JCN Open Access LETTER TO THE EDITOR

pISSN 1738-6586 / eISSN 2005-5013 / J Clin Neurol 2019;15(3):404-406 / https://doi.org/10.3988/jcn.2019.15.3.404

Recurrent Guillain-Barré Syndrome with Anti-GT1a and Anti-GQ1b Ganglioside Antibodies

Jihyeon Hwang[a]
Ye-Ji Kwon[a]
Jong Kuk Kim[b,c]
Nam Jun Kim[c]
Seol-Hee Baek[a]

[a]Department of Neurology, Korea University Anam Hospital, Korea University College of Medicine, Seoul, Korea
[b]Department of Neurology, Dong-A University College of Medicine, Busan, Korea
[c]Peripheral Neuropathy Research Center, Dong-A University College of Medicine, Busan, Korea

Dear Editor,

Recurrent Guillain-Barré syndrome (GBS) is a rare condition that reportedly appears in 2–5% of patients with GBS.[1] The clinical and pathophysiological characteristics of recurrent GBS have not been fully elucidated. Anti-ganglioside antibodies have been reported in a small number of cases of recurrent GBS. Herein we report on a case of recurrent GBS with anti-GT1a and anti-GQ1b ganglioside antibodies.

A previously healthy 63-year-old man arrived at the emergency room with a 1-day history of dysarthria and right facial sensory change. Additional symptoms of facial diplegia, external ophthalmoplegia, truncal ataxia, and areflexia developed on the third day of admission. The findings of a nerve conduction study (NCS) and CSF analysis were unremarkable. Anti-ganglioside study was performed using the enzyme-linked immunosorbent assay for anti-ganglioside antibodies as described previously,[2] and revealed positivity for anti-GT1a (3+) and anti-GQ1b (1+) IgG antibodies. Intravenous immunoglobulin (IVIG) was administered at 0.4 g/kg/day for 5 days from the sixth day after onset. The symptoms of the patient improved gradually after IVIG termination. At 10 days after the 5-day course of IVIG, his neurological deficits had fully recovered except for the ophthalmoplegia. The remaining ophthalmoplegia improved following bilateral medial rectus muscle resection at 6 months after symptom onset. The patient could perform all of the normal activities of daily living, including driving, after the procedure.

The patient was readmitted 12 months after the first attack complaining of paresthesia in both legs. An upper respiratory infection had appeared 1 week previously. These symptoms progressed from both lower extremities to both upper extremities and his face. A neurological examination revealed truncal ataxia and hyporeflexia in both upper and lower extremities along with paresthesia. A CSF analysis produced unremarkable findings, while a motor NCS revealed prolonged distal motor latency with conduction blocks in the bilateral median and ulnar nerves. A sensory NCS revealed the absence of distal sensory nerve action potentials in the median, ulnar, peroneal, and sural nerves. The anti-ganglioside antibody assay was positive for anti-GT1a (1–2+) and anti-GQ1b (1–2+) IgG antibodies, which were the same antibodies present during the previous attack. A laboratory test indicated that the vitamin levels and thyroid function were within the normal limits. Brain MRI findings were unremarkable. Following a diagnosis of GBS, IVIG was administered at 0.4 g/kg/day for 5 days from the day after symptom onset, but his symptoms worsened. Ophthalmoplegia and quadriplegia developed on the fifth day from symptom onset. His symptoms appeared to gradually improve after IVIG termination, but they worsened again 2 weeks later. A second cycle of IVIG at 0.4 g/kg/day for 5 days was administered from day 20 after symptom onset. The patient had recovered fully at 1 month after the second cycle of IVIG treatment.

This patient had experienced two independent episodes of GBS involving the presence of

Received January 11, 2019
Revised March 5, 2019
Accepted March 5, 2019

Correspondence
Seol-Hee Baek, MD
Department of Neurology, Korea University Anam Hospital, Korea University College of Medicine, 73 Goryeodae-ro, Seongbuk-gu, Seoul 02841, Korea
Tel +82-2-920-6825
Fax +82-2-926-5347
E-mail virgo3318@gmail.com

