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SURGICAL SYMPOSIUM CONTRIBUTION

Achalasia and Epiphrenic Diverticulum

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Abstract Epiphrenic diverticula are a rare disease almost always associated with an underlying motility disorder of the esophagus, such as achalasia. Treatment of any underlying motility disorder must be included in the management of epiphrenic diverticula to prevent postoperative complications and recurrences. Therefore, the goal of this paper is to describe the pathophysiology, clinical presentation, and proper methods of diagnosis and treatment of patients with epiphrenic diverticula. In addition, we aim to provide an overview of the surgical management and discuss the indications for surgery and choice of surgical approach. In general, surgical intervention is favored for symptomatic patients and the optimal surgical approach depends on the size and location of the diverticulum. Surgery is not without seemingly high rates of morbidity when a myotomy is not performed together with the diverticulectomy, even in those with normal manometry. The risk of carcinoma is exceedingly rare and it is usually discovered at later stages; therefore, no surveillance programs have been established in asymptomatic patients with unresected diverticula.

Introduction

Epiphrenic diverticula are pulsion diverticula in which the mucosa and submucosa herniate through the muscular layers in the distal 10 cm of the esophagus [1]. Early treatment of epiphrenic diverticula included resection of the diverticulum with primary closure of the esophagus. However, Belsey and Effler suggested in the '60s that the diverticulum was due to an underlying esophageal motility disorder and proposed that a myotomy be performed together with the diverticulectomy [2, 3]. Today, the pathophysiologic link between the presence of an esophageal motility disorder and the epiphrenic diverticula has been well documented. In fact, numerous studies have shown that the vast majority of patients (75–100 %) with epiphrenic diverticula have achalasia or another esophageal motility disorder such as diffuse esophageal spasm or a nutcracker esophagus [4–7]. These findings have then suggested that such esophageal motor disorders may cause a contractile discoordination between the distal esophagus and the lower esophageal sphincter. Over time, this discoordination could lead to increased intraluminal pressure in the distal esophagus and the development of an outpouching of its mucosal and submucosal layers. Failure to realize the pathophysiologic association between the presence of the diverticula and an underlying motility disorder of the esophagus and failure to include the treatment of the motility disorder into the management of epiphrenic diverticula sets up the stage for dire postoperative complications. By being constantly reminiscent of the pathophysiologic basis of the genesis of

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the diverticula, we therefore aim to describe the clinical presentation and proper methods of diagnosis, and to discuss indications for surgery, choice of surgical approach, and results of thoracoscopic and laparoscopic approaches.

Clinical presentation

As many as 40 % of patients can be asymptomatic and their epiphrenic diverticula are found incidentally [8]. Symptomatic patients commonly complain of dysphagia, regurgitation of undigested food, chest pain, heartburn, nocturnal aspiration, aspiration pneumonia, and in severe cases, weight loss [8, 9]. Because the etiology of the diverticulum is often the underlying motility disorder of the esophagus, most symptoms such as dysphagia, regurgitation, and chest pain may be due to the motility disorder rather than the diverticulum itself [9]. This might be the reason why the size of the diverticulum does not seem to correlate with the severity of symptoms experienced by the patient [9]. Similarly, regurgitation of undigested food, nocturnal aspiration, and aspiration pneumonia, which may be due to the motor discoordination of the esophageal motility disorder, might also be suggestive of a symptomatic diverticulum, but again, no correlation between the size of the diverticulum and the severity of these symptoms has been demonstrated. In addition, when the diverticulum becomes large enough, it may cause dysphagia with resultant weight loss by extrinsic compression of the distal esophagus.

While the vast majority of esophageal diverticula are benign, malignant transformation from chronic inflammation—likely due to stasis and fermentation of food inside the diverticulum—it rarely occurs and may be demonstrated by the worsening regurgitation or odynophagia and hematemesis or hemoptysis [10]. Patients presenting with esophageal carcinoma from their diverticular disease present at late stages and therefore no surveillance program has been established in asymptomatic patients with unresected diverticula. Patients who develop cancer from an epiphrenic diverticulum are typically over 60 years of age, male, have large diverticula, and have endured an extended duration of symptoms [10]. The risk of carcinoma, however, is exceedingly rare. Herbella et al., have estimated that the incidence of cancer from epiphrenic diverticula is 0.6 %, with the majority of patients suffering from squamous cell carcinoma over adenocarcinoma [11].

