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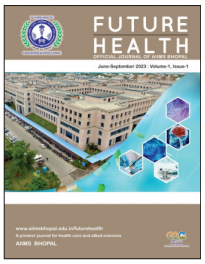

Case Report

Intracranial hypertension in a child with Guillain-Barre syndrome - A rare complication

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ABSTRACT

Guillain-Barre syndrome (GBS) is a rare autoimmune disorder of acute onset causing demyelination primarily in the peripheral nervous system. Central nervous system (CNS) involvement, particularly elevated intracranial pressure (ICP), is an atypical manifestation and is even rarer in children. This case report presents the unique case of an eight-year child with Guillain-Barre syndrome who exhibited signs of raised intracranial pressure. The child presented with progressive weakness, followed by new-onset esotropia, headache, and vomiting. The diagnosis of Guillain-Barre syndrome was made by clinical findings and nerve conduction studies (NCS). With intravenous immunoglobulin (IVIG) therapy and conservative medical management for raised ICP, child showed significant improvement in limb power, but esotropia was persisting. The presence of raised ICP in GBS remains a rare finding with uncertain pathophysiology.

INTRODUCTION

Guillain-Barré syndrome (GBS) is a rare demyelinating disorder of the peripheral nervous system (PNS). It is of autoimmune etiology, characterized by progressive muscle weakness and paralysis. Though the PNS is predominantly involved, the central nervous system (CNS) may be rarely involved. One of the atypical manifestations of GBS is elevated intracranial pressure (ICP), which poses unique diagnostic and therapeutic challenges. Elevated ICP in GBS patients, especially in pediatric cases, is even rarer, with limited documented cases in the literature. This case report presents a unique and intriguing case of a child with GBS who exhibited signs of raised ICP, emphasizing the importance of recognizing this unusual presentation and its implications in clinical management.

CASE REPORT

Clinical description

An 8-year-old male child presented with chief complaints of progressive weakness of bilateral lower limbs, a newly noticed inward deviation of the right eye for 3 days, headache, and vomiting for two days. There was no history of febrile illness or any significant medical illness. The patient had normal sensorium, and there were no abnormal movements. No change in bowel and bladder habits was noted. Antenatal and natal history was uneventful with a smooth perinatal transition. The child was immunized in accordance with the national immunization schedule and was developmentally normal.

The child was found to have right eye esotropia; the motor system examination showed symmetrical weakness in the bilateral upper and lower limbs (Medical Research Council Sum Score of 32), areflexia with mute plantars, and hypotonia in all limbs. No sensory deficit or autonomic dysfunction was noted, and no sign of meningeal irritation could be elicited. The rest of the systemic examination was unremarkable. Modified Erasmus GBS Outcome Score (mEGOS) was 4, and GBS Disability Score at admission was 4.

Management and outcome

Our provisional working diagnosis was GBS, further supported by nerve conduction studies, which suggested the acute motor axonal neuropathy (AMAN) variant. Cerebrospinal fluid (CSF) examination was done on day 7 of the illness, which showed albumin-cytological dissociation, confirming the GBS diagnosis. The presence of new onset esotropia along with headache and vomiting induced the suspicion of raised ICP. The fundus examination showed minimal blurring of disc margins, in view of which neuroimaging of the brain and spine was done. The latter ruled out any space-occupying lesion or parenchymal involvement, consolidating the diagnosis.

The child was given intravenous immunoglobulin (IVIG) 2 g/kg over 5 days. Decongestive measures for raised ICP (head end elevation, osmotic diuresis) and supportive care were initiated and continued from day 1 to day 3 of hospitalization, relieving headache and vomiting.

Serial muscle power charting was done, by day 11 of illness child had nearly regained complete power in all 4 limbs and was ambulatory with an medical research council (MRC) sum score of 44, mEGOS score of 3, and GBS Disability Score of 2. Despite the improvement of power in limbs, the esotropia remained persistent even on a 2-week follow-up. The ophthalmological evaluation showed a normal fundus and no refractive error.

DISCUSSION

The presence of raised ICP in GBS is a rare finding, with a reported incidence of only 4-6% in children, and the underlying pathophysiology remains elusive.[1] Multiple mechanisms have been proposed to explain this finding, the earliest one being the protein absorption theory, which proposed that edema in the spinal nerve rootlets, which occurs in GBS, hampers protein absorption causing increased CSF protein, which deters CSF absorption by the arachnoid granulations, leading to the increase in ICP.[2] Multiple case reports showed a dissociation between CSF protein levels and the development of raised ICP. These include differences in the onset of symptoms and the peak of protein level, and the presence of near-normal protein levels in some patients with raised ICP.[3,4] Another theory put forward speculated that it was cerebral edema, likely due to inflammation, which led to the raised ICP and it was not associated with an increase in CSF, since patients were found to have normal-sized ventricles.[5]

GBS is primarily a T1 helper cell (Th1) mediated disease. However, Th17 and Th22 cells have also been found to have a role in pathogenesis through their cytokines IL-17 and IL-22 and were elevated in GBS patients. This also correlated with disease severity, such as the presence of raised ICP and relapses. IVIG mediates its therapeutic effects by down-regulating these cells and their cytokines.[6]

The current patient had features of raised ICP, which required decongestive measures for symptomatic relief. This was similar to earlier cases, which even required therapeutic lumbar punctures and insertion of the external ventricular drain.[1] An association between the presence of raised ICP and a relapsing course has been noted in earlier cases reported. This was not observed in the current patient on follow-up.[7] Earlier link between obesity and intracranial hypertension in children with GBS was also seen in our patient (body mass index-16.9 kg/m^2) and lies in the overweight category for his age.[1]

Thus, reporting of raised ICP in this pediatric patient adds to the previous knowledge of GBS and insists on awareness and vigilance of the clinician in the presence of relevant clinical manifestations and timely initiation of decongestive measures for prevention of sequelae related to intracranial hypertension.

CONCLUSION

The presence of raised intracranial pressure in Guillain-Barre syndrome remains a rare finding with uncertain pathophysiology. The current case adds to the limited literature on raised intracranial pressure in paediatric Guillain-Barre syndrome, highlighting the importance of recognizing this rare presentation and its implications for clinical management.

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