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SURGICAL SYMPOSIUM CONTRIBUTION

Achalasia and Epiphrenic Diverticulum

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Abstract Esophageal epiphrenic diverticulum (EED) is a pseudodiverticulum of pulsion type located in the distal 10 cm of the esophagus and frequently associated to achalasia. The symptoms and the pathophysiology of achalasia and EED may overlap, leading to the speculation that achalasia may be responsible for the symptoms. Similarly to patients with achalasia without EED, a careful preoperative evaluation is essential in patients with EED. Endoscopy and an esophagram are mandatory in the workup of these patients, while esophageal manometry confirms the associated motility disorder. Treatment is indicated in all patients fit for an operation except those who are asymptomatic with a small EED and no prior history of aspiration. Laparoscopic Heller's myotomy and partial fundoplication is the most adequate therapy. Diverticulectomy must be added to the procedure in large diverticula. Experience with endoscopic therapy is very limited.

Introduction

Achalasia has been associated with diverticular formation in 3.6–7.4 % of cases [1], and achalasia is a concomitant motility disorder in over 60 % of the esophageal epiphrenic diverticulum (EED) [2]. EED found in patients with achalasia is truly a pseudodiverticulum (it lacks the muscular layer) of pulsion type located in the distal 10 cm of the esophagus.

Because of the rarity of EED, it is difficult to extract evidence-based recommendations; however, the management of EED in the setting of achalasia must be certainly tailored to its clinical presentation and pathophysiology.

Clinical presentation and pathophysiology

The symptoms of achalasia and EED may overlap. Both in achalasia [3] and EED [4], the most common complaints are dysphagia and regurgitation. Asymptomatic EED has been reported in over 60 % of cases as an incidental finding [5]. These facts lead to the rational that symptoms may originate in the associated motility disorder—not from the EED per se—found in the majority, if not in all patients [6].

The fear of 2 EED complications may be disquieting, leading some authors to treat asymptomatic patients with EED: (a) risk of cancer and (b) risk of aspiration. The risk of cancer has been reviewed recently [7] and found to be negligible (0.6 %) with old age, male gender, long-standing history, and larger diverticula identified as risk factors. Achalasia does not seem to raise the chance of cancer in

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the EED. Pulmonary symptoms are present in 25–50 % of the patients in EED series [4, 8]; however, the risk of aspiration from food impacted in the diverticulum is difficult to assess as achalasia is also an infrequently recognized cause for aspiration [9]. Achalasia therapy in patients without EED, either endoscopic balloon dilatation [10] or Heller's myotomy [11], seems to ameliorate respiratory symptoms and aspiration.

The pathophysiology of EED is based on intraluminal pressure due to an associated motility disorder with abnormal or absent peristalsis and inability of the lower esophageal sphincter to relax appropriately in response to swallowing. As previously said, a motility disorder has been found in 35–100 % of cases with a predominance of achalasia [6].

Clinical evaluation

Similarly to patients with achalasia without EED, a careful evaluation is essential in patients with EED.

Upper digestive endoscopy

Endoscopy may diagnose the EED (Fig. 1). This test is also useful to rule out malignancy and associated diseases.

Esophagram

The esophagram (barium swallow) provides details on size, shape, neck, location, and distance from the gastroesophageal junction (Fig. 2) [12]. It may assess the degree of esophageal dilatation and eventual associated diseases.

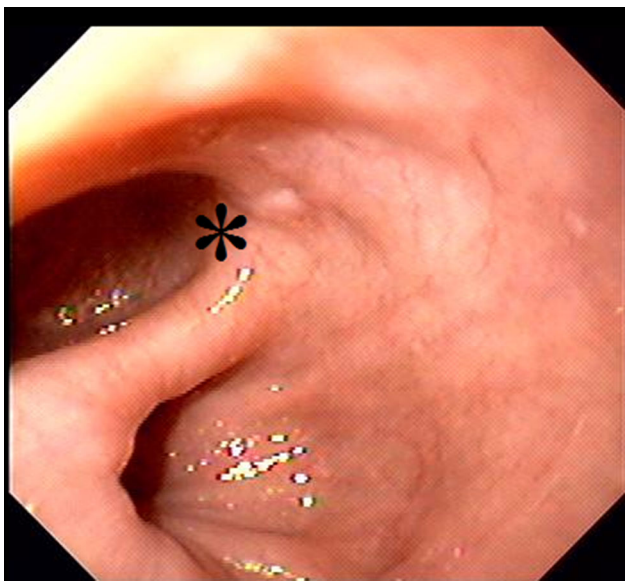

Fig. 1 Endoscopic view of esophageal epiphrenic diverticula in a patient with achalasia. Note the proximity of the diverticulum (*) from the esophagogastric junction. (courtesy Dr. Ramiro Colleoni)

