Transfer Dysphagia Due to Focal Dystonia

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ABSTRACT

Objective The inability to propel a bolus of food successfully from the posterior part of the oral cavity to the oropharynx is defined as transfer dysphagia. The present case series describes the varied presentation of transfer dysphagia due to focal dystonia and highlights the importance of early detection by following up on strong suspicions.

Methods We describe seven cases of transfer dysphagia due to focal dystonia. Transfer dysphagia as a form of focal dystonia may appear as the sole presenting complaint or may present with other forms of focal dystonia.

Results Four out of seven patients had pure transfer dysphagia and had previously been treated for functional dysphagia. A high index of suspicion, barium swallow including videofluoroscopy, associated dystonia in other parts of the body and response to drug therapy with trihexyphenidyl/tetrabenazine helped to confirm the diagnosis.

Conclusion Awareness of these clinical presentations among neurologists and non-neurologists can facilitate an early diagnosis and prevent unnecessary investigations.

Key Words Transfer dysphagia; focal dystonia; eating dystonia; videofluoroscopy; barium swallow.

The process of swallowing involves the movement of the bolus of food inside the oral cavity and then its movement from the oral cavity to the oropharynx. This process requires intricate coordination of the various muscles of the pharynx and oral cavity. The inability to transfer a bolus of food successfully from the posterior part of the oral cavity to the oropharynx is defined as transfer dysphagia.1 Common etiologies of transfer dysphagia can be described as neuromuscular or structural (Table 1). One of the rare causes of transfer dysphagia is focal dystonia involving the oropharyngeal muscles.

MATERIALS & METHODS

This is a retrospective chart review. We discuss seven cases of transfer dysphagia without any local obstruction from patients who presented to the neurology department. All patients had significant weight loss due to dysphagia, and most of them responded to treatment.

The authors confirm that the approval of an institutional review board was not required for this work. During videography, the patient's anonymity was ensured.

RESULTS

Details of the individual cases are described in Table 2. Two representative cases are discussed below in detail.

Case 1
A 55-year-old gentleman with hypertension who was on regular treatment presented with a two-year history of difficulty in swallowing. He did not have any difficulty in biting, chewing or moving the food inside the oral cavity. He only had difficulty in the initial phase of swallowing, which he described as difficulty in moving the bolus down his throat (transfer dys-
Once the bolus moved into the throat, there was no choking, coughing, or nasal regurgitation. There was no associated weakness of muscles of mastication, facial muscle weakness or fatigability. The gag reflex was normal. There were no other involuntary movements in any other parts of the body and there was no history of voice change. There was no history of change in affect, sleep disturbances, etc. A clinical and endoscopic examination of the upper aerodigestive tract did not reveal any abnormalities. An electrophysiological study was performed to rule out any neurogenic changes, thus ruling out bulbar onset motor neuron disease.

In view of the absence of any other findings, the patient was being managed as having functional dysphagia from another center. He was prescribed antidepressants and multivitamins without much benefit and had lost approximately 6 kg of weight due to dysphagia. At our center, the possibility of transfer dysphagia due to focal dystonia was considered. A barium swallow was performed in which the patient was asked to swallow, without chewing, a soaked rasgulla, which is a soft, round Indian sweet made of cheese that is spongy and absorbent. The study showed an impaired voluntary phase of deglutition. The bolus managed to move through the pharynx only after repeated attempts at swallowing. The patient was treated with trihexyphenidyl (THP) (6 mg/day). He reported a 60–70% improvement in his symptoms over the next 3 months.

### Case 4

A 48-year-old gentleman presented with a history of progressive orolingual dystonia followed by intermittent adductor dysphonia and lingual dystonia followed by TD. The patient was managed as having transfer dysphagia due to focal dystonia. An electrophysiological study was performed to rule out any neurogenic changes, thus ruling out bulbar onset motor neuron disease.

