

## CASE REPORTS

# Epiphrenic and middle esophageal diverticula: A rare cause of esophageal dysphagia. Esophageal high resolution manometry findings

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## INTRODUCTION

Esophageal diverticula (ED) constitute a rare pathology, with a prevalence ranging between 0.06% and 4% (1,2). According to its anatomical location it is classified as pharyngo-esophageal, middle esophageal and epiphrenics ED (3). The pathophysiologic mechanism differs in diverticula caused by traction, due to an inflammatory process involving the esophagus, or pulsion, associated with a concomitant motility disorder (4-6).

## CASE REPORT

### Case report 1

A 58-year-old male, without relevant clinical antecedents, presented with progressive esophageal dysphagia in the last 3 months. Two years earlier, he had presented an episode of food impaction. Barium esophagogram (BE) detected a saccular image well delineated of 9 cm, with a hydro-aero level, in the lower esophageal third (Fig. 1A). Thoracic-abdominal computed tomography (CT) corroborated the findings described in the BE (Fig. 1B). The gastroscopy showed a dual lumen image large in size with semi-solid food content inside (Fig. 1C).

### Case report 2

A 78-years-old male patient with no clinical antecedents was forwarded from ear-nose-throat department for pharyngo-laryngeal foreign body sensation to rule out

GERD. The BE revealed the presence of a saccular image in the epiphrenic region (Fig. 1D), which was confirmed endoscopically.

### Case report 3

A 77-year-old male, with clinical antecedents of chronic hepatitis C and hiatal hernia, presenting with progressive esophageal dysphagia over the last 6 months associated with occasional episodes of food regurgitation and heartburn. The BE identified a 3 cm saccular image, located in the last 10 cm of the esophagus and a sliding hiatal hernia (Fig. 2A). Upper endoscopy confirmed the presence of a diverticulum containing food inside, located 30 cm from the incisors (Fig. 2B).

### Case report 4

A 68-year-old woman, diagnosed with systemic lupus erythematosus (SLE), presenting with progressive esophageal dysphagia over the last two months, with frequent episodes of food regurgitation and 10 kg weight loss. The thoracic CT showed esophageal lumen dilatation at the height of the carina, with a diameter of 5.2 x 3.1 cm and food content inside. The EB detected a saccular image of 4.5 x 3.7 cm in the middle esophagus that compressed the esophageal lumen diminishing the intra-luminal esophageal caliber with slowing of the barium pass at this location (Fig. 2C). An upper endoscopy identified a large esophageal diverticulum located 25 cm from the incisors with food inside. The esophageal biopsies were consistent with amyloidosis AA.

Received: 03-09-2014  
Accepted: 29-09-2014

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Ciriza-de-los-Ríos C, Canga-Rodríguez-Valcárcel F, Modesto Dutari-Valdés J, Castel-de-Lucas I. Epiphrenic and middle esophageal diverticula: a rare cause of esophageal dysphagia. Esophageal high resolution manometry findings. *Rev Esp Enferm Dig* 2015;107:316-321.