Diagnostic testing

The diagnostic workup includes barium esophagogram, upper endoscopy, and esophageal manometry [1].

Barium esophagogram is typically the first diagnostic test performed. Not only are the findings diagnostic, but also a contrast esophagogram can provide useful information for surgical planning, including the location of the diverticulum (left or right chest and distance from the diaphragmatic hiatus), diameter of its pouch, as well as the length and width of its neck [1]. A barium esophagogram can also show any abnormalities of the gastroesophageal junction, such as hiatal hernias or lesions suspicious for a malignant process. Furthermore, disordered contractions of the distal esophagus, such as a bird's beak from achalasia, a corkscrew esophagus from diffuse esophageal spasm, or pathologic tertiary contractions might also be seen on esophagogram, which can prove useful in addition to the information gathered from esophageal manometry.

Upper endoscopy is used to evaluate the presence of mucosal lesions within a large diverticulum and to search for any additional pathology in the upper gastrointestinal tract, such as esophageal and gastric ulcers, Barrett's esophagus, or esophagitis, which may overlap the clinical presentation. The advantage of performing an upper endoscopy after the contrast study of the esophagus, when possible, is that the presence of the esophageal diverticulum detected on barium esophagogram may alert the provider performing the endoscopy and to avoid blindly intubating and perforating the diverticulum.

Esophageal manometry is usually performed to identify and confirm the presence of an underlying motility disorder. Some may argue, however, that manometry has only an academic role, as its results would not alter the patient's management, should one assume that almost, if not all, epiphrenic diverticula are caused by an underlying esophageal motility disorder [1]. Yet, some argue that the documentation of any existing esophageal dysmotility is fundamental to determine with certainty any underlying motility disorder. Although the identification of the esophageal dysmotility is very important and reassuring about the treatment plan proposed to the patient, normal manometry results should not be used to influence the surgical management [1]. In fact, in a few cases, due to the episodic nature of some motility disorders (or the inability of conventional manometry to detect subtle but important motor disorders of the esophagus), normal manometry results do not necessarily exclude the presence of dysmotility.

Indications for surgery

Most patients with epiphrenic diverticula are asymptomatic. When dysphagia and regurgitation are mild and infrequent and respiratory complications are absent, surgical treatment is generally not indicated [8]. Treatment

of epiphrenic diverticula is usually reserved for symptomatic patients who complain of invalidating dysphagia and regurgitation, or for those who have had episodes of aspiration from large diverticula [8]. The size of the diverticulum is not an indication for surgery per se, although spontaneous rupture has been documented in very few patients with large diverticula [12]. Patient selection is paramount because surgical treatment of patients with epiphrenic diverticula carries a significant morbidity mainly due to leak from the staple line after the diverticulectomy. Zaninotto et al. compared the outcomes of 22 patients with epiphrenic diverticula (median follow-up of 53 months) with those of 19 patients who were managed non-operatively (median follow-up of 46 months)—only 3/19 patients received esophageal dilatations—and found that none of the patients died for reasons related to their diverticulum and that symptoms improved in all operated patients and, to a lesser extent, also in all non-operated patients [12]. However, four patients complained of new-onset heartburn and regurgitation with esophagitis and/or positive pH-monitoring and three patients had persistent dysphagia or regurgitation and were dissatisfied with the results of the operation. Zaninotto et al. concluded that surgery is an effective treatment but that a conservative management can be safely adopted in patients with minimal symptoms and small epiphrenic diverticula [12].

Choice of surgical approach

The management of epiphrenic diverticula requires addressing the underlying motility disorder with a cardiomyotomy accompanied by a partial fundoplication to prevent postoperative reflux, and addressing the diverticulum.

