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Guillain Barre Syndrome with Miller Fisher Variant: A Case Report

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ABSTRACT

Introduction: Guillain Barre Syndrome (GBS), once considered a single clinical entity, is now recognized as a heterogeneous syndrome with several variants, with varied presentations and complaints. A case of GBS with Miller Fisher Syndrome (MFS), which has rarely been reported in India, is described in this case report. **Case report**: A 5 year old female presented to us complaining of double vision since 4 days and difficulty walking since past 2 days. The patient also had a history of fever 20 days prior to admission. There was paralysis of the sixth cranial nerve bilaterally (ophthalmoplegia), which resulted in convergent strabismus. Poor articulation of words was observed (dysarthria). Muscle strength was noted as 3/5 for all four limbs and mild gait unsteadiness was observed, with tendency to fall on either side. Nerve conduction study was suggestive of mild early generalised demyelinating neuropathy of the right ulnar nerve. The clinical examination of the patient pointed towards the diagnosis of Miller Fisher syndrome, a rare variant of Guillain Barre Syndrome. Patient showed slow regression of motor deficit with intravenous immunoglobulins (IVIG), administered at 2 gm/kg. **Conclusion**: Diagnosing MFS required high clinical suspicion as in many cases all symptoms may not appear at the same time. This case report demonstrates that clinicians should be aware of rare variants of GBS.

Keywords: Guillain Barre syndrome; Campylobacter jejuni; antiganglioside antibodies; intravenous immunoglobulin treatment.

INTRODUCTION

Guillain-Barre syndrome (GBS) has an overall incidence of 1 to 2 cases per 100,000 per year. The classical clinical features of GBS include progressive, symmetric muscle weakness accompanied by absent or depressed deep tendon reflexes. Historically, GBS was considered a single clinical entity. However, it is now recognized as a heterogeneous syndrome with several variants, with varied presentations and complaints. The initial diagnosis of GBS is based on the clinical presentation of the patient. A case of GBS with Miller Fisher variant, which has rarely been reported in India, is described in this case report.

CASE REPORT

A 5 year old female presented to our hospital complaining of double vision since 4 days and difficulty walking since past 2 days. On elaborating, the patient gave history of pins and needles sensations in her lower limb. This was followed by similar sensations in the upper limb. This later progressed to difficulty in wearing footwear in a span of 3 days and resulted in difficulty walking since past 2 days. Double vision or diplopia occurred when looking either to the left or to the right. The patient also had a history of fever 20 days prior to admission. There were no complaints of unconsciousness, convulsions, hearing problems, tinnitus, vertigo, difficulty swallowing, or hoarseness of voice. The patient refused having respiratory infection, diarrhoea or tick bite in recent past. The patient had no bladder or bowel incontinence. Patient's past medical history and family history were insignificant.

On examination, the vital signs were within normal limits. The patient was alert, awake and oriented to time, place and person. There was paralysis of the sixth cranial nerve bilaterally (ophthalmoplegia), which resulted in convergent strabismus. Both the pupils were round, regular and slowly reactive to light. Poor articulation of words was observed (dysarthria). No evidence of seventh cranial nerve paralysis was found on physical examination. Muscle strength was noted as 3/5 for all four limbs. On asking the patient to walk, mild gait unsteadiness was observed, with tendency to fall on either side. Both lower limbs had areflexia as well. Patient's complete blood count, metabolic panel and toxicology screens were negative. Imaging studies

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like chest x-ray and magnetic resonance imaging (MRI) revealed no pathology. Nerve conduction study was suggestive of mild early generalised demyelinating neuropathy of the right ulnar nerve. Cerebrospinal fluid analysis was normal. The clinical examination of the patient pointed towards the diagnosis of Miller Fisher syndrome, a rare variant of Guillain Barre Syndrome. Nerve conduction studies supported our diagnosis. The patient was treated with intravenous immunoglobulins (IVIG), administered at 2 gm/kg over 2 days. Patient showed slow regression of motor deficit. Physical rehabilitation was advised for the patient.

DISCUSSION

In 1916, Guillain, Barre, and Strohl proposed that the main clinical features of GBS are motor weakness, areflexia, paresthesias with minor sensory loss, and increased protein in CSF without pleocytosis. All age groups are affected by GBS, however the incidence increases by approximately 20% with every 10-year increase in age beyond the first decade of life. Additionally, the incidence is slightly greater in males than in females. Our understanding of the underlying pathology was greatly enhanced when multifocal inflammatory demyelination of spinal roots and peripheral nerves was described. Miller-Fisher syndrome (MFS), identified in 1956, is characterized by ophthalmoplegia, ataxia, and decreased or absent reflexes and accounts for about 5% of cases. Acute ataxic neuropathy is a similar disorder with ataxia and areflexia but without ophthalmoplegia. MFS being a variant of Guillain-Barre syndrome is debatable as some authors believe that it is a form of brainstem encephalitis. The clinical distinction between MFS and brainstem encephalitis can be difficult. Clinically, disturbances of sensorium, multiple cranial nerve palsies, an abnormal EEG, or prolongation of the inter-peak latencies of the brainstem auditory evoked response would suggest brainstem encephalitis. Majority of patients present with diplopia followed by gait and limb ataxia. Studies have pointed out that ataxia is attributed to a mismatch between proprioceptive input from the muscle spindles and the kinesthetic information from joint receptors. Although motor strength is usually preserved in MFS, in some cases overlap of clinical picture with typical GBS seems to occur when some patients progress to develop quadriparesis.

Motor conduction studies, F-wave latencies, and needle electromyography are usually normal. A wide range of ocular signs in MFS are possible which include complete ophthalmoplegia, dilated and unreactive pupils, or external ophthalmoparesis with or without ptosis. Cranial nerves other than ocular motor nerves may also be affected. Most patients have increased CSF protein without pleocytosis one week after the onset of the disease. Brain MRI is usually normal, though in some cases it may show brainstem lesions or gadolinium enhancement of ocular motor nerves. Studies have shown that in about 20% patients of MFS, the disease followed Campylobacter jejuni infection and in 8% fit ollowed Haemophilus influenzae infection. Serum IgG antibodies to the ganglioside GQ1b, are detected in 98% of patients with MFS in the acute phase. These findings suggest that the antibodies are disease specific and related to the pathogenesis, which might not be completely understood by us. Other antibodies such as anti-GT1a, anti-GD3, and anti-GD1b have also been associated with MFS but with a lesser frequency.

With improvements in modern critical care, the outcomes in GBS have also improved considerably, including a reduction in the mortality rate from 33% before the introduction of positive-pressure ventilation to the current rate of approximately 1–5%. Clinically it has been seen that the symptoms usually resolve in 2 to 3 months of immunoglobulin therapy.

CONCLUSION

In diagnosing our patient with MFS required high clinical suspicion, because in many cases all symptoms may not appear at the same time. This case report demonstrates that clinicians should be aware of rare variants of GBS, which can be easily missed otherwise. Missing the diagnosis of this potentially treatable disease can be fatal for the patient.

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