Supplemental Data

Table e-1: Detailed diagnostic results

Test	After 3 weeks of illness	After 3 weeks of therapy
Routine blood test	<i>Pathological:</i> Na ⁺ 130 mmol/l ↓ [136-145], Cl ⁻ 86 mmol/l ↓ [98-107] <i>Normal:</i> electrolytes, blood count, parameters for hemolysis, liver/kidney function, inflammation, and coagulation parameters.	No abnormalities
Serological analy- sis	Normal/negative IgM/IgG for: CMV, EBV, HIV, HSV, Enterovirus, Coxsackie, Influenza, Parvo B19, and Hanta Virus, Borrelia burgdorferi, Brucella ceti, Mycoplasma pneumoniae, Bartonella henselae, Francisella tularensis, Treponema pallidum, Cryptococcosis, Toxoplasmosis, and Leptospirosis	-
Endocrine analysis	Pathological: norepinephrine 1970 ng/l ↑↑ [165-460], adrenalin 317 ng/l ↑↑ [30-90], free metanephrine 107 pg/mg ↑ [7.9-87.9], free normetanephrine 144 pg/ml ↑ [<101.9] Normal: hypothalamic-pituitary-thyroid (HPT) axis, hypothalamic-pituitary-adrenal (HPA) axis, fluid deprivation test with normal ADH regulation and ability to concentrate urine, excluding central or renal diabetes insipidus.	No abnormalities
Toxicology	Normal: Urine analysis including drug screening	-
Bone marrow examination	Normal: Bone marrow aspiration and biopsy	-
Auto-antibodies	Pathological: Serum: anti-VGKC (on RIA) 283 pmol/l ↑ [<85], after 14 days: 207 pmol/l ↑ [<85], anti-CASPR2 1:10,000 ↑↑ [negative] on CBA* CSF: positive signal in the hippocampal and cerebellar regions of mouse brain on TBA (Fig. 1), anti-CASPR2 1:10 ↑ on CBA* [negative], antibody specificity index (ASI) 0,08 [<1,5]: no intrathecal synthesis (cell count/lactate/glucose normal, protein 1350mg/l ↑ [<150-450], albumin 801mg/l, no oligoclonal bands) Normal: Serum: ANA, ENA, anti-dsDNA, p-/c-ANCA, anti-TPO, anti-Tg, anti-TRAK, anti-phospholipid, anti-pituitary, anti-VGKC (on RIA), anti-neuronal IgG panel (Hu, Ro, ANNA-3, Yo, Tr/DNER, Myelin, Ma/Ta, GAD65, anti-Amphiphysin, anti-Aquaporin-4, anti-NMDA, AMPA, GABA _B -R, LGI1, ZIC4, DPPX, Glycin-R, mGluR1, mGluR5, ARHGAP26, ITPR1, CARPVIII, Homer3, MOG, Recoverin, Neurochondrin, GluRD2, Flotillin, IgLON5) on a commercial cell-based assay (CBA*)	Pathological: Serum: anti-CASPR2 1:32, anti-LGI1 1:10 [negative] CSF: present, albeit decreased signal on TBA (Fig. 1). Normal: CSF: anti-neuronal IgG panel on CBA*
Imaging	Pathological: Abdominal sonography showing constipation, faecal impaction and residual urine after micturition. Normal: cerebral MRI with MRA, spinal, thoracic, and abdominal MRI, chest X-ray, ECG, echocardiography, thyroid ultrasound.	-
Electrophysiology	Pathological: EMG with and increased rate of polyphasic potentials of increased duration as a sign for neuropathy; fasciculations and spontaneous bursts of motor unit discharges as a sign of denervation, but no doublet, triplet or multiplet discharges.	Pathological: Nerve conduction studies with signs of distal demyelinating neuropathy

If not mentioned otherwise, the laboratory investigations were performed at the clinical laboratory associated with the Charité (Labor Berlin, Charité Vivantes GmbH, Berlin); * performed at EUROIMMUN Biochip-Mosaic, indirect immunofluorescence (IFT) (Clinical-Immunological Laboratory, Prof. Dr. med. W. Stöcker, Lübeck, Germany). Normal values/ranges are given in square brackets [].