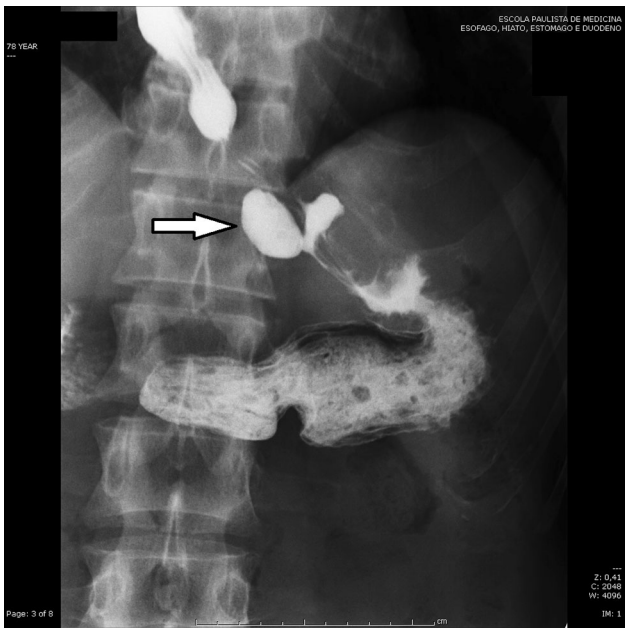

Fig. 2 Esophagram view of esophageal epiphrenic diverticula in a patient with achalasia (*arrow*) (courtesy Dr. Priscila Salvadori)

EED is usually located in the right side. Curiously, it is the opposite site of spontaneous rupture of the esophagus (Boerhaave syndrome) where a weak point has been demonstrated at the margin of contact between clasp and oblique fibers on the left side of cadavers [13].

Esophageal function tests

Esophageal manometry is the gold standard for the diagnosis of achalasia; however, for those authors who believe that a motor disorder is always present, esophageal manometry is not performed routinely other than for research or academic purposes (Fig. 3). In addition, sometimes the catheter must be placed through the gastroesophageal junction either under radiologic or endoscopic guidance.

Ambulatory pH monitoring is seldom used since gastroesophageal reflux disease is rarely associated to EED.

Indications for treatment

The indications for the treatment of EED are not unanimous. While some authors believe that only symptomatic and large diverticula must be treated [12], others believe that the risk for aspiration prompts treatment [14]. In the setting of achalasia, the association of EED with a primary motility disorders with a well-defined therapy must be

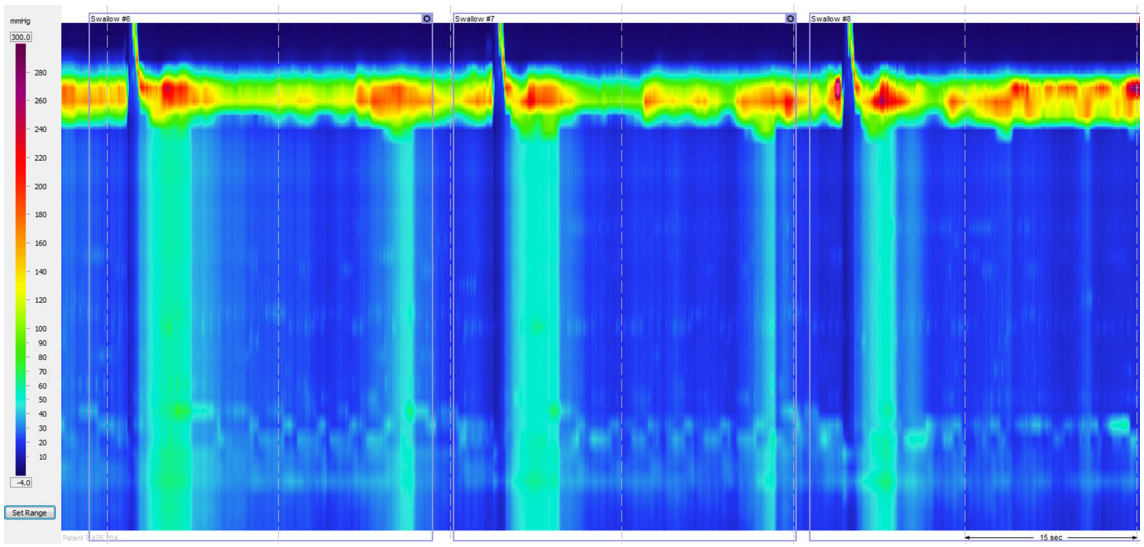

Fig. 3 High-resolution manometry plots of a patient with achalasia and esophageal epiphrenic diverticula

