Annals of Otology, Rhinology & Laryngology
2014, Vol. 123(9) 605–608

DOI: 10.1177/0003489414525588
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Dysphagia Due to Inclusion Body Myositis: Case Presentation and Review of the Literature

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Abstract

Objective: This report aimed to present a case of inclusion body myositis presenting with dysphagia and to review the literature.

Methods: Case report and literature review.

Results: Inclusion body myositis is a chronic progressive acquired myopathy, uniquely distinguished by its selective muscle involvement, normal or moderately elevated muscle enzyme concentrations, and a progressive corticosteroid-resistant course. Compared to other inflammatory myopathies, the esophagus is the most commonly involved organ. Specifically, upper esophageal sphincter dysfunction often occurs. Dysphagia may be the only symptom at the time of presentation.

Conclusion: Unlike other inflammatory myopathies, dysphagia in inclusion body myositis is steroid resistant. Management can be difficult. The otolaryngologist must consider underlying neuromuscular processes when evaluating the patient presenting with oropharyngeal dysphagia.

Keywords

inclusion body myositis, dysphagia, myopathy, myotomy, flexible endoscopic evaluation of swallowing

Introduction

Inclusion body myositis (IBM) is a chronic progressive acquired myopathy, uniquely distinguished by its selective muscle involvement, normal or moderately elevated muscle enzyme concentrations, and a progressive corticosteroid-resistant course. Compared to other inflammatory myopathies, the esophagus is the most commonly involved organ. Specifically, upper esophageal sphincter dysfunction often occurs.[1] Dysphagia may be the only symptom at the time of presentation.[2] Unlike other inflammatory myopathies, dysphagia in IBM is steroid resistant. Management can be difficult. In this article, we present a case of IBM presenting with dysphagia and review the literature. The otolaryngologist must consider underlying neuromuscular processes when evaluating the patient presenting with oropharyngeal dysphagia.

Case Presentation

A 78-year-old Caucasian female with hypertension and a distant history of smoking presented with lower extremity weakness, dysphagia, and a 15-pound weight loss for approximately 1 year. Prior to the otolaryngologic evaluation, an esophagram suggested slight aspiration and presence of a cricopharyngeal bar. Physical examination was remarkable for a weak, breathy voice and reduced laryngeal elevation but normal oromotor function. Functional endoscopic evaluation of swallowing with sensory testing revealed reduced pharyngeal squeeze bilaterally and elevated sensory thresholds. Severe residue in the vallecula and hypopharynx with pureed and solid consistencies was noted (Figure 1), which cleared slowly with multiple swallows and liquid washes. Although 1 episode of premature spillage and penetration with thin liquids was observed, no aspiration occurred during the examination.

A neurologic consultation was ordered as a systemic neurologic process was suspected, particularly given her lower extremity weakness. Neurological evaluation was negative. However, a creatine phosphokinase level measured approximately 500, and a rheumatologic disorder was considered. Referral to a rheumatologist was made.

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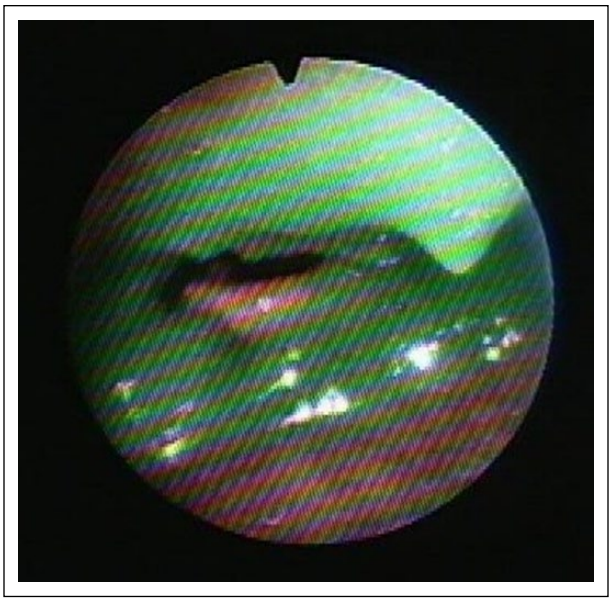

Figure 1. Flexible endoscopic evaluation of swallowing reveals severe residue in the vallecula with pureed consistencies.

