

A Case of Unilateral Recurrent Nerve Palsy with Chronic Inflammatory Demyelinating Polyradiculoneuropathy

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a disease in which peripheral sensory and motor nerves of the four limbs are impaired due to autoimmune mechanism-induced demyelinating changes through a 2-month or longer chronic course. The incidence of complication by cranial neuropathy has been reported to be 15%, but there have been very few reports on disorder of the vagus nerve and its branch, the recurrent nerve. We report a patient who developed left recurrent nerve palsy with CIDP. The patient was a 48-year-old male. The disease developed as progressive muscle weakness and numbness of the four limbs 3 years before and was diagnosed as CIDP. The symptoms had been improved by high-dose intravenous gamma-globulin therapy. However, from 2 months before he became aware of breathy hoarseness, and bilateral decreased grip strength and sensory disturbance of the upper and lower limbs recurred and progressed. On laryngoscopy disorder of left vocal fold movement and glottal closure incompetence during phonation were observed, and neurogenic changes were detected in the left thyroarytenoid muscle by needle electromyography for the intrinsic laryngeal muscles. High-dose intravenous gamma-globulin therapy was performed and left vocal fold movement recovered with recovery of bilateral grip strength and sensory disturbance of the upper and lower limbs, and phonation was also normalized. (*J Nippon Med Sch* 2022; 89: 562–567)

Key words: chronic inflammatory demyelinating polyradiculoneuropathy, recurrent nerve palsy, hoarseness, electromyography

Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an autoimmune demyelinating neuropathy in which left-right symmetric muscle weakness and sensory disturbance of the four limbs develop as the main symptoms through slowly progressing or repeating recurrence and remission over a 2-month or longer course^{1,2}. Abnormal autoimmune reaction occurs targeting the myelin sheath of peripheral nerves. The presence of autoantibodies against the glycolipid component of the myelin sheath binding protein of Ranvier's node has

been noted in some cases, but an autoantibody specific to this disease has not yet been discovered^{1,2}. The frequency of cranial nerve disorder has been reported to be 15% and there are many reports of facial nerve and oculomotor nerve disorders^{3–6}. Cases of trigeminal nerve and hypoglossal nerve disorders have been reported^{3–6}, but recurrent nerve disorder has been rarely reported and only bilateral disorder cases have been reported^{7,8}. We report a patient complicated by unilateral recurrent nerve palsy (left side) in recurrent CIDP.

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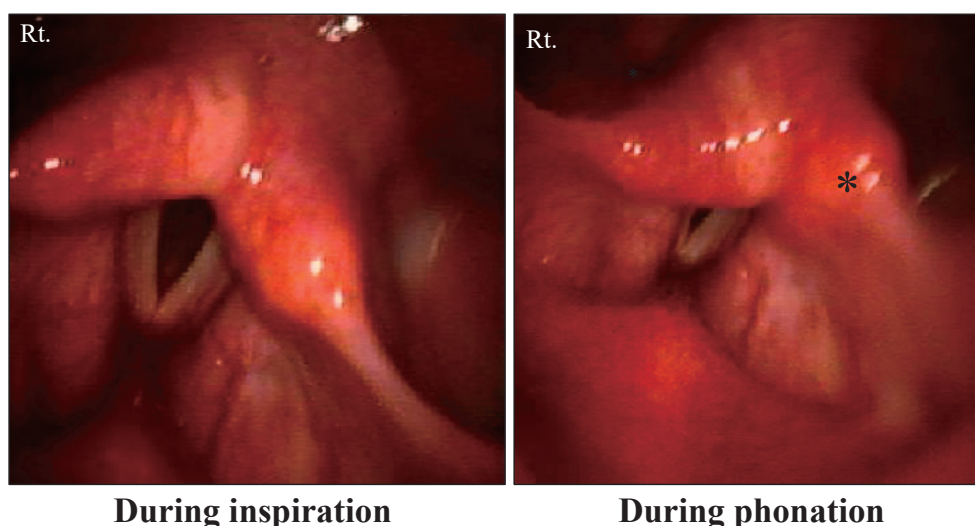


Fig. 1 Laryngoscopic findings before high dose intravenous gamma-globulin therapy. Laryngoscopic examination revealed vocal fold palsy with fasciculation-like movement on the left side (*). Glottal closure incompetence was seen during phonation.

