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Guillain-Barré Syndrome with multiple cranial neuropathies

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ABSTRACT

Background. Guillain-Barré Syndrome (GBS) is an autoimmune reaction against peripheral nerves that manifests clinically as acute polyradiculoneuropathy. Classic sensory-motor, pure motor, paraparesis, pharyngeal-cervical-brachial, Bickerstaff brainstem encephalitis, pure sensory, bilateral facial palsy with paraesthesia, and Miller-Fisher syndrome are all known variants of GBS. This case report seeks to describe a case of GBS with involvement of the cranial nerves.

Case report. A 39-year-old right-handed Balinese woman presented with LMN (Lower Motor Neuron) type paraparesis with ascending paralysis and involvement of cranial nerves III through XII. The results of the LCS (Liquor Cerebro-Spinal) demonstrate albuminocytological dissociation. According to the patient's EMNG (electromyoneurography) findings, Guillain-Barré Syndrome was of the AMAN (acute motor axonal neuropathy) variety. The patient was diagnosed with multiple cranial neuropathies and GBS of the AMAN type. EGRIS (Erasmus GBS Respiratory Insufficiency Score) was 3, and EGOS (Erasmus GBS Obstructive Sleep Apnea Score) was also 3. Following IVIG (Intravenous Immunoglobulin) treatment, the patient exhibited clinical improvement (Hughes score decreased from 4 to 2).

Discussion. The patient exhibits clinical manifestations of flaccid tetraparesis and multiple cranial nerve paresis. The clinical condition of the patient did not meet the criteria for GBS clinical variation. Symptoms of flaccid tetraparesis that do not involve the senses are classified as the motor-only variant. This patient's differential diagnosis includes the possibility of MFS and ophthalmoplegia. This patient, however, had flaccid tetraparesis and no ataxia. Electromyoneurography (EMNG) examination produces AMAN-type GBS. The patient was diagnosed with one of the clinical variants of GBS, AMAN-type GBS with multiple cranial neuropathies.

Conclusion. A case of GBS of the AMAN type accompanied by multiple cranial neuropathies has been reported.

Keywords: GBS type AMAN, GBS variant, multiple cranial neuropathy

INTRODUCTION

Guillain-Barré Syndrome (GBS) is an autoimmune response to peripheral nerves that manifests as acute polyradiculoneuropathy. Ascending paralysis in the extremities, areflexia/hyporeflexia, and hypotonia are classic GBS symptoms. Classic sensory-motor, pure motor, paraparesis, pharyngeal-cervical-brachial, Bickerstaff brainstem encephalitis, pure sensory, bilateral facial palsy with paraesthesia, and Miller-Fisher syndrome are known clinical variants of GBS. The prevalence of GBS increases with age (age 50 to 79) and male gender (2.4 incidents per 100,000) [1].

MFS, pharyngeal-cervical-brachial, Bickerstaff brainstem encephalitis, and bilateral facial palsy with paraesthesia are clinical variants of GBS with involvement of cranial nerves. Those clinical variations are inapplicable to the current scenario. Therefore, the purpose of this case report is to describe a case of AMAN-type GBS with involvement of the cranial nerves.

CASE REPORT

A 39-year-old Balinese female patient presented with complaints of paralysis in all four extremities. The patient reported that he was presently experiencing weakness in all four limbs, with a greater de-

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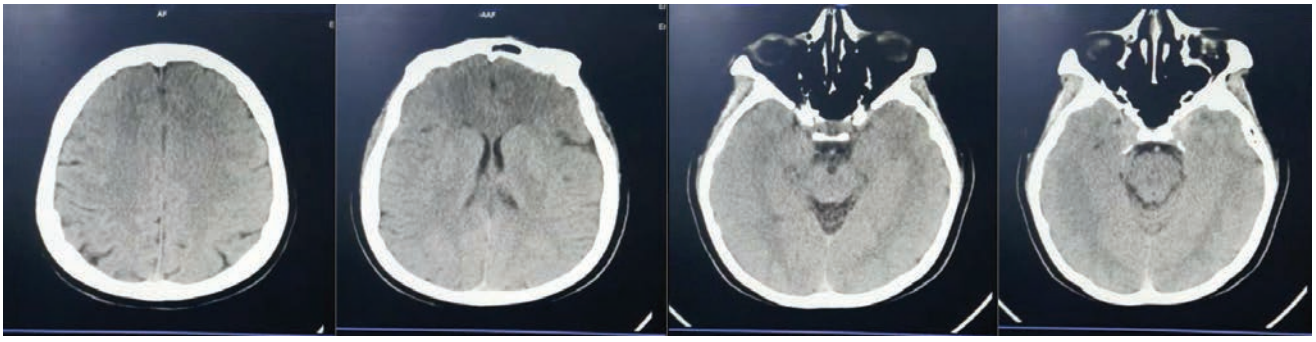


