an acute polyradiculoneuropathy characterised by flaccid weakness associated with decreased reflexes and cytoalbuminologic dissociation in the cerebrospinal fluid (CSF).[1] It is a heterogeneous disorder with diverse clinical presentation ranging from weakness limited to certain regions of the body to tetraparesis with autonomic dysfunction and respiratory failure.[2] Symptoms that are restricted to certain body parts are termed 'GBS variants' or 'topographical variants'. These include Miller Fisher syndrome, Bickerstaff brainstem encephalitis, pharyngeal cervical brachial variant, multiple cranial neuropathy variant, acute ataxic variant, facial diplegia with paraesthesic variant, acute pandysautonomia and the paraparetic variant of GBS.[3–7]

This report describes a case of rapid onset asymmetric paraparesis with predominant involvement of distal muscles with areflexia during the immediate postpartum period after complete recovery from epidural anaesthesia. This case highlights the importance of considering paraparetic variant of GBS when a patient complains of paraparesis. Failure to do so can lead to delay in diagnosis and consequent delay in initiation of specific treatment that can negatively impact outcome.

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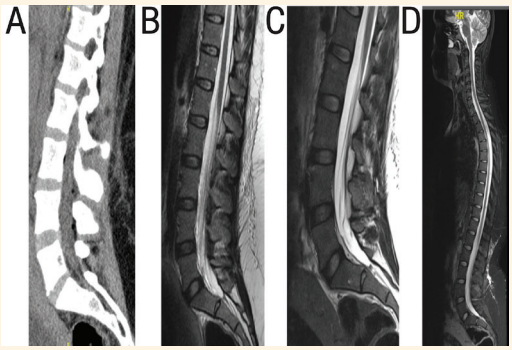

Figure 1: Imaging of the spine of a 29-year-old female patient with a paraparetic variant of Guillain-Barré syndrome. **A:** Computed tomography scan of the lumbosacral spine showing an absence of hyperdense signals. **B & C:** Magnetic resonance imaging (MRI) scans of the lumbosacral spine (**B**) on the day of symptom onset and (**C**) one week later showing no acute infarction or herniated disc. **D:** MRI of the whole spine was grossly unremarkable.

Case Report

A 29-year-old female patient presented to the King Fahd Hospital of the University, Al Khobar, Saudi Arabia, in 2017 with weakness in her lower extremities. She had no known chronic illnesses and was admitted to the hospital for delivery at 38 weeks of pregnancy. She had a history of two Caesarean sections and had given birth to a total of three children. Her family history was unremarkable and she did not have any allergies. At the time of admission, she was taking multivitamins. Her routine blood workup at the time of admission was within normal limits. She delivered a healthy baby girl through an uneventful Caesarean section under epidural anaesthesia, which was given at lumbar (L) 4/L5 level using 10 mg of bupivicaine (0.3%) and 2.5 mL of fentanyl. Her post-delivery recovery was uneventful and she regained full strength of her lower extremities as well as bladder function. The following morning, she complained of weakness in her lower extremities. The weakness started from her feet and worsened over time. Within a few hours, the weakness had progressed to the extent that she was not able to stand or walk unaided. It was associated with urinary retention. There was no fever or pain in the back or lower extremities and her arms were also unaffected. She had had an upper respiratory tract infection one week prior to admission. However, there was no history of recent immunisation or gastrointestinal infection.

Upon physical examination, she was found to have a good build and was able to lie comfortably on the bed with a urinary catheter in place. Her vital signs were stable, her higher mental functions were intact and her extraocular movements were perfect. She had normal speech and facial symmetry. Her gag reflex was intact and the power of her neck flexion was normal. Motor examination of her upper extremities was unremarkable for bulk, tone, power and reflexes. A bulk of her lower extremities was normal but both lower extremities were flaccid. On the Modified Research Council (MRC) scale, the power of her left hip flexion was 3/5 and the power of her right hip flexion was 2/5. Her left hip extension was 4/5 and her right hip extension was 3/5. Her knee flexion was 3/5 on the left side and 2/5 on the right side. The knee extension was 2/5 on both sides, whereas the plantar flexion and dorsiflexion was 0/5 bilaterally. Deep tendon reflexes could not be elicited even with reinforcement in lower extremities. The plantar reflex was equivocal bilaterally. All sensory modalities were found to be intact and the saddle anaesthesia as well as

