

# Carcinoma Arising from an Epiphrenic Diverticulum: A Frequently Misdiagnosed Disease

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**Epiphrenic diverticulum is rare, comprising of about 10% of all esophageal diverticula. Carcinoma arising within such a diverticulum is even less common. We report on two patients with squamous cell carcinoma arising from an epiphrenic diverticulum. Some features of malignant change related to the epiphrenic diverticula were misdiagnosed or missed in our cases and other reports due to lack of clinical experience. It is important to exclude the possibility of malignancy as this is vital for surgical planning. Although the prognosis for patients with malignancy arising from an epiphrenic diverticulum is generally poor, a high index of suspicion may increase the chance of cure. (Ann Thorac Cardiovasc Surg 2007; 13: 110–3)**

**Key words:** esophageal diverticulum, epiphrenic diverticulum, esophageal carcinoma, squamous cell carcinoma

## Introduction

Diverticulum of the lower esophagus, or epiphrenic diverticulum, usually occurs in the lower third or distal 10 cm of the esophagus and is caused by elevated intra-esophageal pressure. Less than 100 cases of esophageal carcinoma arising from or within an epiphrenic diverticulum have been reported in the English medical literature, mainly as case reports. These patients may present clinical symptoms and signs mimicking those associated with other distal esophageal motility disorders, including hiatal hernia, diffuse esophageal spasm, achalasia and reflux esophagitis and can be misdiagnosed even with imaging studies. We report two cases of esophageal carcinoma arising from an epiphrenic diverticulum and discuss the diagnostic challenge to clinicians.

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Received July 31, 2006; accepted for publication August 18, 2006.

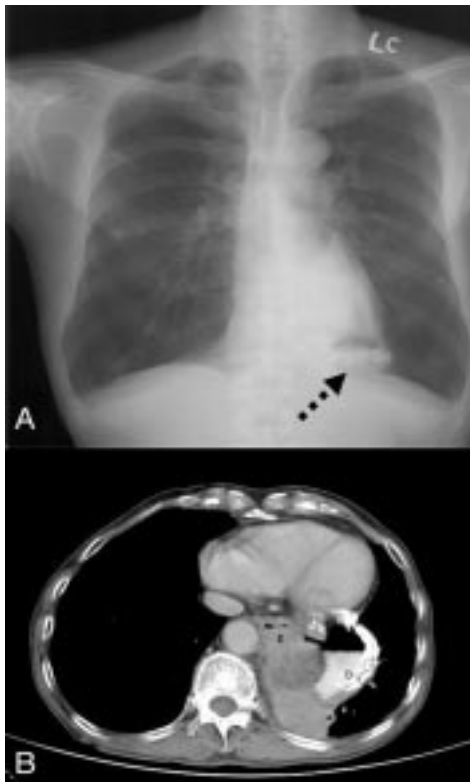
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## Case Reports

### Patient 1 (see Fig. 1, A and B)

A 70-year-old man with a history of smoking and alcohol consumption was admitted to our hospital after experiencing dysphagia and a 13 kg unintentional weight loss over three months. An upper gastrointestinal (UGI) endoscopy and computed tomography (CT) were performed which suggested a hiatal hernia with a gastric cardiac tumor inside. In addition, there was a mass lesion over the left lower lung field on the chest radiograph.

With a provisional diagnosis of gastric cardiac cancer within a sliding hiatal hernia with pulmonary invasion of the left lower lobe, we performed a left thoraco-abdominal incision which disclosed a large tumor arising within the diverticulum of the lower third of the esophagus with direct invasion of the left lower lung and mediastinum. A combined resection including esophagus and left lower lobe followed by retrosternal gastric tube reconstruction was performed. Pathology revealed an epiphrenic diverticulum (5×3×2 cm<sup>3</sup>) located just above the cardio-esophageal junction with moderately differentiated squamous cell carcinoma (8×7×6 cm<sup>3</sup>) extending into the wall of the esophagus with partially calcified capsules. In addition, a moderately differentiated squamous cell carcinoma invaded through the visceral pleura to the lung parenchyma.

