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A Case of Severe Refractory Residual Sensory-neural Hearing Loss in Guillain–Barre Syndrome: An Unexplored Entity

ABSTRACT

Guillain–Barre syndrome (GBS) is an acute areflexic rapidly progressive polyradiculoneuropathy with albumin-cytological dissociation. Cochlear nerve demyelination is a rare cause of hearing loss in GBS patients. Here, we are reporting a 28-year-old man with GBS with bilateral facial and acoustic neuropathy. On neurological examination, he had bifacial weakness, bilateral hypoacusia, generalized areflexia, and sensory motor deficits in the distal limbs. The nerve conduction tests showed the evidence of the demyelinating polyneuropathy. A pure-tone audiogram showed bilateral sensorineural hearing loss of 40–50 dB. Thus, auditory dysfunction in GBS patients must be evaluated particularly in the setting of facial palsy.

Key words: Demyelination, Guillain–Barré syndrome, Refractory, Sensorineural hearing loss

INTRODUCTION

Guillain–Barre syndrome (GBS) is the most common cause of acute flaccid paralysis all over world.[1] It is known to be associated with cranial nerve involvement. Most commonly affected nerve in GBS patients is facial nerve 70%.[1] About 20–60% cases develop bilateral facial paresis during the course of GBS.[2] The involvement of cochlear nerve which causing sensorineural hearing loss (SNHL) is very rare in patients with GBS.[3] The electrodiagnostic studies are described previously in all these patients.[4-7] Cochlear nerve demyelination can be refractory and residual hearing impairment can be the consequence.

CASE REPORT

A 28-year-old man, with no comorbidities, presented with complaints of sudden onset difficulty in walking and paresthesia in both hands and feet for 3 days. He had prior cough and cold before 8 days. Two days later, he complained of difficulty in swallowing and speaking. There was tinnitus with hearing impairment in both the ears. There was no hisotry of bowel and bladder dysfunction. No history of breathing difficulty or fever.

On examination, his higher mental functions were normal. Both pupils were equal and reacting to light. Corneal and conjunctival reflexes were present. Speech was slurred. He had right eye ptosis and bilateral facial weakness. A slight to mild degree of weakness was present in all four limbs (modified Medical Research Council scale of Grade 4). He had generalized areflexia and both plantar reflexes were flexor. A severe dysesthesia was present in the hands and feet. Touch, pain, and temperature sensations were impaired distally in all four limbs. Vibration sense was impaired till both malleoli in the lower limbs. Joint position sense was impaired until both ankle joints in both lower limbs. Vibration and joint position senses in the upper limb were normal. Romberg/s test was positive. Rest of the systemic examination was normal. No signs of autonomic dysfunction were present.

His routine blood investigations were normal. Serum creatinine was borderline – 1.3. CPK was 395. ANA/IF was negative. p-ANCA and c-ANCA were negative. ANA blot was positive for anti-Ro 52kD. Acetylcholine receptor antibody was negative. Complement levels were normal. Chest X-ray and electrocardiograph were normal. At 5 days after clinical onset (day 5), a cerebrospinal fluid study exhibited protein of 54 mg/dL, 18 mononuclear cells/mm^3 with lymphocytic pleocytosis and normal cytology. No malignant cells were present. Urine test for porphobilinogen was negative. Magnetic resonance imaging (MRI) brain with contrast showed that no significant abnormality in brain parenchyma with no abnormal enhancement was found in the 7th–8th nerve complex and inner ear structures. HRCT of petrous temporal bone was normal. MRI whole spine screening showed straightening of cervical lordotic curvature.

