



Contents lists available at ScienceDirect

## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

# Case report of robotic dor fundoplication for scleroderma esophagus with aperistalsis on manometry



Alonso Andrade\*, Matthew K. Folstein, Brian R. Davis

Department of Surgery, Texas Tech University Health Sciences Center, El Paso, TX, United States

## ARTICLE INFO

## Article history:

Received 16 February 2017

Received in revised form 13 June 2017

Accepted 13 June 2017

Available online 16 June 2017

## Keywords:

Robotic surgery

Esophagitis

Gastroesophageal reflux

Scleroderma

Minimally invasive surgery

Case report

## ABSTRACT

Scleroderma is a systemic disease of collagen deposition resulting in fibrosis of small arteries and arterioles. It commonly affects the skin, lungs, and gastrointestinal tract. The most common site of GI tract involvement is the esophagus. We present the case report of a 44 year old female with scleroderma esophagus and severe reflux which was successfully treated with robotic dor fundoplication. Because of the wide variety of symptoms with which this problem can present, a tailored approach taking into consideration the patient's symptomatology and findings during diagnostic work-up was implemented with good results. The patient exhibited complete resolution of symptoms at short term follow up. Robotic dor fundoplication is an effective option for patients with scleroderma esophagus and no evidence of hiatal hernia or esophageal shortening.

© 2017 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Scleroderma is a systemic disease of collagen deposition resulting in fibrosis of small arteries and arterioles. The most common site of involvement for scleroderma in the gastrointestinal tract is the esophagus. Up to eighty percent of patients with scleroderma and GI tract involvement present with either gastroesophageal reflux or dysphagia within two years of diagnosis. Evidence of erosive esophagitis, strictures, or Barrett's metaplasia is commonly seen in these patients. The operative management for GERD or dysphagia in the setting of scleroderma must be tailored to the unique needs of the patient based on the functional and mechanical abnormalities identified during their workup. In accordance with the SCARE criteria [7], we present the case of a 44 year old woman with reflux esophagitis and aperistalsis on manometry due to scleroderma successfully managed with Robotic Dor Fundoplication and complete resolution of symptoms at short-interval follow-up.

## 2. Presentation of case

A 44 year old woman with twenty-five-year history of GERD refractory to high dose proton pump inhibitors presents for evaluation of reflux. Patient reported sleeping on a wedge to help alleviate nighttime symptoms. Her medical history is notorious for a diagnosis of scleroderma and pulmonary fibrosis six years prior to current

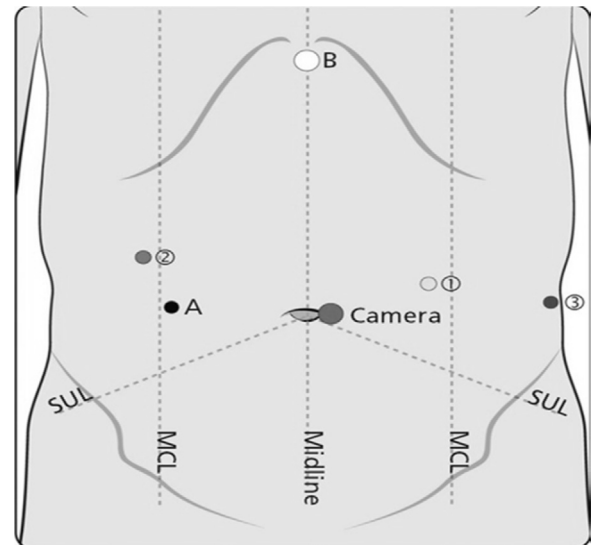
presentation for which she has received intermittent pulse steroids. A recent esophagogastroduodenoscopy demonstrated islands of salmon colored mucosa with Z-line biopsies positive for Barrett's esophagus. As part of her preoperative evaluation she underwent repeat endoscopy, barium swallow, and high resolution manometry. The repeat endoscopy confirmed the absence of a hiatal hernia and was negative for retained food. Evaluation of relaxation of the distal esophagus to differentiate achalasia and scleroderma was also done. The patient underwent a barium swallow to confirm diagnosis and evaluate position of GE junction (Fig. 1). In addition the patient had a high resolution manometry study to confirm aperistalsis and assist in creating a tailored operative approach to address her symptoms (Fig. 2). Of note, the patient had the appearance of achalasia on barium swallow, but showed aperistalsis on manometry and, paradoxically, low LES pressure. Upon completion of these studies, it was determined that the patient suffered from gastroesophageal reflux disease, barrett's metaplasia, and esophageal scleroderma. After informed consent was obtained, the patient underwent a Robotic Dor Fundoplication to alleviate the gastroesophageal reflux and prevent long-term complications with dysphagia (Fig. 3). Intraoperative endoscopy was performed to evaluate the wrap (Fig. 4). Total OR time including anesthesia, robot docking, and endoscopy was 150 min, with a total EBL of 15 mL. The operation and postoperative course were uneventful. The patient was discharged on post-operative day one with a clear liquid diet and subjective description of symptom resolution while in the recumbent position. At one month follow up the patient was tolerating a soft-mechanical diet and reported subjective resolution of all symptoms with cessation of PPI. The patient did not

