AN UNUSUAL PRESENTATION OF GBS: CASE REPORT AND LITERATURE REVIEW

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ABSRACT

Guillain-Barre syndrome (GBS), also known as Landry paralysis is an acute idiopathic polyneuritis, believed to be immunologically mediated. It usually presents as a demyelinating neuropathy with ascending weakness, however, many clinical variants have been well documented in the medical literature, and variants involving the cranial nerves or pure motor involvement with axonal injury have also been described. We report a case of a 50 year old patient who initially presented to the ER with hemiparesis and cranial nerve palsies simulating a cerebrovascular event. Based on neurological examination, CSF analysis and needle EMG finding a diagnosis of GBS was made.

INTRODUCTION

Guillain-Barre syndrome (GBS) was first reported by Landry in 1859 and later detailed by Guillain, Barré and Strohl, in 1916. The disease has become well-known internationally under the name of GuillainBarré Syndrome1.The disease is assumed to be autoimmune and operated by a preceding infection, most of the time respiratory or gastrointestinal infections2.Generally infections by microorganisms such as Campylobacter jejuni, CMV, Mycoplasma pneumonia, or influenza virus exist several weeks prior to approximately two thirds of GBS cases2. A small percentage of patients develop GBS after immunization with quadrivalent meningococcal polysaccharide conjugate vaccine (MCV4), following surgery or trauma, and bone-marrow transplantation3. It was associated with tumor necrosis factor alpha antagonist therapy in one report 4. In another report it was associated with the use of isotretinoin in two patients, however, a definite causal link couldn't be identified5. GBS has also been linked to other autoimmune diseases and malignancy such as systemic lupus erythematosus, and Hodgkin lymphoma6.Although the classic description of GBS is that of a demyelinating neuropathy with ascending weakness, many clinical variants have been well documented in the medical literature, and variants involving the cranial nerves or pure motor involvement and axonal injury have also been described7. We describe a rare case of GBS presenting as hemiparesis.

CASE REPORT

A 50 year old female known hypertensive for 3 years fairly controlled with medication, presented with h/o sudden onset, generalized moderate intensity headache, and progressiveweakness of right side of the body which was followed 2 days later by double vision (while looking towards left) and facial asymmetry. With the clinical suspicion of stroke her MRI brain was done at a local hospital and she was then referred to our hospital. On presentation to our hospital she had developed new onset left sided weakness 6 days later in addition to the persisting right sided weakness, facial asymmetry and diplopia. There was history of diarrhea 3 weeks prior to onset of weakness. She denied any history of fever, dysphagia, nasal regurgitation, urinary or fecal incontinence, backache, neck pain, photophobia, altered sensorium, arthralgias, oral ulcers or photosensitivity. Examination revealed an overweight female conscious, oriented intime, place and person with a respiratory rate of 20/min without use of accessory muscles. Her pulse was regular 70 bpm and BP was 130/90 mmHg. Her higher mental functions were intact. Motor examination revealed normal bulk withhypotonia in all four extremities with power of 3/5 in both upper extremities and 2/5 in both lower extremities (both proximally and distally). She had generalized areflexia with mute planter response. There was facial diplegia (bilateral LMN type facial palsy) and bilateral 6thnerve paresis (left > right), rest of the cranial nerves were intact and symmetric. Cerebellar examination wasn't possible owing to patient's quadriparesis and pin prick and proprioception was intact. Fundoscopic examination revealed grade 1 hypertensive changes. Chest was clear to auscultation.Examination of CVS and GI system was unremarkable. Laboratory parameters showed normal complete blood picture and metabolic panel. Her ESR was 18, ANA, RA factor and hepatitis serology werenegative. MRI brain