Case Report

Patient: A 48-year-old male

Chief complaint: Breathly hoarseness

Familial medical history: None in particular

Past medical history: Hypertension

History of present illness: The patient became aware of numbness of the bilateral dorsum of the hand and fingertip 3 years before. Since the symptoms gradually aggravated and he recognized decreased grip strength of the bilateral hands 7 months later, he visited the neurology department of Nippon Medical School Main Hospital. On the first examination, bilateral biceps reflex, triceps reflex, brachioradialis reflex, patellar reflex, and Achilles tendon reflex were lost and grip strength was markedly reduced (right: 15 kg, left: 1.5 kg; normal grip strength for 45-49 year-old Japanese male was 45.29 kg⁹). Since delayed conduction velocity and conduction block of the bilateral median nerves and sural nerves were noted on peripheral nerve conduction study (NCS), neurogenic changes were pointed out by needle electromyography of the upper and lower limb muscles, whereas no abnormality was detected by head and spinal MRI, the patient was diagnosed with CIDP. High-dose intravenous gamma-globulin therapy (human immunoglobulin 27.5 g/day × 5 days) was performed. Upper limb deep tendon reflex recovered and bilateral grip strength improved to 25 and 17 kg on the right and left sides, respectively. However, the lower limb deep tendon reflex remained lost. After the first therapy, the patient had not gone to hospital at regular interval. Thus, we could not take any

administration to maintain his condition induced by CIDP after the first therapy. The course was observed thereafter, but the patient became aware of voice weakness 1 year before and began to suffer from breathy hoarseness 2 months before. And the patient recognized muscle weakness of the four limbs again around the same time. This was diagnosed as recurrence of CIDP and the patient was re-admitted to the hospital. After admission, the patient was referred to the otorhinolaryngology department for close investigation of the cause of hoarseness.

Neurological findings on re-admission: Bilateral biceps/triceps reflexes, brachioradialis reflex, patellar reflex, and Achilles tendon reflex were lost. Superficial sensation and thermal nociception were normal, but the sense of vibration was reduced in the bilateral lower limbs. Grip strength was reduced (right: 21 kg, left: 6 kg). In addition, left-dominant crural muscular atrophy was noted in the bilateral upper limbs and the patient could not stand on tiptoes.

Physical findings: Breath sound and intestinal peristaltic sound were favorable and no abnormality was observed on abdominal palpation or percussion.

Otorhinolaryngological findings: On laryngoscopy (Fig. 1), the left vocal fold stopped at a paramedian position and fasciculation-like minute movement was noted in the left arytenoid region during inspiration. Glottal closure incompetence was noted during phonation, showing breathy hoarseness.