\* Corresponding author.

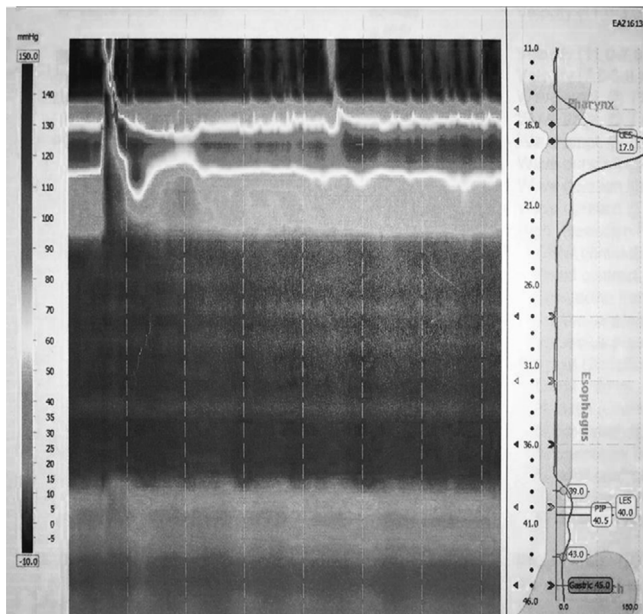
E-mail address: [alonso.andrade.f@gmail.com](mailto:alonso.andrade.f@gmail.com) (A. Andrade).



**Fig. 1.** Barium swallow showing normal deglutition, Dilated smooth esophagus with a smooth stenosis at the EG junction. Retained food in the esophagus with slow passage of contrast into esophagus with decreased peristalsis. Findings consistent with achalasia.



**Fig. 3.** Courtesy of DaVinci Online Community. Standard Procedure Card for foregut surgery. For our Robotic Dor Fundoplication Camera port, Port 2 and Port 1 were used. Port B was used for a Nathanson liver retractor. The Assistant Port A and Port 3 were not used.



**Fig. 2.** High resolution esophageal manometry showing low LES pressures and aperistalsis. Mean LES pressure: 5 mmHg (abnormal <15%), LES Percent Relaxation: 56%, Percent Failed Propulsive Swallows: 100%, and Contraction Amplitudes: <20% (very low).

complain of dysphagia or gas bloating. As of the time of submission of this manuscript the patient has not had additional imaging or endoscopy performed, but she has remained asymptomatic in relation to her preoperative complaints.

**3. Discussion**

Patients with Scleroderma can develop various systemic complications from their disease process. Scleroderma most commonly affects the skin, lungs, and gastrointestinal tract. Of these,



**Fig. 4.** Intraoperative endoscopy showing no evidence of injury to esophagus, no evidence of narrowing at GE junction. Shown here is a retroflex view with GE junction valve consistent with fundoplication.

esophageal pathology can be challenging for the clinician to manage due to the various presentations which can occur [1,2]. The differential diagnosis for a patient with aperistaltic esophagus includes achalasia, scleroderma, eosinophilic esophagitis, and carcinoma of the gastroesophageal junction or gastric fundus. The operative management of these pathologies can be very different, so a meticulous pre-operative evaluation must be carried out and a “tailored approach” is recommended. Even within the subset of patients with scleroderma esophagus, the various existing surgical options and lack of a standard operative approach speaks to the varied presentation and the importance of having a tailored approach to these patients. Patients with scleroderma esophagus can present with any combination of findings including esophageal shortening, barrett’s esophagus, peptic stricture, and aperistaltic esophagus among others [1–6]. The existing surgical options include esophagectomy, collis gastroplasty with fundoplication, roux-en-Y gastric bypass, and the various complete and partial fundoplications [1–6]. Some studies have shown improved quality of life and symptomatic resolution with Roux-en-Y gastric