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Motor and sensory nerve conduction studies were performed on day 7. Distal motor latencies were markedly prolonged in the median, ulnar, peroneal, and tibial nerves, as shown in Table 1. The amplitudes of compound muscle action potentials in the peroneal and sural 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, ulnar, peroneal, and tibial nerves. F-wave was significantly prolonged in latency in the median nerve (36.5 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 (40 m/s). These electrophysiological findings supported the diagnosis of acute inflammatory demyelinating polyneuropathy (AIDP). Pure-tone audiogram revealed SHL of 50.2 dB in the right ear and 48.4 dB in the left ear. Serum antiganglioside Ig G antibody panel was negative. Serum antiganglioside IgM antibody panel was positive for GM3 antibody. The patient was diagnosed with GBS (AIDP) with bifacial and bilateral cochlear nerves involvement. The patient was admitted and treated with intravenous immunoglobulin 2 g/kg divided over 5 days.

At 2 months after admission, the patient had 60–70% improvement in weakness in both upper and lower limbs. He was able to walk without support. However, bilateral tinnitus with hearing impairment and facial weakness persisted. Repeat Electromyography and Nerve conduction study done after 2 months show prolonged blink reflex (demyelinating range), absent soleus "H" reflex and attenuated lower limb SNAPs. No evidence of active axon loss in both upper and lower limbs was present. Residual hearing impairment can be quite disturbing.

DISCUSSION

GBS is usually associated with symmetrical weakness of the limbs which reach a plateau within 4 weeks.[1] Sensory-neural hearing loss is a very rare manifestation and complication of GBS.[8] There have been ten similar studies of GBS associated with hearing loss.[9] Bilateral SNHL occurred in 9 (90%) out ten patients. Simultaneous facial nerve palsy was found in 7 (70%) patients. Dysesthesias in distal limbs found in four patients (40%). In GBS patients with SNHL, numbness in the distal extremities and facial nerve palsy was the most common neurological profile. Nine patients from previous studies underwent nerve conduction studies. The electrophysiological findings suggested demyelinating type in five patients and axonal variety in four patients.[9] Five patients received steroid therapy. Three patients were treated with IVIG. The prognosis of SNHL was good in nine adults (90%) and poor in one child (10%). Therefore, the good recovery from SNHL and abnormal ABR speculated the possibility that demyelinating damage to the cochlear nerve might play a major role in the pathogenesis of SNHL. Serum antiganglioside antibodies were measured in three previous patients. Anti-GM1 ganglioside antibodies were not detected in any of those patients. Our patient had Anti-GM3 IgM antibody seropositivity.

Interestingly in patients with autoimmune ear disease including SNHL and Meniere's disease, serum antibodies to sialyl-I ganglioside, or sulfo-glucuronosyl glycolipids were found.[10] 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. Further, immunological studies are, therefore, required to elucidate whether distinct anti-glycolipid antibodies exist in GBS patients with SNHL. MRI with contrast is able to distinguish the underlying pathology of inner ear lesions; thus, it is commonly performed in patients with SNHL. Further, insight into labyrinthine pathology has been provided by recent technological advancements of MRI, especially the use of gadolinium contrast, refinement of its resolution, and the application of special sequences. Combined 3D T2-weighted SPACE and gadolinium-enhanced 3D FLASH sequences have benefits for the depiction of the facial and cochlear nerve lesions.[9] These electrophysiological changes indicated that demyelination in the facial and acoustic nerves, rather than axonal degeneration, was responsible for the SNHL in this patient.

Table 1: Latency and velocity comparison of nerves

Nerve	Latency ms	Velocity m/s
Right median	5.3	34
Right ulnar	4.8	36
Right peroneal	7.0	25
Right tibial	7.1	27

CONCLUSION

GBS with treatment refractory residual SNHL is quite rare. We have, herein, highlighted the electrophysiological changes and treatment-resistant nature of hearing loss in a GBS patient with facial and cochlear nerve involvement. MRI can be very useful in detecting lesions involving 7th and 8th cranial nerve, but in our case, MRI was also normal. Thus, MRI negative cranial nerve palsy must be kept in mind in such scenario. Auditory dysfunction must be evaluated in patients of GBS particularly when there is simultaneous other cranial nerve involvement.

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