Laryngeal tomography (Fig. 2): During phonation, the

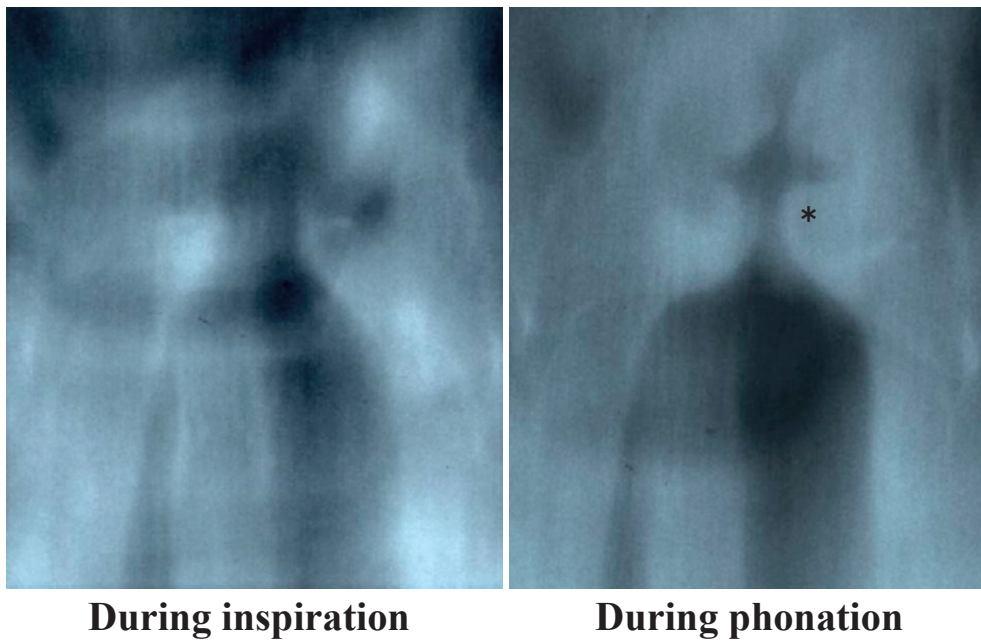


Fig. 2 Tomography of the larynx before treatment. During phonation, the position of the vocal fold on the left side (*) was higher than that on the right side with glottal closure. Atrophic change and enlarged ventricle were not seen on the left vocal fold.

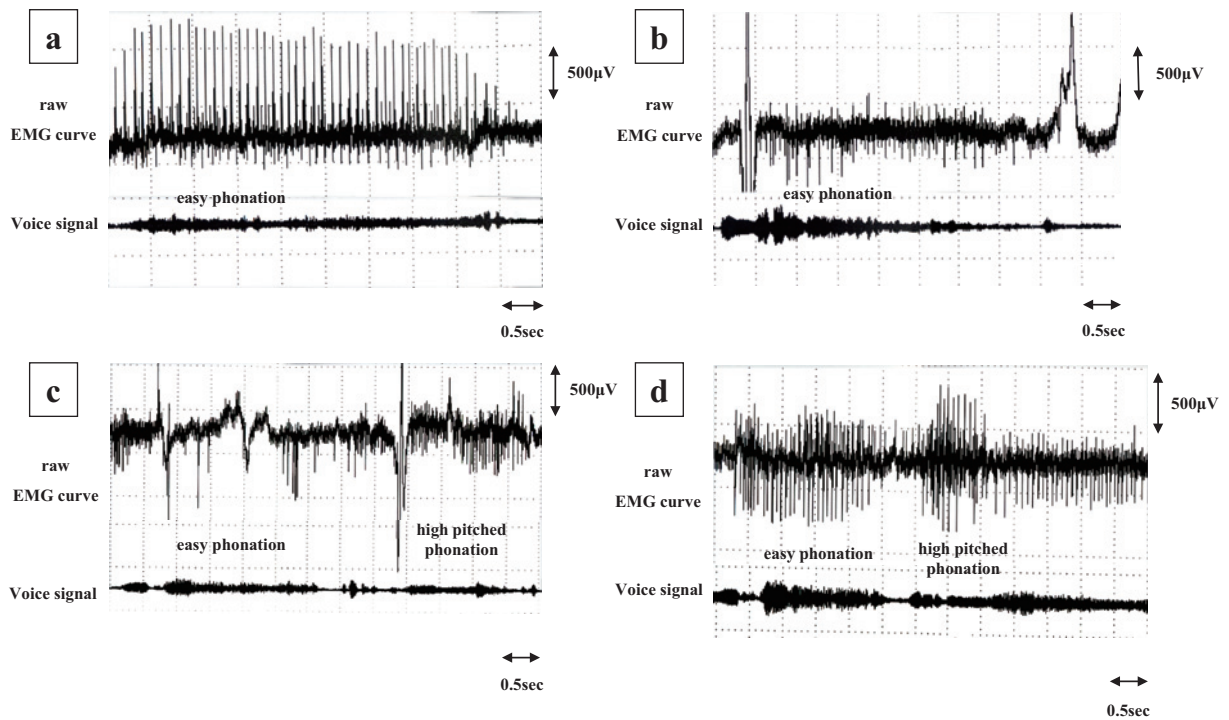


Fig. 3 Raw electromyographic curves for the intralaryngeal muscles. Electromyographic examinations revealed a neurogenic pattern with high amplitude and decreased NMU for the thyroarytenoid muscle on the left side during phonation (a). However, the thyroarytenoid muscle on the right side (b) and the cricothyroid muscles on the both sides (c, on the left side; d, on the right side) did not show any abnormal pattern.

level of the left vocal fold was higher than the level of the right vocal fold, showing glottal closure incompe-

tence, but no atrophy of the left vocal fold or enlargement of laryngeal ventricle was pointed out.

