

# Acute attack of recurrent esophageal duplication cyst in an adult: case report and literature review

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## Introduction

Esophageal duplication cyst (EDC) is an uncommon, usually asymptomatic congenital benign esophageal tumor that was first described by Blassium in 1711 (1). The etiology of EDC is a failure of vacuolization in the primitive foregut during the fourth to eighth week of embryonic development (2). It accounts for 10–15% of all alimentary tract duplications, but only 0.5–2.5% of esophageal tumors (3). Almost 80% of EDC becomes symptomatic and is discovered in early childhood, but in a few patients, EDC remains asymptomatic until adulthood (2). It can usually be cured by surgical resection (4,5), and only one recurrent case has been reported (6) (PubMed database). Furthermore, only five cases (Table 1) of acute onset of EDC in adults have been described in the literature (2,3,5,7,8) (PubMed database). Here, we describe a case of postoperative recurrent EDC accompanied by acute symptoms.

## Case presentation

A 54-year-old man was admitted to the emergency department of Beijing Friendship Hospital on November 14, 2014, with complaints of acute chest pain and severe dysphagia. He stated he had mild dysphagia for nearly 1 week, and then the dysphagia was suddenly aggravated by intermittent nausea and vomiting such that he could not eat and drink. We used morphine to help relieve the patient from persistent chest pain and parenteral alimentation to support nutrition. Past medical history showed previous surgical enucleation for EDC (Table 2) at the Beijing

Tongren Hospital in 1982.

On physical examination, a previous surgical incision scar of approximately 40 cm was observed on the posterior wall of the left chest. Blood test revealed a significantly elevated level of leukocytosis ( $10.9 \times 10^9/L$ ) and neutrophil percentage (87.2%). The other meaningful results were hemoglobin (113 g/L) and platelet ( $98 \times 10^9/L$ ) concentrations. A chest CT showed a  $9.1 \times 4.7 \times 17$  cm<sup>3</sup> soft tissue mass in the lower-middle esophagus associated with upper esophageal dilatation and air-fluid level. The main bronchi, pulmonary artery, and pulmonary vein were all compressed, and pneumonia with a small amount of right pleural effusion was present (Figure 1A,B,C,D). Endoscopic ultrasound (EUS) revealed hypoechoic lesions originating from the esophageal muscularis propria with unclear margins (Figure 1E,F).

Due to uncertain diagnosis and life-threatening symptoms, a right-side exploratory thoracotomy was undertaken, and a large cystic-solid mass with high tension located in the lower-middle esophagus was found. The lesions occupied nearly the entire length of the esophagus and were adhered to trachea, venae azygos, pericardium, and inferior pulmonary veins. We then made a small incision from the protruded part of cyst and aspirated approximately 200 mL of dark-brown clotted blood, which was immediately followed by a quick drop in the previously elevated tension. However, multiple cysts and inflammatory dense adhesions between the cyst wall and the esophageal muscle layer were found. Therefore, we performed a midline laparotomy and removed the diseased esophagus (Figure 2A), making a tubular gastric and gastroesophageal

**Table 1** Published case reports of acute esophageal duplication cyst in adult

Case	Reference	Year	Age	Sex	Symptoms	Complication	Location	Size (cm)	Treatment
1	Gatzinsky P <i>et al.</i> (7)	1978	34	M	Rapidly developed dysphagia, severe retrosternal pain	3-cm long rupture, massive bleeding	Right lower part of the esophagus	Grape-fruit size (after rupture)	Thoracotomy, esophagectomy
2	Nakahara K <i>et al.</i> (8)	1990	36	F	Acute dysphagia and dyspnea	Rupture	Middle mediastinum	8×8	Thoracotomy, enucleation
3	Neo EL <i>et al.</i> (5)	2004	32	M	Acute chest pain and dyspnea	Rupture	Right posteromedial hemithorax	12×9×8	Thoracotomy, enucleation
4	Pisello F <i>et al.</i> (3)	2009	46	M	Acute dysphagia	Hemorrhage	Posterior wall of the middle esophagus	6×8.5	Thoracotomy, enucleation
5	Wang D <i>et al.</i> (2)	2015	52	F	Rapidly progressing chest pain and dysphagia	Inflammatory dense adhesions	Lower part of the esophagus	10×6×4	Thoracotomy, esophagectomy

M, male; F, female.

