Refractory Dysphonia Due to Isolated Cricothyroid Muscle Dystonia

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Summary: Objective. To demonstrate the utility of electromyography (EMG) in the evaluation and management of treatment-resistant dysphonia.

Method. We report a case of refractory dysphonia in which EMG was used to identify and treat isolated cricothyroid (CT) dystonia.

Results. The patient, a healthy 43-year-old woman, presented with 9 months of progressive hoarseness. Her symptoms were present across vocal tasks but were particularly bothersome while dictating. On presentation, her voice was rated grade 3, roughness 3, breathiness 1, asthenia 0, and strain 3 (G3R3B1A0S3). Videostroboscopy was remarkable for hyperfunction. Voice therapy was not beneficial despite appropriate effort. Microdirect laryngoscopy revealed no evidence of structural pathology. The patient was referred for EMG because of her normal examination and failure to improve with therapy. The CT muscle demonstrated an increased latency of 750 ms in all vocal tasks. One month after CT injection with 3 units of botulinum toxin (BTX), her voice was improved. Perceptual voice evaluation was rated G1R1B0A0S1. Voice Handicap Index improved from 87 to 35.

Conclusions. In the absence of structural pathology, EMG can be a useful adjunct in the diagnosis of dysphonia that persists despite adequate trials of voice therapy. To our knowledge, this is the only report of laryngeal dystonia due to isolated CT dysfunction successfully treated with BTX.

Key Words: Laryngeal dystonia–Spasmodic dysphonia–Cricothyroid muscle–Laryngeal electromyography.

INTRODUCTION

Laryngeal dystonia, or spasmodic dysphonia (SD), is included among the movement disorders classified as idiopathic focal or regional dystonias. These disorders are characterized by sustained or repetitive involuntary muscle contractions of a single or related group of muscles.[1] SD patients are evaluated and divided into clinical subtypes on the basis of perceptual voice assessment and examination findings. Initial treatment with botulinum toxin (BTX) is directed toward the predominate muscle responsible for the clinical subtype.[2] Adductor spasmodic dysphonia (ADSD) is treated by thyroarytenoid (TA) injection, and abductor spasmodic dysphonia (ABSD) with posterior cricoarytenoid (PCA) injection.

Laryngeal electromyography (EMG) studies suggest that SD is actually a disorder of mixed abductor and adductor dysfunction (MXD SD) with a predominant clinical phenotype.[3] Fine-wire EMG studies of the intrinsic laryngeal muscles have demonstrated that the lateral cricoarytenoid (LCA) and interarytenoid (IA) muscles can contribute to SD.[4] When EMG was used to evaluate patients in whom TA injection had failed, injecting an overactive IA in addition to the traditional TA injection resulted in improved symptom control.[5] The authors concluded that EMG could be used to generate a "road map" by which to design targeted therapy in patients with refractory SD. More recently, authors have suggested that EMG can be a useful adjunct in differentiating SD from muscle tension dysphonia in challenging patients who exhibit clinical feature of both disorders.[6]

We present a case of recalcitrant dysphonia in a healthy woman. Diagnostic fine-wire EMG provided critical information in identifying what we believe to be the first reported case of isolated cricothyroid (CT) dystonia, allowing us to design a successful regimen of CT muscle injection with BTX.

REPORT OF A CASE

The patient, a healthy 43-year-old woman, was referred for a second opinion regarding her 9-month history of gradually progressive dysphonia. She associated the onset of her voice problem with an upper respiratory tract infection that resolved spontaneously after 5–7 days. Subsequently, she began to experience intermittent periods of dysphonia and effortful voicing. Eventually, she had an episode from which her voice never recovered.

At the time of our evaluation, the patient had not experienced normal voicing in 4 months. She complained of fluctuating vocal quality that would "choke off," elevated pitch that made her "sound like Minnie Mouse," and difficulty talking on the phone. She had been previously diagnosed with vocal fold nodules, but a period of voice rest and voice therapy did not provide a substantial improvement in either vocal quality or voicing effort.

Voice Handicap Index (VHI) at her initial evaluation was 87. The patient reported high vocal demands related to her work as an urgent care physician. Specifically, she relied on her voice for interaction with patients and families, consulting with medical colleagues in person and by phone, teaching, and dictating. She was using a voice amplifier while speaking in a whisper

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voice in effort to maintain voicing throughout her shifts. She was concerned that voicing in this manner interfered with her ability to present herself professionally and to communicate effectively during critical patient care situations.

Perceptual voice evaluation was performed by the senior author. In addition to conversational voice, the patient provided samples of sustained vowels, the Rainbow passage, and the CAPE-V sentences. Her voice was rated grade 3, roughness 3, breathiness 1, asthenia 0, and strain 3 (G3R3B1A0S3). The pitch was noted to be markedly elevated for gender, with a limited range. No tremor or voice breaks were noted. Her videostroboscopic examination was remarkable for mild laryngeal hyperfunction in the lateral dimension. Mucosal wave amplitudes were globally reduced bilaterally. The wave was rarely periodic. No mucosal or submucosal masses or lesions were noted. Acoustic evaluation demonstrated a maximum phonation time (MPT) of 11 seconds. Fundamental frequency (F_0) was 287 Hz, with a minimum pitch (F_{min}) of 151 Hz and a maximum pitch (F_{max}) of 650 Hz (range 499 Hz). The unusual nature of her vocal quality, coupled with the lack of mucosal findings, was suggestive of a functional dysphonia, and the patient was referred for another trial of voice therapy.