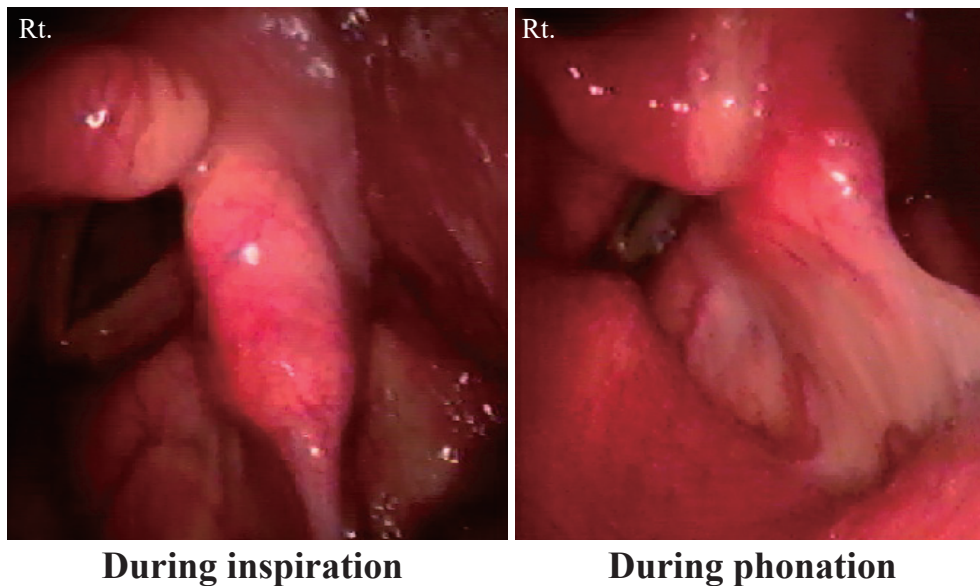


Fig. 4 Laryngoscopic findings after high dose intravenous gamma-globulin therapy. After intravenous immunoglobulin therapy, vocal fold palsy on the left side was improved immediately.

Electromyography of the intrinsic laryngeal muscles

(Fig. 3): The bilateral thyroarytenoid muscles and cricothyroid muscle were approached from the anterior neck skin through the cricothyroid space¹⁰. Regarding interference waves of the left thyroarytenoid muscle, the neuromuscular unit (NMU) decreased during phonation, the amplitude was high, and the duration prolonged (Fig. 3a). Electromyographic interference waves of the left cricothyroid muscle, right thyroarytenoid muscle, and right cricoarytenoid muscle were normal (Fig. 3b~d).

Videofluoroscopy of swallowing: Swallowing movement was normal without aspiration nor was there abnormality in esophageal peristalsis.

Enhanced neck and chest CT: No particular abnormal finding was noted.

Based on the above findings, it was suggested that left vocal fold movement was impaired due to left recurrent nerve palsy and induced breathy hoarseness. Since no abnormality was observed on neck and chest CT, CIDP-induced disorder of the left recurrent nerve was suspected. High-dose intravenous gamma-globulin therapy was then performed to treat recurrence of CIDP at the neurology department, which improved bilateral grip strength (right: 21→24 kg, left: 6→16 kg) after 2 weeks and breathy hoarseness also improved. Thus, laryngoscopy was performed and improvement of left vocal fold movement and resolution of glottal closure incompetence were confirmed (Fig. 4). Based on these, the condition

was diagnosed as CIDP-induced left recurrent nerve palsy. No recurrence has been noted for 5 years thereafter.

Discussion

Regarding CIDP-induced recurrent nerve palsy, only bilateral disorder of vocal folds movement has been reported and no unilateral disorder, such as that observed in this patient, has previously been reported^{7,8}. In addition, in previous reported cases, recurrent nerve palsy was diagnosed based on only laryngoscopic findings of vocal fold movement disorder. This is the initial report in which recurrent nerve neuropathy was clarified by needle electromyography of the intrinsic laryngeal muscle. The intrinsic laryngeal muscles involved in vocal fold movement include the cricothyroid muscle innervated by the external branch of the superior laryngeal nerve and the thyroarytenoid muscle, lateral cricoarytenoid muscle, interarytenoid muscle, and posterior cricoarytenoid muscle innervated by the recurrent nerve. Both the external branch of the superior laryngeal nerve and recurrent nerve branch from the vagus nerve, but the superior laryngeal nerve branches from the inferior ganglion of the vagus nerve and divides to the internal and external branches at the hyoid bone level. The internal branch consists of sensory fibers to the hypopharynx and superior glottic region, and the external branch consists of motor fibers of the cricothyroid muscle. The recurrent nerve branches in the superior mediastinum, and the left

