Sudden Deafness and Facial Diplegia in Guillain-Barré Syndrome: Radiological Depiction of Facial and Acoustic Nerve Lesions

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Abstract

We herein report a 26-year-old man with Guillain-Barré Syndrome (GBS) coexisting facial nerve palsy (FP) and deafness. He developed deafness, facial weakness, and limb weakness and numbness. Neurological examination showed facial diplegia, bilateral hypoacusia, areflexia and sensorimotor deficits in the distal limbs. The nerve conduction study findings supported the diagnosis of the demyelinating polyneuropathy. An audiogram revealed sensorineural hearing loss of 40-50 dB. Auditory brainstem responses disclosed no elicitation of waves I to IV on both sides. Magnetic resonance imaging depicted abnormal enhancement in bilateral facial and acoustic nerves. Physicians should pay more attention to auditory dysfunction in GBS patients with FP.

Key words: Guillain-Barré Syndrome, sensorineural hearing loss, facial nerve palsy, gadolinium-enhanced FLASH imaging, T2-weighted SPACE imaging

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Introduction

Cranial nerve involvement is known to occur in patients with Guillain-Barré syndrome (GBS). The facial nerve is affected most commonly in GBS patients (1-6). During the course of GBS, 20-60% of patients develop bilateral facial nerve paresis (1, 2). A radiological lesion of the facial nerve has been reported in several patients with GBS or Miller Fisher syndrome (3-6). The frequency of sensorineural hearing loss (SHL) was extremely rare in patients with GBS. The electrophysiological studies were described previously in these patients (7-14). However, there have been no previous reports about the radiological depiction of the acoustic nerve lesion in GBS patients. We herein highlight the radiological and electrophysiological changes in a patient with GBS who had facial diplegia and bilateral SHL.

Case Report

A 26-year-old man noticed weakness and numbness in his hands and feet. He had no symptoms of antecedent infection. Four days later, bilateral facial weakness and deafness were present. He was admitted to our department. Physical examination was normal. Neurological examination showed a moderate to severe degree of facial diplegia and bilateral hypoacusia, predominantly on the right side. The other cranial nerves were normal. A slight to mild degree of weakness existed in the distal muscles of the four extremities (Medical Research Council scale of grade 4). Deep tendon reflexes were not elicited in the four extremities, and the plantar responses were flexor. A severe degree of dysesthesia was present in the hands and feet. Superficial, vibration and proprioception sensations were decreased mildly in the

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Figure 1. Serial changes of ABR findings. ABR on Day 9. (A) Waves I to IV were not elicited, and the peak latencies of wave V were delayed on both sides. (B) ABR on Day 19. Waves I and II were not elicited and the peak latencies of waves III, IV and IV were delayed on the right side. The peak latencies of all five waves were delayed on the left side. (C) ABR on Day 34. The peak latencies of waves I, II, III and V were delayed and wave IV was not elicited on the right side. The peak latencies of waves I, II and III were delayed on the left side. The peak latencies of waves IV and V were normal. The mean peak and interpeak latencies (standard deviation) in sex-matched controls: I of 1.70 (0.17) msec, II of 2.93 (0.19), III of 3.90 (0.20), IV of 5.03 (0.27), V of 5.71 (0.27), I-III of 2.10 (0.17), III-V of 1.92 (0.17), I-V of 4.03 (0.23).

distal portion of four extremities. The remaining functions were normal, including the vestibulo-cerebellar system. Routine laboratory studies were not remarkable with the exception of mild hyponatremia (133 mEq/L). Serum antibodies to gangliosides GM1, GD1a, GD1b, GQ1b and GT1a were not detected. Pathogen tests for *Campylobacter jejuni*, *Mycoplasma pneumoniae*, cytomegalovirus, Epstein-Barr virus, rubella virus and herpes simplex virus were negative. Chest X-ray, electrocardiography and carotid ultrasonography were normal. At 6 days after clinical onset (Day 6), a cerebrospinal fluid study exhibited protein of 157 mg/dL, 19 mononuclear cells/mm³ and normal cytology. Myelin basic protein and oligoclonal immunoglobulin G band were not detected.

Motor and sensory nerve conduction studies were performed on Day 10. Distal motor latencies were markedly prolonged in the median (5.2 ms), ulnar (4.9 ms), peroneal (7.1 ms) and tibial nerves (7.0 ms). The amplitudes of compound muscle action potentials in the peroneal and tibial nerves were decreased, whereas they remained within the normal ranges in the median and ulnar nerves. Motor nerve conduction velocity was decreased markedly in the median (32 m/s), ulnar (38 m/s), peroneal (27 m/s) and tibial nerves (26 m/s). F-wave was significantly prolonged in latency in the median nerve (35.5 ms), and not elicited in the tibial nerve. The amplitudes of sensory nerve action potentials were normal in the median, ulnar and sural nerves. Sensory nerve conduction velocity was normal in the median and sural nerves, and mildly reduced in the ulnar nerve (41 m/s). These electrophysiological findings supported the diagnosis of acute inflammatory demyelinating polyneuropathy (AIDP). Audiogram revealed SHL of 59.2 dB in the right