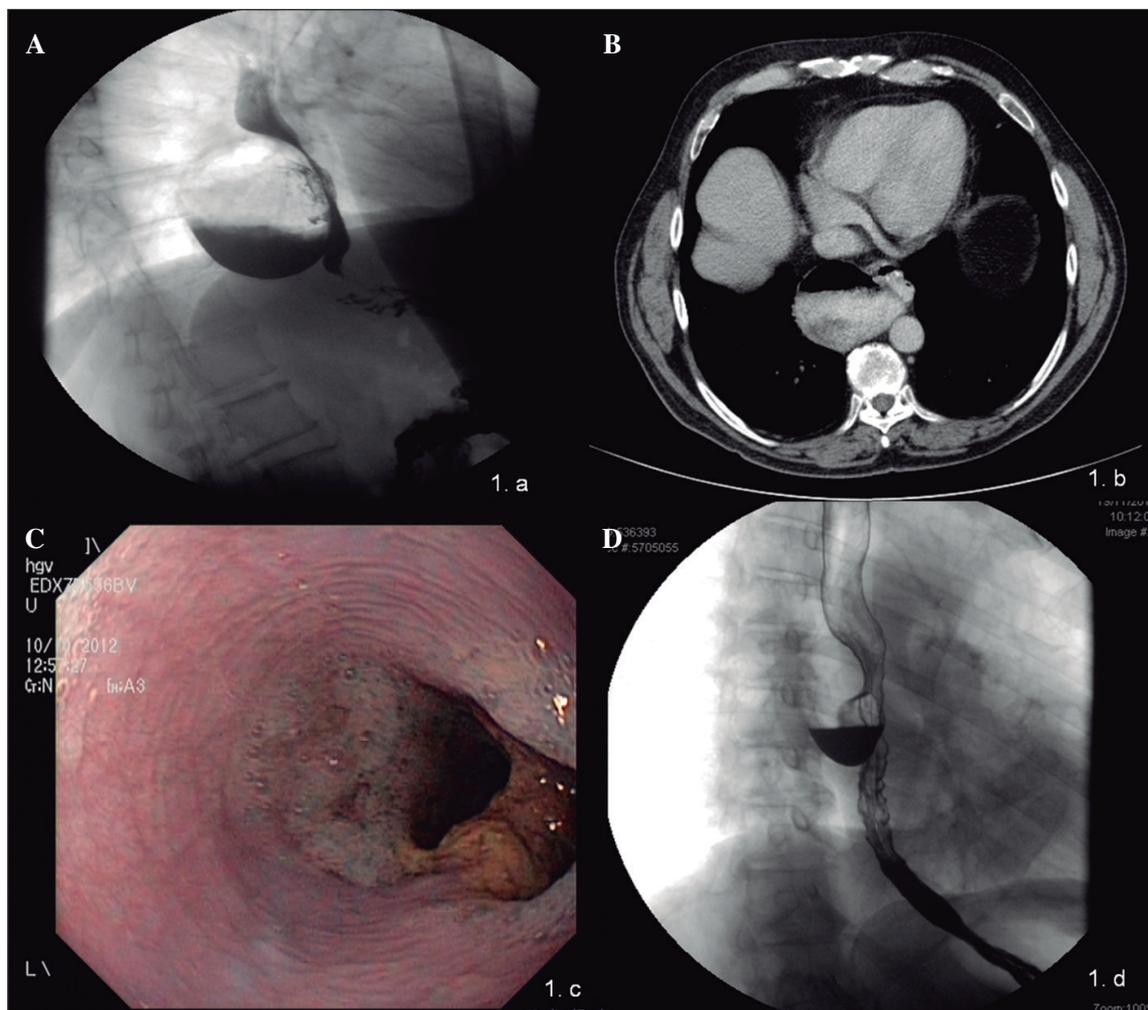


Fig. 1. A. Case 1. Saccular image of 9 cm, with a hydro-aero level in the epiphrenic region. B. Case 1. Large diverticulum dependent on the distal esophagus with mild proximal esophageal dilatation. C. Case 1. Endoscopic image of double lumen with food remains inside. D. Case 2. A saccular image filled with barium is observed adjacent to the wall of the distal esophagus.

### Case report 5

A 68-year-old woman with chronic renal failure antecedents presented with progressive esophageal dysphagia to solids and liquids over two years and bronchoaspiration episodes. EB detected tertiary contractile activity in the lower esophagus and an ED in the inferior and right posterior side of 2.9 x 2.8 cm of diameter (Fig. 3A) this was confirmed with endoscopic examination.

### Caso report 6

A 68-year-old woman with antecedents of total thyroidectomy for endothoracic multinodular goiter, pill induced esophagitis in 2011, ED (Fig. 3B) and small hiatal hernia. She presented with dysphagia, mainly to liquids that provoked her admission into hospital for bronchoaspiration on 3 occasions. In the last admission we confirmed the

ED located at 28 cm from the incisors by upper endoscopy (Fig. 3C).