side goes round below the aortic arch and right subclavian artery, ascends the tracheoesophageal groove, and innervates the intrinsic laryngeal muscles. On needle electromyography of the intrinsic laryngeal muscles, both thyroarytenoid muscle and cricothyroid muscle show muscle activity during phonation, but cricothyroid muscle activity is increased during the phonation of higher pitched sound than natural pitch sound, based on which the two muscles could be identified, and the neuropathic region could be presumed by needle electromyography of the two muscles¹⁰. In this patient, neurogenic changes were observed in the left thyroarytenoid muscle on needle electromyography, normal interference waveforms were seen in the left cricothyroid muscle, and electromyographic interference waves of the right thyroarytenoid muscle and cricothyroid muscle were normal, suggesting that disorder occurred at more peripheral level to branching of the external superior laryngeal nerve from the left vagus nerve. In addition, decreased peristaltic movement of the esophagus suggesting disorder of the vagus nerve trunk was not observed by video-fluoroscopic findings for swallowing and no finding suggesting gastrointestinal motility disorder was noted on physical examination, so that disorder may have occurred after branching of the recurrent nerve from the vagus nerve. It is considered that unlike uniform disorder of the peripheral nerve root over the distal end in diabetic neuropathy, Charcot-Marie-Tooth disease, and amyloidosis causing peripheral nerve disorder, in CIDP-induced disorder, patchy multifocal lesions are formed during peripheral nerve distribution and residual myelinated nerve fibers in the same nerve bundle show heterogeneous distribution, being characteristics¹¹. Therefore, in the present patient, CIDP-induced formation of demyelinated lesions may have occurred in the left recurrent nerve, which is a vagus nerve branch, and impaired left vocal fold movement. Since no denervation potential, such as fibrillation voltage, was detected on thyroarytenoid muscle electromyography and left vocal fold movement was recovered by high-dose intravenous gamma-globulin therapy in this patient, the recurrent nerve disorder might not have reached an irreversible level, being incomplete paralysis. The characteristics of complete unilateral recurrent nerve paralysis are atrophy of the vocal fold and enlargement of laryngeal ventricle on the paralyzed side, and visualization of the vocal fold on the paralyzed side at a level higher than the level of the healthy side during phonation on laryngeal tomography¹². In this patient, the position of the left vocal fold

was higher than the right vocal fold during phonation on laryngeal tomography, but no vocal fold atrophy or enlargement of the laryngeal ventricle was noted, so that the laryngeal tomography findings also suggest that the left recurrent nerve disorder was not irreversible and it was in a state of incomplete paralysis.

Cruccu et al.¹³ and Kokubun et al.¹⁴ performed electrophysiological examination of the trigeminal nerve and facial nerve in CIDP patients and observed that subclinical-level nerve disorder developed at a high rate. Similarly, in recurrent nerve palsy, even though symptoms suggesting recurrent nerve palsy, such as breathy hoarseness and swallowing disorder, are absent, it is possible that patients with subclinical disorder could be seen on laryngoscopy and electromyography of the intrinsic laryngeal muscle. Only left recurrent nerve disorder was observed in this patient, but previously reported cases were bilateral, so that the bilateral recurrent nerves might be impaired as CIDP repeats recurrence and remission in the future. If bilateral recurrent nerve disorder occurs, glottic stenosis develops due to bilateral vocal folds movement disorder and may induce dyspnea, inspiratory stridor, and swallowing disorder, to which attention should be paid. When CIDP-induced peripheral nerve disorder progressed to an irreversible level, and treatment, such as thyroplasty, to improve the phonation function of unilateral recurrent nerve palsy-induced phonation disorder is performed, recurrent nerve disorder may occur on the opposite side due to CIDP in the future, for which confirmation of the presence or absence of nerve disorder in the intrinsic laryngeal muscle, considered the healthy side, by needle electromyography might be important.

Although CIDP is still a disease of unknown cause and advancement of immunological and neurological studies is awaited, investigation of the development of CIDP-induced recurrent nerve disorder by laryngoscopy and electromyography of the intrinsic laryngeal muscle may also be warranted.

Conflict of Interest: The authors declare no conflict of interest.

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