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Atypical Clinical Features of Guillain Barré syndrome

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ABSTRACT

Guillain-Barré syndrome (GBS) is a heterogeneous group of acute immune-mediated polyradiculoneuropathy. The cardinal clinical features of GBS include progressive and symmetric muscle weakness accompanied by the absence of deep tendon reflexes as well as acute tetraplegia and respiratory failure. In addition, more benign or atypical forms might also occur. Atypical GBS is a heterogeneous disorder with various clinical presentations. We reported a case of a 29-year-old woman presenting with a complaint of sudden weakness in all four extremities during the last 10 days, preceded by a history of upper respiratory tract infection 3 weeks prior. She experienced paresthesia in all palms and feet. The patient also had difficulty urinating and experienced constipation for 1 week. On physical examination, peripheral tetraparesis and segmental ecteroceptive sensory disturbances were found at the 5th level of myelo-cervical segment and below. Neurological exam showed reduced deep tendon reflexes in lower extremities. In addition, urinary retention and constipation were found. She was suspected of having myelitis based on clinical symptoms. Serial EMG examination showed demyelinating polyradiculoneuropathy. Cerebrospinal fluid examination revealed cytoalbumin dissociation suggesting GBS. Cervicothoracic MRI imaging showed no abnormalities. The patient was diagnosed with GBS that clinically improved without receiving any IVIG or plasmapheresis therapy.

Keywords: Atypical, GBS, Guillain Barré syndrome, case report, Indonesia

Introduction

Guillain-Barre' syndrome (GBS) is an acute polyradiculopathy syndrome presented with various clinical manifestations.[1,2] A molecular mimicry attacks the peripheral nervous system responsible for the pathogenesis of this immune system-mediated polyneuropathy.[2] The classical feature of GBS is an acute ascending weakness accompanied by the absence of deep tendon reflexes. Respiratory failure requiring intensive care also occurs in 20-30% of patients. However, other patients showed more benign and uncommon clinical features, which is usually referred to as atypical GBS. Atypical GBS is characterized by local or regional involvement of motor and sensory axons of the peripheral nerves and autonomic nervous system. Typical presentations of GBS include acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motoric axonal neuropathy (AMAN) and acute motor-sensory axonal neuropathy (AMSAN). However, unusual presentations also reported in GBS such as cranial nerve involvement, pharyngo-cervical-brachial, cranial polyneuritis, as well as other presentations such as acute pandysautonomic and acute sensory neuropathy.[3-7]The clinical manifestations of atypical GBS often overlap with those of other diseases in early onset, thus making diagnosis more difficult. In initial onset, serial EMG and sometimes MRI are required to rule out the differential diagnosis if the clinical presentation is atypical.

In this case report, we discuss a patient with atypical GBS as reported previously. We report a case with clinical manifestations resembling myelopathy, yet serial EMG results and cerebrospinal fluid examination suggested a diagnosis of GBS. We describe clinical

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manifestations, medical examinations, management, and the differential diagnosis that should be considered when encountering this certain case.

Case Illustration

A 29-year-old woman presented with a complaint of sudden weakness in all four extremities for 10 days. The weakness of the legs became worse making the patient incapable of walking. She felt numbness in her hands and feet. She also had fever, cough and runny nose that recovered without medication. She had been unable to urinate for 2 days before her admission to the hospital. She denied any history of diarrhea, previous immunizations or surgery. The patient was not pregnant. She had no fever, swallowing difficulty, facial flushing or slurred speech. She had never experienced any loss of consciousness or change in behavior. She also had no history of low back pain, chronic cough, trauma, or diabetes mellitus. The patient claimed that she had never experienced anything like this before.

