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Pediatric Guillain-Barre Syndrome presenting with hyperreflexia and opsoclonus myoclonus

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ABSTRACT

Guillain-Barre Syndrome is an acute inflammatory polyneuropathy most commonly characterized by rapidly progressive, essentially symmetrical weakness with hyporeflexia or areflexia. The most common underlying subtypes of GB syndrome include Acute Inflammatory Demyelinating Polyneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN) and Acute Motor and Sensory Axonal Neuropathy (AMSAN). Association of hyperreflexia with GBS is a rare entity and seen very rarely in Indian population. Here we describe a patient with AMSAN/Miller Fisher variant of GB syndrome who presented with exaggerated reflexes.

Keywords: Guillain Barre Syndrome, hyperreflexia, Miller Fisher, neuropathy

1. INTRODUCTION

Guillain-Barré syndrome (GBS) is an acute autoimmune polyradiculoneuropathy, presenting as areflexic, flaccid paralysis with variable sensory disturbances and elevated cerebrospinal fluid (CSF) protein without pleocytosis (Baheti et al., 2010). Even though hyporeflexia or areflexia is associated with GBS, a patient presenting with hyperreflexia does not exclude the possibility of a GBS variant. Patients presenting with hyperreflexia often have other characteristic features of GBS like symmetrical ascending weakness, involvement of cranial nerves, involvement of optic nerve or sensory disturbances. We report one such rare presentation of a patient with motor polyneuropathy and exaggerated reflexes.

2. CASE REPORT

A 5-year-old male child presented with history of pain in lower limbs and tendency to sway on standing up, with inability to walk since 2 days and inability to sit up from sleeping position since 1 day. Patient had history of mild grade fever associated with maculopapular rash lasting for 7 days, prior to the weakness. The fever had resolved before current symptom onset. Patient first developed weakness of lower limb which progressed on day two

of illness to inability to sit up from sleeping position. On third day of illness upper limbs were affected and by day five of illness, it had progressed to inability to hold up the neck. He then developed difficulty in swallowing. Patient had no complaints of loss of touch or pain sensation. There was no history of headache or convulsions. Past history revealed that he also had one episode of febrile convulsion at the age of 2 years. There was no significant family history.

On examination patient was afebrile and vitals were appropriate for age. There was no evidence of sinus bradycardia or hypertension hence ruling out raised intracranial pressure. General physical examination revealed no significant findings. Higher mental functions were within normal limits. Cranial nerve examination revealed a loss of gag reflex. All other cranial nerves were within normal limits. Motor examination revealed hypotonia of all 4 limbs with lower limbs more than upper limbs. There was asymmetrical presentation at time of disease. Power was 3/5 in all 4 limbs. Deep tendon reflexes were 3+ with exaggerated ankle and knee jerk on day of admission and gradually progressed to 1+ on day 5 of illness. Superficial reflexes were present throughout the course of the illness. Plantar reflex on both lower limbs were extensor. He was also observed to have horizontal and vertical nystagmus suggestive of Opsoclonus. As the child had an asymmetrical presentation, MRI brain with spine screening was done to rule out space occupying lesion or any cord compression, which was suggestive of normal report (Figure 1).