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### Table 1. Common etiologies of transfer dysphagia

| Etiologies                                                                 | Neurological causes                                      | Structural (mechanical) causes                          |
|----------------------------------------------------------------------------|----------------------------------------------------------|--------------------------------------------------------|
| Stroke                                                                    | Cervical myelitis                                        | Carcinoma                                               |
| Parkinsonism                                                              | Amyotrophic lateral sclerosis                            | Prior surgery or radiation therapy                      |
| Focal dystonia                                                            | Post-poli syndrome                                       | Local injury/infection                                  |
| Multiple sclerosis                                                        | Lower cranial nerve palsy                               | Osteophytes and other spinal disorders                  |
| CNS tumors                                                                | Myasthenia                                               | Proximal esophageal web                                 |
|                                                                            | Polymyositis/dermatomyositis                             | Thyromegaly                                             |
|                                                                            | Muscular dystrophy/myotonic dystrophy                    | Zenker’s diverticulum                                   |

### Table 2. Summary of patients with TD

| Patient No. | Age/Sex | Symptom duration (months) | Course of illness | Weight loss (kg) | Type | UGIE | Barium swallow (including videofluoroscopy) | Management | Improvement (%) |
|-------------|---------|---------------------------|-------------------|------------------|------|------|--------------------------------------------|------------|----------------|
| 1           | 55/M    | 24                        | TD was the only symptom | 6                | Pure TD | N   | Impaired phase of deglutition | THP, 6 mg/day | 60–70          |
| 2           | 74/F    | 36                        | TD was later followed by development of spasmody dysphonia | 5 | TD Plus | N | N | THP, TBZ | BoNT for dysphonia | 50 |
| 3           | 60/M    | 24                        | OMD, followed by TD followed by spasmody dysphonia and blepharospasm | 3 | TD Plus | Not done | Not done | THP, TBZ | BoNT for torticollis | 60 |
| 4           | 48/M    | 48                        | Blepharospasm followed by intermittent adductor dysphonia and lingual dystonia followed by TD | 5 | TD Plus | Not done | Impaired phase of deglutition | THP, TBZ | 50 |
| 5           | 40/M    | 12                        | TD followed by jaw closing dystonia | 5 | TD Plus | Not done | Impaired phase of deglutition | THP, TBZ | 50 |
| 6           | 68/F    | 72                        | TD followed by orolingual dystonia | 5 | TD Plus | N | N | THP, TBZ | 60–70          |
| 7           | 66/M    | 3                         | TD as the only symptom | 8 | Pure TD | Not done | N | CBZ, Baclofen | Awaited       |

BoNT: botulinum toxin, CBZ: carbamazepine, N: normal, OMD: oromandibular dystonia, TBZ: tetrabenazine, TD: transfer dysphagia, THP: trihexyphenidyl, UGIE: upper gastrointestinal endoscopy.
ry of blepharospasm for 4 years and a sensation of choking around the throat for 2 years with episodes of intermittent stridor lasting for a few seconds. In the last year, he developed an involuntary protrusion of the tongue out of the mouth, especially during eating. Despite this lingual dystonia, he did not have any difficulty moving the food inside the oral cavity or retaining the bolus of food inside the oral cavity. However, he did have difficulty pushing the bolus of food down his throat. For the same reason, he switched to a semisolid diet and had lost 5 kg over 6 months. A video fluoroscopic barium swallow study similar to the other patient was performed. The study revealed a delay in only the initial part of swallowing (Supplementary Video 1 in the online-only Data Supplement). The patient was started on THP and tetrabenazine (TBZ), after which there was a 50% improvement in swallowing, and he gained 3 kg over 2 months.

The other 5 cases had similar presentations and courses of illness; the details of which are listed in Table 2.

**DISCUSSION**

Eating dysfunction or feeding dystonia have been described previously with oromandibular dystonia (OMD) and cervical dystonia.\(^2\) Such dysfunction may be due to abnormal involuntary movements of the lips, tongue, muscles of mastication, facial muscles or pharyngeal muscles.\(^6\) Difficulty in swallowing in patients with cervical dystonia may be part of the disease or may present as a complication of treatment after peripheral denervation.\(^5,7\) Diagnosis in such cases is usually unambiguous and straightforward.

