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Case report

Bronchopulmonary malformation of the foregut communicating with the distal Oesophagus in a form of an Epiphrenic diverticulum. An exceptional cause of epiphrenic diverticulum in adults: Case report

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ABSTRACT

Introduction: Epiphrenic diverticula are rare entities, generally asymptomatic. Most symptomatic ones occur in the form of dysphagia and are related to primary hypertensive oesophageal motor disorders of the distal oesophagus or the lower oesophageal sphincter. Exceptionally, they have been associated with congenital abnormalities in the formation of the primitive foregut.

Case presentation: An 85-year-old male patient with symptoms of dysphagia was diagnosed with epiphrenic diverticulum. Functional studies revealed associated oesophageal motility disorders. A transhiatal laparoscopic approach was used to perform a diverticulectomy, myotomy, and partial anterior fundoplication. The histological study of the diverticulectomy specimen was compatible with extralobar pulmonary sequestration communicating with the distal oesophagus.

Discussion: Bronchopulmonary malformations are very rare entities. A complete preoperative study of an epiphrenic diverticulum facilitates a correct differential diagnosis and decision-making. Endoscopy with biopsies can help define its etiology, rule out malignancy, and even change the therapeutic approach. The surgical approach depends on the morphological characteristics of the diverticulum, the surgical technique to be performed, and the surgical team experience. Intraoperative endoscopic resources facilitate surgical manoeuvres and guarantee the safety of the procedure.

Conclusion: Bronchopulmonary malformation of the foregut communicating with the distal oesophagus in the form of an epiphrenic diverticulum is an extremely rare condition. However, it should be included in the differential diagnosis of an epiphrenic diverticulum. Transhiatal laparoscopic resection under intraoperative endoscopic control is safe and effective for treating this entity.

1. Introduction

Oesophageal diverticula are rare entities with an incidence of 0.06 to 4 %, with epiphrenic diverticula representing 15 % of the total. They remain asymptomatic in >90 % of cases. When they are symptomatic, the predominant symptoms include dysphagia, halitosis, heartburn and regurgitation. They may present complications such as pleural fistula, haemorrhage, perforation, and a greater risk with malignancy of the oesophageal epithelium due to chronic food retention. Up to 70–90 % of epiphrenic diverticulum are related to primary oesophageal motor disorders of the distal oesophagus or lower oesophageal sphincter, such as achalasia, hypertonia of the lower oesophageal sphincter or distal oesophageal spasm, and are considered pulsion diverticula. Exceptionally, they have been described in association with congenital alterations in the formation of the primitive foregut during embryological development [1–5]. This work has been published according to the SCARE criteria [6].

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2. Case presentation

We present the case of an 85-year-old male Caucasian European race patient with a medical history of hypertension, diabetes, and hypothyroidism. He was being treated with oral antidiabetic drugs, thyroid hormone supplements, and antihypertensive medication. The patient complained of dysphagia predominantly to solids for several months, with frequent heartburn not controlled with proton pump inhibitors, regurgitation of undigested food and weight loss of about 5 kg. The endoscopic study showed a large distal oesophageal diverticulum (Fig. 1). The study was complemented with a barium esophagogram and a thoracoabdominal tomography showing a large epiphrenic oesophageal diverticulum, 5 cm from the esophagogastric junction (Fig. 2). In addition, oesophageal functional studies revealed an associated oesophageal motor disorder (Fig. 3). The patient was scheduled for surgical resection of the diverticulum through a transhiatal laparoscopic approach, associating myotomy and anti-reflux surgery.

The procedure was performed in the antitrendelemburg position. After inserting the trocars into the peritoneal cavity, a liver retractor was used to expose the hiatal area. The diaphragmatic pillars and the esophagogastric junction were dissected. Anterior frenotomy was used to access the lower mediastinum and dissect the distal oesophagus. Fig. 4 shows the steps of the surgical procedure performed under endoscopic control, which ensures its suitability and safety, and confirms the location of the diverticulum, wholly adhered to the right pleura, which was opened to begin its dissection. The diverticular pouch was dissected from its partial pleural covering and mediastinal tissue until it became completely independent.