Biopsy of the left quadriceps muscle was performed. Microscopic findings included type I and type II angulated atrophic fibers, which reflect mild denervation and reinnervation, and multifocal endomysial inflammatory foci of T-cell lymphocytes and macrophages. In addition, 62 cytochrome oxidase-negative fibers in 40 square-millimeters of muscle were identified. Trichrome stain revealed ragged red fibers, cytoplasmic bodies, and rimmed vacuoles. Immunohistochemical staining with TDP-43 and ubiquitin identified inclusion bodies, and MHC1 staining showed many fibers with sarcolemmal staining, a feature of inflammatory myopathy. A diagnosis of inclusion body myositis was made.

Discussion

Inclusion body myositis is a chronic progressive myopathy. The etiology is idiopathic, however, the disease is considered "acquired" because it is most commonly seen in patients older than 50 years.[3-5] Inclusion body myositis is generally characterized by an insidious onset and a slow, painless progression of proximal and distal muscle weakness.[1,6,7] Muscle histopathology shows rimmed vacuoles, groups of atrophic angular fibers, and endomysial mononuclear cell infiltrations.[8,9] Unlike most inflammatory myopathies, IBM is uniquely distinguished by its selective muscle involvement of the finger flexors and/or quadriceps femoris, normal or moderately elevated muscle enzyme concentrations, and a progressive corticosteroid-resistant course.

The esophagus is the most common organ involved in IBM.[5] Dysphagia is reported to occur in 38% to 84% of all inflammatory myopathy cases,[4,6,10] but it is most common in IBM.[8,11,12] Dysphagia is often the presenting symptom in IBM. Fifty percent of patients with IBM in 1 study were reported to have dysphagia as the presenting symptom.[8] In another study, 10% of IBM patients presented with dysphagia at the onset of disease, and 40% had dysphagia at the time of diagnosis.[5] Inclusion body myositis is more common in men[4,10]; however, dysphagia is more commonly the initial presenting symptom in women.[10,13] In a study of 26 patients with IBM, dysphagia was the initial symptom in 11 of these patients, 9 of which were female.[8]

The mechanism of dysphagia in IBM resembles that in other inflammatory myopathies and is related to inadequate pharyngeal contraction, poor relaxation of the cricopharyngeus muscle, and reduced hyolaryngeal elevation.[1,8] Videofluoroscopy (VF) often demonstrates hypopharyngeal and vallecular stasis, and distention of the hypopharynx.[12,14] Murata et al[1] studied 10 patients with IBM, and all of the patients complaining of dysphagia demonstrated insufficient upper esophageal sphincter relaxation on VF. Oh et al[8] observed the most common VF abnormalities to be residual pharyngeal pooling, tongue base weakness, airway penetration, reduced pharyngeal constrictor contraction, cricopharyngeal muscle dysfunction, and impaired laryngeal elevation.

The consequences of dysphagia are numerous, including nutritional deficits, weight loss, aspiration pneumonia, the need for modified food consistencies and non-oral feeding, decreased quality of life, and poor prognosis.[9,13,15,16] Prognosis is worse in patients with IBM than in those with either polymyositis or dermatomyositis,[17] likely because of the increased risk of aspiration pneumonia and poor response to therapy.[9,18]

For otolaryngologists, IBM has increased significance because of its steroid-resistant dysphagia[14] and the resultant challenges of management. Multiple treatment modalities have been used to treat dysphagia secondary to IBM, including conservative measures, such as swallowing therapy and dietary modifications.[5] Intravenous immunoglobulin (IVIg) therapy and balloon dilation therapy have also been employed.[9] Surgical intervention has been suggested when conservative therapies fail or when severe weight loss and malnutrition are apparent.[14]

The Mendelsohn maneuver is a noninvasive therapy option for patients with dysphagia and involves volitional prolongation of laryngeal elevation during midswallow. It is used most commonly by patients who have weak and poorly coordinated pharyngeal swallow.[19] In several studies with IBM, the Mendelsohn maneuver was useful for helping some of the patients maintain a stable weight and continue eating without aspiration.[10,13] The usefulness of the Mendelsohn maneuver suggests that hyolaryngeal elevation is compromised in some patients with IBM. This is likely because of involvement of the suprahyoid musculature.[20]

Intravenous immunoglobulin therapy has also been shown to be effective for patients with dysphagia secondary to IBM, however, this treatment does not significantly improve IBM-related limb muscle weakness.[4] Cherin et al[4] followed 4 patients with dysphagia secondary to IBM who were treated with IVIg, and swallowing improvement was noted after the third IVIg treatment. In another study of 8 patients by Oh et al,[13] 3 reported benefit from IVIg therapy.

