마비성 장폐색증으로 발현한 길랑-바레 증후군 소아 1예

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A Child with Guillain-Barré Syndrome Presenting Paralytic Ileus

Guillain-Barré syndrome (GBS) is characterized by ascending symmetric paralysis, paresthesia, and autonomic dysfunction. Autonomic dysfunctions develop in two-thirds of the patients, and urinary retention and paralytic ileus usually develop in severe and advanced cases. There has been no pediatric case with paralytic ileus as a presenting symptom of GBS. Reported herein is a case of GBS presenting vomiting as an initial symptom. A 28-month-old girl was brought to the emergency room due to a 2-day history of vomiting. She vomited multiple times 1 day before the visit, and had only single voiding on admission day. Her abdomen was distended, with decreased bowel sound. Intravenous fluid was given under the diagnosis with acute gastroenteritis with dehydration. The abdominal computed tomography revealed severe paralytic ileus and urinary bladder distention. After having two seizures, she developed respiratory failure necessitating ventilator care. On day 8 after admission, motor weakness with areflexia was noticed. The cerebrospinal fluid analysis showed elevated proteins (80 mg/dL) with no white blood cells (0/mm³). The nerve conduction study showed axonal-type peripheral polyneuropathy. GBS was diagnosed. During the follow-up, the patient was found to have motor weakness in the lower extremities. Paralytic ileus with protracted vomiting can be an initial presentation of GBS in children. Autonomic dysfunction with GBS can be considered in a differential diagnosis in vomiting patients with unclear etiologies.

Key Words: Guillain-Barré syndrome, Vomiting, Ileus

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Introduction

Guillain-Barré syndrome (GBS) encompasses distal paresthesias, symmetric progressive ascending paralysis and diminished tendon stretch reflexes without sensory loss secondary to a postulated immune mediated mechanism involving the patchy destruction of peripheral nerve myelin^{1,2)}. There are several subtypes of GBS, including acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), and acute motor sensory axonal neuropathy. Compared to AIDP, AMAN tends to have prominent motor symptoms with severe fulminant weakness and little sensory loss, and more rapid progression and slow and poor recovery³⁾. Most patients have motor weakness as the initial presenting symptom. Autonomic dysfunctions develop in two-thirds of

the patients^{1,2)}. Dysautonomia is a common and feared finding in severe GBS. The symptoms range from exceptional blood pressure fluctuations and unexplained cardiac arrhythmia to less common signs like bowel or urinary tract involvement. Among them, urinary retention and paralytic ileus are known to develop in severe and advanced cases^{4,5)}. Paralytic ileus as a sign for the involvement of the intestinal autonomic nervous system occurs in only a small fraction of GBS patients, and is rarely reported as a presenting symptom of GBS^{5,6)}. So far, there has been no pediatric case report of paralytic ileus as the first manifestation of GBS. Reported herein is a child with GBS misdiagnosed as acute gastroenteritis (AGE) because of severe vomiting with dehydration at the initial presentation.

Case Report

A 28-month-old girl was brought to the emergency room due to a 2-day history of vomiting. She had mild upper respiratory infection 5 days prior to the visit, and vomited multiple times and had fever 1 day before the visit. On admission day, her vomiting

worsened, and she had poor oral intake and only single voiding. Her heart rate was 132 beats/min, her respiratory rate was 30 breaths/min, her body temperature was 38.6°C, and her blood pressure was 90/60 mmHg. She was alert but had dried lips and oral mucous membrane. And her skin turgor was decreased. Her abdomen was distended, with decreased bowel sound, and was tympanic on percussion. The results of initial laboratory tests were unremarkable except for mildly elevated blood urea nitrogen with 26.9 mg/dL (normal range: 8–23 mg/dL). She was rehydrated with intravenous fluid after being diagnosed with AGE with dehydration. Six hours after her visit, she had seizures twice. During the events, she showed eyeball deviation upward, with salivation but without convulsive movements. Ten hours after her visit, the abdominal distention was aggravated, and tachypnea with oxygen desaturation developed. The chest X-ray showed increased haziness in both lung fields, and the abdominal computed tomography revealed severe paralytic ileus and urinary bladder distention (Fig. 1). She was transferred to the intensive care unit for ventilator care, and was given intravenous antibiotics, antiviral agents, intravenous immunoglobulin (IVIG), and steroids after being diagnosed with febrile seizure,