**Table 2** Summary of the patient's clinical data

Year	Age	Symptoms	Primary check	Treatment	Cystic contents	Pathology	Hospital
1982	22	Chest pain	Barium examination showed a 3×3.5 cm <sup>2</sup> round-like mass on the right side of the esophageal wall at the location of 7–10 cm from the diaphragm	Left posterolateral incision, enucleation	White colloidal substances (10 mL)	Esophageal duplication cyst	Beijing Tongren Hospital
2014	54	Acute chest pain and severe dysphagia	Chest computed tomography showed a 9.1×4.7×17 cm <sup>3</sup> soft tissue mass in the lower-middle esophagus	Right posterolateral and abdominal median incision, esophagectomy	Dark-brownish blood clots (>200 mL)	Recurrent esophageal duplication cyst	Beijing Friendship Hospital

anastomosis at the top of the chest. Postoperative pathology confirmed the diagnosis of recurrent EDC (*Figure 2B,C*), which was consistent with the pathological diagnosis 32 years ago (*Figure 2D,E*).

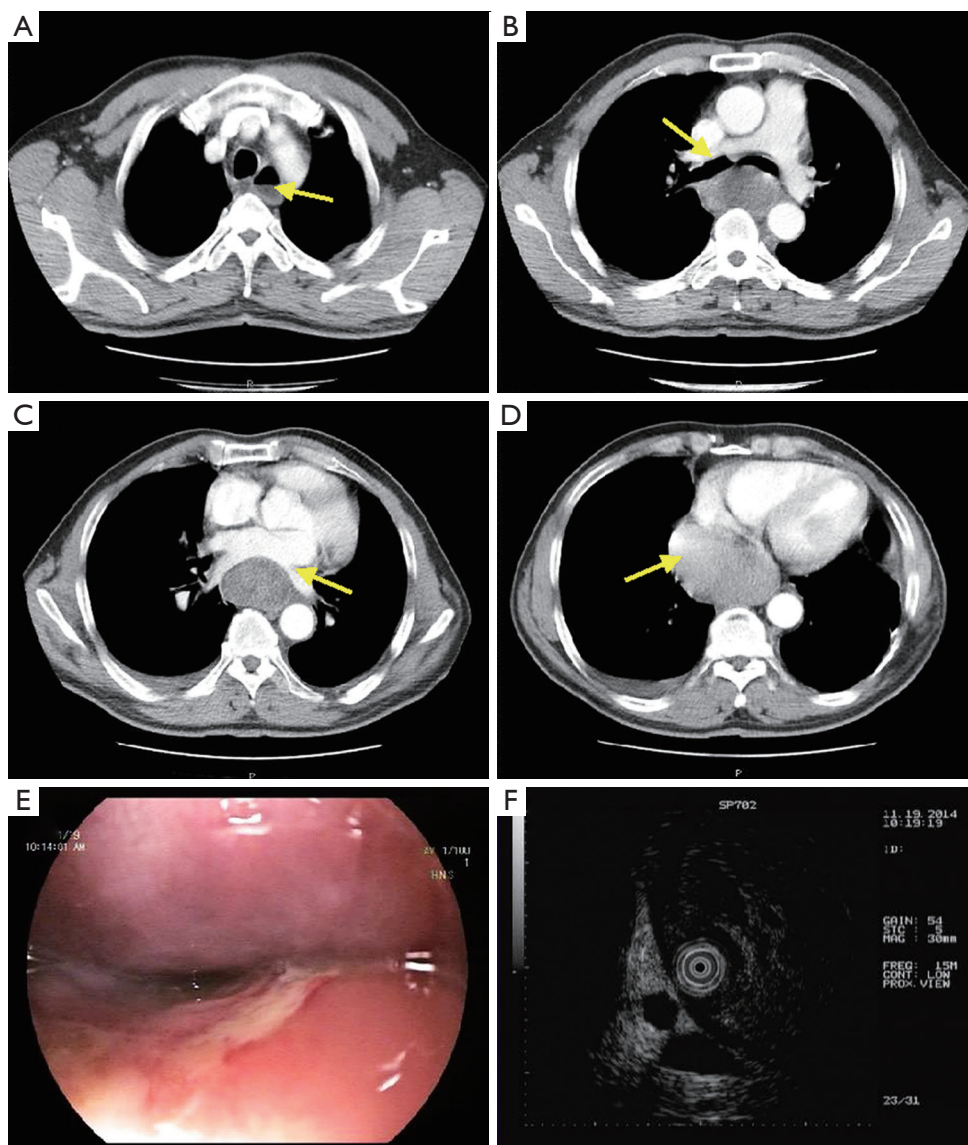
Postoperative barium esophagography showed no complications. The patient was discharged with an uneventful recovery on the 15th postoperative day. During the 36 months of follow-up, only slight acid reflux esophagitis was observed, and the symptoms improved after taking omeprazole (20 mg/d).

