Clinical Case Report

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Increased intracranial pressure in Guillain–Barré syndrome

A case report

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Abstract

Rationale: Guillain–Barré syndrome (GBS) is an inflammatory autoimmune demyelinating polyneuropathy that affects most of the peripheral nervous system. Papilledema and raised intracranial pressure (ICP) are seen in some patients, and are thought to be associated with elevated cerebrospinal fluid (CSF) protein—though CSF protein levels are normal in some patients, thus the specific mechanisms remain unclear. Interleukin (IL)-17 levels are elevated in the CSF and plasma in GBS patients, and elevated IL-17 in the CSF of patients with idiopathic intracranial hypertension has been reported. Intravenous immunoglobulin (IVIG) exerts therapeutic effects by downregulating IL-17 in GBS patients.

Patient concerns: Herein we describe a case of a 14-year-old girl who initially presented with relapsing limb weakness.

Diagnoses: Magnetic resonance imaging revealed an enlarged ventricle, electromyography, and nerve conduction studies were suggestive of polyradiculopathy, and lumbar puncture revealed elevated ICP with normal cells and elevated protein values.

Interventions: She was treated with IVIG 0.4 g/kg per day for 5 days.

Outcomes: At a 6-month follow-up there had been no recurrence.

Lessons subsections: In GBS patients who have a relapsing course and develop papilledema with possible immunological disturbance, an accurate early diagnosis in conjunction with the prompt initiation of immunotherapy may improve clinical symptoms and the prognosis.

Abbreviations: CSF = cerebrospinal fluid, EMG = electromyography, GBS = Guillain–Barré syndrome, ICP = intracranial pressure, IL = interleukin, IVIG = intravenous immunoglobulin, MRI = magnetic resonance imaging.

Keywords: GBS, Guillain–Barré syndrome, IL-17, intravenous immunoglobulin, IVIG

1. Introduction

Guillain–Barré syndrome (GBS) is a self-limiting inflammatory autoimmune demyelinating polyneuropathy that affects most of the peripheral nervous system.[1] Papilledema and increased intracranial pressure (ICP) are seen in some patients,[2–5] and are thought to be associated with elevated cerebrospinal fluid (CSF) protein[5] though CSF levels are normal in some patients,[4,6] thus the specific mechanisms involved remain unclear.[3,7] Herein we describe a case of a 14-year-old girl who initially presented with relapsed limb weakness. Electromyography (EMG) and nerve conduction studies were suggestive of polyradiculopathy, and lumbar puncture revealed elevated ICP with normal cells and elevated protein levels. She was treated with intravenous immunoglobulin (IVIG) 0.4 g/kg per day for 5 days, and at a 6-month follow-up there had been no recurrence. We reviewed the literature pertaining to GBS with intracranial hypertension, to explore the specific mechanisms involved and alert clinicians that an accurate early diagnosis in conjunction with prompt initiation of immunotherapy may alleviate clinical symptoms and improve outcomes.

2. Case report

A 14-year-old girl was admitted to hospital on August 21, 2017 with a 7-day history of progressive weakness and aching in both legs, which had spread to her arms 3 days after initial onset in the legs. Evaluation and history taking at the time of admission suggested generally good health with no significant past medical events or signs of infection. Physical examination revealed weakness in the distal extremities, and areflexia and hyperalgesia between T12 and L2. Magnetic resonance imaging (MRI) revealed that the volume of the ventricle was 16.7 mL. EMG and nerve conduction studies revealed a reduction in the amplitude of muscle action potentials, slowed conduction velocity, prolonged F response waves in median nerves, and absent F response waves in peroneal nerves, consistent with the GBS. Subsequent lumbar puncture revealed a CSF pressure of 190 mmH_2O, and the CSF contained normal cells (2×10^6/L) and an increased protein level (1171.8 mg/L). She was treated with IVIG 0.4 g/kg per day for 5 days, and the weakness gradually improved. One month from symptom onset the patient had fully

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recovered, and she was attending school regularly. One week later she suffered weakness again and her clinical condition gradually worsened. On October 22, 2017 she was admitted to our hospital. Physical examination revealed papilledema, lumbar puncture revealed a CSF pressure of 290 mmH_2O, and the CSF contained normal cells (6×10^6/L) but an increased protein level (1128.4 mg/L). There was no loss of vision, and neither serum nor CSF interleukin (IL) levels were investigated. EMG and nerve conduction studies revealed a reduction in the amplitude of muscle action potentials, slowed conduction velocity, and an absence of F response waves in the motor nerves of limbs (peroneal, tibial, ulnar, and median). MRI and magnetic resonance venography results were normal, with a ventricle volume of 18.2 mL. IVIG 0.4 g/kg per day for 5 days was again administered. Ten days later she had fully recovered, and she has remained asymptomatic over the subsequent 6 months. The publication of this case report was approved by the Ethics Committee of the First Affiliated Hospital of Xinxiang Medical University, China, and written informed consent for its publication was obtained from the subject and her legal guardian in this study.

3. Discussion

GBS is an acute polyneuropathy associated with a variable degree of weakness that reaches maximal severity within 4 weeks. It is most commonly preceded by an infection, and generally runs a monophasic course.[1] The current patient had no definite preceding infection, fully recovered with IVIG treatment, and had a recurrence of limb weakness 2 months after the initial onset of the syndrome. EMG, nerve conduction studies, and lumbar puncture results supported a diagnosis of GBS, and her CSF pressure was 290 mmH_2O.

A number of cases of GBS with papilledema and subsequently increased CSF pressure have been reported since 1936,[2–9] and edema of the spinal nerve rootlets seen in GBS has been implicated in causing decreased absorption of proteins,[6,8] which is thought to contribute to increased ICP.[5] Notably however, some cases of GBS have been reported in which there was increased ICP but normal CSF protein levels.[4,6,9] It is well known that increases in the volume of brain tissue, CSF, or blood can all lead to a critical increase in ICP, and in some cases secondary hydrocephalus and loss of vision occur necessitating surgical intervention.[3,6,9] The current patient had a relapsing course and developed papilledema with an enlarged ventricle. This is concordant with Reid and Draper,[3] who suggested that these symptoms may be caused by immunological disturbances that result in an accumulation of CSF. The specific mechanisms involved remain unclear.

Edwards et al[10] reported that patients with idiopathic intracranial hypertension had increased levels of IL-17 in the CSF. In a recent study IL-17 levels were substantially elevated in both the CSF and plasma in GBS patients, and IVIG evidently exerts its therapeutic effects in GBS patients by downregulating IL-17.[11] The current patient had a relapsing course and developed substantially increased ICP and mildly elevated CSF protein, and in the 6 months after treatment with IVIG alone there was no symptom recurrence or visual impairment, which is concordant with a previous report.[3] Long-term follow-up observation and further studies are required.

4. Conclusion

In summary, we have described the case above to alert clinicians that elevated ICP can be seen in GBS, and an accurate early diagnosis in conjunction with the prompt initiation of immunotherapy may alleviate clinical symptoms and improve outcomes. A randomized controlled trial and systematic studies of treatments are required to confirm our observations.

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