Once the procedure was finished, a right endothoracic drain was placed. Oral intake of liquids and crushed foods was initiated at 24 h. The endothoracic drain was removed on the second day, and the patient was discharged on the fifth postoperative day. One year after surgery, the patient has no dysphagia or reflux and has regained weight. The histopathological study of the diverticulectomy specimen reported lung tissue with remnants of pleural tissue and a bronquiolo, compatible with extralobar pulmonary sequestration communicating with the distal oesophagus (Fig. 5).

3. Discussion

As most epiphrenic diverticula are related to primary motor disorders, a preoperative assessment, including endoscopy, imaging studies, and oesophageal functional tests, is crucial to defining the etiology of the diverticulum and individualizing the treatment.

The histopathological presence of alveolar tissue in the diverticulectomy specimen indicates the diagnosis of a bronchopulmonary malformation during the development of the primitive foregut in its separation from the respiratory system. The literature describes different etiopathogenic hypotheses, definitions and classifications of congenital developmental abnormalities of the foregut [7–9]. The exact sequence of events leading to them is unknown. However, there is a broad spectrum of pulmonary developmental abnormalities depending on the gestational age and the level of the tracheobronchial tree in which the embryogenesis disorder occurs.

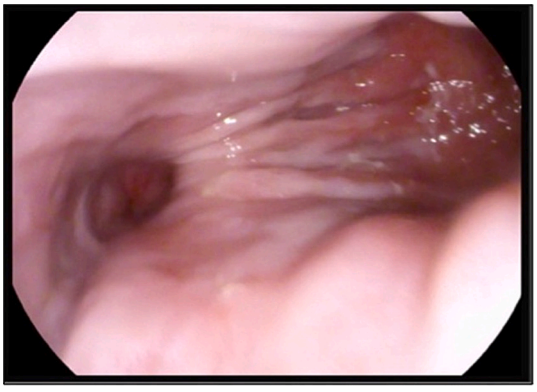

Fig. 1. Endoscopy. Narrowing of the oesophageal lumen secondary to a large diverticular cavity, with mucosal erosions.

Pulmonary sequestration is defined as a developmental abnormality characterized by non-functioning lung tissue without a regular connection to the tracheobronchial tree and an abnormal blood supply. It accounts for 1–6 % of all lung malformations. When it forms before pleural development, lung tissue develops adjacent to normal tissue, constituting an intralobar pulmonary sequestration, with vascularization and drainage through pulmonary branches. If it develops later, independent lung tissue will form, with partial or complete pleural lining and vascularization from the systemic circulation, constituting an extralobar pulmonary sequestration [10]. In the most extensive review on pulmonary sequestrations, published in China with 2625 cases, 83.95 % were intralobar sequestrations, and only 16.05 % were extralobar, with bilateral sequestrations being exceptionally rare. Extralobar sequestrations can be located at the supradiaphragmatic level (neck, mediastinum, pericardial), thus being misdiagnosed as pulmonary masses, bronchogenic cysts, or mediastinal tumours [11]. When located intra-abdominally, they can simulate gastric or oesophageal duplication cysts or even adrenal masses [12,13].

Foregut bronchopulmonary malformation is a rare variant of pulmonary sequestration in which the sequestered tissue is connected to the gastrointestinal tract. This connection can occur in both types of pulmonary sequestration [14]. When an isolated part of the lung communicates with the distal oesophagus, it is considered a group III malformation, according to Srikanth's classification.

The clinical presentation will depend on the level of communication between the respiratory and digestive systems. Asymptomatic forms, more frequent in adults, are discovered incidentally in imaging tests [11,15]. Symptomatic forms, more common in neonates or paediatric age, manifest as recurrent pneumonia, lung abscesses, chronic cough, expectoration, respiratory difficulty and haemoptysis [7–9]. The most severe malformation form is the oesophageal lung, characterized by a hypoplastic lung, communicating with the oesophagus through a rudimentary bronchus lined by respiratory epithelium, with pulmonary vascular supply. It is usually associated with other congenital oesophageal or cardiac anomalies [15,16]. The presentation as a diverticulum is much more exceptional, with only few isolated cases described in the literature [1–5].