The six patients described underwent a high-resolution manometry (HRM) (Manoscan; Given) that identified a concomitant esophageal motor disorder (EMD) in all cases. The manometric parameters for all of the patients are shown in table I; the reference values used were the current Chicago classification (7). The patient in the first case presented hypercontractile contractions at the esophageal segments S2 and S3, being compatible with the manometric diagnosis of hypercontractile esophagus ("jackhammer" type) (Fig. 4). The HRM of the second patient showed a distal esophageal spasm, with involvement at the level of the S3 segment (Fig. 5A). In the third case, the HRM revealed a EMD weak peristalsis with small breaks in the isobaric contour of 20 mmHg (50% of contractions); also the waves had a prolonged duration and morphology of double and triple peaks in the esophageal segment S3, in 56% and 11% of the swallows, respectively (Fig. 5B).

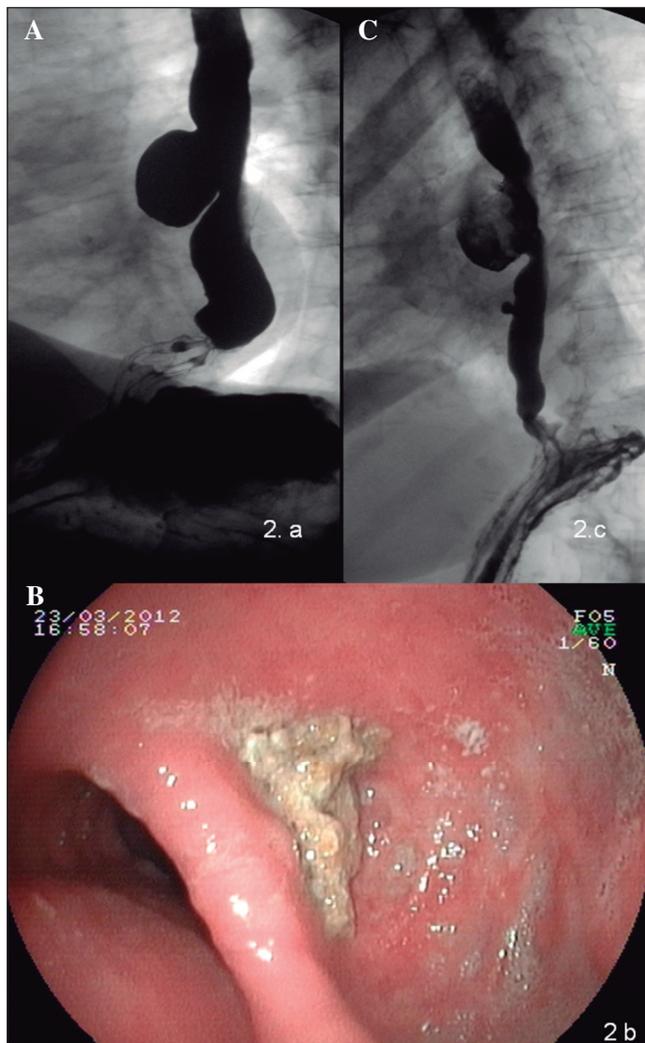


Fig. 2. A. Case 3. Lower third esophageal diverticulum. Sliding hiatal hernia is observed distally. B. Case 3. Food remains stuck on wide diverticulum located 32 cm from the incisors. C. Case 4. Middle esophageal diverticulum image on the posterolateral side. Another small diverticulum is observed caudal to the first.

