Lewis-Sumner Syndrome Manifesting as Unilateral Vocal Fold Paresis and Laryngeal Fasciculations

*,†Jérôme R. Lechien and *,‡Camille Finck, *†Mons and ‡Liège, Belgium

Summary: Lewis-Sumner syndrome (LSS) is a rare disease characterized by asymmetrical and multifocal mononeuropathy commonly located in the upper limbs. Some rare cases affecting cranial nerve have been described, but LSS is unknown to affect especially laryngeal nerves. This paper presents the first case of unilateral vocal fold paresis caused by an LSS in a 59-year-old man complaining of dysphonia, breathy voice, and vocal fatigue. Epidemiology, clinical features, diagnosis, and treatment will be described.

Key Words: Lewis-Sumner–Laryngeal–Paresis–Dysphonia–Neuropathy.

INTRODUCTION

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is the most prevalent subgroup of chronic neuropathies affecting 1–9 cases per 100 000 population per year.[1] Many phenotypic variants of CIDP exist and begin with peripheral motor and sensory nerve dysfunction. Among the different variants, Lewis-Sumner syndrome (LSS) or multifocal acquired demyelinating sensory and motor neuropathy affects 6%–15% of CIDP patients.[2] This rare affection is characterized by asymmetrical and multifocal mononeuropathy usually located in the upper limbs.[1] In this paper, we report a unique case of a neuropathy of the recurrent laryngeal nerve (RLN) caused by LSS. To our knowledge, this report is the first case describing an isolated unilateral vocal fold paresis related to LSS.

CASE REPORT

A 59-year-old man was referred to the Department of Laryngology and Voice Rehabilitation for a few months history of dysphonia, breathy voice, and vocal fatigue. Medical history included a transient ischemic attack of the right thalamus 8 months earlier and a hospitalization for a history of progressive multifocal sensory deficits affecting the periphery of the mouth and the left upper and lower limbs. The patient had not undergone any previous surgery or endotracheal tube intubation. The perceptual voice quality assessment showed a bitonality in the patient's voice (diplophonia). The neurologic physical investigation revealed a left-side hypoesthesia of the entire hemicorpus, a disturbed sensory perception of changes in thermoalgesic stimuli of all four extremities and the left hemiface, and an areflexia of the four limbs. Videolaryngostroboscopy examination showed a left vocal fold paresis with reduced abduction, a slight anteriorization of the left arytenoid cartilage, and fasciculations in the left aryepiglottic fold posteriorly corresponding to the natural position of the lateral cricoarytenoid muscle (Supplementary Video S1 [at 13 seconds and 31 seconds]). Laryngoscopic assessment of the palatal and pharyngeal function was normal. The neurologic examination also excluded other cranial nerve involvement. Additional voice quality evaluations, described in Table 1, reported subjective and objective (aerodynamic and acoustic) alterations. No compressive pathology was detected along the recurrent laryngeal branches or the vagus nerves in computed tomography and magnetic resonance imaging. Holter monitoring and cardiac ultrasonography did not reveal any significant disorder. Electrophysiological motor conduction studies revealed multiple nerve conduction blocks also present in the left upper and lower limbs. The patient refused the laryngeal electrophysiological study. Blood immunologic studies reported antiganglioside antibodies (anti-GM1) and cryoglobulinemia (IgM type). The clinical course, blood tests, and the electrophysiological studies led to the diagnosis of a unilateral left vocal fold paresis caused by a multifocal acquired demyelinating sensory and motor neuropathy, also called LSS. The follow-up of the patient was unremarkable and the voice quality did not improve 1 year after the first consultation. A medialization of the vocal cord was proposed but not accepted by the patient.

DISCUSSION

In this paper, we describe a case of LSS presenting as a unilateral hypomobility of left vocal fold secondary to a left RLN paresis. Since the first report of LSS,[3] approximately 100 cases have been described worldwide, with only 20% of the patients having cranial nerve involvement.[1,4] The most common cranial nerves affected by this disease are II, V, VII, IX, X, and XII.[4] Precisely, two cases of X palsy related to LSS were described, but neither case had isolated RLN palsy.[5,6] Regarding the high rate (30%–50%) of asymptomatic lesions of the RLN,[7] we believe that the occurrence of RLN palsy, in the context of LSS, is probably underestimated.

In both cases of vagus nerve or RLN damage, the palsy can be attributed to the mechanism by which the LSS originates, a chronic destruction of myelin sheaths by antiganglioside antibodies leading to neural edema, loss of myelin, or axonal disruption.[1] Overall, LSS diagnosis is based on some neurologic symptoms (ie, asymmetric hand and arm weakness, pain, and paresthesias in the hands and fingers) and signs (ie, areflexia, thermoalgesic perception disorders, and hypoesthesia), found on complete neurologic examination.[1,4] Our patient presented classical neurologic complaints but was mostly disturbed by the voice

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From the *Laboratory of Phonetics, Faculty of Psychology, Research Institute for Language Sciences and Technology, University of Mons (UMONS), Mons, Belgium; †Laboratory of Anatomy and Cell Biology, Faculty of Medicine, Research Institute for Health Sciences and Technology, University of Mons (UMONS), Mons, Belgium; and the ‡Department of Otolaryngology and Head and Neck Surgery, CHU de Liège, Liège, Belgium.
Address correspondence and reprint requests to Jérôme R. Lechien, Department of Otolaryngology and Head and Neck Surgery, CHU de Liège, Domaine du Sart Tilman, 1, B4000 Liège, Belgium. E-mail: Jerome.lechien@umons.ac.be
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TABLE 1.
Voice Assessment of the Patient

