Review Article

Esophageal duplication cysts: a clinical practice review

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Abstract: Esophageal duplication represents one of the most common types of bronchopulmonary foregut malformations. These rare congenital anomalies occur secondary to embryological aberrations between the 4th and 8th weeks of gestation. In order to be classified as an esophageal cyst a mediastinal cyst must have a close proximity with the esophagus, be lined by alimentary (squamous epithelium) or tracheobronchial mucosa and covered by two smooth muscle layers. These rare anomalies are often asymptomatic during adulthood. However, they can cause symptoms in early childhood, generally during the first 2 years of life. Variations in location, size, presence or absence of heterotopic mucosa, will dictate the clinical presentation. Dysphagia, food impaction, persistent cough and chest pain are common clinical presentations. Imaging studies including esophagram, computed tomography (CT) and magnetic resonance imaging (MRI) can provide key findings to reach the diagnosis. Nonetheless, endoscopic evaluation, particularly endoscopic ultrasound (EUS) is the most valuable tool to determine whether this lesion is cystic versus solid and or if there are abnormal mucosal findings. Needle biopsies are controversial but can help with drainage and to rule out malignant transformation. Therapeutic options include endoluminal drainage. However, more definitive therapies include surgical excision. Open and minimally invasive (laparoscopic and thoracoscopic) techniques have been demonstrated to be safe and effective at completely removing these lesions. Recently, robotic-assisted resections have gained more attention with case reports and series reporting excellent outcomes.

Keywords: Esophagus; congenital; duplication cyst; thoracic surgery

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Introduction

Esophageal duplication cysts are rare congenital anomalies that constitute between 0.5% and 2.5% of all esophageal masses (1). It is estimated that the incidence of esophageal duplication cysts is one in 8,200; they are twice as likely to occur in males when compared to females (2,3). The exact etiology of alimentary tract duplications is not well understood. However, it is thought to occur between the 4th and 8th weeks of development as a result of failure of intrauterine vacuolization of the esophagus (1). The duplication cysts lay within the esophageal wall, falling under the category of intramural lesions and need to be distinguished from esophageal malignancies, leiomyomas, lipomas, and gastrointestinal stromal tumors (GISTs). Esophageal duplications cyst can be lined by alimentary (squamous epithelium) or tracheobronchial mucosa regardless of where they are located. They have an envelope of smooth muscle and share a common wall with the esophagus (2,4). Up to one-third of esophageal duplication cysts contain heterotopic gastric mucosa; heterotopic pancreatic mucosa and mucosa consistent with Peyer's patches have also been described (5,6). Approximately 80%

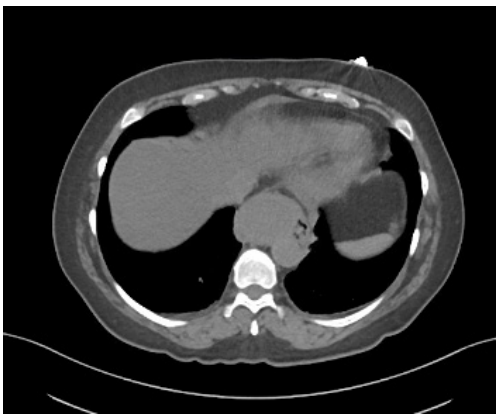

Figure 1 CT axial view demonstrated complex duplication cyst of the esophagus with extrinsic compression of esophageal lumen. CT, computed tomography.

of esophageal duplication cysts do not communicate with the esophageal lumen, whereas tubular duplications do have a direct communication with the lumen.

Clinical presentation

Patients with esophageal duplication cysts often remain asymptomatic, however those that become symptomatic usually present during childhood (1). During adulthood most esophageal duplication cysts are found incidentally while patients are undergoing work-up for unrelated conditions. Esophageal duplication cysts are most likely to be found in the lower third of the esophagus and the remaining are found in the upper/middle third of the esophagus (7). Clinical presentation varies depending on the location of the cyst. Those located within the upper esophagus are associated with respiratory manifestations including, dyspnea, stridor, respiratory distress, mass effect, retrosternal pain and cough. Whereas those located in the lower third of the esophagus are mainly associated with dysphagia, esophageal stenosis, food impaction and emesis. Although rare, hematemesis has been reported in the presence of heterotopic gastric mucosa. Additionally, cardiac arrhythmias, bleeding and rupture with complicated mediastinitis have also been reported in the literature (8). Due to the clockwise rotation of the stomach during embryogenesis, esophageal duplication cysts most commonly lay on the right lateral aspect of the esophagus (2).

