Pharyngeal Dystonia Mimicking Spasmodic Dysphonia

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Summary: Objective. The aim of this study was to describe the presentation of pharyngeal dystonia (PD), which can occur as a focal or segmental dystonia with a primarily pharyngeal involvement for the discussion of treatment methods for controlling consequent symptoms. PD is specific to speech-related tasks.

Methods. A retrospective medical record review of four patients with PD was performed.

Results. All patients were initially misdiagnosed with adductor spasmodic dysphonia and failed standard treatment with botulinum toxin type A (BTX). On laryngoscopy, the patients were discovered to have segmental or focal dystonia primarily affecting the pharyngeal musculature contributing to their vocal manifestations. A novel treatment regimen was designed, which involved directing BTX injections into the muscles involved in spasmodic valving at the oropharyngeal level. After titrating to an optimal dose, all patients showed improvement in their voice and speech with only mild dysphagia. These patients have maintained favorable results with repeat injections at 6- to 12-week intervals.

Conclusions. PD, or dystonia with predominant pharyngeal involvement, is a rare entity with vocal manifestations that are not well described. It can be easily mistaken for spasmodic dysphonia. PD is specific to speech-related tasks. A novel method of BTX injections into the involved muscles results in a significant improvement in voice without significant dysphagia.

Key Words: Dystonia–Dysphonia–Segmental dystonia–Speech-language disorder–Spasmodic dysphonia.

INTRODUCTION

Adult-onset focal and segmental dystonias affect a limited region of the body and can be task specific.[1,2] Spasmodic dysphonia (SD) is a focal dystonia characterized by laryngeal muscle spasms during phonation that is commonly the source of voice breaks.[3] Adductor SD is the most common type of SD and is caused by overactivity of the thyroarytenoid muscles. Consequent hyperadduction of the vocal folds results in a strained-strangled voice quality that tends to occur with vowel-laden phrases. On the other hand, abductor SD is much less common and is caused by an overactivity of the posterior cricoarytenoid muscles, resulting in an excessive opening of the vocal cords and breathy voice breaks. Mixed SD can sometimes occur when laryngeal adductor and abductor muscles are simultaneously involved.[4]

In this paper, we describe a rare, poorly described form of dystonia that most significantly and predominately affects the pharyngeal musculature, known as pharyngeal dystonia (PD). This distinct entity can exist as either an isolated focal or segmental dystonia and could easily be mistaken for more common dystonias of this anatomic region. In this report, we describe four patients who presented with prominent voice breaks mimicking SD because of effortful speech and a constricted or strained-strangled voice. However, flexible laryngoscopy readily revealed spasms primarily of the pharyngeal rather than the laryngeal muscles leading to their symptoms. These patients exhibit oropharyngeal spasms during connected speech resulting in a distinctive-sounding dysphonia characterized by dystonic breaks principally with vowels and nasals that are best characterized by the voice of the cartoon character "Donald Duck." All patients were evaluated by a board-certified neurologist to rule out systematic neurologic disease as a potential etiology. Finally, an effective treatment regimen is also described in our report, which involves botulinum toxin type A (BTX) injections directly into the muscles involved in spasmodic valving at the oropharyngeal level. This method allows for chemodenervation of the affected muscles with only mild dysphagic side effects.

METHODS

A retrospective medical record review of four patients with PD was performed over an 11-year period. Clinical notes and laryngeal videostroboscopies were reviewed. Patients were evaluated based only on available data in the patient files, which included the Voice Handicap Index-10 (VHI-10) score, the Voice-Related Quality of Life (V-RQOL) score, and the Consensus

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Auditory Perceptual Evaluation of Voice (CAPE-V) score. The anchors of the V-RQOL are 0 (lowest quality of life) and 100 (highest quality of life), and the anchors of the CAPE-V are 0 (normal voice) and 100 (most impaired).

CASE REPORTS

Case 1

A 56-year-old man presented to our clinic in August 2001, 2 months after developing acute-onset dysphonia following an upper respiratory infection. Laryngeal symptoms were the presenting and sole complaint of the disorder. Symptoms were progressive and had no clear etiology. The physical examination revealed a strained-strangled speech quality, worse with vowels and nasals, but noted across all speech-related tasks. Flexible laryngoscopy revealed a hyperadduction of the supraglottis and vocal folds. Given these findings, a preliminary diagnosis of SD was rendered. After an initial course of voice therapy failed, the patient underwent electromyogram-guided injections of 1.25 units of Botox (BTX) into the thyroarytenoid-lateral cricoarytenoid (TA/LCA) complex. Following injection, the patient experienced an expected breathy voice. Unfortunately, there was limited improvement in the voice. Flexible laryngoscopy after BTX injections revealed a reduction in supraglottic spasms, vocal fold bowing from chemodenervation, and persistent spasms involving the oropharyngeal musculature with medial contraction of the pharyngeal walls and posterior displacement of the base of the tongue that was most prominent with vowels and nasals (Table 1).

TABLE 1.
Voice Findings, Voice Scores, and Laryngoscopy Findings at Baseline and After Treatment

	Baseline			Treatment		
Case	Voice Findings	Voice Scores	Laryngoscopy Findings	Optimal Treatment	Voice Outcomes	Side Effects
1	Strained-strangled speech, worse with vowels and nasals		Hyperadduction of the supraglottis and vocal folds; oropharyngeal spasms with pharyngeal wall contraction and posterior displacement of the tongue	15 units of BTX superficially to PEF bilaterally	Significant improvement.	Mild dysphagia
2	Pitch and voice breaks, reduced projection, increased vocal effort and strain, vocal fatigue	VHI-10|10	Oropharyngeal spasms with pharyngeal wall contraction and posterior displacement of the base of the tongue; mild glottic and supraglottic compression	20 units of BTX to PEF bilaterally	VHI-10|40	Dysphagia for solids managed with diet
3	Articulatory imprecision on lingual alveolar, lingual dental, palatal, and velar sound; hyponasal voice, irregular interruptions of sound	V-RQOL|23 CAPE-V|58 Global Voice Rating|3/7	Posterior retrusion of the tongue base, pharyngeal constriction of PEF	10 units of BTX to PEF and palatopharyngeus	V-RQOL|70 CAPE-V|35 Global Voice Rating|7/7	No dysphagia
4	Significant hyponasal quality, fluctuation in volume, and intermittent vocal disruptions	V-RQOL|12.5	Dystonic contractions of the pharyngeal, suprahyoid, and tongue musculature	15 units of BTX to PEF bilaterally	V-RQOL|37.5	No dysphagia

Abbreviations: BTX, botulinum toxin type A; CAPE-V, Consensus Auditory Perceptual Evaluation of Voice; PEF, pharyngoepiglottic fold; VHI-10, Voice Handicap Index-10; V-RQOL, Voice-Related Quality of Life.