Voice Quality Assessment	Units	Results
Subjective voice assessment		
VHI	—	35
VHIf	—	8
VHIe	—	8
VHIp	—	19
Aerodynamic		
Maximum phonation time	s	7.33
Phonatory quotient	mL/s	409.28
Mean sound pressure level	dB	83.45
Maximum sound pressure level	dB	92.30
PSGE lowest	cmH_20	6.94
PSGE conversational	cmH_20	18.74
PSGE highest	cmH_20	32.44
Lowest intensity	dB	58
Highest intensity	dB	86
Acoustic		
F0 lowest	Hz	73.42
F0 highest	Hz	293.66
Mean F0	Hz	125.96
Range F0	Hz	220.24
Shimmer percent	—	0.218
Jitter percent	—	0.951
DSI	—	−1.293

Abbreviations: B, decibels; DSI, dysphonia severity index; F0, fundamental frequency; Hz, Hertz; mL/s, milliliters per second; s, second; PSGE, estimated subglottic pressure; VHIf/e/p: functional/emotional/physic voice handicap index.

impairment that led to the consultation in the ear, nose, and throat department. The dysphonia, breathiness, and the findings found on videolaryngostroboscopy, including incomplete glottic closure, were related to the adductory weakness of the left vocal cord. Hence, early head and neck examination and videolaryngostroboscopic evaluation by an otolaryngologist remain important to rule out other causes of RLN palsy. In addition, evaluation of the palatal and pharyngeal function is also important to assess possible neurologic involvement of the other cephalad branches of the vagus nerve. Electrophysiological study of other peripheral nerves is the most reliable approach to differentiate LSS from other neuropathies.[1] Electromyography study in LSS usually shows multiple, asymmetric conduction blocks commonly located in the upper and lower limbs.[1] All of these findings were identified in our patient. Lastly, immunologic studies may support the diagnosis with the presence of antiganglioside antibodies (anti-GM1), whereas cerebrospinal fluid can contain elevated proteins.[8]

It is possible that the hypomobility of the left vocal fold could be secondary to a left cricoarytenoid joint ankylosis. We did not test for this by trying to passively move the arytenoid cartilage either under local or general anesthesia. However, the lack of a history of intubation trauma with postoperative dysphonia, or a viral upper respiratory infection preceding the patient's present and continual dysphonia, speaks against a primary ankylosis. This coupled with clinical observation of fasciculations of the left lateral cricoarytenoid muscle region that supports a primary neurologic disease as the underlying cause of this man's vocal fold hypomobility.

In the present case, and after the completion of the videolaryngostroboscopic examination, computed tomography and magnetic resonance imaging of the head, neck, and chest were used to exclude other possible causes of RLN palsy. Common diseases include cancers of the head and neck, thyroid and parathyroid glands, and lung, and any postsurgical iatrogenic trauma related to thoracic or cervical surgery. Cancers and iatrogenic trauma account for 33.4% and 31.5%, respectively, of vocal fold paresis.[9] In 10%–27% of cases, unilateral vocal folds palsy remains idiopathic.[9] Less frequent etiologies include inflammatory diseases and central nervous system disorders (ie, stroke) corresponding to 13.2% and 6.4% of cases, respectively.[9] Little epidemiologic data exist on other neurologic conditions that may negatively impact the integrity of the RLN. These etiologies include diabetic neuropathy, multiple sclerosis, Guillain-Barré syndrome, vagal nerve stimulation for seizure control, lateral amyotrophic sclerosis, and idiopathic neuritis.[7,10] Other anecdotal causes have been described such as Miller-Fischer syndrome and vitamin B12 deficiency.[11]

Current LSS treatment includes intravenous immunoglobulin and, in some cases, rituximab or infliximab.[8,12] To date, we have no data suggesting a potential neurologic recovery of the laryngeal palsy with the conventional treatment. Therefore, in the cases with disabling hoarseness or aspiration, a medialization of the affected vocal fold may be discussed with the patient earlier in the course of their disease.

CONCLUSION

Although some involvement of cranial nerves have been described in LSS,[3] vocal fold paresis and fasciculations caused by laryngeal recurrent nerve involvement are uncommon manifestations of this disease. Thus, to our knowledge, we report the first case of LSS with isolated RLN involvement. Recurrent laryngeal neuropathy should be suspected in all LSS patients presenting with a new onset of dysphonia. Videolaryngostroboscopic examination and neurophysiological study may clarify the cause of the dysphonia and, according to the complaints of the patient and the potential repercussions on quality of life, an adequate treatment may be proposed early on to rehabilitate the larynx.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found online at doi:10.1016/j.jvoice.2017.05.020.

REFERENCES

1. Mathey EK, Park SB, Hughes RA, et al. Chronic inflammatory demyelinating polyradiculoneuropathy: from pathology to phenotype. *J Neurol Neurosurg Psychiatry*. 2015;86:973–985.
2. Busby M, Donaghy M. Chronic dysimmune neuropathy. A subclassification based upon the clinical features of 102 patients. *J Neurol*. 2003;250:714–724.
3. Lewis RA, Sumner AJ, Brown MJ, et al. Multifocal demyelinating neuropathy with persistent conduction block. *Neurology*. 1982;32:958–964.
4. Verschueren A, Azulay JP, Attarian S, et al. Lewis-Sumner syndrome and multifocal motor neuropathy. *Muscle Nerve*. 2005;31:88–94.