Physical examination revealed flaccid tetraparesis with normal physiological and without any pathological reflexes. Motoric strength was 1/5 for her left and right quadriceps femoris as well as iliopsoas; 2/5 for the hamstring, gastrocnemius, and tibialis anterior; and 4/5 for the upper extremities. Segmental exteroceptive sensory disturbances were found at the 5thlevel of myelo-cervicalsegment and below. Urinaryretention and constipation were also found. Cranial nerves were within normal limits. From cerebrospinal fluid examination, the cell counts were 7 mg/dL consisting predominantly of mononuclear cells. An increase amount of protein around 77.94 mg/dl was also observed. EMG-NCV examination was performed twice. The first one was performed on day 12 of the onset weakness. The result revealed a decreased CMAP amplitude and a prolonged F-wave latency of the right tibial nerve, but no response of the left tibial nerve CMAP (Figure 1). It also showed bilateral H-wave loss (Figure 1). On the second examination, 28 days after the onset weakness, a prolonged latency of the left and right tibial N-waves was observed, but no response to the H-wave, suggesting a demyelinating polyradiculoneuropathy. Cervicothoracic MRI was also performed and did not exhibit any abnormalities (Figure 2). The patient showed improvement with supportive and neurotrophic treatments. The diagnosis was performed based on Brighton level 1 criteria (Table 1).

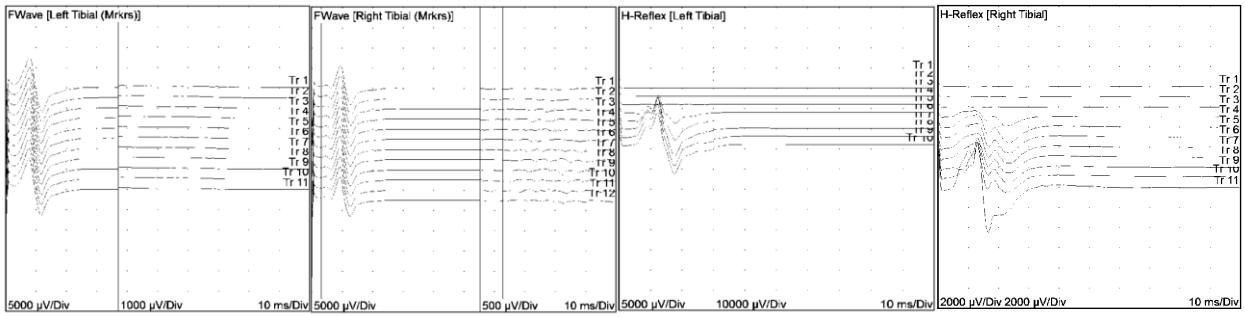

Figure 1. Prolonged F-wave seen in EMG-NCV and no response on H-wave (right panel)

Discussion

GBS is an acute polyneuropathy after infectious events affecting the motor system, sensory nerves and sometimes the autonomic system.[8] The frequency of previous infections in GBS caused by *Campylobacter jejuni* organisms is higher compared to other microorganisms in Asian countries. Clinical variations of GBS include Miller Fisher syndrome, Bickerstaff's

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brain stem encephalitis (BBE), pharyngeal-cervical-brachial motor variant, paraparetic motor variant, Pure sensory ataxic, and pandysautonomic.[8] The cardinal clinical features of GBS include progressive symmetric muscle weakness accompanied by absent or diminished deep tendon reflexes.[9]Lessthan 30% ofpatients suffer from acutetetraplegia and respiratoryfailure, while others showed morebenignoratypicalforms. Atypical GBSis a heterogeneous disorder exhibiting variousclinical presentations.[2]In GBSgroup, localor regional involvementof motor and sensoryaxonsoftheperipheralnerves and autonomicnervoussystem were uncommon.[3] Atypical GBS syndromes have been reported several times, including in 8 children in Iran[3] and 6 children in Croatia.[4]

