SHORT REPORT

Two species of antiganglioside antibodies in a patient with a pharyngeal-cervical-brachial variant of Guillain-Barré syndrome

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Abstract
A patient with a pharyngeal-cervical-brachial variant of Guillain-Barré syndrome had anti-GT1a and anti-GD1a antibodies (IgG) in the serum. The activities of anti-GT1a antibodies were stronger than anti-GD1a antibodies and their activities declined later in the clinical course. These two different antibodies bound independently to each ganglioside in an absorption study with polystyrene beads coated with GT1a or GD1a.

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Antiganglioside antibodies have been detected in patients with Guillain-Barré syndrome, although it is not clear whether they have a specific role in the pathogenesis of Guillain-Barré syndrome. Here we report a patient with a pharyngeal-cervical-brachial variant of Guillain-Barré syndrome, who had two antibodies in his serum, one against GT1a, a minor ganglioside species, and the other against GD1a.

Case report
A forty year old man was admitted for dysphagia on 25 June 1991. He had been treated with phenytoin and phenobarbitone for epilepsy since he was 18 years old. From 6 June, he experienced diarrhoea two or three times a day. He had difficulty in swallowing on 22 June. On the next day, he felt weakness in his neck and both upper limbs. The day before admission, weakness of the proximal regions of both lower limbs developed.

On admission, weakness of the bilateral facial muscle, dysphagia, and dysarthria were present. The strength of neck flexion and the upper limbs were 3/5 (MRC grade), and 4/5 in the lower limbs. Gripping strength was 7 kg on the right and 0 kg on the left. Deep reflexes were normal and pathological reflexes were not elicited. Sensory disturbance and ataxia were not seen. Routine laboratory examinations were normal, including complete blood count, blood chemistries, and immunological examinations. Protein concentration in CSF was 350 mg/l with normal cellularity.

Clostridium jejuni. Motor nerve conduction velocities were normal in the right median, ulnar, and peroneal nerves, but compound muscle action potentials were 2-01 mV at the proximal and 4-39 mV at the distal portion of the ulnar nerve, and 0-366 mV and 0-834 mV in the peroneal nerve, respectively. F Waves of the right median and ulnar nerves had normal latencies, but could not be elicited in the right peroneal nerve.

Three days after admission, diplopia appeared with aggravated weakness of facial and limb muscles. He was treated with double filtration plasmapheresis on the 7th, 9th, 11th, 14th, and 16th days. The strength of all limbs, but not facial and neck muscles, improved to 4-5/5 with grip strength of 9.5 kg in the right and 6.0 kg in the left after plasmapheresis. Deep reflexes were slightly exaggerated. Protein concentration in CSF was normal on the 52nd day. Follow up motor nerve conduction velocity studies showed that compound action potentials of the ulnar and peroneal nerves were normalised by the 24th day. He was discharged 60 days after admission with almost full recovery except for the facial and neck muscles. The strength of his neck muscles had fully recovered by three months and his facial muscles by six months after discharge.

Methods
ENZYME LINKED IMMUNOSORBENT ASSAY (ELISA)
Antigenic gangliosides, which included GM3, GM2, GM1a, GD1a, GD1b, GT1b, and GQ1b, were prepared from bovine brains by Q sepharose column chromatography according to the method of Hirabayashi et al. GT1a was a gift from Dr S Ando (Tokyo Metropolitan Institute of Gerontology).

The ELISA was carried out by the method of Higashi et al. The antigenic solution contained 20 pmol of each ganglioside. After blocking with 5% bovine serum albumin-phosphate buffered saline (BSA-PBS), triplicate samples of serum were diluted 1:100 with 0-25% BSA-PBS and incubated for two hours at room temperature. Goat anti-human IgM or IgG antibody conjugated to horse radish peroxidase (Jackson Immunoresearch Laboratories, Inc, West Grove, PA, USA) diluted 1:1000 with 0-25% BSA-PBS was used as the second antibody.
We detected serum, beads. The polystyrene open patient. An highest microplate spectrophotometer after incubation of the beads, not beads body of the beads and 0.02% solution adding the temperature, and 0.000, 0.02% dilution, as bodies in hand, other was serum on to serum activities antibodies GTla antibodies GTla IgG syndrome, anti-GQlb Guillain-Barre syndrome, anti-GDla antibodies anti-GDla antibodies GTla IgG 1:100.

**Discussion**

In 1986, Ropper reported three patients who, like ours, developed prominent weakness of the pharyngeal-cervical-brachial regions without either lower limb weakness or sensory disturbance, and labelled it "pharyngeal-cervical-brachial weakness resembling botulism or diphtheria." Conduction block in the ulnar and peroneal nerves in our patient was compatible with postinfectious demyelinating polyneuropathy.

From our results it is suggested that the detected antibodies, anti-GTla and anti-GDla, participated in the pathological process of this patient, and that these two different antibodies independently reacted with GTla and GDla. Ilyas et al detected two kinds of anti-ganglioside antibodies in two patients with Guillain-Barré syndrome. Their antibodies, however, reacted with the common structure of GDla and GTlb, GalNAC-Gal-NeuAc-NeuAc. To our knowledge, there is no other report of a patient who has two antibodies reacting independently with a different ganglioside without sharing a common structure.

Anti-GDla antibody was reported in patients with Guillain-Barré syndrome by Ilyas et al and Yuki et al. These antibodies were also detected in a patient with motor neuropathy with monoclonal IgM protein. On the other hand, the existence of anti-GTla IgG antibodies was reported in patients with Miller-Fisher syndrome, although such patients mostly have anti-GQlb IgG antibodies. GTla was first detected in the human brain by Ando et al, and later in the human spinal cord and cauda equina. GTla is a minor ganglioside, forming 1-8% of the total gangliosides of human cerebral white matter, 2-2% of human cerebral grey matter, and 0-5% of the human spinal cord, respectively.

We thank Dr S Ando (Tokyo Metropolitan Institute of Gerontology) for his gracious gift of GTla.

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