The treatment of an underlying motility disorder such as achalasia has been well codified [13]. The length of the cardiomyotomy and the choice of fundoplication have been extensively studied. The cardiomyotomy usually extends for 2–3 cm onto the gastric wall [14]. A fundoplication is always added to prevent postoperative reflux, because when this step is omitted, the incidence of reflux is 48 %, versus 9.5 % when a Dor fundoplication is added to the myotomy [15]. As far as the type of partial fundoplication, a Dor or a Toupet fundoplication work equally well to relieve dysphagia and to provide control of postoperative reflux [16]. Conversely, a Nissen fundoplication is contraindicated [17]. In addition, the current recommendations from the Society of American Gastrointestinal and Endoscopic Surgeons advocate only a partial fundoplication—the specific type is left to the surgeon's preference—to prevent reflux [18].

If the treatment of the motility disorder underlying the epiphrenic diverticula has been well codified, the appropriate method to address the diverticulum itself is still unclear. Allaix et al. analyzed the outcomes of 13 patients with achalasia and epiphrenic diverticula who underwent laparoscopic myotomy and Dor fundoplication: 6 of which underwent also a diverticulectomy, whereas in 7 patients the diverticulum was left in place because it was too small (3 patients) or for technical reasons (4 patients) [19]. Allaix et al. found that all patients, even those who underwent a myotomy without diverticulectomy, had resolution of their symptoms. Allaix et al. then challenged the notion that all diverticula need to be excised, especially the small ones, and argued that the underlying motility disorder rather than the diverticulum, independent of its size, may be responsible for the symptoms experienced by the patients and therefore it should be addressed regardless of the diverticulectomy [19].

Up until the 1990s, the transthoracic approach through a right thoracotomy (most diverticula arise from the right side of the esophagus) was the standard of care. This approach ensured optimal visualization and access to the distal esophagus and provided the best exposure for the resection of the diverticulum and for oversewing the esophageal musculature over the staple line after the diverticulectomy, and allowed a contralateral distal esophageal cardiomyotomy. However, a right thoracotomy did not allow the addition of a partial fundoplication to control postoperative reflux after the cardiomyotomy.

With advances in minimally invasive operative techniques, laparoscopy has also become a reasonable alternative to open surgery, and it is now considered the approach of choice in most cases [7, 20–26]. The advantages of laparoscopic approach are related to avoiding a thoracotomy, which is a source of significant pain postoperatively as well as discomfort associated with the chest tube. A thoracoscopic approach can also prolong hospital stay and requires intubation with a double lumen endotracheal tube or bronchial block by the anesthesiologist, as it requires one-lung ventilation [20, 23, 24]. Other advantages of the laparoscopic approach include an easier application of the endostapler to transect the diverticula—the endostapler needs in fact to be applied longitudinally, along the major axis of the esophagus—and greater ease in performing both the cardiomyotomy onto the stomach wall and a partial fundoplication. However, these advantages may be of limited application in patients with larger diverticula, a long distance between the neck of the diverticulum and the hiatus (usually about 10 cm), and the presence of dense adhesions between the diverticulum and the adjacent mediastinal structures, making the dissection, application of the stapler, and approximation of the muscle layers more difficult

laparoscopically [1, 20, 23, 24]. In these circumstances, video-assisted thoracoscopic surgery (VATS) may be more appropriately the approach of choice [21].

Results of thoracoscopic and laparoscopic approaches

The most common complication from either surgical approach is leakage from the staple line after diverticulectomy, with resultant severe complications including sepsis, pneumonia, empyema, and abscess formation. Performing an appropriate myotomy is crucial to obtain resolution of symptoms when an esophageal motor disorder is identified and to eliminate the risk of a leak. When the diverticulectomy is performed without a myotomy, the staple line is subjected to the same motor discoordination that caused the pulsion diverticula initially. To be effective, the esophageal myotomy should be made contralateral to the diverticulum and should extend 5–8 cm above the gastroesophageal junction and not less than 3 cm below the gastroesophageal junction, onto the anterior gastric wall. Vagal nerve injury or transection can also occur, particularly with aggressive mediastinal dissection.

Currently, there are no studies comparing the outcomes of laparoscopic and thoracoscopic approaches, and given the limited number of cases and the variety of surgical techniques and measured outcomes, it is difficult to make a quantitative conclusion about the superiority of one procedure over the other. The results of laparoscopic and thoracoscopic operations for epiphrenic diverticula are summarized in Tables 1 and 2 [27]. These data show that the incidence of complications is low, mortality rates range from 0 to 10 %, which are comparable with those of open approaches and morbidity rates are similar between the two approaches, ranging from 0 to 33 %. Therefore, both laparoscopic and thoracoscopic treatment strategies have been shown to be very effective surgical modalities, each one having its own advantages and disadvantages and clear indications.

