CASE REPORT

Steakhouse Syndrome in Myotonic Dystrophy

Nobuhiko Ogasawara, Kenichiro Sato, Michiko Tsutsumiuchi, Mami Kanzaki and Yoshikazu Uesaka

Abstract:
A 70-year-old man with myotonic dystrophy (MD) showed repetitive vomiting and decreased food ingestion. These symptoms were caused by acute mass of steak impaction occluding the esophagus, known as “steakhouse syndrome,” which may have occurred in response to esophageal functional changes following gastrointestinal involvement due to MD pathology. The occluding food was successfully removed endoscopically, and his symptoms resolved without relapse. Our case suggests that MD patients can present with “steakhouse syndrome” due to bolus food impaction occluding the esophagus as one of their gastrointestinal manifestations, which underscores the need for its consideration in MD patients presenting with similar symptoms.

Key words: myotonic dystrophy, esophageal occlusion, food bolus impaction, steakhouse syndrome, dysphagia

(Intern Med 56: 3179-3181, 2017) (DOI: 10.2169/internalmedicine.9185-17)

Introduction
Myotonic dystrophy (MD) is the most common type of muscular dystrophy and presents not only with skeletal muscle weakness and wasting but often with several other disorders, including cardiac arrhythmia, diabetes mellitus, cataracts, cognitive change, and gastrointestinal manifestations (1). The gastrointestinal symptoms can present as dysphagia, enlargement of the upper esophagus, gastroesophageal reflux disease, abdominal pain, constipation, diarrhea (2-4), or rarely, pseudo-obstruction of the intestine (5, 6).

We herein report a case of an old man with MD who presented, in addition to these gastrointestinal comorbidities, with acute esophageal obstruction due to food mass impaction, or “steakhouse syndrome” (7, 8).

Case Report
A 70-year-old man with late-onset MD type 1 (1) visited our outpatient clinic because of repetitive vomiting, exacerbated difficulty in swallowing, and a decrease in food ingestion that had emerged 2 days before his visit. He had been diagnosed with MD at 65 years of age because of myotonic symptoms, elevated serum creatinine kinase levels, myotonic discharges found in electromyography, and a positive familial history of genetically confirmed MD type 1 in his young son. Despite weakness, his basic activities of daily living were well retained, and he had no restrictive ventilation impairment. He had used laxatives for his constipation. He also had a moderate level of dysphagia, which he himself tended to disregard.

His current symptoms emerged after eating steak, but he had no abdominal pain or diarrhea. He experienced vomiting only when he tried to eat or drink anything. Esophageal obstruction due to mechanical lesions or congestion of food in his esophagus was suspected following computed tomography (Figure A and B; indicated using yellow arrowheads). An insufficiently masticated mass of steak that was occluding the esophagus was identified and endoscopically removed using net forceps (Figure C-E). No mechanical obstructive lesions, such as malignancy or herniation, were found on the esophageal lining, and his symptoms resolved without relapse. He also simultaneously acquired aspiration pneumonia, which was also successfully treated. He was ultimately diagnosed with steakhouse syndrome.
Discussion

Our patient exhibited “steakhouse syndrome” with repetitive vomiting and decreased food ingestion due to food bolus impaction of the esophagus (7, 8). This syndrome can be caused by various etiologies: esophageal mechanical narrowing due to reasons such as esophageal carcinoma, diverticulum or hiatal hernia (7, 8), or esophageal motility disturbances, including esophageal achalasia, diffuse esophageal spasm, esophagogastric junction outflow obstruction, and Nutcracker esophagus (7, 8). Although there was no apparent evidence of esophageal mechanical narrowing, we were unable to perform esophageal manometry or esophagogram to further narrow down the differential diagnoses regarding the functional causes of the syndrome. However, we suspect that the steakhouse syndrome in our case may have been due to the functional changes in the esophagus often reported in MD patients: an impaired lower esophageal sphincter function, a decreased or absent esophageal peristaltic function (9, 10), and decreased esophageal pressure (9) in addition to weakened mastication (1) and a decreased pharyngeal function (4). These esophageal motility changes are induced by the pathological esophageal muscle involvement due to the thinning of the striatal muscle layer in the upper esophagus and the smooth muscle layer in the middle and lower esophagus, as observed in previous pathological studies (9, 10).

This case suggested that MD patients can present with steakhouse syndrome due to food bolus impaction of the esophagus as one of their gastrointestinal manifestations. Although MD patients often present with symptoms such as aspiration pneumonia due to dysphagia or rarely with repetitive vomiting due to pseudo-obstruction of the intestine (2-4), it is important to consider the possibility of esophageal obstruction due to food congestion. This is nec-
ecessary in the management of the dietary intake in MD patients or when treating MD patients with gastrointestinal symptoms. The cognitive and personality changes that are occasionally observed in MD patients (1), such as in our patient who tended to disregard his dysphagia, may also contribute to this type of event. In addition, if the food incompletely occludes the esophagus, these symptoms might occur as subacute or chronic onset, which can result in a delayed recognition of the underlying cause of the symptoms.

In conclusion, our experience with the present case suggested that MD patients can present with steakhouse syndrome due to food bolus impaction occluding the esophagus as one of their gastrointestinal manifestations, underscoring the need for the consideration of this syndrome in MD patients presenting with similar symptoms.

The authors state that they have no Conflict of Interest (COI).

References

1. Bird TD. Myotonic Dystrophy Type 1. GeneReviews© [Internet]. 1999 Sep 17 [cited 2017 Feb. 1]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK1165/ 2. Rönnblom A, Forsberg H, Danielsson A. Gastrointestinal symptoms in myotonic dystrophy. Scand J Gastroenterol 31: 654-657, 1996.
3. Modolell I, Mearin F, Baudet JS, Gámez J, Cervera C, Malagelada JR. Pharyngo-esophageal motility disturbances in patients with myotonic dystrophy. Scand J Gastroenterol 34: 878-882, 1999.
4. Bellini M, Biagi S, Stasi C, et al. Gastrointestinal manifestations in myotonic muscular dystrophy. World J Gastroenterol 12: 1821-1828, 2006.
5. Saito T, Miyai I, Matsumura T, Nozaki S, Kang J. [A case of myotonic dystrophy with intestinal pseudo-obstruction syndrome]. Rinsho Shinkeigaku 39: 1064-1066, 1999 (in Japanese, Abstract in English).
6. Glaser AM, Johnston JH, Gleason WA, Rhoads JM. Myotonic dystrophy as a cause of colonic pseudoobstruction: not just another constipated child. Clin Case Rep 3: 424-426, 2015.
7. Stadler J, Hölscher AH, Feussner H, Dittler J, Siewert JR. The “steakhouse syndrome”. Primary and definitive diagnosis and therapy. Surg Endosc 3: 195-198, 1989.
8. Kita K, Nagatsuna M, Fujinami H, Yamashiro S. Steakhouse syndrome: a case report. Gen Med 12: 83-84, 2011.
9. Kaida K, Kono S, Komiya T, Kawai M. [Imaging and pathological studies on the esophageal dysfunction in patients with myotonic dystrophy]. Rinsho Shinkeigaku 36: 300-305, 1996 (in Japanese, Abstract in English).
10. Kono S, Kaida K, Kawai M. [Esophageal dilatation in a patient with myotonic dystrophy: a correlative study on CT and pathologic findings]. Rinsho Shinkeigaku 38: 238-241, 1998 (in Japanese, Abstract in English).

The Internal Medicine is an Open Access article distributed under the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view the details of this license, please visit (https://creativecommons.org/licenses/by-nc-nd/4.0/).