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Case Report

A case of Guillain–Barré syndrome with meningeal irritation

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Abstract

Here, we report a 5-year-old girl with Guillain–Barré syndrome who presented with a chief complaint of pain in the extremities, which was followed by neck stiffness. Bladder dysfunction was found, which required catheterization. Magnetic resonance imaging revealed marked enhancement of the nerve roots in the cauda equina on T1-weighted imaging after gadolinium injection, and nerve conduction studies led to a diagnosis of Guillain–Barré syndrome. Her symptoms improved after intravenous immunoglobulin therapy, but her neck stiffness remained 16 days after admission. Four weeks after admission, she could walk without support. As patients with signs of meningeal irritation may be diagnosed with other diseases, such as meningitis, it is important to recognize atypical cases of pediatric Guillain–Barré syndrome to achieve early diagnosis and treatment.
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Keywords: Guillain–Barré syndrome; Acute inflammatory demyelinating polyneuropathy; Neck stiffness; Meningeal irritation

1. Introduction

Guillain–Barré syndrome (GBS) represents an acute, immune-mediated attack on the peripheral nervous system that leads to flaccid paralysis and can be divided into various subtypes, including acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), Miller Fisher syndrome, and Bickerstaff brainstem encephalitis. The diagnosis of GBS is relatively easy in patients with typical clinical and neurophysiological findings. However, children presenting with atypical features, such as meningeal irritation have also been reported [1–4]. Here, we report the clinical course and neurophysiological and neuroimaging findings of a patient with GBS who had an atypical presentation, including signs of meningeal irritation.

2. Case report

A previously healthy 5-year old girl was admitted to our hospital due to neck stiffness that began 1 day before admission. She suffered from acute otitis media eleven days before admission. She recognized pain in the lower extremities 6 days before admission and was unable to walk 1 day before admission. On admission, she was alert but could not stand. The results of head computed tomography (CT) were normal. Analysis of her cerebrospinal fluid (CSF) showed cell counts of 11/μl and 133 mg/dl protein. One day after admission, neck stiffness was continuous and hoarseness was also present. The straight leg raising test was also positive.

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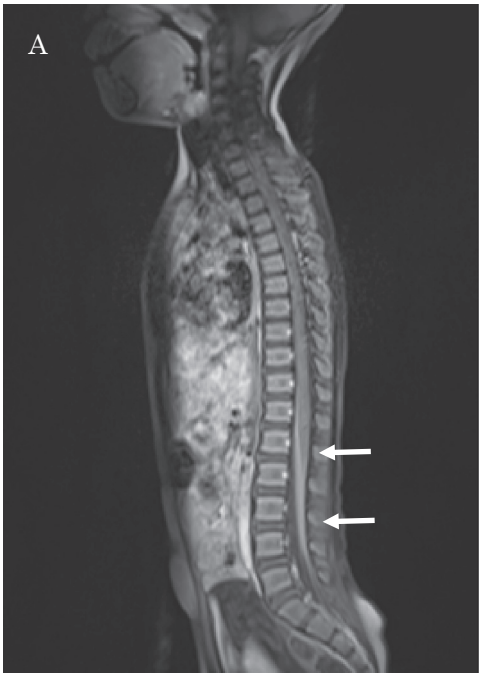
Neurological examinations revealed that the deep tendon reflexes (DTRs) were absent in the lower extremities and decreased in the upper extremities; manual muscle testing indicated grade 4 muscle strength for the upper and lower extremities. She showed reduced sensation in the lower extremities and bladder dysfunction necessitating catheterization. Although the magnetic resonance imaging (MRI) findings were normal without enhancement of the total spine and brain, T1-weighted imaging after gadolinium injection showed marked enhancement of the nerve roots in the cauda equina (Fig. 1), which suggested the possibility of GBS. Neurophysiological studies indicated a diagnosis of GBS with AIDP (Table 1). The IgG and IgM antibodies against the gangliosides GM1, GM2, GM3, GD1a, GD1b, GD3, GT1b, GQ1b, Gla-C, GalNAc-GD1a, and GD1a/GD1b were all negative. In addition, stool culture yielded no growth of *Campylobacter jejuni*, and antibodies against cytomegalovirus, Epstein-Barr virus, and mycoplasma were negative. She was successfully treated with intravenous immunoglobulin (400 mg/kg for 5 days) (Fig. 2).



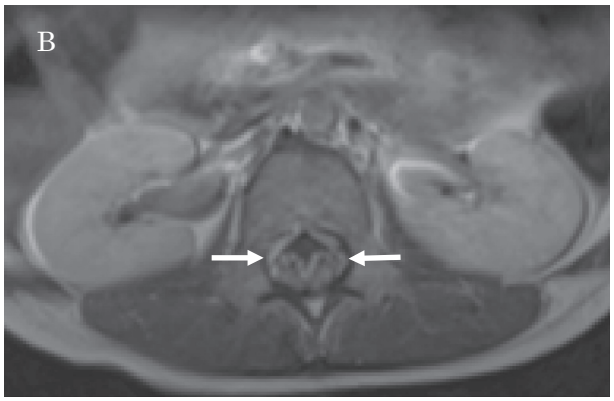


Fig. 1. Sagittal T1-weighted (TR600/TE11) (A) and axial T1-weighted (TR513/TE11) (B) magnetic resonance images after gadolinium injection showed marked enhancement of the cauda equina and ventral nerve roots (arrows), consistent with GBS [Baran GA et al. AJR Am J Roentgenol 1993;161:161–3].

3. Discussion

The diagnosis of GBS is relatively easy in patients with typical clinical and neurophysiological findings of motor and sensory neuropathy. Childhood cases of GBS presenting with meningeal irritation as an accompanying sign have been previously reported as atypical cases [3,4]. Nishimoto et al. reported similar features in two AMAN cases (our case was classified as AIDP) [3]. In another previous study, 16 of 46 patients [1], or 38% of children aged <6 years old [2], who were diagnosed with GBS, also exhibited neck stiffness. Although signs of meningeal irritation may be highly indicative of meningitis, one study reported that meningitis was diagnosed as bacterial or aseptic in only 30% and 13%, respectively, of children exhibiting signs of meningeal irritation [5], suggesting that the differential diagnosis of meningeal irritation should not be limited to meningitis. In our case, the clinical features, including absent DTR, decreased muscle strength, and pain in the extremities, suggested a diagnosis of GBS after meningitis was excluded because there were no significant increases in the CSF cell counts. Even if meningeal irritation were found, other clinical and neurological findings might be important to prevent misdiagnosis. GBS with central nervous system (CNS) lesions, which can present with meningeal irritation, has been reported only rarely [6,7] and Bickerstaff's brainstem encephalitis, characterized by progressive ophthalmoplegia, ataxia, disturbances in consciousness, pyramidal signs, extensor plantar responses, and long-tract sensory disturbance, is considered to overlap with the axonal GBS subtype [8]. The clinical and imaging findings were not supportive of CNS involvement or a GBS variant, such as Bickerstaff brainstem encephalitis.

Limb pain was the initial symptom in 53.4% and 24% of GBS patients aged >5 years and <5 years old, respectively [9]. Lower limb pain, which was the first symptom in our case and appeared 5 days before neck stiffness, may be important for the early diagnosis of GBS. The patient described here also had bladder dysfunction, which required catheterization, with no spinal lesion in repeated spine MRI. Watson et al. reported that bladder dysfunction requiring catheterization was found in 4 of 27 children with GBS [10].

Although the mechanisms underlying the signs of meningeal irritation in GBS patients are unknown, neck

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