FIGURE 1. The density of gray and white matter, the gyrus and cerebral sulcus, the ventricular system, and the subarachnoid space were all within normal limits, as determined by a head CT without contrast. There was no obstruction or deviation of the paranasal sinuses or nasal septum

gree of weakness in the shoulder blade and groin. Five days before being admitted to the hospital, the patient awoke feeling weak in both extremities. The next day, both hands were feeble, and two days later, the entire body was weak. The patient is still able to walk but begins to stumble as a result of his or her frailty, which worsens until he or she is unable to stand or walk. After experiencing weakness in both limbs and legs, the patient complains that his voice changes to dysarthria, that the dysarthria is becoming heavier, and that his right eye begins to experience double vision. The next morning, the patient's family brought him to the local hospital due to generalized weakness, a visage pulled to the right, and a left eyelid that drooped more than the right. On the fourth day of treatment at the local hospital, it was reported that the patient had trouble swallowing (especially water) and rapidly choked. Other complaints, including worsening migraines, vomiting, tingling/thickness/heat sensation, and seizures, are denied.

This is the first time the patient has experienced this symptom. Approximately one week prior to this initial complaint, the patient complained of wheezing and fever, but the temperature was not measured and resolved with over-the-counter medication. Urination and feces production are within normal parameters.

The patient had no previous history of hypertension, type 2 diabetes, heart disease, stroke, spinal or cranial trauma, or cancer. The patient at the hospital had previously received intravenous mecobalamin 500 mg every 12 hours, intravenous methylprednisolone 62.5 mg every 12 hours, intravenous folic acid 1 mg every 12 hours, and intravenous citicoline 500 mg every 12 hours because of cranial nerve paresis, which led to the suspicion of a vascular event. Figure 1 depicts the findings of a head CT without contrast at the referring hospital, which revealed no infarction, bleeding, or intracranial abnormalities. According to the patient, the complaint has not changed despite receiving treatment, and no family members have experienced the same issue. In 2022, the COVID-19 vaccine was administered as the previous vaccination.

Vital signs were within normal limits upon physical examination. On neurological examination, consciousness was compos mentis (Glasgow Coma Scale (GCS) E4V5M6), left ptosis, extra et left ophthalmoplegia, left VII nerve paresis left intranuclear, IX and X nerve lesions, left XI nerve paresis (slight), XII nerve paresis left intranuclear, flaccid tetraparesis, areflexia, and sensibility and autonomic within normal limits.

Flaccid tetraparesis with ascending paralysis, involvement of the cranial nerves, and a history of upper respiratory tract infection lead to acute polyradiculoneuropathy with suspicion of GBS, a clinical variant of MFS, based on the patient's history and physical examination. Included in the follow-up examinations were a complete blood count, a lumbar puncture, and an EMNG (Electromyoneurography). Leukocytosis determined by routine blood tests (Leukocytes: 24.12 x 103/ L, neutrophils: 22.2 x 103/ L, lymphocytes: 0.98 x 103/ L). The patient underwent a chest X-ray to determine the source of the infection, which revealed pneumonia and right pleural effusion (Figure 2). With albuminocytological dissociation (Cells: 4/mm; Protein: 46.5 mg/dL), a lumbar puncture yielded an opening pressure of 18mmHg (Cells: 4/mm; Protein: 46.5 mg/dL). Figure 3 and Table 1 demonstrates that the patient's EMNG revealed mixed-type polyneuropathy, supporting the appearance of AMAN-type GBS (Acute Motor Axonal Neuropathy).