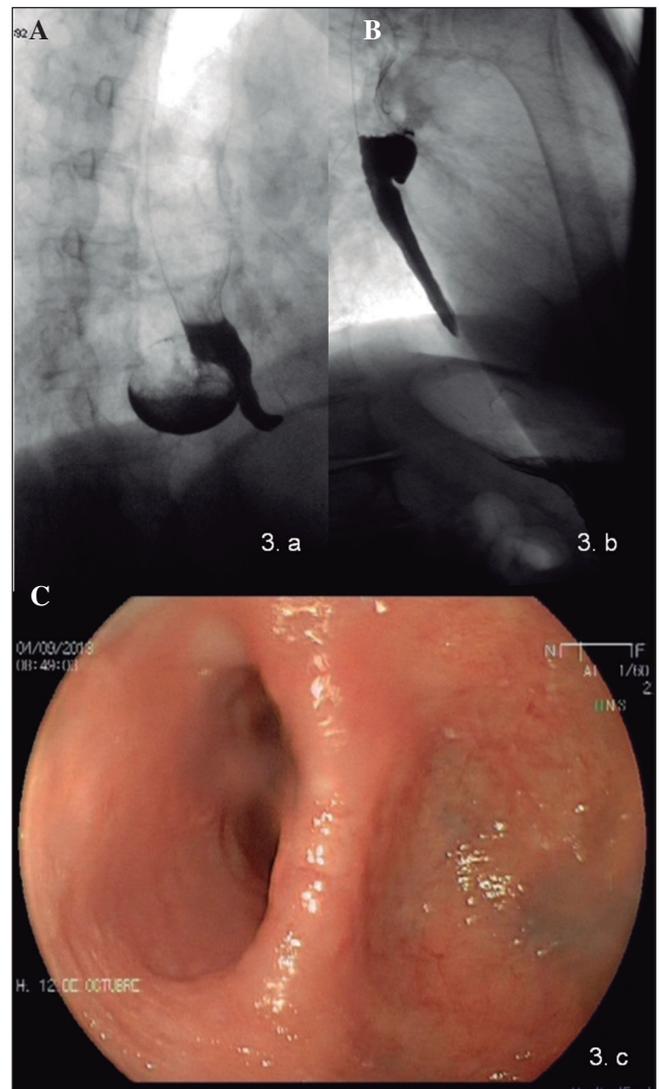


Fig. 3. A. Case 5. Lower esophageal diverticulum image located in the right posterior side. B. Case 6. Middle esophageal diverticulum. C. Case 6. Endoscopic image of the same diverticulum (figure 3B) located at 28 cm from incisors.

The patient in the 4<sup>th</sup> case had outflow obstruction of the esophagogastric junction (EGJ) with features of achalasia subtype III (spastic) of probable secondary origin (Fig. 6 A and B); also, great duration of esophageal waves were observed in the S2 segment (about 30 cm of the nostril).

The patient in the 5<sup>th</sup> case had a hypercontractile (“jackhammer”) motor disorder as well, with high amplitude waves in the segment S2 and particularly in S3 (Fig. 7). The 6<sup>th</sup> case patient, an EMD was observed consisting of outflow obstruction of the EGJ, as well as alteration in the contractile sequence in the anatomic area of the diverticulum where enlargement of the contraction was observed (between segments S2-S3) (Fig. 6 C and D).

The segment predominantly affected in the HRM, coincided with the location of the ED observed in the radiologi-

cal and endoscopic studies in all cases. Following the completion of the HRM, five of the six patients were referred for assessment for surgical treatment. The first case patient was operated by diverticulectomy that was complicated with esophagopleural fistula that was treated with endoscopic placement of a fully covered self expanding metal stent (WallFlex®).

The second and third case patients decided to continue a conservative management, after rejecting the option for surgical treatment due to the morbidity and mortality associated with it.

The fourth case patient presented an episode of upper gastrointestinal bleeding. A large clot attached to the wall of the diverticulum was detected in the urgent gastroscopy. This accelerated the decision for surgical treatment of the

**Table I. HRM findings in patients with esophageal diverticula**

Parameter	Normal value <sup>a</sup>	Case 1 (♂ 58 y) (epiphrenic diverticulum)	Case 2 (♂ 78 y) (epiphrenic diverticulum)	Case 3 (♂ 77 y) (epiphrenic diverticulum)	Case 4 (♂ 68 y) (middle esophageal diverticulum)	Case 5 (♂ 68 y) (epiphrenic diverticulum)	Case 6 (♂ 86 y) (epiphrenic diverticulum)
EGJ resting pressure (mmHg)	10-32	27.3	40.8	8.9	18.4	21.6	17.2
IRP-4s (s)	< 15	12	14	11,5	15	8,4	16,1
DCI (mmHg.cm.s)	< 5,000	7,103	2,770.8	906.8	3,103.5	8,059.3	1,993.8
CFV (cm/s)	< 9	5.2	4.4	4.9	6.8	5	3.9
Distal latency (s)	> 4.5	6	4.9	6	5	5.9	5.8
Intrabolus pressure (mmHg)	< 17	20.5	47.4	14.7	24	19.9	28.5
% Peristaltic waves	90-100%	100	80	90	20	100	90
% Failed waves	0-10%	0	0	0	0	0	0
% Failed waves with panpressurization	0-10%	0	0	10	80	0	0
% Premature	0-10%	0	20	0	20	0	0
% Small peristaltic defect	< 30	0	0	50	0	0	0
Manometric diagnosis		Hypercontractile (jackhammer)	Distal spasm	Weak peristalsis with small breaks	EGJ Outflow obstruction and achalasia type III (spastic)	Hypercontractile (jackhammer)	EGJ outflow obstruction