Fig. 1. The axial (A, B) and coronal (C) abdominal computed tomography at the initial visit showed gas dilatation of multiple segments of the bowel loop, and urinary bladder distention.

sepsis, pneumonia, and acute respiratory failure. The serum antibody to *Mycoplasma pneumonia* was positive with titer of 5.73 (normal range: <0.8) at initial visit, and the titer decreased to 2.43 at 1 month later. Initial stool rotavirus antigen was negative and stool culture for *Campylobacter jejunii* was negative. On day 4 after admission, efforts were made to wean the patient from the ventilator after she showed improvement in her respiratory symptoms and chest X-ray findings, but the weaning was unsuccessful due to ventilation failure. On day 8 after admission, motor weakness with areflexia was noticed. Her motor strength was Medical Research Council (MRC) grade 0/5, and there were no deep tendon reflexes in her lower extremities. The cerebrospinal fluid analysis showed elevated proteins (80 mg/dL) with no white blood cells (0/mm³). The results of the nerve conduction studies (NCS) were compatible with axonal-loss-dominant peripheral polyneuropathy. The NCS showed that there was no sensory nerve action potential (SNAP) on both sural, right superficial peroneal nerves, and a low amplitude of compound motor action potential (CMAP) on the right median, ulnar, both peroneal, and tibial nerve. GBS was diagnosed, and IVIG was administered for 5 days. On day 18 after admission, the patient was weaned from the ventilator, and on day 40 after admission, subtle movements in her lower extremities were observed. Self-defecation was impossible, however, until the 32nd day after admission. Thereafter, the abdominal distension was improved. The Foley catheter was removed on day 34. On day 43 after admission, deep tendon reflexes started to appear. The patient's neurologic conditions improved with rehabilitative treatment. During the 9-year follow-up, however, motor weakness resulting in waddling gait remained as a sequela.

Discussion

Beyond motor weakness, autonomic nervous system may be prominent during the course of GBS. Autonomic manifestations most likely result from sympathetic and parasympathetic failure or overreactivity^{1,7)}. The symptoms include cardiac arrhythmia, blood pressure fluctuations, and abnormal hemodynamic responses to drugs, sweating abnormalities, pupillary abnormalities, and bladder and bowel dysfunction^{1,2,4)}. Autonomic dysfunction has been associated with an increased likelihood of intubation, suggesting that these clinical findings are indicative of a more severe disease¹⁾. There was one pediatric study reporting autonomic dysfunction of GBS in South Korea. Coe⁸⁾ reported that 9/132 (6.8%) of the patients had autonomic nerve dysfunction like arrhythmia, bradycardia, dysuria, and hypertension, which was a

lower incidence compared to the other reports^{1,2,9)}. Adynamic ileus has been estimated to occur in 2-9% of GBS patients. It is not known whether ileus is an immune-mediated dysautonomia or due to causes such as prolonged immobilization. According to the literature, ileus was almost as common due to immobilization. This category of ileus may be due to the stress of mechanical ventilation and immobility¹⁰⁾. Burns et al.¹⁰⁾ showed that adynamic ileus developed in 17/114 (15%) of the patients with severe GBS, and it was more common with immobilization. None, however, had ileus as the initial presentation of the disease¹⁰⁾. In GBS, paralytic ileus can occur due to the involvement of the sympathetic and parasympathetic fibers⁶⁾. In this type of ileus, immune attack may impair the balance between sympathetic and parasympathetic tone. This is supported by autopsy findings in GBS, where mononuclear inflammatory cell infiltration and demyelination of somatic, parasympathetic, and sympathetic fibers have been observed¹⁰⁾. This rare involvement of the autonomic nervous system is usually seen in severe cases and at advanced stages of the disease, but it is rarely reported as an early symptom of GBS⁵⁾. There are few case reports about paralytic ileus as the presenting symptom of GBS in adult patients^{5,6)}, but in the pediatric population, there has been no case report of paralytic ileus as the initial presentation of GBS. Hong et al.¹¹⁾ reported an 8-year-old boy with visceral autonomic neuropathy complicated by GBS, but his gastroparesis was a late complication of GBS during the recovery phase, not a presenting manifestation, as in the case of the patient herein. The case reported herein also had urinary retention at the initial visit, besides paralytic ileus. As the patient had the clinical features of protracted vomiting with abdominal distention, and decreased urination, it was initially thought that she had AGE with severe dehydration. Even after finding urinary retention on the abdominal CT, a neurologic condition like GBS could not be suspected. When her motor weakness became prominent, however, a neurogenic underlying condition was started to be investigated. Urinary retention is usually seen in neurologic disease or bladder outlet obstructive lesion, not with AGE with dehydration. Therefore, urinary retention might be a critical clue to the final diagnosis of the case reported herein at the beginning of the illness. With regard to the subtypes of GBS, autonomic dysfunction frequently affects patients with AIDP, but it rarely occurs in those with AMAN³⁾. Also, the patterns of autonomic involvement are qualitatively different between AIDP and AMAN⁷⁾. The bladder dysfunction developed in 50% of the patients with axonal GBS, contrary to 21% of those with classic GBS. Ryuji et al.¹²⁾ reported a 62-year-old woman with axonal GBS who simultaneously developed weakness and urinary retention, but without cardiovascular dysfunction. Two