Table 1: Motor nerve conduction study results of a 29-year-old female patient with a paraparetic variant of Guillain-Barré syndrome

Nerve test location	Latency in ms			Amplitude in mV			Conduction velocity in m/s		
	First study	Second study	Third study	First study	Second study	Third study	First study	Second study	Third study
Left peroneal nerve									
Fibula head	4.2	10.3	10.5	7.0	5.5	5.7	-	-	-
Popliteal *fossa*	6.2	20.1	15.7	6.1	1.0	1.9	45	26	30
Left tibial nerve									
Ankle	3.3	13.5	18.4	23	20.0	21.5	-	-	-
Popliteal *fossa*	7.0	18.0	17.9	18.2	5.2	10.8	41	30	27.6
Right peroneal nerve									
Ankle	6.0	11.7	13.9	5.8	4.5	6.6	-	-	-
Fibula head	8.0	12.8	15.5	5.3	1.3	2.6	39	21	26
Right tibial nerve									
Ankle	4.3	6.4	19.3	18.2	16.3	19.5	-	-	-
Popliteal *fossa*	8.6	10.3	11.5	12.9	6.4	9.7	43	27	25

Table 2: Sensory nerve conduction study results of a 29-year-old female patient with a paraparetic variant of Guillain-Barré syndrome

Sensory nerve test location	Onset latency in ms			Peak latency in ms			Amplitude in μV			Conduction velocity in m/s		
	First study	Second study	Third study	First study	Second study	Third study	First study	Second study	Third study	First study	Second study	Third study
Right sural nerve in the ankle	2.3	1.8	2.6	3.2	2.1	3.6	12.5	10.2	9.7	52	50	56

Table 3: F-wave study results of a 29-year-old female patient with a paraparetic variant of Guillain-Barré syndrome

Nerve test location	Motor distal latency in ms			F-wave latency in ms		
	First study	Second study	Third study	First study	Second study	Third study
Right peroneal nerve	3.2	5.5	7.5	NR	59.0	63.1

NR = not recordable

a sensory level were also absent. Signs of any cerebellar dysfunction were absent in the arms and could not be examined in lower extremities. The rest of the systemic examination was unremarkable.

Laboratory investigations including serum electrolyte, potassium, magnesium and phosphate levels as well as the coagulation profile, were within normal limits. The C-reactive protein level was 0.9 mg/dL (normal range: 0.05–0.3 mg/dL). A computed tomography scan of the lumbosacral spine was immediately performed which excluded an epidural haematoma [Figure 1A]. In addition, a magnetic resonance imaging scan of the entire spine did not show any findings suggestive of transverse myelitis, herniated intervertebral disc, spinal cord compression from a haematoma, spinal cord infraction, demyelinating disorder, a tumour, arteriovenous malformation or arachnoiditis of an infective or chemical nature [Figure 1B–D]. As imaging did not reveal any aetiology of her symptoms, a lumbar puncture was performed. CSF studies showed a protein level of 270 mg/dL (normal range: 15–45 mg/dL) and a white blood cell count of 22 k/μL (normal range: 0–5 k/μL) with 92% lymphocytes (normal range: 40–80%). Next, the nerve conduction studies (NCS) were performed on the first day of the symptoms' onset and repeated after one and two weeks [Tables 1–3]. This showed a prolongation of F-wave latencies, significant delay in distal latencies and decreased conduction velocities with conduction block and temporal dispersion. Sensory NCS were normal. NCS of her arms were also normal. Features of NCS were suggestive of a demyelinating disorder.

A diagnosis of the paraparetic variant of GBS was made based on the rapid onset of areflexic paraparesis, cytoalbuminologic dissociation in CSF, demyelinating features on the NCS and absence of any other explanation of her symptoms on the remainder of investigations, including imaging. The patient was administered intravenous immunoglobulin for five days. She started to regain power in her lower extremities after one week and was able to move with support after two weeks. She had almost completely recovered, was able to walk unaided and could return to her daily activities after two months.