The patient was diagnosed with GBS of the AMAN type with multiple cranial neuropathies, based on clinical and supporting evidence. Hughes score: 4 (bedridden or dependent on a wheelchair), Erasmus GBS Score (EGRIS): 3 (moderate risk), and Erasmus GBS Outcome Score (EGOS): 3 (risk 0.5% of patients unable to walk independently within six months).

The patient was treated with intravenous immunoglobulin (IVIG) at a dose of 0.4 mg/kg/day for five consecutive days. The patient was discharged with a Hughes score of 2 (able to walk >10 meters but unable to sprint).

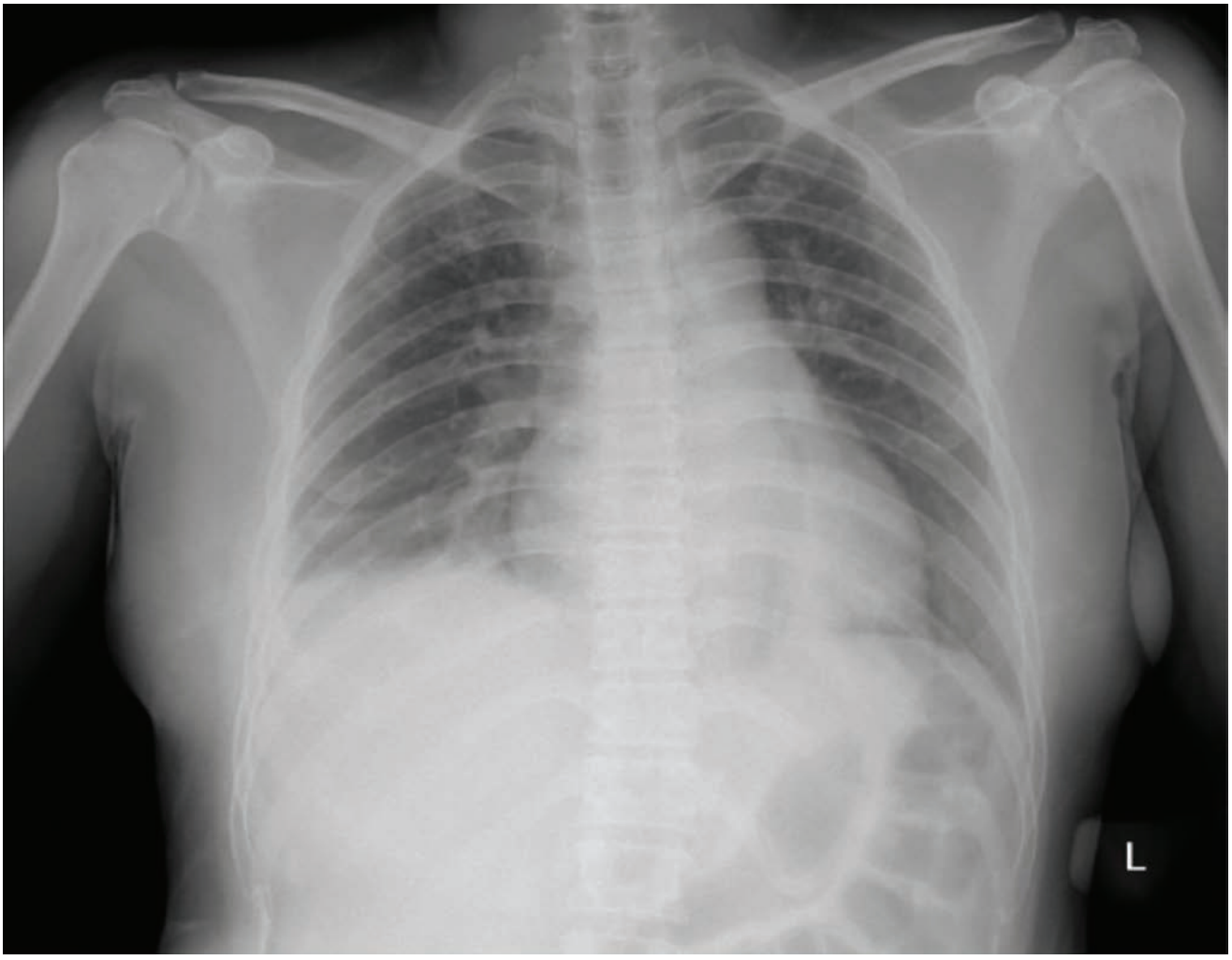
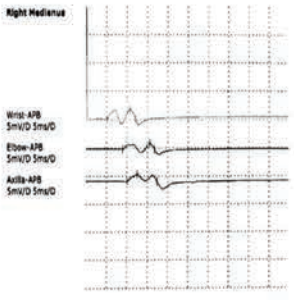