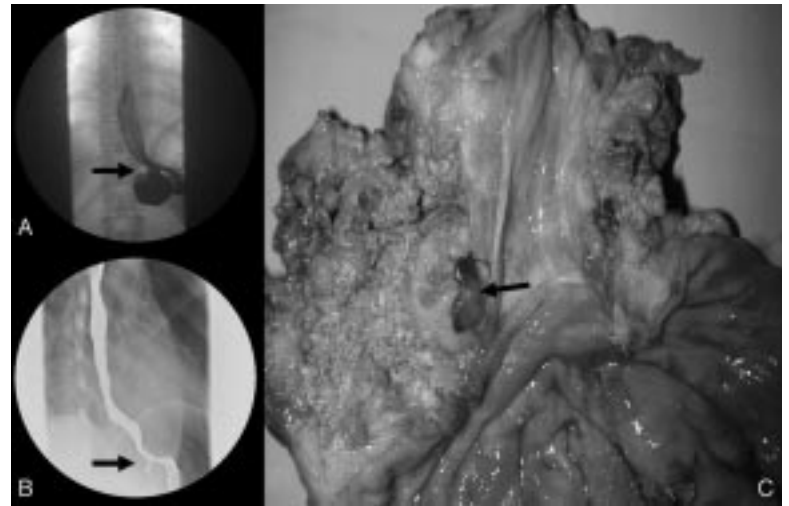


**Fig. 1.** Patient 1.

**A:** Chest roentgenogram shows a mass lesion with air-fluid level at the left retropericardial area (arrow).

**B:** CT section close to the diaphragm shows a diverticulum (D), with thickened and rough walls and an air-fluid-filled level, on the left side of the esophagus (E) connected with the esophagus through a narrow neck. An esophageal cancer arises from the diverticulum, invading through the left lower lung parenchyma.

Postoperatively, the patient developed respiratory failure due to pneumonia. This required prolonged ventilator support. A tracheostomy was performed on postoperative day 21. Subsequent bronchoscopy showed tumor growth at the middle portion of the left main bronchus with 80 percent obstruction on postoperative day 125. Follow-up mediastinal CT on postoperative day 162 further demonstrated a large necrotic tumor arising from the subcarinal region with extension to the left lower mediastinum and encroachment of the left pulmonary artery. The patient died on postoperative day 197 from respiratory failure due to total collapse of the remaining portion of the left lung.



**Fig. 2.** Patient 2.

**A:** Barium esophagram examination 2 years ago showing one diverticula on the right side of the lower esophagus.

**B:** Swallowing study before the current operation. The diverticulum orifice to the lower esophagus was opacified, but the diverticulum was not opacified.

**C:** Gross appearance of the 7×4.5×2 cm<sup>3</sup> resected tumor with large and irregular shape arising from the diverticulum, invading the adventitia and involving the E–C junction and the proximal cardia.

The neck of the diverticulum is indicated by arrows.

### **Patient 2 (see Fig. 2, A–C)**

A 34-year-old woman was admitted to our hospital because of progressive pyrosis and dysphagia. The patient had experienced intermittent abdominal fullness, postprandial emesis and foreign body sensation over the throat for 2 years. Three months prior to the admission, she had an esophagogastroscopy in another hospital, which defined a large esophageal diverticulum with a wide neck above the esophagogastric junction projecting into the right chest. A barium swallow examination in our hospital demonstrated a 4 cm epiphrenic diverticulum. Esophageal manometry study revealed a hypertensive low esophageal sphincter. She declined surgical intervention but received medical therapy.

Three months before the present admission, she presented with progressive pyrosis, left upper abdominal pain radiating to her back solid food dysphagia, regurgitation of undigested foods and a 15 kg unintentional weight loss. Endoscopy revealed the narrow orifice of the previous esophageal diverticulum at the right side and adherent food particles inside the diverticulum which allowed a thorough view inside. However, the endoscope could not pass the tight cardia. An upper GI barium study again disclosed a lower opacified orifice of the diverticulum.

With the diagnosis of simple epiphrenic diverticulum, we performed a left thoracotomy for planned lower esophageal myotomy and diverticulectomy. At exploration, a tumor mass was palpated at the base of the diverticulum that extended into the right chest, with local extension into the retroperitoneum and possible invasion to the inferior vena cava. A diagnosis of squamous cell carcinoma was confirmed by frozen section pathology. By extending the incision to the abdomen, palliative distal esophagectomy, proximal subtotal gastrectomy and intrathoracic esophagogastrectomy were performed through laparotomy and left thoracotomy. Pathology revealed an epiphrenic diverticulum with moderately differentiated squamous cell carcinoma ( $7 \times 4.5 \times 2 \text{ cm}^3$ ) extending into the wall of the esophagus, stomach, and surrounding soft tissue but the lymph nodes were tumor free. The patient's postoperative course was complicated by pneumonia delaying discharge until postoperative day 21 and she was referred for chemotherapy. Currently, she is still alive without disease recurrence 544 days after the surgery.