## Discussion

The predilection site of EDC is the right posteroinferior mediastinum or lower-middle esophagus (3). Most EDCs present in isolation, while some develop as multiple cysts (9). Cysts of different sizes and locations may cause various symptoms such as cough, recurrent pneumonitis, chest pain, dysphagia, and dyspnea (2,3,7). As the cysts continue

to develop, it might become acutely enlarged, leading to life-threatening complications such as rupture, obstruction, hemorrhage, and malignancy (2,3,5,7,8,10).

Preoperative diagnosis of EDC usually needs a combination of various imaging techniques. Chest roentgenology, barium examination, and chest CT are the basic examinations performed before EDC surgery. Magnetic resonance imaging (MRI) has high diagnostic value in identifying cystic or solid tumors, especially for cystic fluid. Endoscopic ultrasound (EUS) can effectively evaluate these lesions and show if the cysts are anechoic or hypoechoic. In EUS, thin-walled cystic structures are the most common sign of EDC (9). EUS-guided fine needle aspiration biopsy can offer a quick and accurate qualitative diagnosis, but it may increase the risk of future surgery (11). Histopathological diagnose of EDC should follow Palmer's three criteria (12): (I) adhere to or in the esophageal wall; (II) covered by gastrointestinal tract epithelium such as squamous, columnar, cuboid, pseudostratified, or ciliated



**Figure 1** Chest CT demonstrated a  $9.1 \times 4.7 \times 17 \text{ cm}^3$  soft tissue mass in the lower-middle esophagus (D) with upper esophageal dilatation and air-fluid level (A). The bronchi and pulmonary artery/vein (B and C) were compressed. EUS revealed hypoechoic lesions associated with unclear margins (E and F).

