Antigangliosides antibodies anti-GQ1b associated with Small fiber neuropathy: a case report

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Background:

Small fiber neuropathy (SFN) selectively affects small diameter sensory and/or autonomic axons. Pain or autonomic dysfunctions are the most common symptoms. Among different causes SFN occurs in several autoimmune diseases and autoantibodies against neuronal proteins may play a role in SFN pathophysiology Anti-GQ1b has been associated with Miller Fisher syndrome, Bickerstaff brainstem encephalitis, acute ophthalmoplegia, pharyngeal-cervical-brachial weakness and peripheral neuropathy involving large fibers: anti-GQ1b antibody syndrome. An isolated SFN associated with anti-GQ1b antibodies has not previously reported.

Case report:

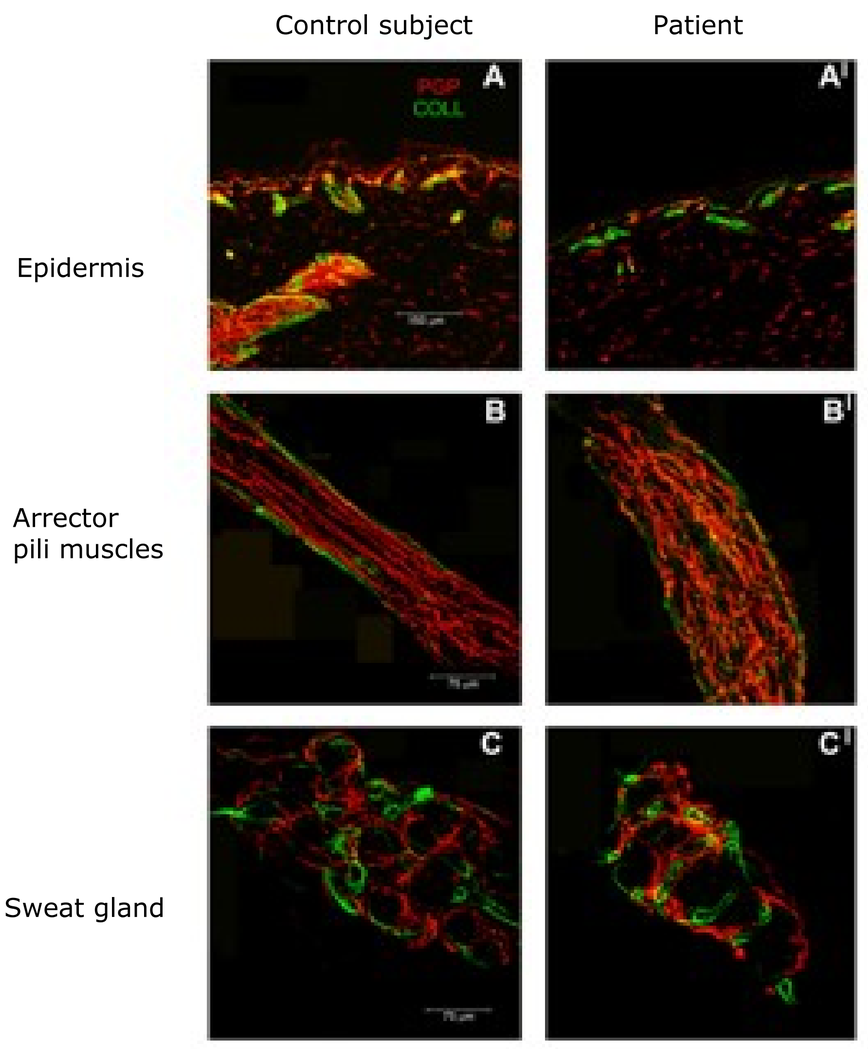
We described a 45-year-old woman complaining of a two-year history of tingling or burning sensation in the arms and legs, with nocturnal exacerbation. She did not complain ophthalmoplegia, ataxia or additional central nervous system symtpoms. Anamnesis revealed an autoimmune disorders involving several organs such as HLA-B27 negative psoriatic arthritis and complement C3 deficiency, membrano-proliferative glomerulonephritis and sicca syndrome without evidence of Sjogren's syndrome (labial salivary gland biopsy was negative).

***Neurological examination** did not disclose ophthalmoplegia, signs of central nervous dysfunctions or peripheral large nerve fiber abnormalities.*

The patient undergo to: 1)laboratory screening; 2) skin biopsies from distal leg and thigh to evaluate somatic (epidermal) and autonomic (dermal annexes) innervations; 3) motor (Tibial nerve bilaterally) and sensory (sural nerve bilaterally) conduction velocity studies to evaluate large peripheral nerve fiber.

- *Laboratory analysis disclosed **IgM antigangliosides antibodies anti-GQ1b (highly positive) and anti-GD1b.***
- *Skin biopsy revealed **decreased innervation** in the epidermal layer with preserved innervation of dermal annexes such as arrector pili muscles and sweat glands consistent with a somatic SFN (Figure 1).*
- *Motor and sensory conduction velocities were normal excluding a large nerve fiber involvement.*

This finding likely explain sensory dysfunctions complained by the patient. Consequently a trial with duloxetine (60 mg daily) was started but stopped after four weeks because of a severe constipation. The patient refused other medical treatments in this phase.



LEGEND FOR FIGURE
A confocal study of somatic and autonomic patterns of innervation in the patient and a control subject.
Leg somatic and autonomic innervations disclosed by confocal microscope (x20 for epidermis and x40 for muscle arrector pilorum and sweat gland) in the patient and an age-matched
control subject. Nerve fibers are marked in red by a pan-neuronal marker PGP 9.5 whereas the collagen staining is shown in green.
Epidermis: free-ending PGP immunoreactive fibers are evident in the epidermis of the control whereas the patient showed a marked decrease of such fibers.
The muscle arrector pilorum showed a rich density of fibers running in a longitudinal and wavy pattern in the control subject but also in the patient.
Sweat gland: the innervation appeared abundant both in the control and patient.

Conclusions:

This is the first report of SFN associated with antigangliosides antibodies anti-GQ1b. Our report expand the spectrum of antigangliosides antibodies anti-GQ1b which may include selective SFN without involvement of peripheral large nerve fiber or central nervous system dysfunctions.

References

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