The suspected diagnosis is clinical and radiological, defining the morphology of the fistulous tract. The diagnosis is confirmed by CT angiography and magnetic resonance angiography, showing the artery that supplies it and its venous drainage. The patient in our clinical case began to show symptoms of dysphagia at 85 years of age, associated with pathological reflux due to food stasis in the distal oesophagus and weight loss of 5 kg. Complementary examinations showed an epiphrenic pulsion diverticulum without associated masses or pulmonary fistulization. CT angiography was not performed to demonstrate the origin of the circulation of this malformation, since this entity was not suspected in the preoperative study. However, the complete dissection of the extralobar pulmonary sequestration from the lower mediastinum up to its communication with the distal oesophagus leads us to believe that the vascular supply and venous drainage came from the systemic circulation.

Malignancy of the epithelium of an epiphrenic diverticulum is rare (0.6 % of the cases) [17]. The percentage of malignancy of this type of malformation is unknown. Biopsies of the diverticular mucosa were not taken in our patient because malignancy was not suspected, which could have given us a clue to its origin. However, endoscopic follow-up of the diverticulum with biopsies should be performed periodically, even after resection, to control the remnant [18].

The indication for treatment of the diverticulum is based on

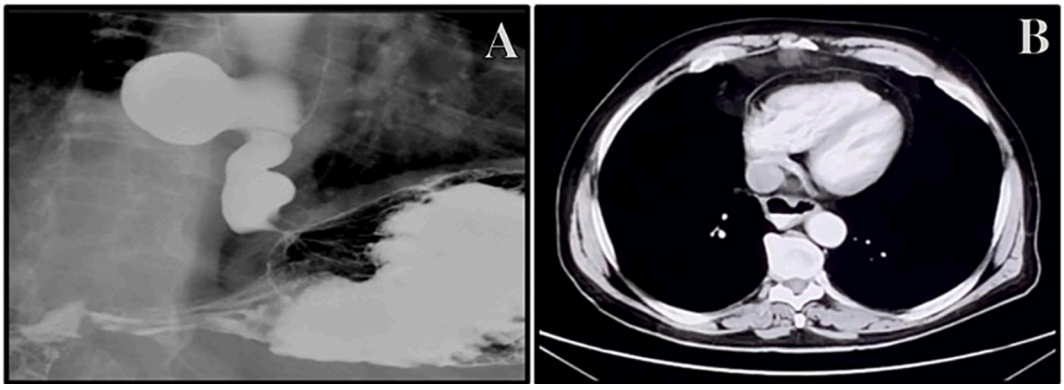

Fig. 2. A) Barium oesophagram: diverticulum of 4 cm to 5 cm from the esophagogastric junction. This causes significant diverticular-oesophageal reflux without associated stenosis. B) Thoracoabdominal tomography: distal oesophageal diverticulum without contrast defects or associated masses, possible oesophageal pulsion diverticulum.