<sup>a</sup>Based on the Chicago classification 2012. EGJ: Esophagogastric junction; IRP-4s: Integrated residual pressure within 4 seconds; DCI: Distal contractile integral; CFV: Contractile front velocity.

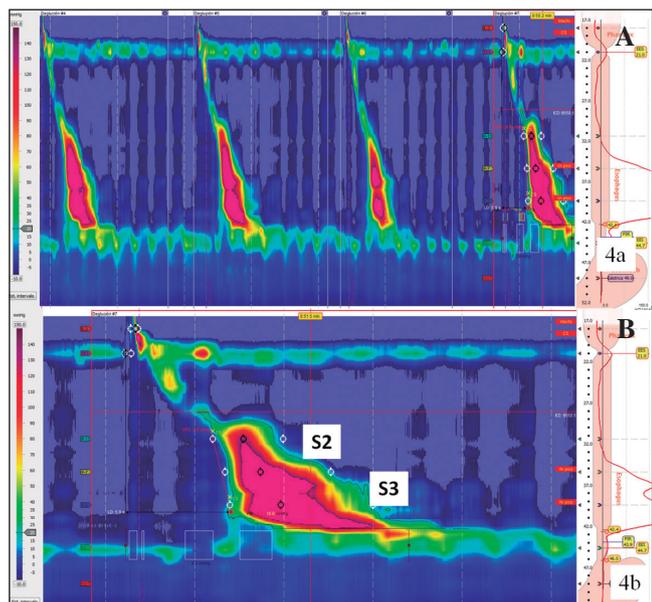


Fig. 4. A. Case 1. HRM topographic plot in which hypercontractile waves (DCI > 8,000 mmHg.cm.s) can be observed diagnosing for hypercontractile esophagus ("jackhammer"). B. Detailed liquid swallow in which the hypercontractile pattern is observed in segments S2 and S3.

diverticulum, once the episode of bleeding had surpassed. However, in the days after the bleeding event, the patient developed aspiration pneumonia and finally died.

The 5<sup>th</sup> case patient was treated surgically by performing thoroscopic diverticulectomy and myotomy. The postoperative was torpid, complicated with mediastinitis caused by esophagopleural fistula that was treated with endoscopic placement of a fully covered self expanding metal stent (WallFlex®).

The 6<sup>th</sup> case patient was treated with endoscopic injection of botulinum toxin at the level of the EGJ to solve the functional outflow obstruction. This treatment was considered the best option considering the advanced age of the patient, improving the symptoms.

## DISCUSSION

The majority of diverticula are asymptomatic and are incidentally diagnosed during the investigation of lesser related symptoms. In 25% of the cases, the ED can be symptomatic, dysphagia being one of the predominant manifestations (8), as a result of the dimensions of the