considered for the decision making. The literature omits the management of asymptomatic patients with achalasia. We believe that the clinical scenario of the absence of symptoms in patients without prior history of aspiration and a non-dilated esophagus obviates the need for treatment. Therefore, while in patients with a small EED treatment can be precluded, all others should be treated.

Treatment

The motility disorder

Laparoscopic Heller's myotomy is probably the best available therapy for achalasia, with long-lasting good outcomes with a success rate >90 % [15]. It is questionable if this procedure is adequate in dilated end-stage esophagi [16, 17]; however, EED is quite rare in dilated esophagi probably due to its atonic, non-spastic, nature.

Some technical points may be peculiar for the performance of a myotomy in patients with EED. First, if resection of the EED is not planned due to small size of the pouch, the myotomy must extend from the stomach upward all the way to the neck of the diverticulum. In most cases, the EED will simply vanish after the esophageal and gastric mucosa herniates through the muscular breach. Second, if a resection of the EED is planned, a myotomy on the opposite side of the pouch allows reinforcement closure of the muscular layer over the resection line.

A partial fundoplication should follow the myotomy, since the incidence of reflux is very high after myotomy alone [18]. A total fundoplication is associated to worse results for dysphagia relief [19]. An anterior or posterior partial fundoplication seems to provide similar control of reflux after the myotomy [20, 21].

Experience with endoscopic forceful balloon dilatation of the cardia is very limited.

The diverticulum

Dissection and treatment of the pouch access have traditionally been performed through a left thoracotomy [6]. Rosati et al., in 1998 started treating these patients through a laparoscopic approach [22]. Recently, most series opt for this via, as it allows good visualization of the distal esophagus and the EED and make the confection of a fundoplication easier [6]. The upper portion of the EED may be difficult to visualize and access; however, with proper downward traction and dissection of the adhesions, the upper pole can come to a comfortable level. The use of articulated staplers makes the procedure easier. It may be more difficult to approximate the muscle layers laparoscopically when the diverticular neck is high in the mediastinum [12].

As previously mentioned, EED has a neglectable risk for cancer and symptoms, and aspiration may be attributed to the underlying achalasia. These assumptions, added to the risk for complications, made some authors to hinder the addition of a diverticulectomy to the myotomy unnecessary [23].

Outcomes

Laparoscopic myotomy, diverticulectomy, and fundoplication bring symptom relief in 85–100 % of the cases with

a high rate of complications: leaks (8–23 %), pulmonary complications (8–10 %), and mortality ranging from 0 to 7 % [6, 24]. Laparoscopic approach seems to bring better results with lower morbidity and mortality, although most series deal with a small number of cases due to the rarity of the disease [12]. A significant series with 23 patients (21 laparoscopic) and 2 years follow-up showed a 92 % relief in dysphagia with a fistula index of 4 and 4 % mortality [25]. This can be compared to the open thoracic approach when excellent/good results were obtained in 76 % of 113 patients followed up for 7 years with a leak rate of 5 % and a mortality of 9 % [26]

Zaninotto et al. [27] reported symptomatic improvement in 2 patients unfit for surgery with achalasia and EED treated with endoscopic dilatation in a median follow-up of 46 months. There is no report of peroral endoscopic myotomy (POEM).

Little is known about the outcome of patients in whom a myotomy and fundoplication are performed without a diverticulectomy. Allaix et al. [23] reported good relief of symptoms in 7 patients at 11 months of follow-up.

Conclusions

EED is a rare disease that may complicate achalasia. The treatment of EED must be based on the physiopathology and clinical presentation of the disease. It is linked to a high index of morbidity, even in the hands of experienced esophageal surgeons. Achalasia should be treated via laparoscopic Heller's myotomy except in the absence of symptoms in patients without prior history of aspiration and a non-dilated esophagus. Small and medium EED can be left in place.

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