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OVERLAP SYNDROME OF MILLER-FISHER SYNDROME (MFS) WITH PHARYNGEAL-CERVICAL-BRACHIAL (PCB) VARIANT-GUILLAIN BARRE SYNDROME (GBS)

Neurology

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ABSTRACT

Guillain-Barre syndrome (GBS) and Miller Fisher syndrome (MFS) can present with overlapping features. A 28-year-old gentleman presented with complaint of dull neck pain, diplopia, difficulty in puffing up his cheeks and tremors of both hands. On examination there was bilateral gaze palsy, dysphagia, marked neck weakness, bilateral upper limbs weakness (greater than lower limbs), nasal intonation, arefelxia and ataxia. A diagnosis of pharyngeal-cervical-brachial variant of GBS with feature of Miller Fischer Syndrome was made after excluding all other possible differentials, based on cerebrospinal fluid analysis and nerve conduction study (NCV). The patient improved following five cycles of plasma exchange. We report this case that presented as atypical variant of GBS with features of both Miller Fisher Syndrome and Pharyangocervical variant.

KEYWORDS

INTRODUCTION –

Guillain-Barre Syndrome (GBS) can manifest as several clinical subtypes with various subsets of clinical features. The most-classical feature of GBS is acute progressive and ascending limb weakness after a preceding infection. Miller Fisher syndrome (MFS) is a representative focal variant of GBS characterized by acute ophthalmoparesis, ataxia, and areflexia. [1] The pharyngeal-cervical-brachial variant (PCB) as another well-known but rare focal variant that presents with acute bulbar palsy along with arm and neck weakness.[2] Many of these subtypes appear in different combinations as well as those that occur as individual localized types, which makes it difficult to clearly classify them as specific subtypes. [1-3] MFS can appear simultaneously with other types of GBS and appear as an overlap syndrome: MFS/PCB or MFS/PCB-GBS in which diagnosis and treatment becomes difficult. [3] We present a case of overlap syndrome of MFS/PCB-GBS, who was diagnosed on basis of clinical symptoms and NCV study and treated successfully with plasmapheresis.

Case Presentation –

A 28-year-old male presented with dull neck pain for 10 days, diplopia for 1 day and numbness around mouth since 1 day. There was no history of trauma, canned food consumption, headache, fever, vomiting or tick bite. He had history of diarrhea and myalgia 10 days back. On neurological examination patient was conscious, alert, oriented with normal higher mental functions. Patient had left 6th cranial nerve and bilateral 7th nerve palsy. {{Figure – 1}} He had normal motor and sensory examination in both upper limbs and lower limbs with preserved deep tendon reflexes and bilateral flexor plantar reflexes.

Nerve conduction studies and blink reflex were normal on initial presentation to the hospital. MRI brain and cervical spine was also normal. On the basis of above findings diagnosis of Miller Fischer Syndrome was made and treatment was started with Injection Methyl-prednisolone along with supportive care. After 1-day patient's weakness progressed with involvement of neck flexors to Medical Research Council [MRC] grade 3, in upper extremities (both Medical Research Council [MRC] grade 3) and with sparing of lower extremities. Neurological examination revealed bilateral complete 6th nerve palsy, dysarthria, and dysphagia. {{Figure-2}} His bilateral facial weakness (7th nerve palsy) also began to progress slowly during the day, making it difficult for him to wrinkle, blink, and puff up his cheeks.