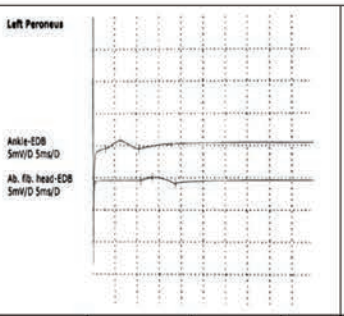
FIGURE 2. AP chest X-ray reveals a homogeneous covering at the base of the right hemithorax and a pointed left sheath closing the right pleural sinus

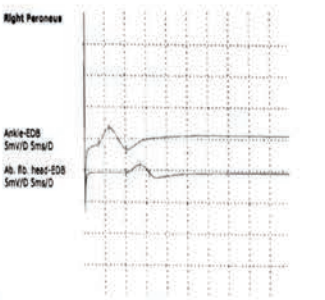
TABLE 1. Interpretation of electromyoneurography reveals a mixed polyneuropathy, supporting the presence of GBS (Acute Motor Axonal Neuropathy) of the AMAN variety

Compound Muscle Action Potential (CMAP)
• Right Median Nerve: prolonged distal latency, decreased amplitude, nerve conduction velocity normal • Right Radialis Nerve: prolonged distal latency, decreased amplitude, decreased nerve conduction velocity. • Right Ulnaris Nerve: normal distal latency, normal amplitude, normal nerve conduction velocity. • Right Peroneus Nerve: normal distal latency, normal amplitude, normal nerve conduction velocity. • Left Peroneus Nerve: normal distal latency, decreased amplitude, decreased nerve conduction velocity. • Right Tibialis Nerve: normal distal latency, normal amplitude, decreased nerve conduction velocity. • Left Tibialis Nerve: normal distal latency, normal amplitude, decreased nerve conduction velocity. • Right Tibialis F-Response: F-M latency < 56ms, persistence > 50%. • Left Tibialis F-Response: F-M latency < 56ms, persistence > 50%. • Right Median F-Response: F-M latency < 32ms, persistence > 50%. • Right Ulnaris F-Response: F-M latency < 32ms, persistence > 50%. • Left H-Reflex: No response. • Right H-Reflex: No response. • Distal CMAP amplitude < 80% of the lower limit of normal on the right median nerve, right radial nerve, and left peroneal nerve. • Conduction block with a proximal CMAP amplitude ratio: distal CMAP < 0.7 in the left peroneal nerve, right peroneus nerve, right ulna nerve, and right radial nerve.
Sensory Nerve Action Potential (SNAP)
• Right Median Nerve: Normal distal latency, normal amplitude, nerve conduction velocity normal • Right Radial Nerve: Prolonged distal latency, decreased amplitude, nerve conduction velocity normal • Right Ulna Nerve and Sural Nerve: Normal distal latency, normal amplitude, nerve conduction velocity normal • Left Sural Nerve: Normal distal latency, normal amplitude, nerve conduction velocity normal

