CIDP-like onset of axonal polyneuropathy

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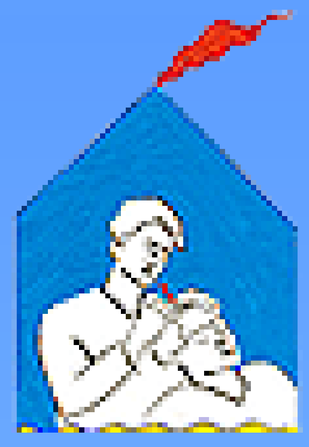

Introduction

Axonal polyneuropathies are a large group of diseases involving the axon of the peripheral nerve with clinical findings of motor and sensitive involvement with progressive course. They may have different etiologies requiring comprehensive clinical, laboratory and instrumental analyses. We present an atypical case of asymmetric chronic axonal polyneuropathy with clinical and cerebrospinal fluid (CSF) evidences CIDP–like (*Chronic Inflammatory Demyelinating Polyneuropathy)* associated with serum positivity of antibodies to the ganglioside GD1a (Fig.1)

Presentation and history

A 42-year-old Caucasian man with insidious onset of low back pain unresponsive to common non steroidal anti-inflammatory drugs, in following months reported progressive abnormal gait and hypoesthesia of the lower limbs.
His medical history and the family history were unremarkable. He was referred to our department for a complete evaluation, including neurological examination, neurophysiological tests, spine MRI and lumbar puncture.

Neurological examination

Steppage; the muscle bulk of the left lower limb was hypotrophic; the muscle strength, assessed by the Medical Research Council (MRC) scale, of the bilateral upper limbs was normal; the right quadriceps femoris, tibialis anterior, gastrocnemius, gluteus maximus and the left biceps femoris were weak (MRC 4); left quadriceps femoris and tibialis anterior were more impaired (MRC 3); tendon reflexes in the left lower limb were absent; examination of the sensory system revealed hypoesthesia of the left lower limb, in the lateral side of the right leg and hypopallesthesia of both lower limbs.

Electroneurography & Electromyography

Marked axonopathy predominantly motor of the lower limbs

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Axonal Polyneuropathy

Blood values

Routine (abnormal): CPK 634 U/l, ESR 16 mm/h
- *TSH, FT3, FT4*: normal
- *ANA, ENA, ANCA, ANTI-MAG*: negative
- *Anti-Onconeural Antibodies:* negative

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- *Antiganglioside Antibodies:*

Positivity for IgG anti-GD1a

CSF analysis

Phisical: clear and colorless
Glucose: normal
Protein: 173 mg/dl (20-50)
Cell count: 2 cells/μl (0-3)

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Albumin-Cytological Dissociation

Therapy

Intravenous Immunoglobulin therapy (0.4 g/kg/day) for 5 days
with partial improvement of motor symptoms.

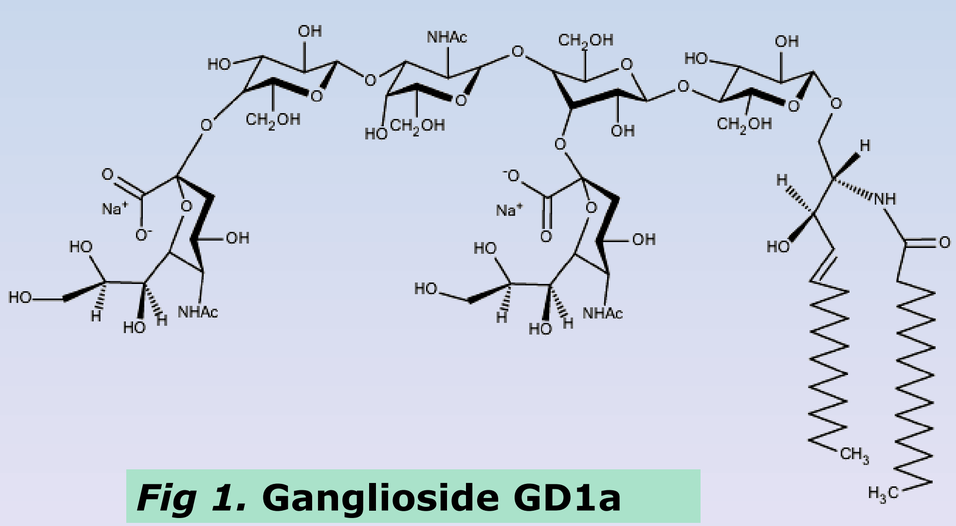

Fig 1. **Ganglioside GD1a**

Discussion & Conclusions

This is an unusual presentation of chronic axonal polyneuropathy. The onset of symptoms was not typical not only for presentation with low back sciatica, but also for the progressive rapid worsening. This clinical picture is more frequent in demyelinating disease rather than in exclusive axonal involvement, as we observe in the neurophysiologic tests and CSF analysis in our case; anti-GD1a positivity, usually associated with Acute Motor Axonal Neuropathy, was present. This case extends the range of neuropathies associated with anti-gangliosides antibodies.

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