## Discussion

The diagnosis of epiphrenic diverticulum requires a high degree of suspicion regarding both the symptoms and imaging studies, including barium swallow and UGI endoscopy. The incidence of cancer arising within the esophageal diverticulum is between 0.3% and 3%.<sup>1)</sup> The pathogenesis of carcinomatous change in the esophageal diverticulum may be caused by chronic irritation by food, inflammation, repeated injury or hyperplasia of the epithelium within or close to the diverticulum.<sup>2)</sup> These tumors are usually diagnosed in the advanced stage because of difficulties in preoperative diagnosis.

It is estimated that 30–40% of epiphrenic diverticula are asymptomatic.<sup>3)</sup> The most common symptoms, include dysphagia, weight loss, and substernal pain, are related to the complications arising from the diverticulum, such as inflammation, perforation, hemorrhage, stricture formation and malignant changes within or around the diverticulum.<sup>4)</sup> In our second case, the initial symptoms of intermittent abdominal fullness, postprandial emesis and foreign body sensation over the throat for 2 years might simply be due to the epiphrenic diverticulum. However, such cases should alert clinicians to the consideration of other complications arising from the diverticula when progressive dysphagia and unintentional weight loss are noted.

In general, barium swallow provides clues to any associated motility disorder, which serves as a baseline if the patient is asymptomatic, and may detect other lesions, such as cancer or stricture causing symptoms. In the second case, we concluded that progressive reduction in the size of the epiphrenic diverticulum, filling defect or irregularity of stenosis of the lumen of the esophagus near the diverticulum seen in the esophagogram should not be attributed to retention of food but rather be investigated to rule out malignant change. In addition, esophagoscopy allows careful inspection and biopsy of the esophageal mucosa and should be performed to exclude malignancy.<sup>4)</sup> In both of our patients, the esophagoscopy was performed but did not provide the precise clinical diagnosis before surgery. In the first case, the esophageal cancer within the epiphrenic diverticulum was interpreted as a cardiac cancer within the sliding hiatal hernia. In the second case, biopsy was not performed because the tumor was concealed by food materials within the diverticulum.

Concerning computed tomography, it should be performed selectively for more precise definition of a mass lesion arising from a perforated or malignant diverticulum and plays an important role in surgical management. In a typical case, a pulsion diverticulum appears as a well-defined, thin-walled, air-filled structure with smooth internal and external surfaces and is sharply separated from surrounding structures.<sup>5)</sup> It may contain air-fluid level and there may be communication with the esophagus.<sup>5)</sup> However, while the inner and external surfaces of the epiphrenic diverticulum, as in our first case, may be thickened and become irregular due to malignant change, it is often misdiagnosed as a gastric cardiac cancer in the sliding hiatal hernia. In our second case, since the possibility of malignant change within the epiphrenic diverticulum was not considered, preoperative CT was not done. Despite such investigations, the presence of a pulsion diverticulum and related motility disorder may occasionally remain undiagnosed until surgical exploration.<sup>6)</sup>

The preoperative diagnosis considerably influences the surgical planning. If malignant change arising from the epiphrenic diverticulum can be confirmed preoperatively, the procedure of choice would be esophagogastrectomy, rather than simple diverticulectomy.<sup>7)</sup> However, there is no current data that irrefutably confirms one approach (transhiatal or transthoracic) for esophageal carcinoma over another. Schultz et al. assumed that carcinoma arising within an epiphrenic diverticulum behaves as other distal esophageal carcinomas.<sup>4)</sup> The traditional approach to the distal esophagus has been a left thoraco-abdominal

approach with an intrathoracic esophagogastric anastomosis. An alternative approach is the transhiatal esophagectomy with cervical esophagogastric anastomosis.<sup>4)</sup>

The prognosis for patients with carcinoma in the esophageal diverticulum is generally poor.<sup>8)</sup> At diagnosis, the cancer is usually advanced because most patients have long-standing symptoms related to the diverticulum that conceal the presence of malignancy. Additionally, the absence of the muscularis propria surrounding the diverticulum probably allows rapid invasion to the contiguous structures.<sup>1)</sup> The late stage of the disease increases the difficulty of surgical management. The epiphrenic diverticulum is considered analogous to the part of an iceberg that is visible above the water surface. One should always consider the possibility of associated problems instead of treating the epiphrenic diverticulum itself alone. What is needed is a detailed history taking to evaluate these patients and the rare possibility of malignant change should be diagnosed preoperatively.

## Conclusion

Carcinomas arising within an esophageal diverticulum are uncommon. The prognosis is usually poor due to locally advanced invasion of the tumor. High degree of suspicion leads to a correct preoperative diagnosis, and better surgical planning.

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