Using MRI Kira et al. [3] found that 4 of 8 MS patients with hyperprolactinemia had diencephalic hypothalamic lesion(s) contiguous with the third ventricle. All relapsing-remitting MS patients with hyperprolactinemia showed a rise in prolactin levels in the acute stage of the relapse and a decrease during the remission phase. Rader and Lowy [4] found normal serum prolactin levels in 35 MS patients with chronic progressive MS and 19 MS patients with acute exacerbation.

In our patient, over the 6 years of observation, hyperprolactinemia was not of relapsing nature and increased steadily by 30%. Such hyperprolactinemia is mostly caused by tumors. Neuroimaging studies confirmed demyelinating lesion of the hypothalamus and the absence of pituitary prolactinoma. Hyperprolactinemia in our patient was not caused by drugs, endocrine diseases or other causes. The long evolution of hyperprolactinemia, positive TRH loading test, chronic amenorrhea and temporary polydipsia were arguments in favor of a relationship between hyperprolactinemia and an MS lesion in the lower part of the hypothalamus.

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Central Nervous System Involvement in Guillain-Barré-Like Syndrome: Clinical and Magnetic Resonance Imaging Evidence

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Introduction

The Guillain-Barré syndrome (GBS) is a clinical diagnosis associating an ascending progressive paralysis, areflexia, and various degrees of sensory deficits. Spontaneous improvement occurs in about 80% but recurring or chronic presentations have been described. The diagnosis is further supported by an albuminocytologic dissociation in the cerebrospinal fluid (CSF) and electrophysiological abnormalities compatible with peripheral demyelination [1]. Central nervous system (CNS) involvement in otherwise typical GBS patients has been questioned. The Miller Fisher syndrome (MFS) associates ophthalmoplegia, cerebellar ataxia, and areflexia. MFS has been thought to represent a variant of GBS, although it has been questioned whether MFS might represent a central nervous system disorder whereas GBS would represent a disease of the peripheral nervous system.

We treated an unusual patient with GBS who presented features suggestive of CNS involvement both on clinical examination and on magnetic resonance imaging (MRI).

Case Report

In early February 1992, a 31-year-old obese black woman experienced the sudden onset of headaches followed 3 days later by neck stiffness and pain in neck, shoulders and arms. The following day her gait became unsteady and, 2 days later, she experienced painless progressive darkening of vision to total blindness. Past medical history was remarkable for systemic hypertension controlled by diet, and smoking (15 cigarettes per day). Two weeks prior to initial symptoms, her 3-year-old daughter developed a flu-like syndrome with neck stiffness and headaches.

During hospitalization, she developed progressive bilateral ptosis, complete external ophthalmoplegia and a gradual onset of generalized weakness and areflexia. Neck pain worsened as did chest pain that increased with deep breathing. All initial investigations were negative, including brain CT and MRI, cerebral angiography, lumbar puncture, thyroid function, rheumatoid factor, antinuclear antibodies, acetylcholine receptor antibodies, and viral serologies. Westergren erythrocyte sedimentation rate was 19 mm.

Two weeks later she was referred to the University of Miami. On February 24, she was oriented, her memory was good but her speech was slurred. There was generalized weakness, more pronounced in the legs (walking or standing up was not possible) and on the left side. Sensory modalities were preserved for pin prick, light touch and vibration but proprioception was defective in the toes. Bilateral areflexia was present. The patient was totally blind with 8-mm pupils nonreactive to light but reactive to near effort. A left ptosis and a left hypertropia were present. Ocular movements were markedly restricted in all directions, and forcedduction test was negative. There was profound facial diplegia. Fundus examination revealed marked bilateral optic disc swelling with peripapillary splinter hemorrhages. A scan orbital echography revealed distended optic nerve sheaths containing fluid. Lumbar puncture showed an opening pressure of 550 mm water; CSF was clear without cells, proteins were 36 mg/dl (normal <41 mg/dl), glucose was 122 mg/dl. Anticampylobacter and antiglycolipid antibodies were not tested. In addition to the enlarged optic nerves, a brain MRI revealed cerebellar, cerebral and pontine white matter hypertensive signals on T2 compatible with demyelinating or vasculitic process (fig. 1a, b). No lesions had been seen on the previous MRI of February 12. Electromyography did not show evidence of active denervation, and neurography revealed normal distal latency but absent F-waves.