Balloon dilation is a simple, minimally invasive, and low-cost therapeutic method[12] for dysphagia in IBM.[9,17] However, the dysphagia typically recurs.[14] In a study of 3 patients by Murata et al,[11] patients were instructed to perform balloon dilation daily at home. Initially, balloon dilation therapy once a day was sufficient, however, efficacy gradually diminished. Ultimately, it became necessary to perform this therapy several times before each meal as the effect of the dilation diminished after 30 minutes. They suggest that balloon dilation be used only for several days in conjunction with the initiation of IVIg therapy until the effect of IVIg begins or while the patient is waiting for myotomy.

One study looked at balloon dilation after initiation of IVIg therapy compared with IVIg therapy alone. After a single dose of IVIg therapy, treatment efficacy lasted 2 months as represented by reports of symptom improvement and decreases in both hypopharyngeal peak pressure and upper esophageal sphincter contraction. When 3 months of balloon dilation therapy was given after initiating IVIg therapy, it became possible for each patient to eat regular meals for at least 1 year.[21] This study suggests that IVIg and balloon dilation therapies may be more beneficial if administered together or in succession rather than exclusively.

Cricopharyngeal myotomy appears to be effective for dysphagia secondary to IBM,[20] especially when hypertonicity of the cricopharyngeus and impaired relaxation are present.[18] The goal is to eliminate the zone of elevated pressure by transecting extramural muscle fibers between the pharynx and esophagus.[1,9] This could potentially improve flow of bolus into the esophagus by decreasing pressure against which the propulsive force of the pharyngeal constrictors is working. Some effectiveness might be attributable to improved passive flow. Based on a study of 8 patients, Berg et al[15] believe that myotomy can be most beneficial in patients who demonstrate impaired cricopharyngeal relaxation yet have normal pharyngeal motion and pressures. Postmyotomy VF and manometry often do not correlate with symptomatic improvement,[16] which can include improvements in secretions, swallowing, and weight gain.[12,13] Given that a cricopharyngeal bar can be seen on VF due to reduced hyolaryngeal elevation,[20] its effectiveness in such patients may be less certain. It is important for the otolaryngologist to evaluate the pharyngeal phase of swallow carefully to determine what treatment(s) is most likely to help the patient. Cricopharyngeal myotomy should be reserved for cases in which conservative management fails.

The case presentation involved a patient presenting with severe dysphagia, as well as extremity weakness. Flexible endoscopic evaluation of swallowing with sensory testing revealed absent pharyngeal squeeze, poor laryngeal elevation, and significant vallecular and hypopharyngeal residue. Sensory testing was performed as part of the in-office swallowing evaluation prior to our recognizing that this was a rheumatologic disorder. The elevated sensory thresholds are unlikely a result of IBM but rather due to reduced laryngeal adductor reflex sensitivity perhaps due to age or reflux. Flexible endoscopic evaluation of swallowing without sensory testing would have been sufficient for her evaluation. The patient's VF demonstrated a cricopharyngeal bar, which may be a sign of achalasia or poor hyolaryngeal elevation. Such findings in the absence of an obvious explanation (eg, chemotherapy or radiation treatment) should raise suspicion of an underlying systemic process. Neurologic and rheumatologic disease should be considered. Management options were discussed with the patient, however, she declined any further treatment and was subsequently lost to follow-up.

Conclusion

Inclusion body myositis is a chronic progressive acquired myopathy, distinguished by its selective muscle involvement and a progressive corticosteroid-resistant course. Dysphagia is a common symptom in IBM and may be the only symptom at the time of presentation.[2] Neuromuscular and rheumatologic disorders need to be considered in patients presenting with dysphagia, particularly when other systemic symptoms are present. Flexible endoscopic evaluation of swallowing allows the otolaryngologist to get a first-hand look at swallowing function. When findings such as absent pharyngeal squeeze, impaired laryngeal elevation, and significant vallecula and hypopharyngeal stasis are present without another explanation (eg, history of chemoradiation for head and neck cancer), an underlying systemic disorder should be suspected. Collaboration with neurologists and rheumatologists is often warranted. Therapy options for dysphagia secondary to IBM include swallowing therapy, IVIg therapy, balloon dilation, and cricopharyngeal myotomy. Cricopharyngeal myotomy has been reported to be an effective procedure for dysphagia secondary to IBM when more conservative management fails. However, the otolaryngologist should proceed with caution, as there are often multiple elements of the swallowing mechanism affected.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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