Letters to the editor

Isolated ataxia and autonomic dysfunction: a new variant of Guillain-Barré syndrome?

Several variants of Guillain-Barré syndrome have been described including Fisher's syndrome, pure motor Guillain-Barré syndrome, pharyngeal-cervical-brachial weakness, paraparetic pattern, ataxic form, and pure pandysautonomia. The connection between these variants and Guillain-Barré syndrome is based on (1) a close resemblance to some symptoms of the typical disease, (2) overlap phases between typical disease and variant syndromes, (3) electrophysiological and CSF abnormalities that confirm Guillain-Barré syndrome, and (4) exclusion of other, more common, causes of the symptomatology. We report a patient with a possible new variant consisting of progressive ataxia and profound postural hypotension without appreciable weakness or sensory symptoms.

An otherwise healthy 47 year old man developed flu-like symptoms three weeks before admission. He had a history of a bipolar schizo-affective disorder and had been treated for many years with lithium, carbamazepine, and bromoperidol. Five days before admission his gait progressively became unsteady until he could not stand up without support. The day before admission he was seen at the psychiatry outpatient clinic by a consultant neurologist (GT) who concluded that he had a progressive cerebellar syndrome with areflexia of the lower limbs. Brain CT did not show abnormalities of the cerebellum or brainstem. The next day his symptoms worsened, confining him to his bed, and he was admitted to our hospital. Neurological examination of the cranial nerves was unremarkable except for a slight dysarthria. There were no abnormalities of muscle strength or sensory modalities. Finger-nose and heel-knee tests were abnormal showing intention tremor on both sides. He could sit, but not stand, unsupported. He could take a few steps with a wide based gait; the tandem gait could not be performed. Reflexes were slightly decreased in the arms and absent in the legs, and plantar responses were flexor. Serum concentrations of his medication were within the therapeutic range. Routine laboratory examinations did not disclose any abnormalities. The second day after admission he collapsed on sitting up, probably due to postural hypotension resulting from autonomic neuropathy. Sinus bradycardia was recorded on ECG. There were no other signs of autonomic dysfunction. Examination of CSF disclosed a raised protein content of 1.42 g/l with 16 leucocytes/mm³. The CSF/blood albumin ratio was high and the IgG index was slightly raised at 0.89. No oligoclonal bands were present in the CSF. At neurological examination on the second day after admission reflexes of the arms had further decreased and a slight impairment of proximal muscle strength of the legs was noted for the first time. In addition, the patient when specifically asked, complained of paraesthesia in both feet and hands. These findings prompted us to consider the diagnosis of (a variant of) Guillain-Barré syndrome.

Extensive EMG on day 7 disclosed normal conduction velocities and sensory and motor action potentials but absent F waves and delayed H reflexes in the lower limbs without signs of denervation. These findings were thought to be consistent with a proximal nerve lesion and matched early electrophysiological findings in Guillain-Barré syndrome. Serological screening results for neurotropic viruses, Borrelia burgdorferi, Treponema pallidum, and HIV-1 were negative. IgG antibodies against Campylobacter jejuni were detectable in serum but IgM antibodies were absent. Autoantibodies against gangliosides GM1 and GQ1b, and against...
sulphatides were not detectable in serum. Because of the unusual symptomatology for Guillain-Barré syndrome and the absence of pronounced weakness, treatment with intravenous immunoglobulins (IVIgs) was deferred and the patient was kept under close observation. During the next three weeks his ataxia improved slightly and increased dietary salt intake, abdominal compression using a belt, and fludrocortisone was started to alleviate his postural hypotension. However, after five weeks pronounced postural hypotension was still present which severely disabled the patient by preventing him sitting up for more than a few minutes. Therefore it was decided to treat him with IVIg at 0.4 g/kg during five days. During the next week his blood pressure increased and he was able to sit for more than an hour without collapsing. He was subsequently successfully rehabilitated and discharged from our hospital three months after admission while capable of sitting in a wheelchair all day. After eight months he was functioning again at his premorbid level and neurological examination was unremarkable except for the inability to walk in a perfect straight line.

The presentation of this patient with isolated ataxia suggested a cerebellar disorder. The subsequent development of autonomic dysfunction and areflexia with very mild and only short lasting motor and sensory disturbances was suggestive of Guillain-Barré syndrome. Abnormalities in CSF and electrophysiological findings supported this diagnosis. During the full observation period ocular movements were repeatedly tested and always found to be normal. As the patient's postural hypotension did not react to symptomatic treatment, we started treatment with IVIg assuming that his disease was a variant of Guillain-Barré syndrome. After IVIg treatment postural hypotension indeed dramatically improved and did not return. The positive effect of IVIg did not diminish after a few days, suggesting a specific, immunomodulating mode of action rather than a short lasting increase in blood pressure through an increase in blood protein concentrations by high dose IVIg.

A diagnosis of Guillain-Barré syndrome could not be made in our patient because he did not fulfill the diagnostic criteria of the typical syndrome.1 Intact proprioception and vibration sense combined with pronounced ataxia argue against the presence of the sensory form of GBS and against the sensory ganglionopathies as recently reviewed by Dalakas and Quarles.2 A combination of acute sensory and autonomic neuropathy has been previously described in a 26 year old woman after Coxsackie B virus infection.3 However, in that case there was also pronounced impairment of cutaneous sensation and extensive muscle weakness. A Japanese patient has been described with ataxia and orthostatic hypotension after infection with Epstein-Barr virus.4 By contrast with our patient no CSF abnormalities were noted.

We conclude that the unique combination of ataxia without loss of proprioceptive sense and severe postural hypotension as a result of autonomic neuropathy in our patient suggests either a new variant of Guillain-Barré syndrome or a new overlap syndrome of acute pandysautonomia and acute cerebellar ataxia.

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