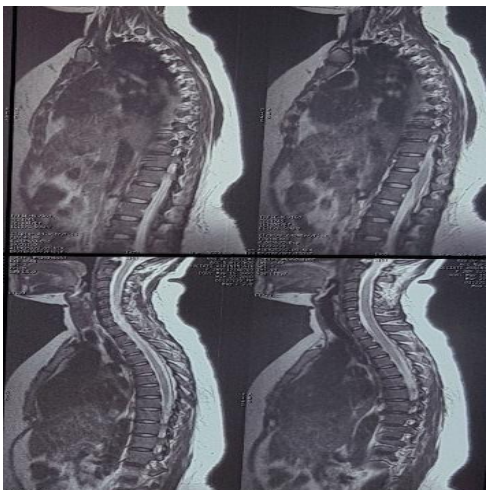

Figure 2. Normal contrast-enhanced cervicothoracic spine MRI

Table 1. Brighton criteria

Diagnostic criteria	Patient
Bilateral and flaccid weakness of limb	+
Decreased or absent deep tendon reflexes in weak limb	+
Monophasic course and time between onset-nadir = 12 hours to 28 days	+
Absence of alternative diagnosis for weakness	+
CSF cell count <50/ml	+
CSF protein concentration >60mg/dL	+
Nerve conduction study findings consistent with one of the subtypes of GBS	+
Level	1

Early symptoms of GBS are numbness, paresthesia, weakness, leg pain, or a combination of these symptoms. The main symptom presents as a progressive and symmetric bilateral limb weakness which persists for 12 hours to 28 days before a plateau phase. The patient in our report experienced tetraparesis with normal deep tendon reflexes, segmental sensory deficits (inconsistent) and autonomic disturbances in the form of urinaryretention and constipation. Her clinical features suggested a myelopathic process in the form of infection or demyelination in the spinal cord. However, due to the peripheral tetraparesis in the initial onset and the history of respiratory tract infection during 3 weeks prior, we did an EMG-NCV test and a lumbar puncture examination. EMG-NCV examination was performed on the 12th day after the onset of weakness.

Lumbar puncture performed at the onset of D13 showed cytoalbuminologic dissociation with

a cell count of 7/ml and a total protein of 77.94 mg/dL. Due to a suspicion of spinal cord process, a cervicothoracic MRI examination was carried out and abnormalities were not detected. A second NCV-EMG examination, performed at the onset of D30, found a prolonged distal F latency leading to the GBS-AIDP. GBS diagnosis was made based on clinical manifestations, CSF analysis, and electrophysiological studies according to Brighton level 1 criteria.

Autonomic dysfunction in GBS, especially bladder dysfunction has been reported. Previous studies have shown that 25% of GBS cases have bladder dysfunction. Therefore, clinical signs of early bladder involvement in GBS are considered an atypical presentation. Here, we report a case of GBS that presented as urinary retention with constipation and progressive limb weakness.[10]A study found that 25% of GBS patients had urinary difficulties, nocturnal urinary frequency incontinence and urinary retention.[11]Another study reported a similar prevalence (27.7%) of bladder dysfunction.[12]

Urodynamic findings include detrusor areflexia, sphincter dysfunction, impaired bladder sensation and/or excess detrusor activity.[12]The pathogenesis of urinary dysfunction in GBS is debatable, but it has been shown that urinary dysfunction in GBS is associated with upper motor neuron hyperactivity. Therefore, both the hyperreflexia and urinary dysfunction seen in this case presumably due to the involvement of inhibitory interneurons in the spinal cord or upper motor neuron.[13] Another study stated that the mechanism of bladder dysfunction may be due to hypoactivity and hyperactivity of the lumbosacral nerve with detrusor hypoactivity, detrusor hyperactivity, and hyperactive sphincter.[12]

In general, the differential diagnosis of GBS is hypokalemic paralysis. Atypical features of GBS, segmental sensory disturbances, and autonomic disturbances e.g. urinaryretention and constipation could obscure the diagnosis of GBS. Tetraparesis with normal deep tendon reflexes, segmental (inconsistent) sensory deficits, and autonomic disturbances e.g. urinaryretention and constipation found in patient could be an atypical presentation of GBS.

Conclusion

The atypical presentations of GBS present as peripheral limb weakness accompanied by segmental exteroceptive sensory disturbances, urinary retention, and constipation. These presentations suggested a myelopathy conditions; however, clinician should consider the possibility of GBS by carefully observed and comprehensively examined the clinical features. The diagnosis should be confirmed by performing nerve conduction tests and CSF analysis and further treatment should be prepared to improve patient outcome.

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