ear and 44.2 dB in the left ear. To assess the auditory brainstem responses (ABR), the auditory stimulus used monaural rarefaction clicks of 0.1 msec duration at an intensity of 90 dB. The unstimulated ear was masked with white noise at 50 dB below the stimulus intensity. The right and left ears were independently stimulated, and Cz-A1 and Cz-A2 (International 10-20 system) recordings were performed. Two separate averages of 1,000 stimuli were superimposed for the analysis. Prolongation of peak latencies or interpeak latencies was defined at ≥ 3 standard deviation of the mean value in sex-matched controls. On Day 9, ABR disclosed no elicitation of bilateral waves I, II, III and IV. The peak latencies of wave V were delayed at 8.09 msec on the right side and 8.01 on the left side (Fig. 1A).

To visualize the fine structure of the facial and acoustic nerves, cochlea and vestibule, three-dimensional (3D) T2-weighted sampling perfection with application optimized contrast using different flip angle evolution (SPACE) imaging and 3D fast low angle shot (FLASH) T1-weighted fat-saturated gradient echo imaging were applied using a 1.5-T scanner (Siemens, Erlangen, Germany). On Day 9, gadolinium-enhanced FLASH imaging displayed abnormal enhancement in bilateral acoustic nerves and, the geniculate ganglions, the intracanalicular and mastoid segments of bilateral facial nerves (Fig. 2-4). Brain and spinal cord magnetic resonance imaging (MRI) was normal.

The patient was diagnosed with GBS (AIDP) accompanied by the involvement of the facial and cochlear nerves. After treatment with intravenous immunoglobulin (IVIg: 500 mg/kg/day for 5 days), his hearing impairment markedly ameliorated. On Day 19, audiogram was normal. The

Figure 2. Axial MRI on Day 9. (A, B) Post-contrast 3D FLASH imaging showed gadolinium enhancement in bilateral facial (VII) and acoustic nerves (VIII). (C, D) 3D T2-weighted SPACE imaging identified the geniculate ganglion (GG), intracanalicular facial nerve (VII) and acoustic nerve (VIII).

Figure 3. Coronal MRI on Day 9. (A, C, E) Post-contrast 3D FLASH imaging showed gadolinium enhancement in bilateral facial (VII) and acoustic nerves (VIII). (B, D, F) 3D T2-weighted SPACE imaging identified the cochlea (Co), geniculate ganglion (GG), intracanalicular facial nerve (VII) and acoustic nerve (VIII).

2nd ABR revealed no elucidation of waves I and II, and the delayed peak latencies of wave III (5.04 msec), IV (5.99) and V (6.74) on the right side. The peak latencies of all five waves (wave I of 2.72 msec, II of 3.85, III of 4.73, IV of 6.16 and V of 6.71) were prolonged on the left side. These interpeak latencies of I-III, III-V and I-V were within the normal ranges (Fig. 1B). On Day 34, follow-up ABR showed no elucidation of wave IV and prolonged peak latencies of waves I (2.42 msec), II (3.91), III (4.78) and V (6.56) on the right side. The peak latencies of waves I (2.41 msec), II (3.48) and III (4.53) were delayed, and the peak latencies of waves IV (5.83) and V (6.38) were within the normal ranges on the left side. The interpeak latencies of I-III, III-V and I-V were within the normal ranges on both sides (Fig. 1C). At three months after admission, the patient had no motor deficits in the four extremities. The right facial nerve paresis and numbness in the hands and feet per-

sisted. No abnormal enhancement was found in the facial and acoustic nerves on MRI.

Discussion

We reported the radiological hallmark and serial changes of ABR in a patient with GBS coexisting facial diplegia and bilateral SHL. SHL has been reported rarely in GBS patients (7-14). Previous cases are summarized in Table. Of 10 patients, bilateral SHL occurred in 9 patients (90%). Seven patients (70%) had simultaneous facial nerve palsy. Four patients (40%) had dysesthesia in the distal limbs. The present patient also had facial diplegia and a severe degree of dysesthesia in the hands and feet. Numbness in the distal extremities and facial nerve palsy were the most common neurological profile in GBS patients with SHL. Eight patients in the previous reports underwent motor and sensory nerve

Figure 4. Gadolinium-enhanced 3D FLASH imaging on Day 9. Axial (A, B) and coronal views (C, D). Widespread enhancement was found in the mastoid segments of bilateral facial nerves (VII).