with contrast was normal. CSF showed cytoprotein dissociation with 5 cells, 77 mg/dl protein, and a normal glucose 65mg/dl(BSR134mg/dl). Nerve Conduction Studies/Electromyography was suggestive of a demyelinating polyneuropathy with secondary axonal degeneration. Left median, peroneal and tibial nerve showed markedly prolonged distal latency with decreased amplitude. Left median and ulnar showed mildly reduced velocity, low normal velocity was seen in left peroneal and normal in left tibial with mild dispersal of response in left tibial nerve. Left ulnar nerve showed normal motor and sensory response with absent response in left median nerve. F waves were absent in all the tested nerves. EMG of upper limb muscles showed reduced recruitment of normal duration and amplitude MUAPs while those of lower limbs showed reduced recruitment of mildly neuropathic MUAPs. On the basis of the above clinical and lab data a diagnosis of GBS was made and plasmapharesis was initiated alongwith supportive treatment. There wasn't much improvement in her power however her shortness of breath improved and she was discharged after 6 sessions of plasmapharesis and referred for rehabilitation. Unfortunately she was lost to follow ups.

DISCUSSION

Guillain-Barré syndrome (GBS), also known as acute idiopathic polyneuritis, is a type of neuromuscular paralysis that has several variants4. In about 75% of cases it is preceded 1,2,7-8 weeks by an acute illness mostly respiratory or gastrointestinal. Camplylobacterjejuni (C. jejuni) infection is, overall, the most common antecedent infection and has been reported in up to 32% of cases. C. jejuniGBS has marked motor axon degeneration, an elevated anti-GM1 antibody, and a delayed and often incomplete recovery. There appears to be an over-representation on of certain strains of C. jejuni suggesting that the lipopolysaccharides of these organisms share ganglioside-like epitopes with peripheral nerves. This molecular mimicry appears to confuse the immune system resulting in a mistaken attack against neural antigens9. Patients usually present with complaints of finger dysesthesias andmuscle weakness of the lower extremities. The weakness may progress over hours to days to involve the arms, truncal muscles, cranial nerves and muscles of respiration. Variants of GBS may present as pure motor dysfunction (AMAN) or acute dysautonomia7. Half of GBS patients have some degree of cranial nerve dysfunction during their illness. Facial weakness is most common, especially if substantial limb weakness is present. Ophthalmoparesis is seen in 10-20% of patients with abducens palsy being most common which is usually bilateral. Acute motor axonal neuropathy (AMAN) is distinguished from AIDP by its involvement of exclusively motor nerves and an electrophysiologic pattern indicating axonal involvement. AMAN is seen commonly in northern China and Japan10,11, usually associated with a preceding Campylobacter jejuni infection. The clinical features and recovery are very similar to those of AIDP12. However, more AMAN patients require assisted ventilation because of impending respiratory failure.
Acute motor-sensory axonal neuropathy (AMSAN) affects both sensory and motor fibers and is thus a more severe form of AMAN 11. Clinically, it resembles AMAN variant with the exception that sensory symptoms are more in AMSAN. There is marked axonal degeneration of both motor and sensory nerve fibers demonstrated by severely reduced or absent motor and sensory responses on nerve conduction studies and extensive denervation on follow-up EMG studies, resulting in delayed and incomplete recovery. Typical presentation of Miller Fischer Syndrome (MFS) is that of ophthalmoplegia with ataxia and areflexia13. About one-quarter of patients who present with MFS will develop some extremity weakness, clearly linking this disorder to GBS. A limited form of MFS presents as cerebellar ataxia and hyporeflexia without ophthalmoplegia. Antibodies against GQ1b are present in 85 to 90 percent of patients with MFS GBS. Elevated antibodies to GQ1b suggest an immune attack against GQ1b gangliosides which are concentrated in the Para nodalregions of extraocular nerves9. Certain features of MFS like ophthalmoplegia and ataxia along with encephalopathy and hyperreflexia characterize another variant of GBS, namely Bickerstaff encephalitis. It is also associated with anti-GQ1b antibodies and thus responds to IVIG or plasma exchange 14,15. The pharyngeal-cervical-brachial variant of GBS is characterized by acute muscular weakness of the oropharynx, neck, and shoulder girdle with swallowing abnormality 16,17. It represents a localized form of axonal GBS 16,17. Facial weakness may be seen. Motor strength and reflexes are typically preserved. This form overlaps with MFS17. Some patients with pharyngeal-cervical-brachial weakness have antibodies against GQ1b, or less often GD1a. It is thought by some experts that MFS, Bickerstaff encephalitis, and pharyngeal-cervical-brachial weakness with anti-GQ1b antibodies constitute overlapping expressions of the anti-GQ1b antibody syndrome 18.Tatsumoto M et al showed in their study that isolated abducents nerve palsy can be categorized as a regional variant of Guillain-Barré syndrome or mild form of Fisher syndrome. They reviewed clinical profiles and laboratory findings for 100 cases of abducens nerve paresis. Tentative diagnoses made by the primary physicians on request of anti-ganglioside antibody testing were abducens nerve palsy (n=68), Fisher syndrome (n=14), acute ophthalmoparesis without ataxia (n=14). Serum anti-GQ1b antibody was positive in 2519. Yoon-Sik Jo, and Sang-Jun Nav have reported an unusual case of pure motor variant of GBS initially presenting with hemiparesis, (a 59 year-old-male presenting with left sided weakness 1 day prior to admission who developed right sided weakness 2 days following admission). Electrophysiological findings were consistent with