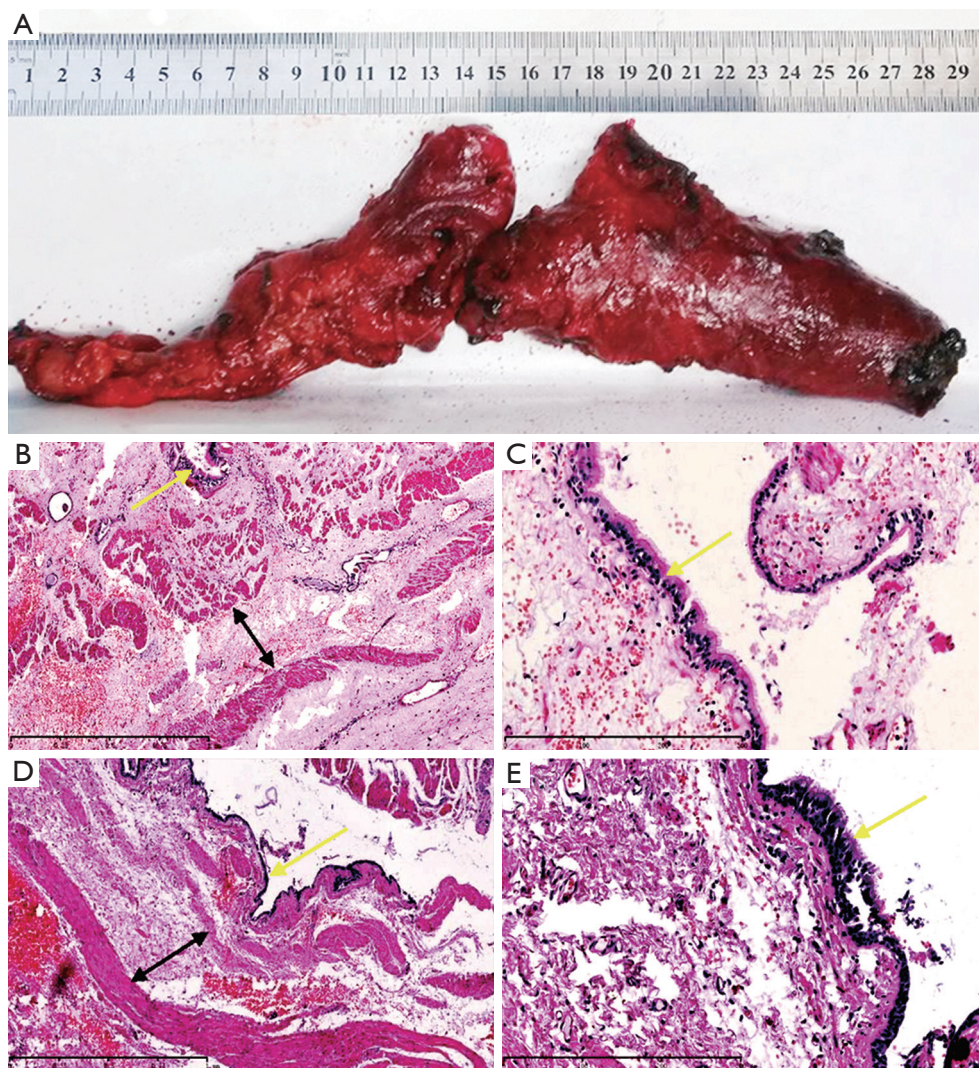
epithelium; and (III) double layer muscularis propria under epithelium.

As the preferred treatment for EDC, surgical resection has the advantages of definitive diagnosis and relieving existing symptoms and can prevent future serious complications. The surgical approach includes thoracotomy and minimally invasive surgery, such as video-assisted thoracoscopic surgery and endoscopic submucosal dissection (13). The reasonable surgical option should be

determined according to the size and location of the cyst and its relation to adjacent tissue and organs.

The preoperative diagnosis in this case was difficult because the conclusions drawn from the various inspections were equivocal. The final pathological diagnosis was recurrent EDC. We suspect the recurrence is attributed to failure of the initial surgery to completely remove the cyst itself or perhaps smaller cysts that might have been adjacent to the resected lesion. It is important to emphasize





**Figure 2** The resected specimen of the postoperative recurrent EDC (A) and pathology (B and C). The patient's first EDC pathology (D and E). (hematoxylin and eosin staining, B and D  $\times 50$ , C and E  $\times 200$ ). Both pathological examinations showed pseudostratified columnar ciliated epithelium (yellow arrow) with two layers of muscularis propria underneath (black arrow).

that an esophageal cyst is not a recurrent disease, such as a malignant tumor, but may have recurrent features when the surgical treatment is incomplete. In addition to the recurrence of EDC, another point of our report is the rapid progression of symptoms. Only five similar cases have been reported (*Table 1*). After reviewing these cases, we can conclude that acute EDC has the following features: (I) acute dysphagia and chest pain are the most common symptoms. Symptoms in the course of disease progression start from mild to acute exacerbation and then are suddenly alleviated due to rupture; (II) large size; (III) secretion or hemorrhage in a cyst, which may be the main

reason why preoperative diagnosis is difficult; (IV) severe complications; and (V) inflammatory adhesions. Given all these characteristics, thoracotomy is advised when treating acute EDC and complete resection should be emphasized during the operation to avoid neglecting the smaller cysts. Meanwhile, for non-ruptured high-tension EDC, fenestrating decompression can help reduce the degree of surgical difficulty.

In summary, after EDC surgery, particular consideration should be given to determining whether the case is one of recurrent EDC where esophageal tumors were rediscovered. Moreover, close follow-up should be emphasized.

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## Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

*Informed Consent:* Written informed consent was obtained from the patients for the publication of this manuscript and the accompanying image.

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