mechanisms are hypothesized for the urinary dysfunction: the inflammation or immune attack of the lumbosacral autonomic fibers, and the immune attack of the inhibitory spinal cord interneurons, which secondarily leads to lumbosacral autonomic hyperactivity¹²⁾. The case reported herein was found to have axonal GBS rather than the demyelinating type through the NCS. Although it is known that autonomic dysfunction is rarely a complication in axonal GBS, the case reported herein had prominent bowel and bladder dysfunction as the first presentation of the disease. The patient's clinical course seemed to be related with the axonal subtype of GBS. Besides urinary retention, her albuminocytologic dissociation on CSF exam can support the paralytic ileus as initial presentation of GBS. It is known that CSF protein concentration is often normal in the first week after onset, but increases in more than 90% of patients by the end of the second week¹³⁾. Because she had seizures with vomiting, benign convulsion associated with mild gastroenteritis (BCAGE) could be one of differential diagnosis initially. However, her high fever and clinical findings suggestive of severe dehydration were thought to be against the diagnosis of BCAGE. Finally as for a preceding infection, *Mycoplasma pneumoniae* seemed to be associated with developing GBS in this patient based on changes in antibody titer over 1 month.

Paralytic ileus with protracted vomiting can be the initial presentation of GBS in children. Autonomic dysfunction with GBS can be considered in a differential diagnosis in vomiting patients with unclear etiologies.

요약

길랑-바레 증후군은 상행성의 대칭성 운동 마비와 이상 감각, 자율 신경계 이상 등을 특징으로 하는 질환이다. 자율신경계 이상이 약 2/3에서 발생하며, 이중 배뇨 장애 및 마비성 장폐색증은 주로 증상이 심한 경우와 병증이 상당히 진행된 단계에서 나타난다고 알려져 있다. 아직까지 소아에서 마비성 장폐색증으로 발현된 길랑-바레 증후군에 대한 증례 보고는 없다. 이에 저자들은 구토가 첫 증상으로 나타난 길랑-바레 증후군 1례를 경험하였기에 보고하고자 한다. 28개월 여아가 2일간의 구토가 있어 내원하였다. 내원 1일전부터 수차례의 구토가 발생하였고, 내원 당일 단 한차례의 소변을 보았다. 진찰상 장음 감소와 심한 복부 팽만이 관찰되었다. 탈수를 동반한 장염으로 진단 후 수액 치료를 하였다. 복부 전산화 단층 촬영에서 심한 장폐색증과 방광 확장이 관찰되었다. 두차례 발작 이후, 기계환기가 필요한 급성 호흡부전이 발생하였다. 입원 8병일째 양 하지의 마비와 함께 심부전

반사가 소실되었다. 뇌척수액 검사에서 단백 세포 해리 소견(백혈구 0 개/ μ L, 단백 80 mg/dL)가 있었고, 신경전달검사상 축삭형 다발성 말 초신경병증 소견을 보였다. 길랑-바레 증후군으로 진단되었다. 외래경과 관찰 시에 하지 근력 약화는 지속되었다. 심한 구토와 마비성 장폐색증은 길랑-바레 증후군의 첫 증상으로 나타날 수 있다. 소아에서 원인이 뚜렷하지 않은 구토의 감별 진단으로 길랑-바레 증후군의 자율신경 이상 증상을 고려해야 할 것이다.

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