ACUTE MOTOR AND SENSORY AXONAL NEUROPATHY WITH REVERSIBLE ACUTE PANDYSAUTONOMIA IN A MULTIPLE SCLEROSIS PATIENT

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Introduction

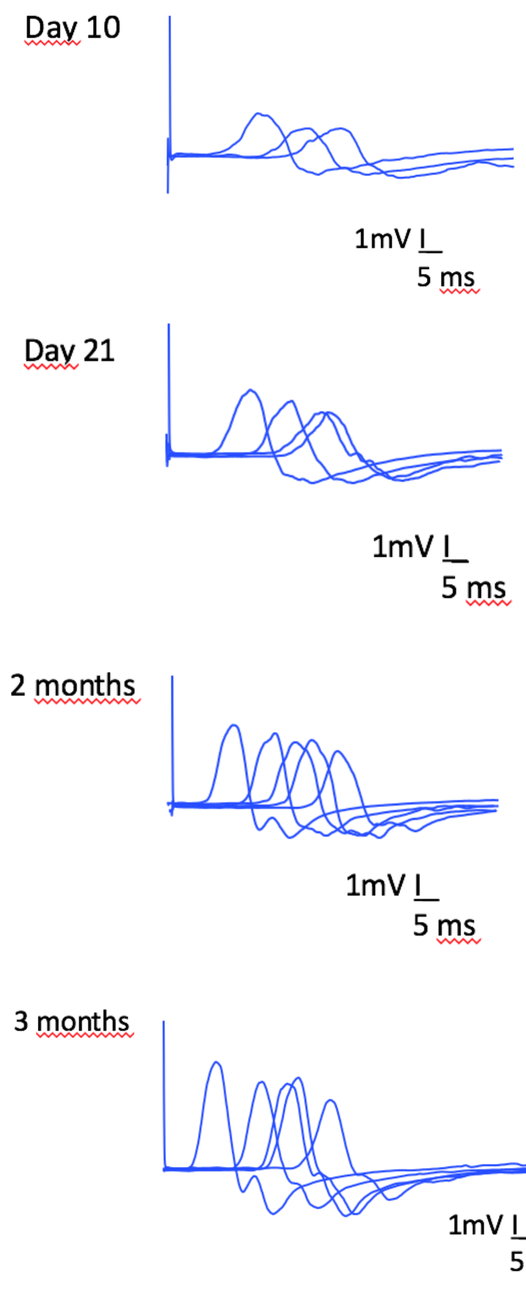
The **acute motor and sensory axonal neuropathy (AMSAN)** is a subtype of Guillain-Barré syndrome (GBS).[1] The acute pandysautonomia is an uncommon variant of GBS.[2] We describe a rare case of AMSAN associated with reversible acute pandysautonomia in a patient with Multiple Sclerosis (MS).

Case report

A 38-year-old male with relapsing-remitting MS in stable phase from four years, developed **acute weakness and paraesthesia to legs, urinary retention, constipation, dryness of mouth** two weeks after an episode of pharyngeal flogosis. His neurological examination showed **flaccid paraplegia, facial diplegia, bilateral Bell's phenomenon, bilateral mydriasis, absent pupillary reactivity and areflexia at the four limbs**. The blood sodium level was 126 mmol/l indicating hyponatraemia. His cerebrospinal fluid (obtained eight days after the onset of the symptoms) showed an albumin-cytological dissociation. The patient's serum was tested for the presence of anti-gangliosides immunoglobulin IgG and IgM antibodies against GM1, GM2, GM3, GM4, GD1a, GD1b, GD2, GD3, GT1a, GT1b, GQ1b and Sulfatides and it was found negative. The brain and spinal cord MRI showed stable MS lesion load, but highlighted **leptomeningeal enhancement of cauda equina and conus medullaris.**

The nerve conduction studies showed an absence of the sensory responses in all nerves except that the Sural nerve; distal CMAP amplitude was severely reduced in all motor nerves. The heart rate variability (HRV) analysis showed a **marked involvement of both parasympathetic and sympathetic systems** (LF= 343 ms^2; HF= 149 ms^2). The patient recieved treatment with slow infusions of hypertonic salin solution because of the symptomatic hyponatraemia. **Intravenous Immunoglobulin** (IVIg) was administered at dose of **0.4 mg/Kg/day** for five days for two consecutive cycles. The patient showed a rapid improvement of dysautonomic symptoms and a full recovery was achieved in 12 weeks. Serial electrophysiological studies demonstrated the presence of reversible conduction failures and a slowly recovery of conduction described in nodo-paranodopathies (*figure*).[3] After three months from onset, the HRV-analysis demonstrated improvement of the autonomic balance (LF=1113 ms^2; HF=547 ms^2).

Serial motor conductions of the right Ulnar nerve



Discussion

This patient showed a complete and dramatic improvement of the severe acute pandysautonomia with administration of IVIg, supporting the evidence that this condition is in fact a variant of GBS. But there aren't reports in litterature that describe an **Acute Pandysautonomia in association with AMSAN variant of GBS** that show fully recovery after IVIg treatment. Besides, our patient showed **hyponatraemia** before treatment with IVIg, and this complication could be part of the dysautonomia resulting in sympathoadrenal dysregulation or an effect of the possible coexisting SIADH. The Hyponatremia involves the 16%-25% of patients with GBS[4], but the incidence in AMSAN remains uncertain.

Conclusions

We describe a rare case of AMSAN associated with acute pandysautonomia. Our report confirms that **serial electrophysiological studies** are needed for diagnosis of GBS subtypes. Moreover the **HRV analysis** is an useful tool for investigating the **autonomic balance** also in patients with Acute Inflammatory Neuropathies. In this case the early recognition of the diagnosis, the immediate IVIg administration and the appropriate symptomatic treatment certainly have improved outcomes.

References:

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XLVII CONGRESSO DELLA SOCIETA' ITALIANA DI NEUROLOGIA
Venezia - 22-25 ottobre 2016