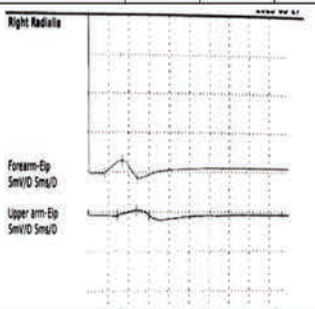
Motoric

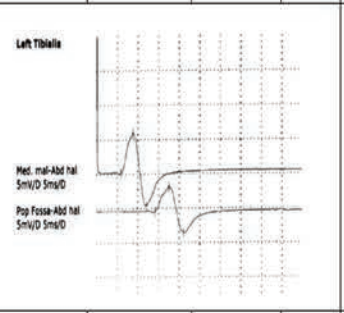


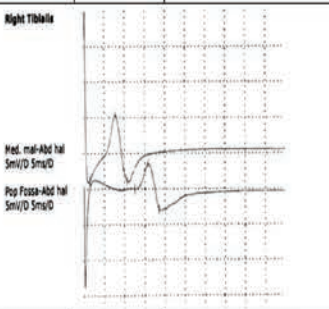




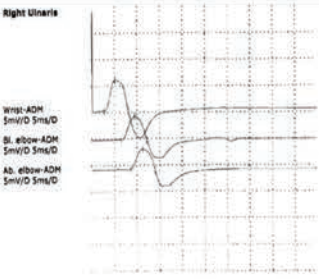
	Lat	Amp	CV		Lat	Amp	CV		Lat	Amp	CV
Wrist	5.13	1.58		Ankle-EDB	2.93	1.22		Ankle-EDB	3.48	2.5	
Elbow-Wrist	8.94	0.98	65.6	Ab.fib head-ankle	10.9	0.5	40.2	Ab.fib head-ankle	10.1	1.24	48.3
Axilla-Elbow	10.1	1.23	94.8								





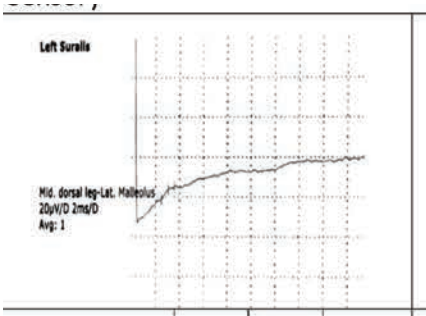


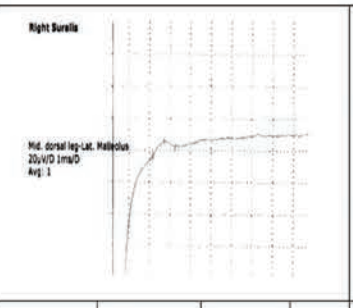
	Lat	Amp	CV		Lat	Amp	CV		Lat	Amp	CV
Forearm-Elp	4.16	1.5		Med. mal-Abd hal	5.9	5.8		Med. mal-Abd hal	1.47	9.2	
Elbow-Wrist	7.25	0.68	45.3	Pop Fossa-Med. mal	14.2	3.5	39.8	Pop Fossa-Med. mal	12.6	3.6	30.5

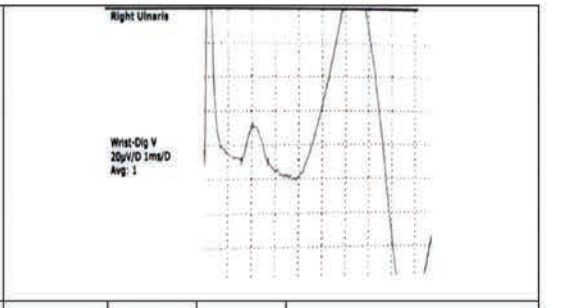


	Lat	Amp	CV
Wrist-ADM	2.79	5.8	
Bl.elbow-Wrist	6.58	4.5	60.7
Ab.elbow-Bl.elbow	8.58	3.9	60.0

