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CASE REPORT

Acute Motor and Sensory Axonal Neuropathy in a Teenager

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ABSTRACT

A 14-year-old girl presented with a one-month history of progressive, ascending weakness in all four limbs, culminating in an inability to ambulate and increasing shortness of breath. Neurological examination revealed right-sided lower motor neuron facial weakness, 4/5 muscle strength in the upper extremities, and 2/5 in the lower extremities. Deep tendon reflexes were diminished. Nerve conduction studies demonstrated a severe axonal sensorimotor polyneuropathy with reduced compound muscle action potentials (CMAPs) and sensory nerve action potentials (SNAPs), consistent with axonal loss. Cerebrospinal fluid analysis revealed albuminocy to logic dissociation, with a protein level of 75 mg/dL and a normal cell count. Brain and spinal MRI were normal. The patient was diagnosed with acute motor and sensory axonal neuropathy (AMSAN), a variant of Guillain-Barré syndrome (GBS), and received a 5-day course of intravenous immunoglobulin (IVIG). She demonstrated gradual improvement and regained independent ambulation eventually. This case highlights the importance of considering GBS in the differential diagnosis of acute flaccid paralysis, even with atypical presentations like facial nerve palsy, and emphasizes the crucial role of electrodiagnostic studies and CSF analysis in confirming the diagnosis.

KEYWORDS:

• Guillain-Barré syndrome • Acute flaccid paralysis • AMSAN • Nerve conduction studies • Lumbar puncture • Adolescent.

INTRODUCTION

Guillain-Barré syndrome (GBS) is an immune-mediated polyradiculoneuropathy characterized by rapidly progressive, often ascending, weakness that can lead to paralysis[1] The annual incidence of GBS is estimated at 1-2 cases per 100,000 individuals.[2] GBS presents with diverse clinical manifestations, requiring a broad differential diagnosis for acute flaccid

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paralysis (AFP).[3] We report this case because it highlights the diagnostic challenge of GBS presenting with atypical features, such as facial nerve palsy, which can mimic other neurological conditions.

CASE PRESENTATION

A 14-year-old girl presented with a one-month history of progressive weakness in all four limbs, progressing to an inability to ambulate independently. She also reported increasing shortness of breath. Four months prior to presentation, she experienced a febrile illness with headache, for which she was prescribed escitalopram and clonazepam. She denied recent travel, unusual exposures, or vaccinations.

CLINICAL FINDINGS

On examination, the patient was alert and oriented but exhibited mild tachypnea. She had right-sided lower motor neuron facial weakness. Muscle strength was graded as 4/5 in the upper extremities and 2/5 in the lower extremities (Medical Research Council scale). Deep tendon reflexes were diminished (1+) at the ankles and knees, with equivocal plantar responses. Sensory examination revealed decreased sensation to light touch and pinprick distally in all limbs.

INVESTIGATIONS

- **Nerve conduction studies (NCS):** Demonstrated a severe axonal sensorimotor polyneuropathy with reduced CMAPs and SNAPs, consistent with axonal loss.
- **Cerebrospinal fluid (CSF) analysis:** Revealed albuminocytologic dissociation, with a protein level of 75 mg/dL and a normal cell count.
- **Brain and spinal MRI:** Normal, excluding structural abnormalities and other neurological conditions.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for AFP in this patient included:

- **Acute disseminated encephalomyelitis (ADEM):** Distinguished by the frequent presence of encephalopathy and multifocal neurological deficits, which were absent in this case, and the normal MRI findings.
- **Myasthenia gravis:** Differentiated by the presence of fatigability, often with fluctuating weakness, and the distinct findings on electrophysiological studies (repetitive nerve stimulation), which were inconsistent with this case.
- **Botulism:** But less likely given the pattern of weakness was 'ascending' not 'descending' and the absence of prominent autonomic dysfunction (e.g., pupillary abnormalities, dry mouth, constipation).
- **Spinal cord lesion (e.g., compression, transverse myelitis):** Excluded by the absence of a sensory level, bowel/bladder dysfunction, and the normal spinal MRI.

DISCUSSION

This case highlights the diagnostic challenges posed by AFP in adolescents. The timely use of NCS and CSF analysis was essential for diagnosing AMSAN and differentiating it from other potential causes. Prompt IVIG treatment likely contributed to the patient's favorable outcome. This case also underscores the importance of recognizing atypical GBS presentations, such as facial nerve palsy.

AMSAN is a less common GBS variant characterized by axonal degeneration of motor and sensory nerves. It is often associated with a more severe clinical course and slower recovery compared to demyelinating GBS subtypes.[11,12] The pathogenesis of AMSAN involves immune-mediated attack on the axonal membrane, leading to axonal degeneration and conduction block.[13]

Facial nerve involvement in GBS is not uncommon, occurring in up to 50% of patients.[4] However, isolated or prominent facial diplegia can sometimes mimic other conditions like Miller Fisher syndrome (MFS) or brainstem lesions.[5] In this case, the presence of limb weakness and the electrodiagnostic findings of a sensorimotor axonal neuropathy pointed towards a GBS variant rather than MFS, which is characterized by ophthalmoplegia, ataxia, and areflexia. The presence of albuminocytologic dissociation in the CSF further supported the diagnosis of GBS.[6]

Several studies have reported cases of GBS with prominent facial nerve involvement. A study by Ropper *et al*. found that patients with facial weakness at onset were more likely to have cranial nerve involvement and a poorer prognosis.[7] Another study by Yuki *et al*. highlighted the association of facial diplegia with anti-GQ1b antibodies, often seen in MFS, but also reported cases of facial diplegia with other GBS subtypes.[8] Our case, however, did not have ophthalmoplegia or ataxia, making MFS less likely.

The patient's response to IVIG was positive, with gradual improvement in muscle strength and eventual regaining of independent ambulation. IVIG is a first-line treatment for GBS, and its efficacy has been demonstrated in several clinical trials.[9,10] However, the response to IVIG can vary, and some patients may require additional treatments like plasma exchange or rehabilitation.

Limitations: This case report is limited by its single-case design, which limits the generalizability of the findings. A larger case series or a retrospective review would provide more information about the clinical spectrum and outcomes of AMSAN in adolescents. Furthermore, while the patient showed improvement, long-term follow-up data would be valuable to assess for any residual neurological deficits.

CONCLUSION

This case report emphasizes the need for a comprehensive approach to evaluating adolescents with AFP. Clinicians should maintain a high index of suspicion for GBS and utilize appropriate investigations, including NCS and CSF analysis, to ensure accurate and timely diagnosis. Early intervention with IVIG can significantly improve patient outcomes and facilitate recovery.

Conflicts of interest: None

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