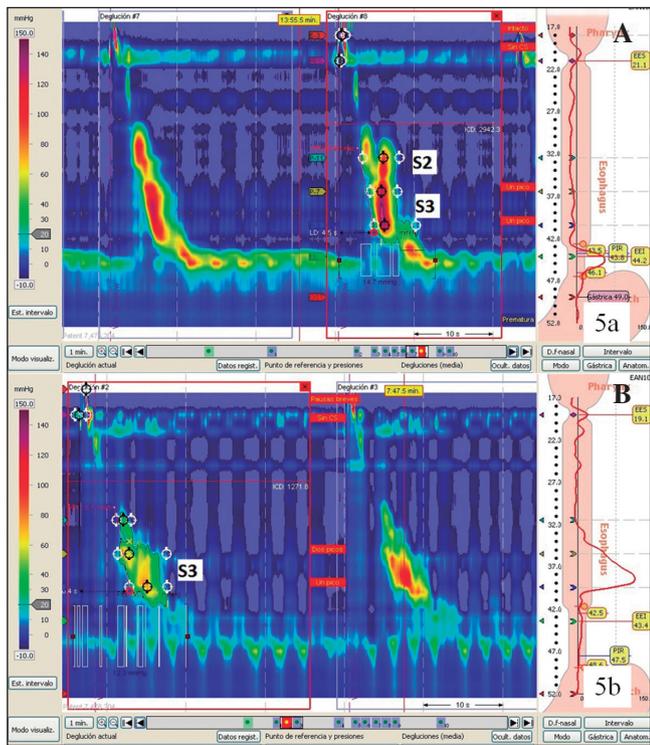


Fig. 5. A. Case 2. HRM topographic plot in which contractions with short distal latency (< 4.5 seconds) can be observed diagnosing for esophageal distal spasm in segments S2 and S3. B. Case 3. HRM topographic plot that shows contractions with peristaltic defects, large duration and double peak morphology in segment S3; also EGJ type III (hiatal hernia).

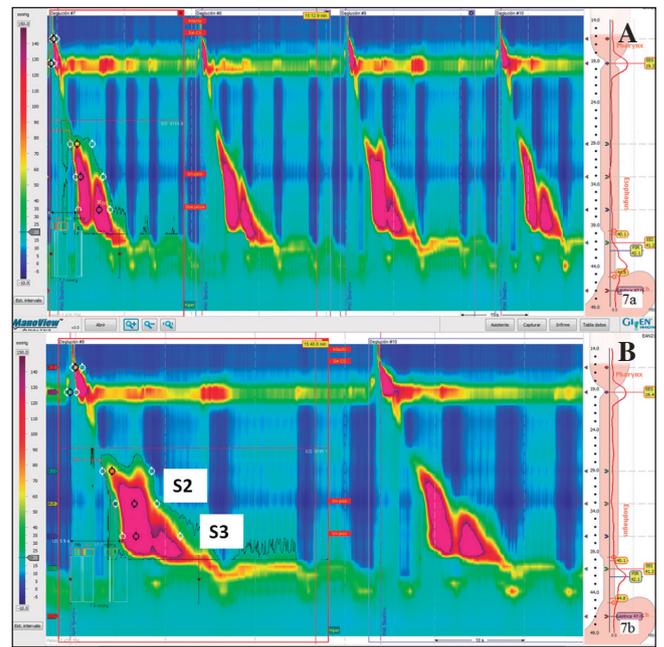


Fig. 7. A. Case 5. HRM topographic plot that shows hypercontractile contractions (DCI > 8,000 mmHg.cm.s) diagnosing for hypercontractile esophagus ("jackhammer"). B. Case 5. Detailed swallows in which the hypercontractile pattern can be observed in segment S2 and particularly in S3 with double peak morphology.