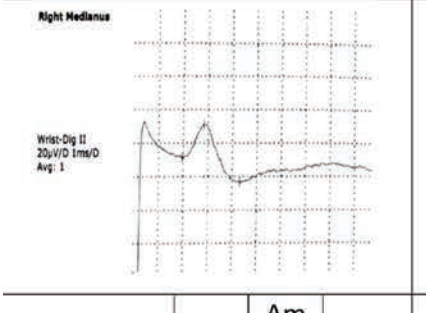
Sensory

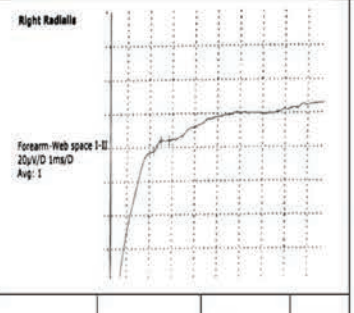






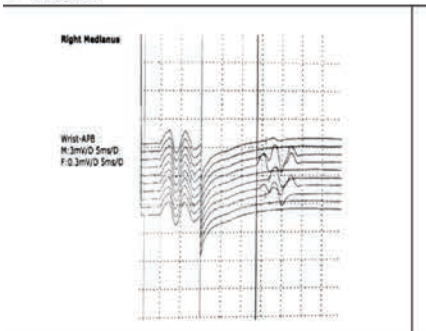
	Lat	Amp	CV		Lat	Amp	CV		Lat	Amp	CV
Mid. dorsal leg-Lat.Malleolus	3.06	1.85	52.6	Mid. dorsal leg-Lat.Malleolus	2.71	6.2	55.6	Wrist-Dig V	2.02	20.9	70.5

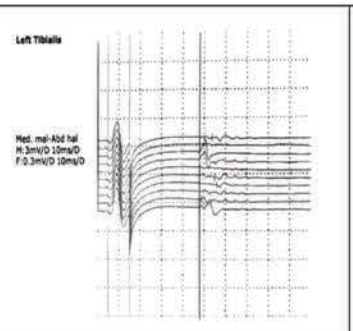


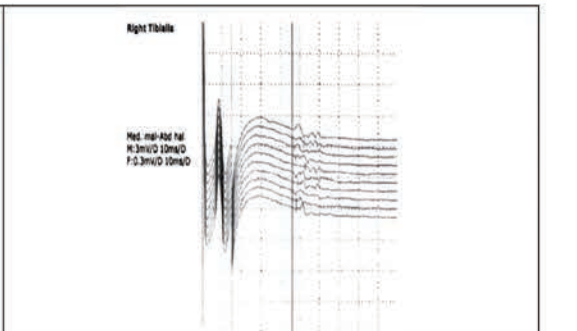


	Lat	Amp	CV		Lat	Amp	CV
Wrist-Dig II	2.83	24.9	66.3	Forearm-Web Space I-II	2.49	3.2	50.2

F-Wave

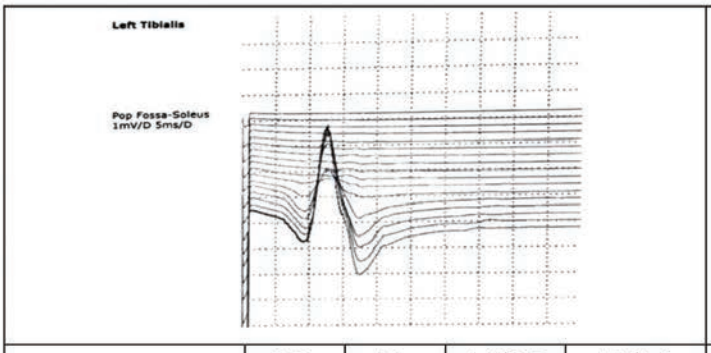


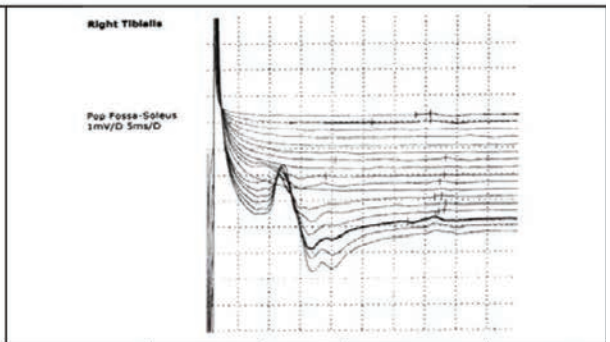




	F-M Lat	Shortest F-Lat	%F		F-M Lat	Shortest F-Lat	%F		F-M Lat	Shortest F-Lat	%F
Wrist-APB	27.8	28.8	60.0	Med. mal-Abd hal	43.1	47.9	70.0	Med. mal-Abd hal	45.1	46.1	>80.0

flex





	M-lat	H-lat	H/M Lat	H/M Amp		M-lat	H-lat	H/M Lat	H/M Amp
Pop Fossa-Soleus	9.8		<32.0		Pop Fossa-Soleus		32.2		<32.0