Conclusions

Esophageal diverticula are almost always due an esophageal motility disorder, such as achalasia. Treatment must aim to address such esophageal motility disorder in addition to a diverticulectomy in most cases. In general, surgical intervention is indicated for symptomatic patients depending on the size and location of the diverticulum, but not without seemingly high rates of morbidity, when the proper techniques are not utilized. The risk of carcinoma is exceedingly rare and it is usually discovered at later stages and no surveillance program has been established in asymptomatic patients with unresected diverticula.

Table 1 Results of VATS for the treatment of esophageal diverticula

Authors (year)	N	Side	Procedures	Months of follow-up (median)	Mortality N	Complications N (%)	Good outcome[a] (%)
Peracchia et al. (1994)	8	Right	Diverticulectomy = 3 (converted to open surgery = 2) Diverticulectomy with preoperative pneumatic dilatation = 5	–	0	Overall = 0	83
van de Peet et al. (2001)	5	Right	Diverticulectomy = 3 (converted to laparoscopy = 1) Diverticulectomy with myotomy = 2	–	0	Overall = 1 (20 %) Leak with abscess/sepsis = 1	–
Champion (2003)	3	Left	Diverticulectomy = 2 Myotomy = unknown Fundoplication = unknown	–	0	Overall	–
Matthews et al. (2003)	1	Right	Diverticulectomy with myotomy = 1	16	0	Overall = 0	–
Fernando et al. (2005)	9	Right	Diverticulectomy = 2 Diverticulectomy with myotomy = 4 Diverticulectomy, myotomy with fundoplication = 2 (combined with laparoscopy) Other = 1	15	0	Overall Leak = 2	–

N Number of patients

[a] Good outcome = significant improvement or resolution of symptoms

Table 2 Results of laparoscopic treatment of esophageal diverticula

Authors (year)	N	Procedures	Months of follow-up (mean/median)	Mortality N (%)	Complications N (%)	Good outcome[a] (%)
Klaus et al. (2003)	10	Diverticulectomy with myotomy = 6 Diverticulectomy = 4	26.4 (mean)	0	Overall = 2 (20 %) Empyema = 1 Leak = 1	–
Fraiji et al. (2003)	6	Diverticulectomy, myotomy with fundoplication = 6	9.3 (mean)	0	Overall = 2 (33 %) Empyema = 1 Ileus = 1	100
Del Genio et al. (2004)	13	Diverticulectomy, myotomy with fundoplication = 13	58 (mean)	1 (8 %)	Overall = 4 (30 %) Leak = 3 Myocardial infarction = 1	100
Tedesco et al. (2005)	7	Diverticulectomy, myotomy with fundoplication = 7	60 (median)	0	Overall = 1 (14 %) Leak with paraesophageal hernia = 1	100
Fernando et al. (2005)	10	Diverticulectomy, myotomy with fundoplication = 10	15 (median)	1 (10 %)	Overall - Leak = 2	–
Zaninotto et al. (2008)	17	Diverticulectomy, myotomy with fundoplication = 14 Diverticulectomy with fundoplication = 3	53 (median)	0	Overall	–
Melman et al. (2009)	13	Diverticulectomy, myotomy with fundoplication = 13	13.6 (mean)	0	Overall = 2 (15 %) Atelectasis = 1 Leak = 1	85
Rosati et al. (2011)	20	Diverticulectomy, myotomy with fundoplication = 20	52 (median)	0	Overall = 1 (5 %) Leak = 1	100
Soares et al. (2011)	18	Diverticulectomy, myotomy with fundoplication = 16 Diverticulectomy with excision of leiomyoma = 1 Diverticulectomy, myotomy with fundoplication and Roux en Y Gastric bypass = 1	45 (median)	1 (5.6 %)	Overall = 5 (28 %) Bleeding = 1 Leak = 1 Pleural effusion = 2 Port site hernia = 1	86

N Number of patients

[a] Good outcome = significant improvement or resolution of symptoms

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