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An atypical case of Guillain-Barré: Overlap of Miller Fisher syndrome with pharyngeal-cervical-brachial variant

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1. Case presentation

The patient was a 30-year-old female. She had developed dysphagia three days prior to admission. Initially, she only had problem swallowing solids but later the problem progressed to liquids, and after that dysphonia was added. She was visited by a first-level doctor who diagnosed her problem as pharyngotonsillitis and treated her with amoxicillin + clavulanate without improvement. Two days later, diplopia was added, which led her to come to our hospital. Upon arrival, the vital signs were recorded as blood pressure 120/70 mmHg, respiratory rate 18 breaths per minute, temperature 35.7 °C, and O2 saturation 98% in room air. She was alert, oriented in three spheres, normoreflexic, had isochoric pupils with ptosis when making a pathetic look, and had preserved motor and sensory function of the facial nerve. She also had dysphonia, inability to swallow, with difficulty to manage secretions. She had normal thorax with adequate ventilatory mechanics, lung fields with adequate air entry and exit, rhythmic precordium without aggregated auscultator phenomena, globular abdomen at the expense of panniculus adiposus, preserved peristalsis, pelvic limbs with 1/5 bilateral loss of strength, preserved sensitivity, bilateral areflexia, thoracic limbs strength 3/5 bilateral, and ataxic gait.

Suspecting Guillain Barré syndrome due to finding areflexia, ophthalmoplegia, ataxia, and progressive weakness with sensory deficit, paraclinical tests were performed in order to evaluate a probable differential diagnosis, in which no abnormality was observed except for leukocytosis (15.05 × 103/mm3). Lumbar puncture was performed, the results of which are presented in table 1 in detail. An Erasmus Guillain-Barré Syndrome Respiratory Insufficiency Score (EGRIS) of 7 points and a Medical Research Council sum score (MRC-SS) of 14 points were calculated. Therefore, it was decided to secure the airway. Thereafter, she was transferred to the intensive care unit where she continued her hospitalization under joint management of the hematology and neurology services. Plasmapheresis was performed in 5 sessions. The nerve conduction study of four extremities reported moderate to severe pure motor polyradiculoneuropathy, characterized by demyelination as the primary pathophysiological mechanism and secondary axonal degeneration, which affected the four extremities in a relatively symmetrical manner, compatible with Guillain-Barré variety acute inflammatory demyelinating polyradiculoneuropathy. During her hospitalization in the intensive care unit, she developed atelectasis as a complication, which was resolved by dynamic changes in ventilatory mechanics. The patient was successfully extubated on the third day of hospital stay. She was classified by the neurology service as Guillain-Barré syndrome, Miller Fisher variant, with pharyngeal-cervical-brachial variant overlap. She remained hospitalized for seven more days, she was discharged from the hospital after 15 days of hospital stay without apparent sequelae.

Later, the patient came to receive medical treatment due to progressive inability to swallow, upon arrival at the emergency room she reported restlessness and inability to walk.

As time passed, she showed significant clinical improvement, until she was completely asymptomatic.

2. Learning points

The Miller Fisher variant occurs in approximately 1-2 cases per 1,000,000 population with a predilection for the male gender (1). They usually present with the triad of ophthalmoplegia, areflexia and ataxia, while facial and pharyngeal weakness occurs in the pharyngeal-cervical-brachial variant (2). But our case presented with palpebral ptosis, in addition to dysphonia, inability to swallow, and inability to manage secretions. Additionally, she had ataxic gait. When making a clinical diagnosis, we found data suggestive of the Miller Fisher variant, as well as the pharyngeal-cervical-brachial variant. We consider it an overlap of these pathological entities. In the literature, we found that overlap of Guillain-Barré variants is not uncommon in the clinical setting; Sekiguchi et al. found overlaps in 50% of Miller fisher syndrome patients, 23% with Pharynx Cervico Brachyeal-Guillain barré syndrome and 15% with conventional Guilllain-Barré syndrome (3). The authors believe that the Guillain-Barre overlapping Miller Fisher and Cervico brachial Pharynx variety is a rare entity, it is necessary to adequately identify it by performing studies to check ganglioside antibodies to GM1, GD1a, and GQ1b. However, we are in a hospital unit where these antibody studies are not yet available, the presence of

Table 1 Details of lumbar puncture sample analysis

Initial Lumbar Puncture	Lumbar Puncture after 5 days
Glucose: 69 mg/dL	Glucose: 89 mg/dL
Proteins: 28 mg/dL	Proteins: 37.8 mg/dL
Cells 1 Cell/mm^3	Cells: 1 Cell/mm^3
Cl: 130 mEq/L	Cl: 121 mEq/L
Crenocytes: negative	Crenocytes: Negative
Pandy: Negative	Pandy: Negative
CSF aspect: Rock water	CSF aspect: Rock Water

CSF: Cerebrospinal fluid

complex anti-ganglioside antibodies may aid the diagnosis of cases in which Differential diagnoses cannot be ruled out.

3. Declarations

3.1. Acknowledgement

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3.2. Authors' contribution

All the authors met the standards of authorship based on the recommendations of the International Committee of Medical Journal Editors.

3.3. Conflict of interest

The authors state that they have no conflicts of interest.

3.4. Funding

Not applicable.

3.5. Consent for publication

Patient's consent for publishing the case with no identifiable personal information was obtained.

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