FIGURE 3. Electromyoneurography

DISCUSSION

GBS is an immune-related disease that manifests as polyradiculoneuropathy in peripheral nerves. Typically, respiratory or gastrointestinal infections precede GBS, but this is not the case. The origin of GBS is still unknown, but the emergence of GBS is triggered by infections, vaccinations, and other immune reactions [2]. Globally, the incidence of GBS ranges from 0.81 to 1.89 per 100,000 individuals per year. GBS is a monophasic disease characterized by progressive and acute limb weakness [3]. GBS is characterized by flaccid tetraparesis, areflexia/hyperreflexia, cytoalbumin dissociation, and sensory disturbances, cranial nerve involvement, consciousness, autonomic dysfunction, and muscular/radicular pain and examination in some variants. The EMNG is necessary for GBS type determination [4-6].

The three most common variants of GBS, according to epidemiology, are classic sensorimotor (30-85%), pure motor (5-70%), and paraparetic (5-10%) [3]. Each variant of GBS has characteristic clinical symptoms. The classic sensorimotor variant is characterized by swiftly progressive symmetrical weakness, areflexia, or hyporeflexia, which peaks in two weeks. If it only affects the motor system, then it is a motor variant; if it only affects the sensory system, then it is a sensory variant. The paraparesis variant is distinguished by paralysis in the lower extremities only. Cranial nerve variants include pharyngeal-cervical-brachial variants, bilateral facial neuropathy with paraesthesia, multiple facial sclerosis, and Bickerstaff brainstem encephalitis. The pharyngeal-cervical-brachial variant includes pharyngeal, cervical, and brachial paralysis. In this instance, there is no cervical disorder, so the pharyngeal-cervical-brachial variant is incompatible. If the facial nerve is affected bilaterally, paresthesia and hyporeflexia are included in the variant of bilateral facial palsy with paresthesia. In this instance, the involvement was limited to the facial nerve. In contrast, the involvement of cranial nerves III-XII was incompatible with bilateral facial palsy. Clinically, MFS demonstrates ophthalmoplegia, ataxia, and areflexia. There was no ataxia in this patient, and MFS could not account for the involvement of other cranial nerves in this patient; therefore, it was not MFS. Bickerstaff brainstem encephalitis is characterized by a combination of classic sensorimotor and MFS symptoms, as well as a decrease in consciousness. In this instance, the patient's level of consciousness is still high, so the diagnosis of variant Bickerstaff brainstem encephalitis does not apply [6,7].

In addition to being a clinical condition, GBS can be classified into several subtypes based on EMNG results. AIDP (acute inflammatory demyelinating polyradiculopathy) is the most prevalent form of demyelinating symptoms and is associated with a more favorable prognosis. AMAN (acute axonal motor neuropathy) is a rare variant characterized by clinical

manifestations of axonal injury, motor involvement, and a worse prognosis for recovery. AMSAN (acute motor sensory axonal polyneuropathy) has a similar pathogenesis to AMAN, with additional sensory involvement symptoms.9 The results of the patient's EMNG, namely polyneuropathy of the mixed type, support the AMAN type of GBS (Acute Motor Axonal Neuropathy) [8,9].

The patient in this case report was diagnosed with AMAN-type GBS with multiple cranial neuropathies based on the above description. IVIG was administered at a dose of 2 gr/kg/day for five days to patients with GBS, resulting in a clinical improvement (the Hughes score decreased from four to two) and an improvement in cranial nerve paresis. Immunotherapy produces outstanding outcomes for motor weakness and multiple cranial nerve paresis [10,11]. This supports the notion that multiple cranial nerve weakness is one of the clinical symptoms of GBS.

CONCLUSION

A case of AMAN-type GBS with multiple cranial neuropathies that improved with immunotherapy has been reported.

Conflict of interest: none declared
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