

Yet Another Variant Of Gbs With Hyperreflexia And Loss Of Pain

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Citation

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Abstract

Guillain Barre Syndrome is an acute demyelinating disorder of spinal roots and peripheral nerves occasionally the cranial nerves due to an immune-mediated disturbance involving the peripheral myelin sheath. It is characterized clinically by acute ascending type of motor weakness of limbs with hypo- or areflexia with preserved sensation and autonomic nervous system. However, other GBS variants like pure acute panautonomia³, Miller Fisher syndrome⁴, polyneuritis cranialis, pharyngeal-cervical-brachial variant⁵, facial diplegia with hyper reflexes⁶ have also been reported. Patient presented with sub acute, progressive, weakness of all four limbs associated with hyper reflexia and loss of pain and temperature, with electrophysiological features suggestive of severe demyelinating neuropathy motor as well as sensory component and responding to IVIG.

INTRODUCTION

Guillain Barre Syndrome is an acute, diffuse demyelinating disorder of spinal roots and peripheral nerves occasionally the cranial nerves due to an immune-mediated disturbance involving the peripheral myelin sheath^{1,2}. It is characterized clinically by acute ascending type of motor weakness of limbs with hypo- or areflexia with preserved sensation and autonomic nervous system. However, other GBS variants like pure acute panautonomia³, Miller Fisher syndrome⁴, polyneuritis cranialis, pharyngeal-cervical-brachial variant⁵, facial diplegia with hyper reflexes⁶ have also been reported. Recently, patients with acute motor paralysis of limbs with hyper tendon reflexes are reported. Such atypical GBS has been called as acute motor axonal neuropathy^{7,8,9}. We present here a patient who had acute quadriplegia and predominant glove-stocking sensory involvement with brisk tendon reflexes.

CASE REPORT

A 39 years old male presented with complaints of weakness of all 4 limbs gradually worsening over a period of 2 weeks associated with numbness of all four limbs and difficulty in walking. There was no associated fever, fatigability, headache, skin rash, dysphagia, diplopia, urinary incontinence.

2 weeks prior to this complaint he had sore throat for which he took antibiotics from local hospital and improved. No h/o

gastroenteritis prior to this complaint. There was no previous h/o similar weakness and no diurnal variation in weakness of limbs. No past medical h/o diabetes or any other systemic illness.

On examination, patient was conscious, cooperative and oriented. Extra ocular movements were normal; no facial muscles weakness and other cranial nerves were normal. Fundus was normal. Motor system examination showed power proximally 4/5 and distally 3/5 in both upper and lower limbs. But the deep tendon reflexes were brisk in all limbs. Plantar reflexes were flexor. No abnormal cerebellar signs noted. Decreased pin prick sensation distally like glove and stocking pattern with intact joint position and vibration sense was noted. No abnormal autonomic manifestations were identified.

Other systemic review was unremarkable. Investigations showed CBC- normal, ESR 15mm/hr. Electrolytes and renal functions were normal. CSF showed protein- 200mg/dL, glucose- 60mg/dL (RBS at the time of lumbar puncture-114mg/dL); cells- 05/cu.mm. Stool culture was negative for Campylobacter Jejuni. Nerve Conduction Studies showed severe demyelinating neuropathy and axonopathy (fig.1 and 2).

Patient was given Immunoglobulin IV (IVIG) 0.4gm/ kg /day for 5 days. He started to improve in his motor power within 3 weeks, but continued to have paresthesia for 6-8

months which partially responded to gabapentin. A year later he was neurologically asymptomatic. The nerve conduction studies were done during follow-up which showed normal motor distal latency, CMAP, CV with normal F wave in tested peripheral nerves. The SNAP of sensory nerves became normal .

DISCUSSION

Guillain- Barre Syndrome is a constellation of sign and symptoms of which areflexia / hypo reflexia is mandatory, but now literature has in enough that even retained tendon reflexes or brisk reflexes are part of Guillain- Barre Syndrome. The diagnosis of GBS in such patients is supported by immunological studies, nerve conduction studies, effective response to immunoglobulin^{1,2}. The nerve conduction studies play a pivotal role. Acute facial diplegia with hyperreflexia has been described as a Guillain- Barre Syndrome variant with normal nerve conduction studies in these reports. Another variant has been described of GBS with brisk and retained tendon reflexes where the electrophysiological study showed motor conduction block neuropathy. In 1991, a mysterious epidemic of paralytic syndrome occurring in Northern China noticed in every summer. Electrophysiology testing of these children showed motor axonal loss with little or no demyelinating features¹⁰. There after many countries including, Mexico, Spain, South America, Japan, South Korea, India and recently, North America have reported such cases of GBS and primary axonal degeneration affecting only the motor fibres called Acute Motor Axonal Neuropathy (AMAN) have reported. Most of these cases are linked most commonly with *Campylobacter jejuni* infection. Though not known exact mechanism, the molecular mimicry is highly possible in AMAN with *C.Jejuni* .

Our patient had clinical features of acute onset quadriparesis with brisk tendon reflexes and glove-stoking type of sensory symptoms and the electrophysiological studies favored the severe demyelination and axonal neuropathy. Our patient is different from the previous reported cases in terms of predominant sensory symptoms and involvement of pin-prick sensation than the touch, vibration and joint position sensations. In literature, a prominent sensory ataxia type of GBS associated with IgG anti-GD1b antibody was described. The sensory ataxia in this reported case and others were due to large myelinated fibres damage. To our surprise

this case had loss of pain with preservation of touch, joint position and vibration sensation. The abnormal amplitude of SNAP was found in our case. Such combination has not reported in literature to the best of our knowledge. Unfortunately we were unable to do serum antibody titre for *C.Jejuni*.

CONCLUSION

Although hypo- or areflexia is necessary for clinical diagnosis of GBS, a lesson should be learned that retained or hyperreflexia of deep tendon reflexes do not exclude the diagnosis of Guillain- Barre syndrome. The sensory examination should be done meticulously to note the wide spectrum of GBS syndromes in our days. The electrophysiological studies play a very important role in differentiating it from other causes.

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