During voice therapy, multiple strategies were used to unload baseline muscle tension and evaluate the patient's ability to improve her voice through modifications in vocal technique. With repeated trials and feedback, she was able to achieve a clear but breathy voice in a more appropriate pitch during sustained phonation and in repeated syllables using a chanting voice. The patient reported that this took significant effort to maintain and repeatedly noted, "this is better, but not my normal voice." During connected speech, pitch breaks and diplophonia again became evident.

Despite the patient's considerable efforts, improved vocal quality was difficult to maintain beyond the syllable level, and we questioned the presence of a subtle structural lesion. When a short course of steroids and strict voice rest provided no appreciable benefit, the patient elected to proceed to surgical evaluation. Operative suspension microlaryngoscopy with subepithelial infusion did not reveal evidence of sulcus, mucosal bridge, or other subtle submucosal pathology. Interestingly, the patient's abnormal vocal quality was noted to persist in the recovery room as the patient emerged from anesthesia.

Because of failed response to voice therapy in the setting of a normal laryngeal examination, we recommended evaluation with EMG. Fine-wire electrodes were placed in the left TA, left CT, and the IA (Figure 1). An electrode was not successfully placed in the LCA. During simple phonation, the latencies for the TA and the IA were noted to be 300 ms. The latency for the CT was noted to be prolonged at 750 ms (Figure 2). Increased CT latency and activity persisted throughout all voicing tasks, to include conversational speech (Figure 3).

Because of the unusual dystonic activity of the CT by EMG, the patient was offered a trial of BTX injection. Approximately 48 hours after injecting 3 units in each CT, the patient reported a pronounced improvement in her voice, with decreased vocal effort and improved (lower) pitch. Perceptual evaluation of her voice at follow-up was rated G1R1B1A0S1. Acoustic analysis revealed an MPT of 10 seconds. F_0 was decreased to 212 Hz (F_{min} = 145 and F_{max} = 518). Her VHI markedly improved to 35. She returned to full professional

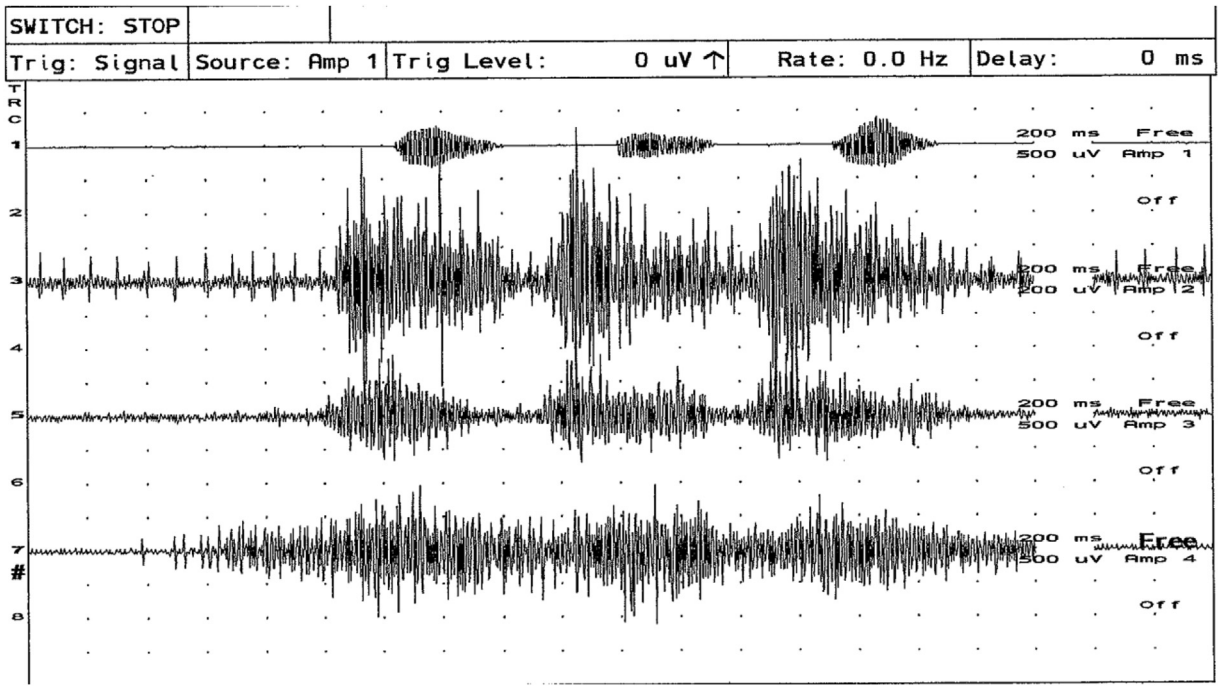

FIGURE 1. Fine-wire EMG. Channel 1 (line 1) = voice, channel 3 (line 2) = TA, channel 5 (line 3) = LCA, and channel 7 (line 4) = CT. Patient was instructed to glide from low pitch to high pitch using a sustained "eee." CT recruitment at higher pitches confirms wire placement in the CT.