Poetic Justice: A Case of Resolved Intractable Hiccups Following POEM for Achalasia

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ABSTRACT

Achalasia is an esophageal motility disorder of impaired lower esophageal sphincter relaxation and absent peristalsis. The presenting symptoms are commonly dysphagia, chest pain, regurgitation, and weight loss. Hiccups have been associated with gastrointestinal diseases but uncommonly associated with achalasia. We present a 62-year-old man with a history of dysphagia, weight loss, and intractable hiccups. High-resolution impedance manometry revealed Type I achalasia, which was treated with per oral endoscopic myotomy. Postoperatively, his dysphagia, weight loss, and intractable hiccups resolved.

INTRODUCTION

Achalasia is an insidious esophageal motility disorder of impaired lower esophageal sphincter relaxation and loss of peristaltic contractions due to loss of myenteric plexus ganglion cells and inhibitory neurons. Achalasia has a reported annual worldwide incidence of 1/100,000 and may be the result of triggered autoimmune response following a viral infection in a genetically susceptible host, although the mechanism remains poorly understood. The much more common and likewise incompletely understood phenomenon of hiccups, or the repetitive contractions of the diaphragm and intercostal muscles, followed by laryngeal closure and characteristic sound, has been found to have a rare association with foregut disorders. Hiccups that persist beyond 1 month are classified as intractable and have been associated with various lesions along the hiccup reflex arc, which involves central or peripheral afferent triggers to a central processing center in the medulla oblongata, with efferent effects to the diaphragm and respiratory muscles. Gastrointestinal disorders that have been linked to intractable hiccups include gastroesophageal reflux disease, esophageal tumors, hiatal hernia, and Helicobacter pylori infection, among others. Pharmacologic treatment of hiccups is typically directed at dopaminergic or γ-aminobutyric acid-ergic receptors, and first-line therapies include baclofen, gabapentin, and pregabalin. Nonpharmacologic treatments including acupuncture, phrenic nerve blockade, and antireflux procedures have been reported in small, nonrandomized trials and case reports; thus, the efficacy of these therapies is limited in their generalizability.

CASE REPORT

A 62-year-old African American man was referred for evaluation of progressive dysphagia to solids and liquids for 2 years. His dysphagia was associated with heartburn, regurgitation, retrosternal pain, 30-lb weight loss, and intractable hiccups that were not responsive to baclofen treatment. Pertinent medical history included daily cigarette use, well-controlled HIV infection, diabetes mellitus, gastroesophageal reflux disease, and posttraumatic stress disorder. Initial evaluation by barium esophagram showed a normal caliber esophagus with marked esophageal dysmotility and severe stasis of contrast in the mid- to distal esophagus. He subsequently underwent an esophagogastroduodenoscopy, which revealed mild superficial erosions in the distal esophagus but was otherwise normal. Biopsies of the esophagus were taken, which revealed mild acute inflammation; no viral or fungal organisms were found, and there was rare to absent intraepithelial eosinophils. High-resolution impedance manometry was then performed, which was notable for absent peristalsis, a mean distal contractile integral DCI of 66 mm Hg s cm, and an elevated median integrated relaxation pressure of 32 mm Hg consistent with Type I achalasia (Figure 1). As shown in Figure 1, the high-resolution impedance manometry topography plot also clearly demonstrated the patient’s ongoing intractable hiccups. His significant
weight loss and symptoms of dysphagia, retrosternal pain, and regurgitation with every meal corresponded to a pretreatment Eckardt score of 12, indicating severe disease, and he was referred for per oral endoscopic myotomy (POEM). His hiccups resolved on postoperative day 1 after POEM. Two weeks later, he had gained weight and achieved clinical remission as reflected by his postoperative Eckardt score of 3.6

DISCUSSION

The association between hiccups and achalasia has rarely been reported. It is possible that the functional esophageal obstruction and subsequent dilation seen in achalasia stimulates the afferent vagal fibers that innervate the distal esophagus, resulting in hiccups.7 One small study reported that 9 of 15 consecutive patients referred for achalasia also had concurrent hiccups. In these patients, dysphagia had been present for at least 6 months, and hiccups were only associated with meals.7 In this series, hiccups improved following treatment for achalasia, which was primarily with pneumatic dilation.7

Over the last decade, POEM has evolved into a mainstream treatment option for achalasia syndromes, and preliminary data from a recent randomized controlled trial suggest a higher year therapeutic success rate compared with pneumatic dilation of 92.2% and 70.0%, respectively.8 According to recently updated American Gastroenterological Association practice recommendations, POEM should be offered as a therapeutic option for achalasia, as it is comparable to laparoscopic Heller myotomy for achalasia Types I and II and is the preferred therapy for Type III achalasia, when a longer myotomy is desired.9

Chronic or intractable hiccups can significantly reduce a patient’s quality of life and can result in poor sleep, suboptimal nutritional status, and weight loss from decreased oral intake.10 This, coupled with the dysphagia associated with achalasia, can result in substantial morbidity, as seen in this patient. In this case, an underlying major esophageal motility disorder was uncovered, and successful treatment of achalasia led to resolution of the associated hiccups.

DISCLOSURES

Author contributions: K. Christian and J. Wellington collected and interpreted data, and wrote and revised the manuscript. R. Kim and S. Quezada revised the manuscript. G. Xie collected and interpreted data, wrote and revised the manuscript, and is the article guarantor.

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