

# Pseudoachalasia Following Nissen Fundoplication

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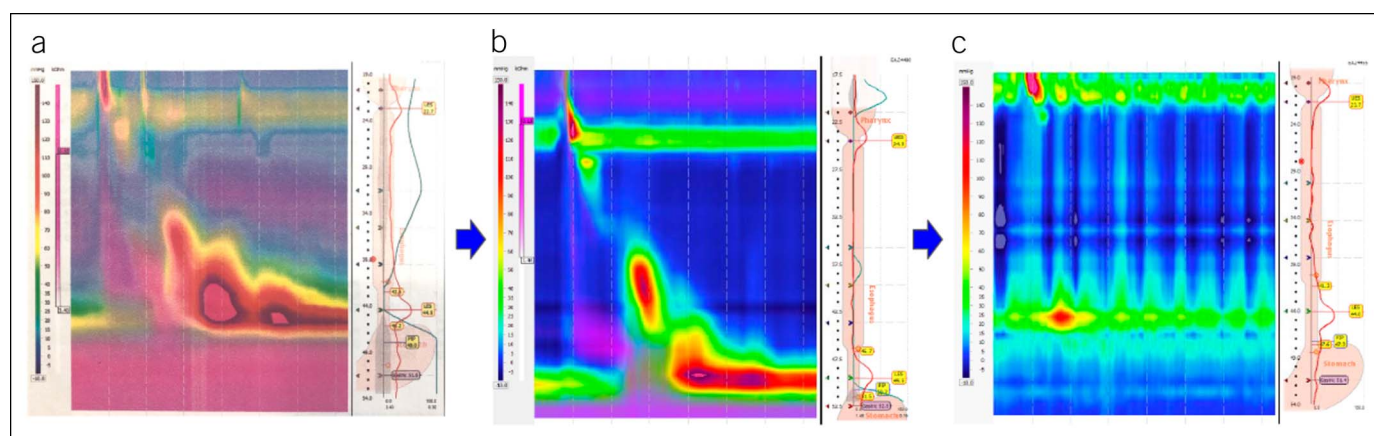
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## ABSTRACT

Pseudoachalasia, clinically indistinct from achalasia in symptoms and high-resolution manometry findings, differs by a secondary etiology with more than half of the occurrences arising from malignancy. Rarely pseudoachalasia presents after surgeries of the esophagus and gastroesophageal junction. This case offers an additional example of pseudoachalasia after Nissen fundoplication; however, it is unique to the literature by documenting complete manometric progression from normal to pseudoachalasia in a single patient. This case serves to highlight the importance of thorough workups in patients with achalasia symptoms and broadens understanding of this disease process by offering manometric findings in an evolutionary phase.

## INTRODUCTION

Pseudoachalasia and achalasia are clinically indistinct in their symptoms and findings on high-resolution manometry (HRM) but differ in etiologies. Achalasia, inflammatory in nature with many proposed etiologies, is defined by the lack of esophageal peristalsis and lower esophageal sphincter (LES) relaxation on HRM without identification of a secondary cause.<sup>1,2</sup> Pseudoachalasia, or secondary achalasia, arises most commonly because of malignancy; however, the literature reports approximately 12% of cases are due to surgeries involving the esophagus and gastroesophageal (GE) junction.<sup>3,4</sup> The multimodal diagnostic workup is the same for both clinical entities and includes a barium esophagram, esophagogastroduodenoscopy (EGD) with biopsies, computed tomography (CT) scan, and HRM.<sup>4,5</sup> There have been several cases of pseudoachalasia reported after Nissen fundoplication, including both early and late development of symptoms.<sup>6,7</sup> We present a case after Nissen fundoplication, unique by documenting progression with HRM, further supporting this clinical entity while augmenting the understanding of the disease progression.



**Figure 1.** (A) High-resolution manometry documenting the progression from an initial normal esophageal motility and lower esophageal sphincter (LES) relaxation prefundoplication. (B) Followed by preserved peristalsis and LES relaxation with intermediary development of distal esophageal spasm after Nissen fundoplication and (C) degeneration to aperistalsis with a lack of LES relaxation consistent with Type I achalasia.

## CASE REPORT

A 66-year-old man presented to the clinic with dysphagia and GE reflux disease symptoms. Initial EGD revealed Los Angeles grade A esophagitis at the GE junction and a 3-cm hiatal hernia, and the patient weighed 144 pounds. The LES was traversed easily without identifiable strictures or masses. The pharyngeal function study was without evidence of aspiration or stasis. A CT emphasized a hiatal hernia, showed no esophageal dilation, esophagogastric junctional (EGJ) narrowing, gastric tumors, or vascular obstructions. He was managed medically with pantoprazole 40 mg twice daily; de-escalating to daily therapy after 8 weeks was not tolerated because of worsening symptoms. Ultimately, he continued twice-daily dosing along with recommended lifestyle and dietary modifications including avoidance of foods which trigger symptoms, elevating the head of his bed, healthy weight maintenance, and avoidance of alcohol and smoking. This was continued in addition to a nightly dose of ranitidine 150 mg that was trialed for the 3 months before a repeat EGD at the age of 68 years for continued symptoms. His hiatal hernia measured 5 cm and Los Angeles grade B esophagitis was present; a progression in size was attributed to recurrent straining from persistent regurgitation and an increase in patient weight to 164.4 pounds. HRM showed normal LES resting pressures, relaxation, and esophageal peristalsis. Laparoscopic Nissen fundoplication was performed at the age of 69 years. Three months postoperative EGD identified a distal esophageal stricture at the level of the wrap; although the regular endoscope could traverse it, he underwent 16–18 mm sequential dilatation. At the age of 72 years, progression of dysphagia, vomiting, and chronic hiccups led to repeat EGD and discovery of an intrinsic 14-mm lower esophageal stricture that was dilated to 18 mm. Repeat HRM revealed intact peristalsis with normal LES relaxation, but on 6 of 10 swallows, there was decreased distal latency (<4.5 seconds) consistent with esophageal spasm (Figure 1). After 4 months of only transient improvement, he was referred to the esophageal disease clinic at the age of 73 years.