Discussion

GBS is an acute polyradiculoneuropathy that has diverse clinical presentations, findings on electrophysiological tests and outcome. GBS is an acute peripheral neuropathy that is thought to be due to an autoimmune reaction triggered by a vaccination or surgical procedure or a preceding infection, mainly respiratory or gastrointestinal.[2] This immunopathogenesis is mainly controlled by four factors: (1) antiganglioside antibodies which can be detected in the serum of approximately half of GBS patients; (2) molecular mimicry and cross-reactivity to nerve gangliosides as in campylobacter-associated GBS cases; (3) complement activation that causes a neurotoxic effect; and (4) the characteristic of the patient themselves such as their immune status and genetic polymorphisms.[8] Clinical presentation is the primary diagnostic criterion along with electrophysiologic studies and CSF analysis. In addition to the typical clinical presentation, many atypical clinical presentations of GBS have been described.[3–7] It is crucial to identify these variants of GBS as early diagnosis of such cases can lead to earlier initiation of specific treatment and monitoring and thus, better outcomes.

The paraparetic variant of GBS is a typical *forme fruste* of this disorder where weakness is mainly limited to the legs. It was first described by Ropper in 1986 as paraparesis with normal power and reflexes in the arms.[3] Pokalkar *et al*. also identified a paraparetic variant of GBS and described it as a variant of GBS with an isolated weakness of the lower limbs with

minimal or no paraesthesia or sensory loss.[5] They found that 7% of their study population with GBS had a paraparetic variant characterised by symmetrical areflexic weakness of the lower extremities without sensory loss. However, one-third of those patients showed clinical or electrophysiologic involvement of the upper extremities. NCS in those patients showed axonopathy in 59% and demyelinating features in 33%.[5] Van Den Berg *et al*. found that 8% of their study population with GBS had a paraparetic variant.[9] Those patients with a paraparetic variant of GBS had mild disease, less cranial nerve involvement and less respiratory failure compared to the GBS patients with quadriparesis. They also identified a delay between symptom onset and initiation of treatment and a lower likelihood of receiving specific treatment (88% with paraparesis versus 97% with quadriparesis; $P = 0.014$).

Bladder involvement was seen in 14% of Van Den Berg *et al*.'s cases, as was seen in the current case.[9] The asymmetrical weakness and predominantly distal distribution in the current case were associated with areflexia and bladder dysfunction, which resembled *cauda equina* syndrome, while the classic GBS is considered to present with a symmetrical distribution of weakness with more gradual onset and infrequent involvement of the bladder. The current patient had a similar cell count in their CSF as 16% of Van Den Berg *et al*.'s cases (CSF cell count range: 10–50).[9] Data from imaging was available only for 30% of their patients and there was no explanation for paraparesis on imaging which is similar to the current case. NCS studies in Van Den Berg *et al*.'s study showed demyelinating features as the most common finding in almost half of their patients; this finding is similar to the current case.[9] One patient in their cohort had clinical and electrophysiologic abnormalities limited to the lower extremities, as was the case in the current patient. A total of 98% of their patients could walk unaided at six months.[9]

There are very few reported cases of the paraparetic variant of GBS with findings restricted to the lower extremities.[10,11] Kimachi *et al*. reported the case of an elderly female patient who developed progressive, symmetrical lower limb weakness after an upper respiratory tract infection sparing the arms and cranial nerves.[10] Similarly, the limb weakness in the current patient was also preceded by an upper respiratory tract infection. CSF showed cytoalbuminologic dissociation and NCS showed axonal type features.[10] Nagashima *et al*. reported the case of an older female patient who developed weakness of the lower extremities three weeks after an episode of fever and diarrhoea.[11] The weakness was restricted to the lower extremities with intact sensation. The NCS showed findings of an axonal variety.[11] Compared to other cases, the current patient was younger and experienced rapid onset asymmetrical paraparesis and demyelinating features on NCS. In the current case, the diagnosis is supported by antecedent upper respiratory tract infection, areflexic flaccid paraparesis, cytoalbuminologic dissociation in CSF, demyelinating features on NCS, normal spine imaging and a monophasic course of illness.

Conclusion

This case highlights that the paraparetic variant of GBS should be considered as one of the differential diagnoses of cauda equina syndrome, if there is no alternative aetiology. Early recognition of GBS is of vital importance with respect to the patient's outcome.

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