Diagnosis

Benign esophageal lesions are often times asymptomatic and incidentally encountered during endoscopic or radiologic evaluation of the esophagus. Nevertheless, they pose challenges in establishing an accurate diagnosis and management plan. In the work-up of an esophageal duplication cyst, plain radiographs may demonstrate a well-defined soft tissue density in close association with the esophagus or widening of the mediastinum. A barium esophagram is an excellent study to delineate the anatomy of the esophagus, assess mucosal irregularities and visualize the location of the duplication cyst in relationship to surrounding organs. The esophagram can show an extrinsic versus and intramural compression on the wall of esophagus. Furthermore, intraluminal communication can also be recognized during this study. We routinely order an esophagram for pre-operative planning and post operatively to have a new baseline of the neo-anatomy post excision.

Computed tomography (CT), preferably, with intravenous contrast assists to delineate thick-walled cystic structures that can have complex versus simple fluid within. Generally, the cysts have smooth contour and low attenuation, with Hounsfield units ranging from 8 to 12 corresponding to water density. However, cysts can be filled with proteinaceous complex material, suffer spontaneous rupture or post instrumentation hemorrhage leading to images with hyper attenuating characteristics and elevated Hounsfield units closer to blood density (*Figure 1*). With the usage of oral contrast, filling defects are sometimes seen, but since duplication cysts are generally submucosal lesions, the lining appears smooth and with normal characteristics. While the differentiation between bronchogenic cysts and esophageal duplications cysts ca be challenging, a close relationship with the esophagus, lack of cartilaginous components and the presence of a double smooth muscle layer favors the diagnosis of an esophageal duplication cyst over bronchogenic cyst.

Further imaging modalities include magnetic resonance imaging (MRI) that shows the cystic nature of the mass as well as high intensity when imaged with T2-weighted sequences, regardless of the nature of the cyst contents. In the pediatric population, radionuclide scanning with Technetium 99m sodium pertechnetate assists with the localization of ectopic gastric mucosa, as the technetium is up taken by the gastric mucosa and this can be visualized

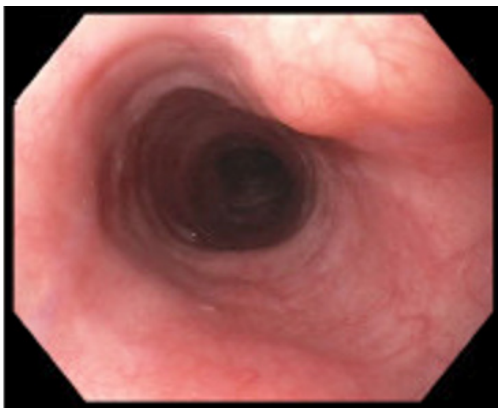

Figure 2 Esophagoscopy demonstrating extrinsic compression of the esophageal wall at 35 cm from the incisors with normal appearing mucosa.

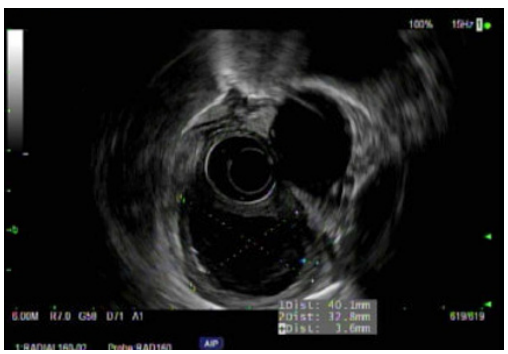

Figure 3 EUS with anechoic cystic lesion in direct contact with the esophagus and thickened wall consistent with esophageal duplication cyst. EUS, endoscopic ultrasound.

with nuclear scans (9).

Positron emission tomography (PET) CT should only be ordered if malignant transformation of the duplication cyst has been confirmed with tissue biopsy. In this case, a PET scan will allow further investigation regarding invasion and whether there is spread to mediastinal or distant structures.