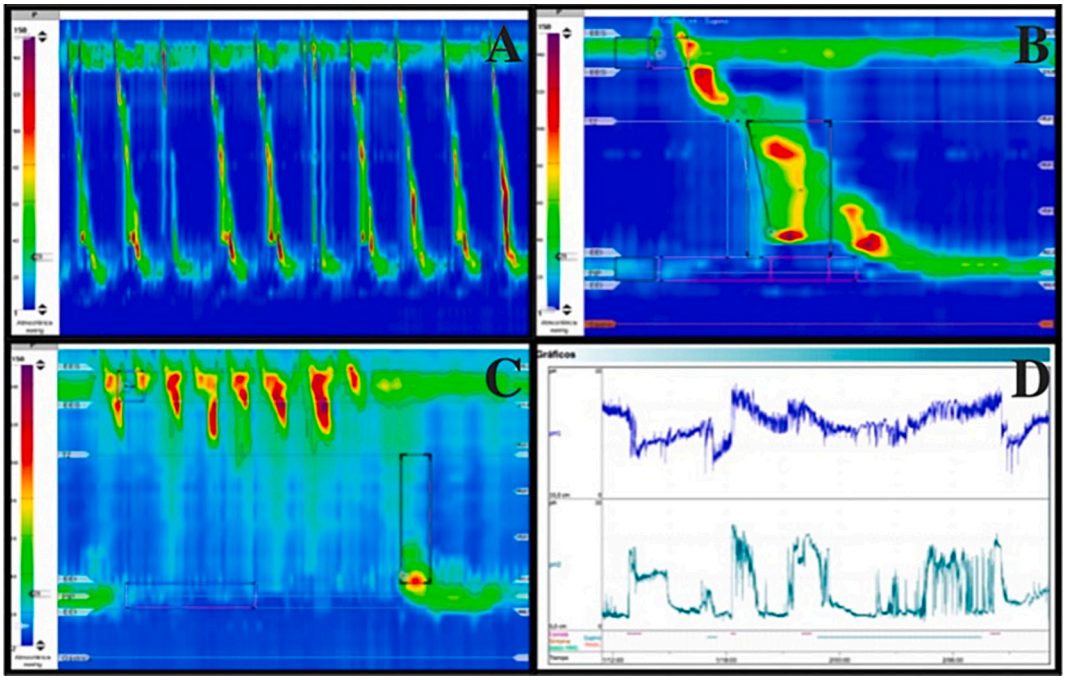

Fig. 3. High resolution oesophageal manometry. A) Pattern of fragmented peristalsis at the level of the epiphrenic ampulla. B) Intrabolus pressure area in each swallow. Short lower oesophageal sphincter with high pressure and adequate relaxation, but its hyperpressive etiology in dysphagia cannot be ruled out. C) Provocation test: Complete relaxation of the lower oesophageal sphincter. Poor oesophageal clearance due to oesophageal pressurization pattern related to food retained in the diverticulum, with hyperpressure of the lower oesophageal sphincter or both. D) Recording of oesophageal pHmetry (24 h) with dual oesophageal and gastric channels: Pathological postprandial oesophageal reflux (Acid Expoxure Time: 6 %, normal <4 %). There are 7 long episodes of pH < 4 in the oesophageal canal, when the gastric pH is not acid, due to poor oesophageal clearance secondary to food retained in the diverticulum.

symthoms rather than size. In our case, persistent dysphagia was the indication for surgery. There is no consensus regarding the most appropriate treatment for epiphrenic diverticula. However, due to the strong association with oesophageal functional alterations, it is recommended to perform diverticulectomy, myotomy from the base of the diverticulum to the stomach and associate a partial fundoplication. Peroral endoscopic myotomy (POEM) with or without associated diverticular septotomy (D-POEM) has up to 95 % success in symptom control. Its main advantage is that it allows the myotomy to be extended to the entire oesophagus. However, it is not considered the first-line treatment, as it does not include an effective anti-reflux technique [19].

The surgical approach choice will depend on factors such as the location and size of the diverticulum, the distance to the esophagogastric junction, the surgical technique to be performed, and the experience of the surgical team. We are familiar with the thoracic approach and have extensive experience in oncological laparoscopic esophagogastric surgery and extended mediastinal dissection to treat giant paraesophageal hernias. The location of the diverticulum about 5 cm from the esophagogastric junction and the existence of manometric alterations led us to opt for a laparoscopic approach. Diverticula located on the right posterolateral side of the oesophagus are accessible through laparoscopy, with a good angle of vision to introduce dissection instruments and endostaplers. Extended extramucosal myotomy including 2 cm of the stomach and reconstruction of the hiatus with a Dor-type anterior fundoplication is more feasible when this approach is used. We must emphasize the importance of the simultaneous endoscopic view to help identify the diverticulum, verify the complete section of the diverticular neck and guarantee the integrity of the oesophageal mucosa after the myotomy.