In the present series, the patients presented with focal dystonia of the pharyngeal muscles. The dystonia was task specific because it occurred only when the patient attempted to swallow. Due to its location, it is not possible to visualize the contraction of specific muscles during swallowing. To facilitate diagnosis, we performed barium swallow using a solid bolus (as described). Videofluoroscopy shows how the bolus was unable to proceed during the first few attempts, and in the final attempt, the bolus moved forward as a whole, thus ruling out any permanent mechanical obstruction. The difficulty in swallowing can be explained by a sustained contraction of the pharyngeal muscles, which is triggered only during swallowing. Additionally, in four patients, there was a contraction of adjacent muscles, namely, OMD, spasmodic dysphonia, etc. The task-specific nature of the problem, associated dystonia and response to anticholinergics point to the diagnosis of dystonia.\(^8\)

We propose a clinical classification of transfer dysphagia due to dystonia.

- Pure transfer dysphagia: transfer dysphagia as the sole presenting feature, as seen in case numbers 1, 5, 6 and 7 (during the initial 5 years). The only problem was task-specific focal dystonia of the pharyngeal muscles leading to dysphagia. Although the possibility of functional dysphagia cannot be completely ruled out, the absence of response to antidepressants and improvement with TBZ/THP was strongly suggestive of dystonia.

- Transfer dysphagia plus: transfer dysphagia associated with other focal dystonia (blepharospasm, OMD, lingual dystonia, etc.), as seen in case numbers 2, 3, 4, 5 and 6 (in number 6, other dystonia developed after 5 years of onset of transfer dysphagia).

Pure transfer dysphagia is more challenging to diagnose, as the only complaint in these patients is difficulty in swallowing, and these patients seldom present to a neurologist early during disease.

During the normal process of swallowing, the tongue pushes the bolus into the oropharynx. The simultaneous relaxation of the pharyngeal muscles facilitates entry into the posterior part, and the bolus is pushed further down into the esophagus. Focal dystonia of pharyngeal muscles temporarily blocks the passage, and the bolus is trapped in the oral cavity due to the simultaneous contraction of the lingual and pharyngeal muscles. In spite of normal neuro-muscular apparatus and no apparent structural obstruction, the patient is unable to swallow.

Transfer dysphagia could be secondary to a local painful cause or may be part of a segmental dystonia involving the craniopharyngeal muscles. Hence, it is important to perform laryngoscopy and gastrointestinal endoscopy in pure transfer dysphagia. Barium swallow (including videofluoroscopy) using a solid bolus (e.g., rasgulla or cotton ball soaked in barium fluid) may be performed to support the diagnosis. In our series, three out of six cases wherein barium swallow was performed revealed an impaired phase of deglutition.
A high index of suspicion by the treating doctor (usually a gastroenterologist, otorhinolaryngologist or neurologist) and by the radiologist in the interpretation of the barium swallow can result in an early neurology referral. A videofluoroscopy examination and review of the fluoroscopic clips with a suspicion of focal dystonia are necessary. A detailed evaluation of other movement disorders and adequate drug trials can lead to a diagnosis.

Transfer dysphagia as a form of focal dystonia may appear as the sole presenting complaint or may present with other forms of focal dystonia. Focal dystonia must be kept in mind before diagnosing a patient with idiopathic or functional dysphagia. A high index of suspicion and an adequate drug trial of sufficient dosage and duration can facilitate an early diagnosis, which could prevent unnecessary investigations.

Supplementary Video Legends

Video 1. Videofluoroscopy image of patient 4 in the pharyngeal phase showing a delayed initiation of swallowing (approximately 16 seconds) followed by a normal passage of barium into the esophagus and stomach without any delay.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.14802/jmd.17081.

Conflicts of Interest

The authors have no financial conflicts of interest.

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