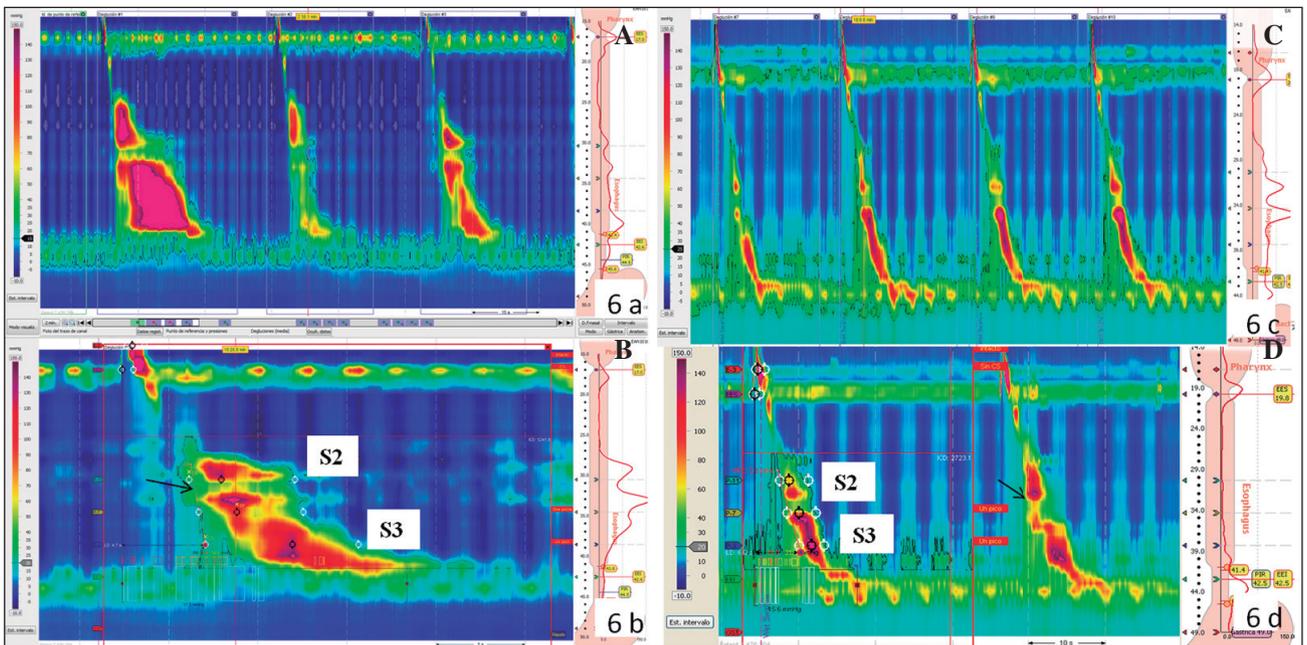


Fig. 6. A. Case 4. HRM topographic plot that shows simultaneous waves with short distal latency (< 4.5 seconds) with features of achalasia type III (spastic) and impaired EGJ relaxation (IRP > 15 mmHg). B. Case 4. Detailed liquid swallow in which alteration of the isobaric contour can be observed, consisting of prolonged duration in the segment S2 in the area of the diverticulum (arrow). C. Case 6. HRM topographic plot in which impaired EGJ relaxation can be observed diagnosing for EGJ outflow obstruction. D. Case 6. Detailed swallows in which alteration in segment S2 can be observed (arrow) similar to case 4, in the anatomic area of the diverticulum.

diverticulum and the concomitant esophageal motor disorder. The complications of ED, such as ulcers and bleeding, are infrequent (3). However, middle esophageal diverticula in particular, have a high prevalence of aspiration (45%), as well as a considerable potential for pulmonary complications that can be life threatening to the patients (15%) (9), as in one of our patients (case 4).

Epiphrenics and medium ED can be associated with a concomitant EMD in 35-100% (10) and 90% (11) cases, respectively. In the case of epiphrenic diverticula it was considered the protrusion of the mucosa and submucosa was caused by the increase in the esophageal pressure secondary to a motor disorder (8).

In our series of patients with ED a motor disorder in the esophageal body or in the EGJ was detected in all of them. Esophageal manometry is the gold standard for the diagnosis of the EMD (3). For many years, the conventional water-perfused manometry was the tool available to diagnose such disorders. However, in approximately 25% of the cases the EMD could pass unnoticed with this diagnostic technique. Today, the implementation of the HRM with the topographic plots obtained allows a higher accuracy in evaluation of the EGJ (12) and detection of segmental peristaltic defects, often unnoticed in the conventional study, though it obtains topographic plots of all of the esophagus by using a large number of sensors (4,13). In this respect, in our series of cases, we found pressure alterations in the anatomic area of the diverticulum, and we also identified the subjacent EMD, mainly spastic or hypercontractile, as well as alterations in the relaxation of the EGJ.

In conclusion, HRM clarifies the pathophysiology of the formation of the ED and can support the need to perform a myotomy added to the reconstructive surgical technique and its extension according to the EMD type and the affected esophageal segments as in the case of the 5<sup>th</sup>

patient, in which surgical diverticulectomy and myotomy were performed.

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