On esophagogastroscopy, duplication cysts appear as submucosal lesions with normal appearing esophageal mucosa (*Figure 2*), differential diagnosis include leiomyoma, GISTs and lipomas (10). Endoscopic ultrasound (EUS) demonstrates homogenous hypoechoic periesophageal multi-layered wall mass. Margins are generally smooth and the muscularis propria of the esophagus is in direct contact with the cyst wall (11).

The role of EUS-guided fine needle aspiration (EUS-FNA) in the diagnosis of esophageal duplication cysts is controversial. Studies have shown an infection rate as high as 14% when EUS-FNA is performed (12,13). EUS-FNA should be reserved for lesions of indeterminate appearance, lesions concerning for malignancy and lesions that appear atypical (*Figure 3*).

Once diagnosis has been confirmed, an open discussion with patients regarding risks and benefits of surgical excision is recommended for a shared decision making approach. Surgery can be considered for both asymptomatic and symptomatic patients given the risk of future complications or malignant transformation (14,15). These lesions do not regress spontaneously and identification of the entire tract is key to ensure adequate resection. The surgical approach can be open, through a laparotomy or thoracotomy, or via minimally-invasive techniques dependent on the skill set of the surgeon. Minimally-invasive approaches have been demonstrated to be safe, feasible and with excellent outcomes (16,17). Given that the majority of esophageal duplication cysts are localized to the distal esophagus, herein we describe our trans-abdominal approach for surgical excision using the robotic platform. Cervical and thoracoscopic approaches are better reserved for upper and mid-esophageal lesions.

Surgical approach

The patient is placed in the operating table with a footboard and all the pressure points adequately padded. Our patients receive preoperative antibiotics with a first generation cephalosporin and deep vein thrombosis prophylaxis routinely. A foley catheter and arterial line are also placed for continuous monitoring of the urine output and blood pressure. We utilize four robotic ports, the first (camera port) is placed 2 cm above and towards the left from the umbilicus. The second port is place in the left hemi abdomen, at the peritoneal reflexion, a third robotic port is placed in between the two prior ones and the fourth robotic arm is placed in the right upper quadrant. An assistant (12 mm) port goes in the left lower quadrant. Finally, a subxiphoid 5 mm port for the liver retractor (picture). the robot is routinely armed with the cadiere forceps (arm 1), camera (arm 2), vessel sealer (arm 3) and the curve bipolar and double fenestrated (interchange in arm 4) (*Figure 4*).

We start by dissecting at the phrenoesophageal membrane to enter the diaphanous plane of dissection. We

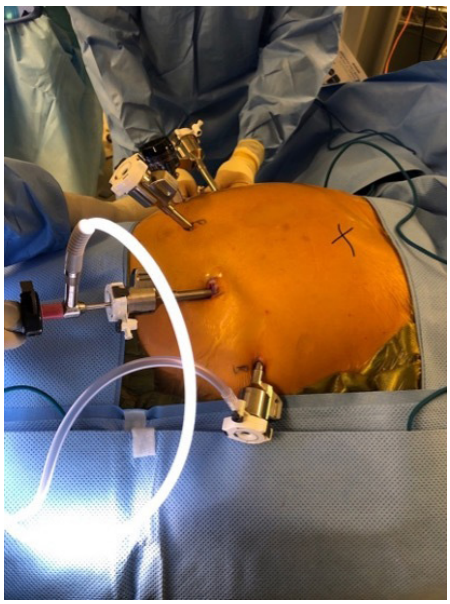

Figure 4 Robotic port placement for trans-hiatal robotic assisted laparoscopic duplication cyst excision and Dor fundoplication.

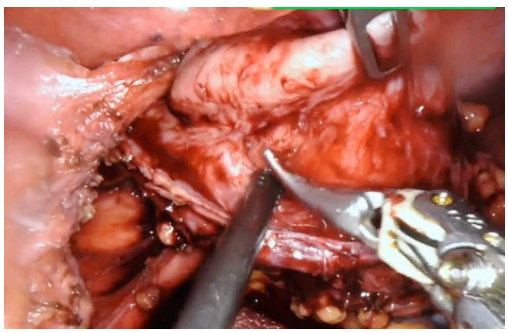

Figure 5 Myotomy and cyst enucleation.