**Fig. 5.** Intraoperative view of completed anterior Dor Fundoplication as seen in surgeon console.

bypass when compared to esophagectomy and standard fundoplication [2]. Still others have shown good success with techniques like the collis gastroplasty and fundoplication [3]. Laparoscopic Nissen fundoplication is a widely accepted technique for managing GERD, but in the setting of scleroderma, there is clearly a concern for post-operative dysphagia and gas bloating, therefore a partial fundoplication was considered optimal in our patient. Due to the complexity and sometimes difficult exposure with the various available laparoscopic procedures for treating the scleroderma esophagus, robotic surgery has emerged as a valuable tool for the surgeon treating these pathologies. The robotic approach has been touted as providing better instrument control, better visualization, and greater ease of use when compared to traditional laparoscopic surgery (Fig. 5). With this in mind, a robotic approach was selected for the management of our patient. The decision to use an anterior fundoplication was based on the ease of performing the procedure and the fact that this operation also obviates the need for extensive dissection in an inflamed or otherwise difficult operative field. In addition, this approach has been shown to be successful in treating reflux and avoid the common problem of dysphagia in the setting of an aperistaltic esophagus.

#### 4. Conclusion

The management of a patient with scleroderma esophagus can be challenging for the clinician. The main learning points from this case are that the clinician must take into consideration the specific signs, symptoms, and findings on diagnostic work-up in order to come up with a tailored surgical approach that will address the problem at hand and minimize the morbidity associated with some of these surgical approaches. In this patient, a complete fundoplication to address the gastroesophageal reflux in the setting of

scleroderma esophagus, can be complicated by dysphagia. Dor and Toupet funduplications can be as effective in preventing reflux as a Nissen fundoplication at 5-year follow up and are associated with decreased risk of dysphagia. Taking advantage of existing technology which allows surgeons to perform more complex procedures is an important tool when managing this patient population. As shown by our patient, the anterior robotic Dor Fundoplication is an effective option for patients with scleroderma esophagus and no evidence of hiatal hernia or esophageal shortening.

#### Conflicts of interest

None.

#### Sources of funding

Department of Surgery, TTUHSC El Paso.

#### Ethical approval

Does not apply.

#### Consent

Consent available upon request.

#### Authors contribution

Brian R. Davis– Study concept and design.

Alonso Andrade– Data collection, analysis, manuscript writing and editing.

Matthew Folstein– Data collection, analysis.

#### Guarantors

Alonso Andrade MD.

Brian R. Davis MD.

#### References

- [1] R. Henderson, F. Pearson, *Surgical Management of esophageal scleroderma*, J. Thorac. Cardiovasc. Surg. 66 (1973) 686.
- [2] M.S. Kent, J.D. Luketich, K. Irshad, et al., Comparison of surgical approaches to recalcitrant gastroesophageal reflux disease in the patient with scleroderma, *Ann. Thorac. Surg.* 84 (2007) 1710–1716.
- [3] M. Orringer, J. Orringer, L. Davich, C. Zarofonotis, Combined Collis gastroplasty-fundoplication operations for scleroderma reflux esophagitis, *Surgery* 90 (1981) 624–630.
- [4] F.G. Pearson, *Esophageal Surgery*, second edition, Churchill Livingstone, Philadelphia, PA, 2002.
- [5] T. Poirer, R. Taillefer, P. Topart, et al., Antireflux operations in patients with scleroderma, *Ann. Thorac. Surg.* 58 (1994) 66.
- [6] D.I. Watson, G.G. Jamieson, J.R. Bessel, P.G. Devitt, Laparoscopic fundoplication in patients with aperistaltic esophagus and gastroesophageal reflux, *Dis. Esophagus* 19 (2006) 94–98.
- [7] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, the SCARE group, The SCARE statement: consensus-based surgical case report guidelines, *Int. J. Surg.* 34 (2016) 180–186.

#### Open Access

This article is published Open Access at [sciencedirect.com](https://www.sciencedirect.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.