Table 1. Anti-ganglioside antibodies in recurrent Guillain-Barré syndrome

No.	Sex	Age, years	Episode	Preceding infection	Clinical manifestations	NCS findings	Ganglioside Abs	Reference
1	F	38	First	Gastroenteritis	Quadriparesis (ascending pattern) and facial diplegia	Motor neuropathy with CB	NA	5
		39	Second	URI	Quadriparesis (descending pattern) and bulbar symptoms	Motor neuropathy with CB	IgG anti-GD1a	
2	M	58	First	None	Cerebellar ataxia, ptosis, and paresthesia	Sensory neuropathy, unclassified	NA	3
		65	Second	URI	Cerebellar ataxia, ptosis, and paresthesia	Sensory neuropathy, unclassified	IgG anti-GD1b and anti-GQ1b	
		68	Third	Arthralgia	Cerebellar ataxia, ptosis, and paresthesia	Sensory neuropathy, unclassified	IgG anti-GD1b and anti-GQ1b	
3	M	29	First	None	Quadriparesis	Motor neuropathy with CB, demyelinating	NA	3
		40	Second	Gastroenteritis	Quadriparesis	Motor neuropathy with CB, demyelinating	IgG anti-GD1b and anti-GM1	
4	F	56	First	None	Distal leg weakness and sensory loss	SM polyneuropathy with CB, demyelinating	NA	3
		68	Second	URI	Distal leg weakness and sensory loss	SM polyneuropathy with CB, demyelinating	NA	
		69	Third	Gastroenteritis	Distal leg weakness and sensory loss	SM polyneuropathy, demyelinating	IgM anti-GM1	
5	M	25	First	URI	Bulbar symptoms, ophthalmoplegia, ataxia, and quadriparesis	SM polyneuropathy, unclassified	IgG GA1 and GSC-Abs	4
		28	Second	URI	Bulbar symptoms, ophthalmoplegia, ataxia, and weakness of the lower extremities	SM polyneuropathy, unclassified	GSC-Abs	
		33	Third	URI	Bulbar symptoms, ophthalmoplegia, quadriparesis, tremor, and disturbed consciousness	SM polyneuropathy, demyelinating	GSC-Abs	
6	M	63	First	None	Bulbar symptoms, ophthalmoplegia, and ataxia	Unremarkable	IgG anti-GT1a and anti-GQ1b	Our case
		64	Second	URI	Bulbar symptom, ophthalmoplegia, ataxia, and quadriparesis	SM polyneuropathy with CB, demyelinating	IgG anti-GT1a and anti-GQ1b	

CB: conduction block, F: female, GSC-Abs: anti-ganglioside complex antibodies, M: male, NA: not applicable, NCS: nerve conduction study, SM: sensorimotor, URI: upper respiratory tract infection.

the same anti-ganglioside antibodies during each attack. A few cases of recurrent GBS with anti-ganglioside antibodies have been reported, and are summarized in Table 1.[3-5] Previous reports of recurrent GBS with anti-ganglioside antibodies have described shorter intervals between the attacks and more-severe neurological deterioration during recurrences. Recurrent GBS is typically reported to present with similar clinical manifestations during different attacks even when different conditions are present prior to the attacks.[3] However, some cases of recurrent GBS with different phenotypes during recurrent attacks have been reported.[4] These findings suggest that genetic or immunological factors are associated with the pathogenesis of recurrent GBS.

In conclusion, the clinical manifestations of recurrent GBS are heterogeneous, and the associated risk factors remain unclear. Genetic and immunological host factors and anti-ganglioside antibodies might affect the clinical manifestations and pathophysiology of recurrent GBS. Further studies are needed to obtain a deeper understanding of the clinical and pathophysiological factors underlying recurrent GBS.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Acknowledgements

This study was supported by the National Research Foundation of Korea (NRF) grant funded by the Korea government (MEST) (No. 2016R1A5A 2007009).

REFERENCES

1. Grand'Maison F, Feasby TE, Hahn AF, Koopman WJ. Recurrent Guillain-Barré syndrome. Clinical and laboratory features. *Brain* 1992;115: 1093-1106.
2. Kim JK, Bae JS, Kim DS, Kusunoki S, Kim JE, Kim JS, et al. Prevalence of anti-ganglioside antibodies and their clinical correlates with Guillain-Barré syndrome in Korea: a nationwide multicenter study. *J Clin Neurol* 2014;10:94-100.
3. Pyun SY, Jeong JH, Bae JS. Recurrent Guillain-Barré syndrome presenting stereotypic manifestations, positive antiganglioside antibodies, and rapid recovery. *Clin Neurol Neurosurg* 2015;139:230-233.
4. Ito H, Hatanaka Y, Fukami Y, Harada Y, Kobayashi R, Okada H, et al. Anti-ganglioside complex antibody profiles in a recurrent complicated case of GQ1b-seronegative Miller Fisher syndrome and Bickerstaff brainstem encephalitis: a case report. *BMC Neurol* 2018;18:72.
5. Yalinay Dikmen P, Emre Oge A. Recurrent GBS with GD1a anti-ganglioside antibodies. *J Peripher Nerv Syst* 2012;17:135-137.