Finally, the pathological study was compatible with a

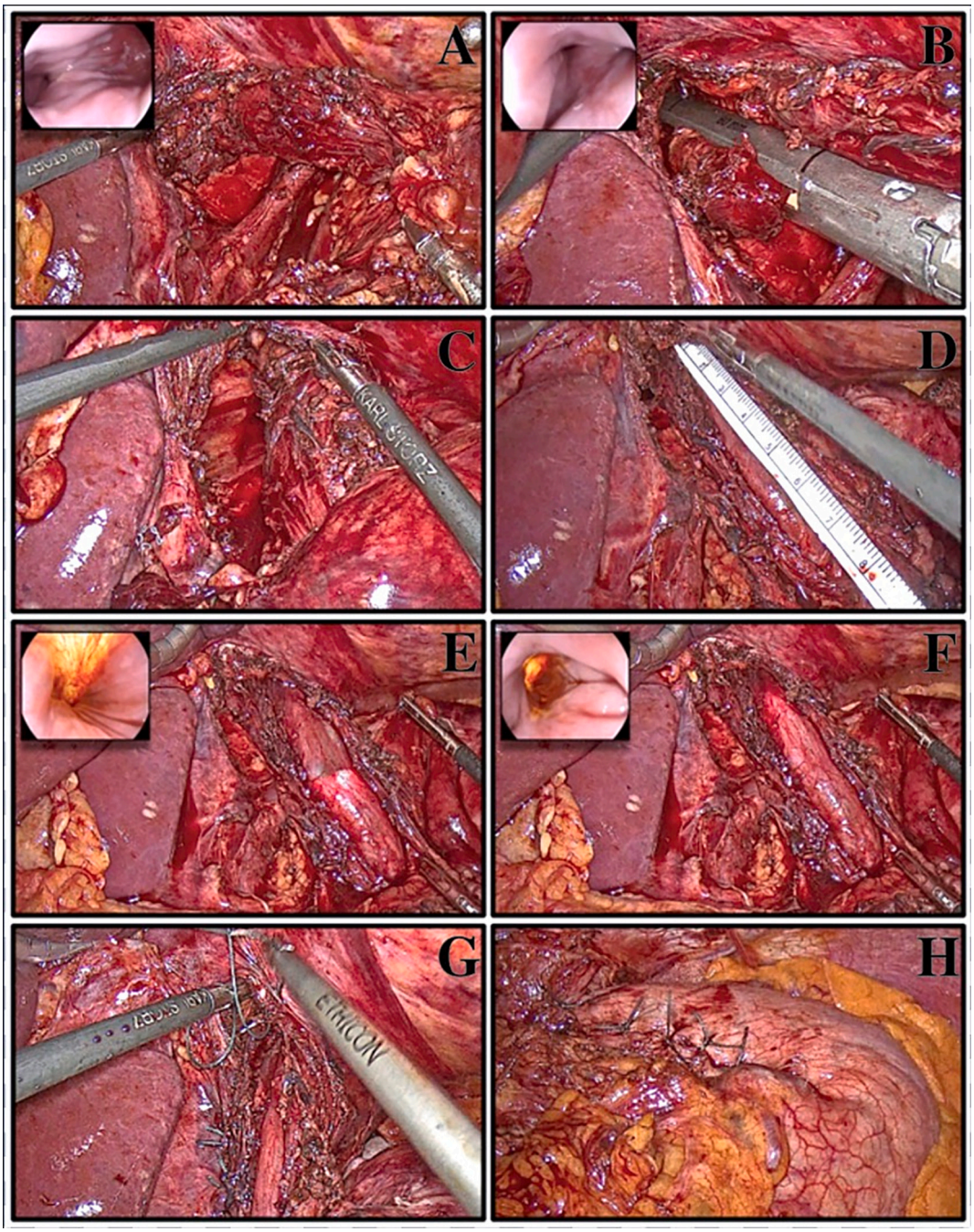

Fig. 4. Surgery with simultaneous endoscopic vision. A) Identification of the oesophageal diverticulum on the right posterolateral side. B) Section of the diverticulum with linear surgical stapler that collapses the diverticular lumen. C) Image of the oesophageal section line after diverticulectomy. D) Measurement of myotomy length (8 cm). E) Endoscopic verification of mucosal integrity after myotomy. F) Endoscopic verification of the complete section of the diverticular neck. G) Reconstruction of the hiatus with 0 polyester sutures. H) Dor type anterior partial fundoplication with 2/0 polyester sutures.

bronchopulmonary malformation communicating with the oesophagus (Srikanth group III) [9]. To the best of our knowledge, isolated cases of epiphrenic diverticulum composed of histological elements of the primordial airway have been published, and all were treated by thoracic approach [1–5]. This is the first case of bronchopulmonary malformation of the foregut communicating with the distal oesophagus in the form of an oesophageal diverticulum treated laparoscopically.

In conclusion, bronchopulmonary malformations of the foregut communicating with the distal oesophagus in the form of an epiphrenic diverticulum are extremely rare. Performing a complete preoperative study of epiphrenic diverticula is essential for a correct differential diagnosis and better decision-making. Endoscopy