Due to severe headaches and bilateral severe optic neuropathy, the patient was treated with intravenous methylprednisolone 250 mg q.i.d. Rapid relief of headaches and neck stiffness were noted over the next week. However, vision did not improve. Decrease in lung vital capacity prompted intubation and ventilation on March 4 for the next 10 days. Plasmapheresis was initiated on March 9 (7 ses-

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Fig. 1. a, b February 26, 1992 (axial magnetic resonance imaging. T2-weighted sequences, TR: 2550, TE: 80): hyperintense signals were visible (arrows) in the cerebellum (a) and the periventricular white matter (b). c, d November 30, 1992 (axial MRI, T2-weighted sequences, TR: 2650, TE: 80): 9 months later, no cerebellar lesion was detectable (c) but some periventricular white matter lesions were still visible, although decreased in intensity (d, arrow).

sions, each 4 liters exchange, last session on March 22) and respiratory function gradually recovered during this period. Progressive recovery of motor and sensory deficits ensued but she remained blind. Ocular motility recovered gradually and optic disc edema subsided over 4 weeks, leaving profound optic atrophy.

Nine months after discharge she was still completely blind but recovered from all other deficits. MRI performed on November 30, 1992, showed disappearance of the cerebellar and pontine white matter abnormal signals, but persistence of the periventricular lesions (fig. 1c, d).

Comments
GBS is thought to be primarily a demyelinating disease of the peripheral nervous system, characterized by ascending weakness, areflexia and ataxia. The MFS, characterized by areflexia, cerebellar ataxia and ophthalmoplegia is a bulbar variant of GBS, with possible central white matter involvement [1–3].

Our patient presented with several unusual, although well described, features of GBS and MFS. Initially severe neck stiffness and headaches were present without a CSF inflammatory reaction. Such associations have indeed been reported to occur in up to 10% of GBS
patients [1]. She subsequently developed almost total ophthalmoplegia, which can be present in up to 15% of GBS patients, and characterizes the Miller-Fisher variant [1]. Three weeks after onset of symptoms bilateral florid optic disc edema was present in our patient, a rare complication of GBS, occurring in 1–3% of large GBS series, usually noted about the 3rd week of the disease [1]. Papilledema is usually associated with high CSF protein content, but some cases have been reported with a normal CSF protein content [4, 5], as in the present case. The mechanism of development of increased intracranial pressure in GBS is thus unclear. Lumbar puncture in our case revealed an opening pressure above 500 mm H₂O, and standardized echography revealed markedly distended optic nerve sheaths. The bilateral optic disc swelling most probably represented papilledema due to increased intracranial pressure. However, total visual loss secondary to papilledema of raised CSF pressure is rare and progresses slowly beginning with peripheral constriction of the visual field [11, 12].

Within 48 h of onset of symptoms our patient became totally blind, suggesting severe optic neuritis as the most likely cause. Although reported as isolated cases [6–10], this is a rare event with an unknown incidence [1]. All such reported cases were of the retrobulbar type and, except for case 1 [6], some degree of visual recovery always occurred. In their monograph, Ropper et al. [1] state that “Optic neuritis remains the only consistent, albeit very uncommon, evidence of inflammation of central nervous system myelin in GBS”. The sudden onset of bilateral visual loss in our patient, in the absence of vascular disturbance to both optic nerve and retina, was likely due to inflammatory optic neuritis. MRI revealed multiple white matter lesions in cerebellum, cerebral hemispheres, and within the pons. These lesions were hyperintense on T2 and proton-density sequences, and enhanced by gadolinium. No such lesions were found on the initial MRI, and improved on follow-up study. Such MRI findings were recently reported in a GBS patient with retrobulbar optic neuritis [10]. Other reports of CNS lesions on CT scan or MRI are scarce. Among 223 MFS cases reported in the world literature, Berlit and Rackicky [13] found abnormal results in 8/66 CT scans and 2/5 MRI studies, and lower medulla lesions disappearing on follow-up MRI study were recently reported by Unishitani et al. [14]. Histologic studies of peripheral nerves in typical GBS patients reveal inflammatory mononuclear infiltrates with a tendency to surround small endoneurial vessels, mostly veins; rarely, clusters of perivascular lymphocytes are found in the brain or spinal cord [15]. Such lesions are very similar both on MRI on pathologically to those present in multiple sclerosis (MS) and acute disseminated encephalomyelitis (ADEM). We postulate that the white matter lesions seen on the MRI of our patient represent perivascular inflammatory infiltrates, as in MS and ADEM, partially resolving over time. Other pathological evidence of CNS involvement has been demonstrated [2, 6, 8–10, 16].

In summary, we report a patient who exhibited a markedly severe form of GBS with both clinical and neuroradiologic evidence of central nervous system involvement. This supports the hypothesis that GBS, MFS, MS and ADEM might be related to common pathogenetic mechanisms with variable expressions in peripheral and central nervous system.

References


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