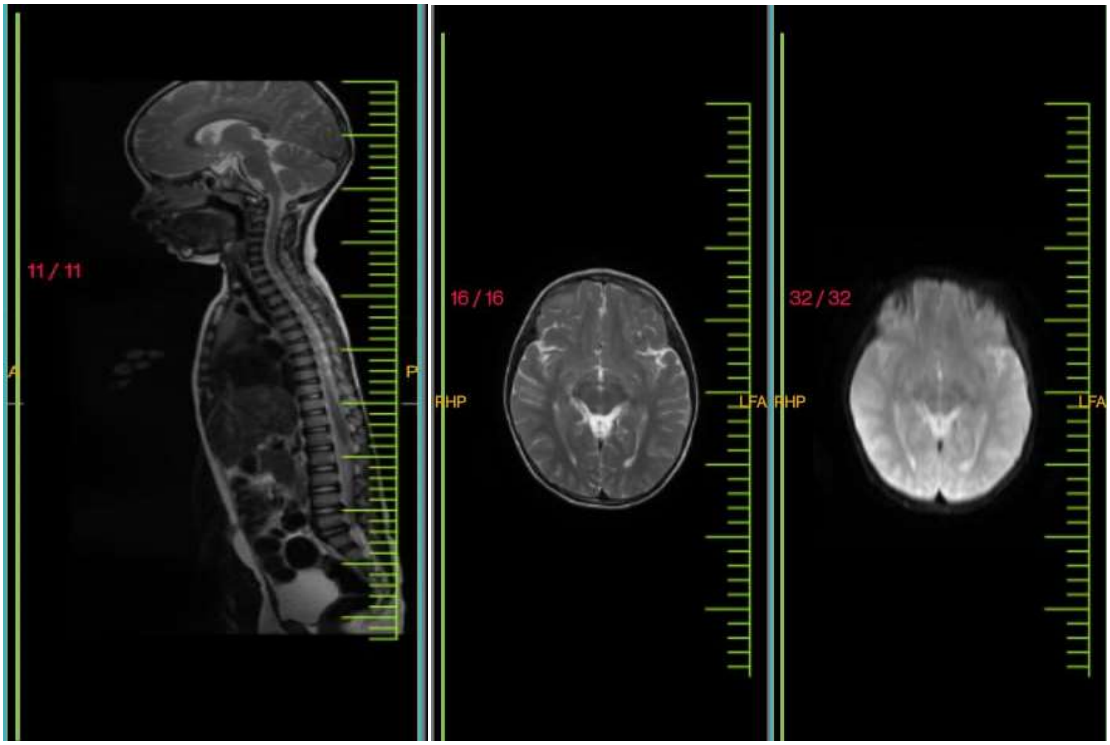

Figure 1 MRI brain with contrast and whole spine screening

NCV was done whose report showed sensory motor polyneuropathy. There was reduced CMAP amplitude with normal distal motor latency and CV in bilateral peroneal nerves. CMAP amplitude, distal motor latency and conduction velocity were within normal limits in right median, ulnar and bilateral tibial nerves. SNAP amplitude could not be elicited in bilateral sural nerves. This was suggestive of AMSAN/Miller Fischer variant of GBS.

Lumbar puncture was done which was s/o CSF protein 27 LDH 54 Glucose 50mg/dl with parallel sugar 130mg/dl. There was 0-1 WBC/hpf which was predominantly mononuclear cells. Blood culture was sent on admission which was sterile. There was no history of diarrhea and stool sample sent for *C. jejuni* came out to be negative. They were not affordable for GM1 antiganglioside antibody titres.

Patient was administered IV Immunoglobulin at 2g/kg over 3 days. Initially the upper limb power improved, gag reflex reappeared. On second day there was improvement of lower limb power with deep tendon reflex 2+ in lower limbs. Patient had pain in lower limbs for which Gabapentin and syrup vitcofol was added. By fourth day patient could stand with support and was accepting well orally. Neurophysiotherapy was continued. Patient was discharged after 2 weeks of IVIG treatment. On discharge patient could walk with support and was accepting feeds per oral.

3. DISCUSSION

Guillain-Barre syndrome is an acute condition. It has many subtypes and clinical presentations. It is classified on a pathologic basis into demyelinating and axonal forms. Axonal GBS has been sub classified into acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN). Although hyporeflexia or areflexia is a cardinal feature of GBS, preserved reflexes or hyperreflexia is not a finding inconsistent with GBS. Hyperreflexia which is a frequent symptom on GBS is usually also associated with a history of pain in the abdomen and loose stools secondary to infection by *Campylobacter jejuni*. These cases also tend to show a positive test for antibodies like IgG anti GM1 ganglioside. Amongst the cases which show hyperreflexia the most common type associated is found to be the AMAN type. In a study around 30% of Japanese and 50% of Chinese patients with AMAN type of GBS showed some degree of exaggerated reflexes during their phase of recovery (Neki et al., 2017). In recent times, axonal variants of GBS show preserved or exaggerated reflexes amongst Asian and European populations. The same study also supported the finding of exaggerated reflexes seen during the recovery phase of GBS (Kuwabara et al., 1999). Although most of the studies are based off Chinese and Japanese populations and this variant is not common in the Indian subcontinent, a few cases have been reported. The incidence in pediatric age group is even more rare (Baheti et al., 2010).