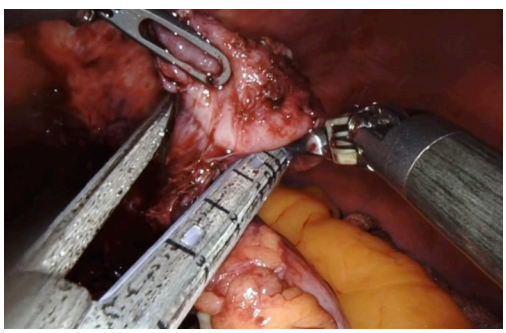

Figure 6 Cyst excision.

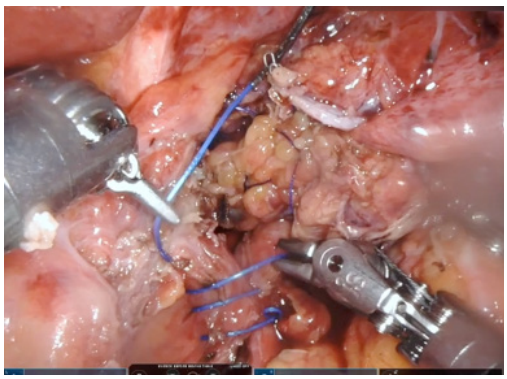

Figure 7 Posterior cruroplasty with running non absorbable 0 barbed suture.

are meticulous at visualizing and preserving both vagus nerves during the dissection. Thereafter, we dissect on the pars flacida towards the right crus preserving the peritoneal lining. We create a retro-esophageal window and utilize a Penrose drain for caudal and lateral traction. Finally, we mobilize the fundus by entering into the lesser sac. We take down the short gastric arteries with a combination of energy and hemostatic clips. We routinely extend our mediastinal circumferential esophageal dissection to the level of the inferior pulmonary veins, providing great exposure to the cystic mass. Thereafter, a myotomy is performed with blunt spreading and low energy. The myotomy is extended at least two centimeters proximally and distally to the cyst. Thereafter the cyst is enucleated with great care to avoid mucosal entry (*Figures 5,6*). Intraoperative evaluation with endoscopy and water and air tight test should be performed. A loose re-approximation of the myotomy is performed with 3.0 barbed absorbable sutures. A posterior cruroplasty with zero non absorbable barbed sutures is performed (*Figure 7*). If there is an associate large anterior defect, then an anterior cruroplasty is also performed. Finally, we perform a Dor fundoplication as it has been previously described (18) (*Figure 8*). If inadvertent mucosal entry were to happen and recognized intraoperatively, primary repair with 3.0 absorbable barbed sutures or interrupted PDS suture can be performed. We routinely perform an esophagram on post operative day #1, then begin patients on a liquid diet with discharge home within 48 hours of the index operation. Their diet is advanced at the follow-up appointment 1 week later.

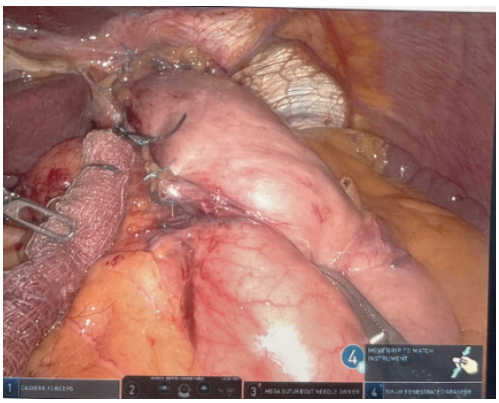

Figure 8 Anterior (Dor) fundoplication.

Summary

Esophageal duplication cysts of the esophagus are infrequent and mostly asymptomatic. However, they can cause complications later in life and shared decision making with patients regarding surgical excision is indicated. Historically open posterolateral thoracotomy has been described as the traditional approach to mediastinal cyst. Despite the adequate visualization and reach that this approach provides, it has been associated with significant post-operative pain and prolonged length of stay in the hospital (19). More recently, a minimally invasive technique including thoracoscopic and laparoscopic approaches have been shown to be safe and successful. At our institution we prefer a robotic assisted approach. The robotic platform not only provides with a three-dimensional view and digital imaging, but also increases dexterity particularly while dissecting and suturing in a reduced space like the hiatus. Moreover, the robotic platform provides the operating surgeon with unparalleled autonomy. One of the greatest advantages that this platform provides is the capacity to control your own camera and obtain the best angle of visualization during the entire dissection. This translates in reduce risk of mucosal injury, improving surgical outcomes (20).

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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