Table. The Present Case and Previous Reports of Guillain-Barré Syndrome (GBS) with Sensorineural Hearing Loss

Authors. (*) reported years	Age/sex	Lesion side	Other CN impairment	ABR findings	Serum antibodies to gangliosides	Treatment	Prognosis of SHL
Sugiyama et al. (7) 1985	38 years/woman	Bilateral	VII, IX, X	No elucidation of waves II to IV.	ND	PSL	Good
Pall et al. (8) 1987	19 years/man	Bilateral	II, III	ND	ND	mPSL	Good
	30 years/man	Right	II, III	ND	PSL	PSL	Good
Nelson et al. (9) 1988	21 years/woman	Bilateral	III, V, VII, IX, X	No elucidation of all five waves.	ND	ND	Good
Nagai et al. (10) 1991	48 years/man	Bilateral	III, VII, IX, X	Unclear waves I to IV.	ND	PSL	Good
				Delayed peak latencies of wave V.			
Herinckx et al. (11) 1995	8 years child	Bilateral	Absence	Delayed peak latencies of waves I, II and III, and no elucidation of waves IV and V on the right side.	ND	ND	Poor
				Normal on the left side.			
Tamura et al. (12) 1997	24 years/man	Bilateral	VII	Delayed peak latencies of waves I, III and V.	ND	mPSL, PSL	Good
				Delayed interpeak I-III latencies.		IVIG	Good
Yano et al. (13) 1999	31 years/man	Bilateral	VII	No elucidation of all five waves.	Negative anti-GM1 IgG		Good
Ueda et al. (14) 2008	31 years/woman	Bilateral	VII, IX, X	Delayed peak latencies of wave I.	Negative	IVIG	Fairly good
Present case	26 years/man	Bilateral	VII	No elucidation of waves I to IV.	Negative	IVIG	Good
				Delayed peak latencies of wave V.			

ABR: auditory brainstem response, CN: cranial nerve, IVIG: intravenous immunoglobulin, mPSL: methylprednisolone, ND: not described, PSL: prednisolone or prednisone
 *Reference number

conduction studies. The electrophysiological findings suggested AIDP in 4 patients and axonal degeneration in 4 patients. ABR was performed in 8 patients, and the results showed no elucidation of all five waves, prolonged peak latencies of waves I to III or prolonged interpeak I-III. These abnormalities of the peripheral segments on ABR were improved (7, 9, 10, 13, 14). The present patient also had the similar pattern. With respect to treatment in 8 patients, 5 patients received steroid therapy. Three patients were treated with IVIG. The prognosis of SHL was good in 9 adults (90%) and poor in one child (10%). Therefore, the good recovery from SHL and abnormal ABR speculated the possibility that demyelinating damage to the cochlear nerve might play a major role in the pathogenesis of SHL.

Since an autoimmune mechanism underlying GBS-associated SHL was suspected, the serum antibodies to gangliosides were measured in three of the previous patients (13, 14). Anti-GM1 ganglioside antibodies were not detected in any of those patients (Table). Interestingly, serum antibodies to sialyl-i ganglioside or sulfoglucuronosyl glycolipids were found in patients with autoimmune inner ear diseases, including SHL and Meniere's disease (15, 16).

A previous study reported that the frequency of these antibodies to peripheral nerve glycosphingolipids did not differ significantly between the sera of GBS patients and control subjects (17). Further immunological studies are therefore needed to elucidate whether distinct anti-glycolipid antibodies exist in GBS patients with SHL.

Sudden onset of hearing loss can be caused by various etiological factors. MRI is commonly performed in patients with SHL because its superior soft tissue contrast is able to distinguish the underlying pathology of inner ear lesions. Recent technological advancements of MRI, especially the use of gadolinium contrast, refinement of its resolution, and the application of special sequences have provided further insight into labyrinthine pathology. 3D SPACE, 3D Fourier transformation-constructive interference in a steady-state, 3D FLASH and 3D spoiled gradient-echo imaging have been conducted in SHL patients (18, 19). Combined 3D T2-weighted SPACE and gadolinium-enhanced 3D FLASH sequences had benefits for the depiction of the facial and acoustic nerve lesions in the present patient. The nerve conduction studies suggested AIDP in the present patient. Abnormal findings of ABR and audiography were markedly

ameliorated after IVIG treatment. These electrophysiological and radiological changes indicated that demyelination in the facial and acoustic nerves, rather than axonal degeneration, was responsible for the SHL in this patient.

In conclusion, we have herein highlighted the radiological and electrophysiological changes in a GBS patient with facial and cochlear nerve involvement. Combined 3D T2-weighted SPACE and gadolinium-enhanced 3D FLASH images were a useful method for detecting lesions in the cranial nerves VII and VIII. Abnormal ABR was reported previously in GBS patients without SHL (20, 21). One of these studies mentioned the presence of facial nerve palsy in three of five GBS patients with abnormal ABR (20). Thus, physicians should pay more attention to auditory dysfunction in GBS patients with facial nerve palsy.

The authors state that they have no Conflict of Interest (COI).

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