Acute motor axonal neuropathy (AMAN) is a pure motor axonal subtype of Guillain-Barré syndrome (GBS) that was identified in the late 1990s. Majority of the patients of GBS amounting to 40-60% in the Asian and South America AMAN is the major subtype. This subtype is also known to progress faster and in a more aggressive manner when compared to demyelinating GBS. The pathogenesis involves variable levels of molecular mimicry between a lipo-oligosaccharide of *Campylobacter jejuni* and human gangliosides. This causes pathological changes at the nodes and paranodes which can be detected in electrophysiology as rapidly reversible nerve conduction slowing or blockade. Autoantibodies that bind to GM1 or GD1a gangliosides at the nodes of Ranvier activate complement and disrupt sodium-channel clusters and axoglial junctions, which leads to nerve conduction failure and muscle weakness (Kuwabara and Yuki, 2013).

A few other cases have been reported in the similar line. Neki et al., (2017) reported one such case where Guillain-Barre syndrome was of acquired predominantly demyelinating (with axonal) type of motor polyradiculoneuropathy but reflexes were exaggerated throughout the course of the disease. There was no cranial nerve involvement and sensory examination was within normal limit. Our case too had no sensory involvement but reflexes were initially exaggerated and later mute.

Baheti et al., (2010) also reported two such cases where a 40-year-old man presented with progressive, ascending flaccid paralysis which was confirmed on nerve conduction study to be of Acute Motor Axonal Neuropathy. This patient responded to 5 days of IVIG. Another study reported by them mentions an 18-year-old girl who presented with acute onset weakness of limbs, lower limbs greater than upper limbs. There was no sensory or autonomic affection. Nerve conduction study showed features suggestive of acute pure motor axonopathic variant of GBS. She was treated with large volume plasma exchange with significant improvement at 2 weeks with only mild dorsiflexor weakness. Our case had a similar presentation of lower limb weakness greater than upper limb weakness on onset. But none of the above-mentioned cases mentions a loss of gag reflex or inability to swallow as part of disease progression.

Incecik et al., (2016) has also described a pediatric case who presented AMSAN type of Guillain-Barre which was also associated with papillitis. Pulse therapy of methylprednisolone was given and IVIG was given once diagnosis was finalized. Patient improved with IVIG and could be discharged. Our case did not present with papillitis but with opsoclonus which also settled with IVIG treatment.

Singhal and Bhat, (2011) also described a pediatric case of a 10-year-old child presenting with asymmetric weakness fitting into AMAN type of GBS and preservation of reflexes. Cranial nerve involvement was seen in this case. The patient improved after administration of both IVIG and IV methylprednisolone. Our case also had similar symmetrical presentation but responded to IVIG and did not require IV methylprednisolone.

4. CONCLUSION

Though the textbook presentation of Guillain-Barre syndrome involves polyneuropathy secondary to inflammation with symmetrical weakness and hyporeflexia or areflexia, hyperreflexia should not mislead the diagnosis of Guillain-Barre Syndrome. Our case showed hyperreflexia with opsoclonus and loss of gag reflex with no sensory involvement. Another differentiating feature was the low CSF protein level which is unusual in cases of Guillain-Barre Syndrome. A nerve conduction study would hence be diagnostic in times of doubt. Hyperreflexia in GBS has been reported many a time and a pediatrician should be aware of such unusual presentations so as to administer timely treatment before the disease progresses. The treatment of choice regardless of presentation is Intravenous Immunoglobulin.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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