pure motor GBS20. This patient presented with hemiparesis developing weakness of the other side few days later with NCS findings supporting diagnosis of GBS, similar to our patient however he didn't have facial diplegia or ophthalmoparesis as seen in our case report moreover the neurophysiology studies showed pure motor GBS while it was suggestive of demyelinating polyneuropathy with secondary axonal degeneration in our patient. Guillain-Barré syndrome (GBS) is generally diagnosed on clinical grounds7. Electromyography (EMG) and nerve conduction studies (NCS) can be very helpful in the diagnosis as well as for prognosis. Abnormalities in NCS that are consistent with demyelination are sensitive and represent specific findings for classic GBS(AIDP wariant). Delayed distal latencies, slowed nerve conduction velocities, temporal dispersion of waveforms, conduction block, prolonged or absent F waves, and prolonged or absent H-reflexes are all findings that support demyelination. Needle EMG may be normal in acute nerve lesions, and it may take 3-4 weeks for fibrillation to develop. In the acute phase, the only needle EMG abnormality may be abnormal motor recruitment, with decreased recruitment and rapid firing motor units in weak muscles. Unfortunately, electrodiagnostic studies can be completely normal inearly GBS and a normal study does not rule out the disease.21,22. Characteristic findings on CSF analysis include albuminocytologic dissociation, which is an elevation in CSF protein (>0.55 g/L) without an elevation in white blood cells. The increase in CSF protein is thought to reflect the widespread inflammation of the nerve roots7. Our patient initially presented with hemiparesis with cranial nerve palsies, a rare presentation of GBS her examination along with electrodiagnostic studies and CSF examination supported the diagnosis of GBS. Chauhan v et al havereported a case of GBS in India presenting as Bell's palsy with crossed hemiparesis who recovered over a month23. Kim EJ and Yuki N from Australia have reported reported an unusual case of GBS who presented with left hemiparesis at the acute phase and right hemifacial weakness at the recovery phase of the hemiparesis24. Acute hemiplegia as a clinical presentation of GBS could be confidently added to the category of atypical presentation of pediatric GBS which constitutes 11.2-24.3% of the whole pediatric GBS presentation reported previously.

The management of patients with Guillain-Barré syndrome (GBS) can be intimidating.25The unpredictable course and potential for rapidly producing life-threatening respiratory failure may prompt admission to an intensive care unit (ICU).Treatment of GBS though challenging, is mainly supportive and is centered at the anticipation and prevention of various complications. Immunomodulation with plasmapheresis and IV immunoglobulin (IVIG) helps to shorten the course of the disease.26-29 Patients with more severe disease and rapid progression of illness require extensive and prolonged rehabilitation and also show delayed and incomplete recovery.

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