with biopsies of the diverticular mucosa may define its etiology, rule out malignancy, and even change the surgical approach. Transhiatal laparoscopic resection is safe and effective for the treatment of this entity. A combined minimally invasive thoracoabdominal approach may help extend the myotomy above the level of the diverticulum.

CRediT authorship contribution statement

MHB: original draft, collect data, writing the paper and editing of photography images.

AMR: original draft, critic review and editing the paper

GTR: critic review writing the paper

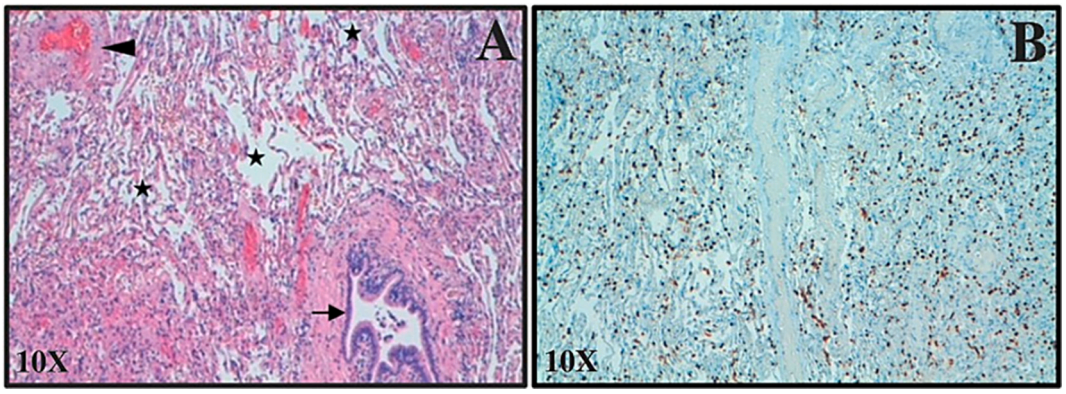

Fig. 5. Pathological anatomy. A) Lung tissue composed of alveolar spaces (star), bronchiole (arrow) and blood vessels (arrowhead) with remnants of pleural covering. HE staining technique. B) Identification of alveolar pneumocytes with nuclear staining (brown dots). TTF1 Immunohistochemistry Technique. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

RJT: parcial writing the paper and editing of some photography images

CMS: critic review writing the paper

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Ethical approval for this case report was given by the "Research Ethics Committee" at our institution.

Guarantor

MHB (first Author)

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This study did not receive any funding.

Declaration of competing interest

The authors declare that they have no competing interests.

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