At his initial visit, he reported vomitus after 90% of meals, difficulty in swallowing pills, and a 15-pound unintentional weight loss over 2 weeks, which prompted stepwise evaluation. A timed barium esophagram (TBE) revealed a high-grade obstruction in the distal esophagus, suggestive of total blocking with only minimal contrast passage (Figure 2). Repeat EGD had a significant resistance to gastric intubation of the scope raising concern for achalasia or extrinsic compression. An abdominal, thoracic, and pelvic CT showed no extrinsic masses or alternative malignancy and suggested esophageal dysmotility associated with a thickened and narrowed lower esophageal wall above the esophagogastric junction. Repeat HRM showed 100% incomplete bolus transit for liquids and complete aperistalsis. In addition, the LES had elevated basal (56.7 mm Hg) and median integrated relaxation (22.4 mm Hg) pressures consistent with Type I achalasia by the Chicago Classifications. Correlating clinical and diagnostic information, he was diagnosed with mechanical EGJ outflow obstruction and development of pseudoachalasia secondary to



**Figure 2.** Timed barium esophagram showing high-grade obstruction.

a tight Nissen fundoplication. He underwent a laparoscopic Nissen removal with Heller myotomy resulting in resolution of dysphagia and sustained positive results at the 4-month follow-up.

## DISCUSSION

This case is unique to the literature by showing a complete manometric progression from normal esophageal motility to pseudoachalasia post-Nissen fundoplication over a 5-year period. Development was attributed to a tight fundoplication with the evolution of aperistalsis, which has been described in the

literature.<sup>6,8</sup> In addition, injury to the myenteric plexus inadvertently during wrap construction or by degeneration from sustained distal esophageal mechanical obstruction is theorized, even the direct vagus nerve injury during surgery has been reported.<sup>9</sup> The proposed pathophysiology of achalasia-type dysmotility is a functional loss of the myenteric plexus ganglion cells in the lower esophagus and LES. The dysfunction of post-ganglionic inhibitory neurons (responsive to nitric oxide and vasoactive intestinal peptide) results in defective excitation and inhibition. Persistent cholinergic activation leading to failed relaxation of the LES, the defining feature of achalasia, may also manifest as hypercontractility and rapid propagation of contractions in the distal esophagus.<sup>10</sup> The intermediary development of esophageal spasms on HRM shown here may offer future clinical value if a similar manometric progression can be documented post-Nissen fundoplication. For instance, it has already been suggested that the removal or loosening of adjustable gastric bands, in which pseudoachalasia has developed, can reverse dysmotility on subsequent HRM in a number of patients.<sup>11,12</sup>

Post-Nissen fundoplication dysphagia, commonly encountered by gastroenterologists and minimally invasive surgeons, can be evaluated with a TBE, which offers insight on esophageal emptying and the location of esophageal outflow obstruction. The TBE was originally designed to measure esophageal emptying in patients with achalasia before and after therapy. The TBE is an objective assessment of the severity of esophageal obstruction and treatment response in any process resulting in poor esophageal emptying, including mechanical EGJ outflow obstruction from a Nissen fundoplication.<sup>13</sup>

Pseudoachalasia remains uncommon, reported in up to only 4.7% of those with symptoms consistent with achalasia.<sup>14</sup> Abubakar et al reported in their review of literature that 12% of reported cases are due to surgeries involving the esophagus and GE junction, whereas 68% are from primary and secondary malignancies. Therefore, it remains important for the clinician to use a multimodal approach targeted at separating primary from secondary achalasia. Pseudoachalasia should be favored and evaluated in those with a more rapid symptom development over the age of 50 years and include a barium esophagram, EGD with biopsies, CT scan, and HRM to distinguish the etiology.<sup>4,5</sup> In addition, amyl nitrite inhalation has been shown to produce relaxation of the LES in achalasia but not in pseudoachalasia.<sup>15</sup> In conclusion, clinicians should understand the components and importance of a broad workup in those with achalasia symptoms. In addition, be aware of the risk, albeit rare, of pseudoachalasia development after antireflux

surgery and cognizant of the possible manometric progression as identified in this patient.

## DISCLOSURES

**Author contributions:** Both authors contributed equally to this manuscript. JW Harbaugh is the article guarantor.

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**Informed consent** was obtained for this case report.

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