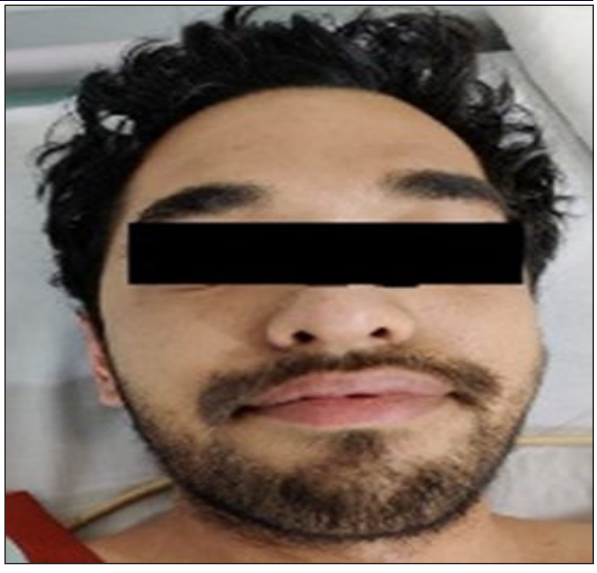

Figure1: Bilateral Seventh Nerve Palsy

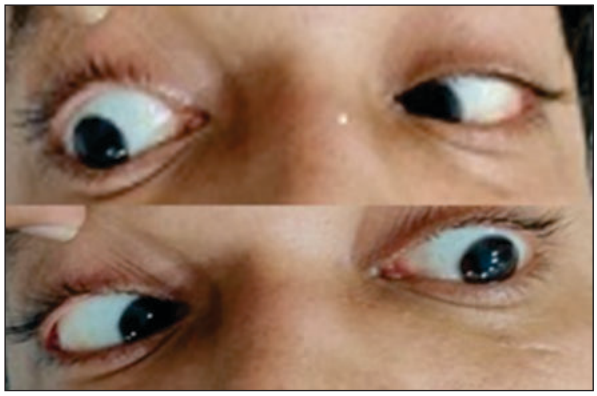

Figure2: Bilateral Sixth Nerve Palsy

Investigations –
Routine blood and urine work up was normal. Autoimmune markers like ANA and dsDNA were negative. Cerebrospinal fluid examination revealed albuminocytologic dissociation. An enzyme-linked immunosorbent assay (ELISA) to detect various antiganglioside antibodies could not be done. Nerve conduction studies performed on the third day after admission revealed a predominant, bilateral facial and brachial sensory and motor axonal neuropathy, and radiculopathies, with relative sparing of the lower limbs, with modest progression compared to the study performed previously. There were slowed median conduction velocities in both forearms and decreased compound motor action potentials (CMAPs) in bilateral median and ulnar nerves, with mild prolonged distal latencies and low normal conduction velocities. Focal conduction blocks were seen for the left ulnar nerve across the elbow. Left ulnar F-wave was absent, and right ulnar F wave was non persistent (these were normal in previous study). Tibial F- waves and H-reflexes were normal in latencies, as in the previous study. Blink reflex study which was normal earlier, now revealed bilateral prolonged R1 latency with relatively preserved R2i and R2c latencies and mild global reduction in amplitudes.

Such peculiar presentations of MFS and PCB restrict the differential diagnosis to very few clinical conditions. Differential diagnosis includes brainstem (Bickerstaff) encephalitis, pharyngeal-cervical-brachial weakness GBS variant, and other neuromuscular autoimmune disorders (which were ruled out with a negative ANA, dsDNA tests).

Treatment and Outcome –
We started the patient on plasmapheresis (PLEX) based on the presence of PCB and MFS. During the treatment, the limb muscle weakness worsened bilaterally to MRC grade 2 in the upper extremities and MRC grade 4+ in the lower extremities after 3 days. His dysarthria, dysphagia, and dyspnea also worsened. There was progressive improvement in leg strength on day 4 of PLEX treatment, followed by increase in strength in upper extremities. After 5 cycles of PLEX patient responded very well and all symptoms improved. Patient was discharged over next few days in stable condition. After 6 months of follow up, patient has no residual neurological deficit.

DISCUSSION –
The present case suggests that the manifestations of overlap between MFS-PCB variants and GBS is an acute autoimmune polyneuropathy that can be classified into various regional variants. [1] Although GBS, MFS, and PCB variants have distinct symptoms and antibody test results, overlapping cases of GBS, MFS, and PCB variants are not uncommon in the clinical setting; Sekiguchi et al, found overlaps in 50% of MFS patients, 23% of PCB-GBS patients and 15% of conventional GBS patients. [3] Previous results suggest that MFS, PCB, and GBS are likely to form a continuous spectrum. [2,3] The regional progression of the MFS spectrum can be either downward or upward, and it was downward in the present case. [3] Clarifying the classification of MFS/PCB, MFS/PCB-GBS, and MFS/GBS is difficult due to the continuity of the disease, and so judgment based on the specific clinical situation in individual patients remains important. [3] The electrophysiologic results of our patient led to a classification as AMSAN (acute motor sensory axonal neuropathy). Previous studies have indicated that the electrophysiologic results for MFS and MFS/GBS were similar for acute motor axonal neuropathy (AMAN). [3] Therefore, both previous cases and the present case indicate that MFS, MFS/GBS, and MFS/PCB-GBS may also occur as a result of AMAN or AMSAN depending on the severity of the disease.

CONCLUSION –
Overlapping cases of GBS, MFS, and PCB variants are not uncommon in the clinical setting; such overlaps have been found in 50% of MFS patients, with 23% PCB-GBS and 15% conventional GBS.

This overlapping case of ASMAN variant of GBS, MFS, and BBE provides further support that these conditions are part of the same spectrum and share a common autoimmune mechanism.

MFS can appear simultaneously with other types of GBS and appear as an overlap syndrome: MFS/PCB or MFS/PCB-GBS in which diagnosis and treatment becomes difficult.

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