Case report

Barrett's adenocarcinoma associated with acquired eventration of the diaphragm

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Abstract:

We report a rare case of Barrett's adenocarcinoma associated with acquired eventration of the diaphragm in a 71-year-old woman. She initially developed dysphagia and epigastric discomfort in May, 1997. On July 9, she was referred to our Department of Surgery at the Ryukyus University Hospital for thorough examination and treatment. Esophageal adenocarcinoma and eventration of the diaphragm were revealed by exhaustive examinations, including chest X-ray, computed tomography, and magnetic resonance imaging, and proximal gastrectomy with reconstruction of jejunal interposition was performed, on August 8. Histologically, the tumor revealed that the adenocarcinoma arose from short-segment Barrett's esophagus (SSBE). It thus appears that eventration of the diaphragm may induce SSBE and Barrett's adenocarcinoma. We therefore recommend that periodic examinations of the esophagus and stomach be performed in patients with eventration of the diaphragm. Barrett's adenocarcinoma associated with acquired eventration of the diaphragm is reported. Patients with eventration of the diaphragm should undergo periodic examinations of the esophagus and stomach.

Key words: Barrett's adenocarcinoma, eventration of the diaphragm, short-segment Barrett's esophagus

Introduction

Eventration of the diaphragm [1] is a comparatively rare condition. It is reported to be found on routine chest X-rays at a frequency of once every 1400 to once over 13000 examinations [2]. Barrett's adenocarcinoma is the most common adenocarcinoma of the esophagus and has recently been reported at an increasing incidence in patients with esophageal adenocarcinoma. To

our knowledge, no case of Barrett's adenocarcinoma associated with eventration of the diaphragm has previously been reported. We report here such a rare case and discuss the association of Barrett's esophagus and eventration of the diaphragm.

Case report

A 71-year-old woman initially presented with epigastralgia and dysphagia in early May, 1997. On July 9, she was referred to our Department of Surgery at the Ryukyus University Hospital to undergo thorough examination and treatment after presenting at a local hospital with worsening of these symptoms.

She had no contributory family history. Her past medical history revealed that she had previously undergone an appendectomy at 38 years of age, and that elevation of the left diaphragm was detected during a regular medical checkup at 44 years of age. However, she had not complained of any symptoms since then.

On physical examination at admission, her respiratory sounds were diminished over the lower left lung field, but otherwise no abnormalities were observed. Chest X-ray revealed elevated left diaphragm, while the heart had shifted toward the right (Fig. 1). Esophagography showed dilatation of the lower esophagus and partial obstruction by a mass with a ragged surface in the lower esophagus. Barium meal stagnated in the esophagus and streamed only slightly into the stomach (Fig. 2). Computed tomography (CT) and magnetic resonance imaging (MRI) revealed increased wall thickness of the abdominal esophagus through the cardia of the stomach and elevated thin diaphragm (Fig. 3). Endoscopic examination showed a large cauliflower-type tumor in the distal esophagus approximately 36cm away from the incisors. The tumor occupied most of the esophageal lumen and the endoscopic fiber could not get through to the anal side of the tumor.

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Endoscopically, the oral side epithelium of the tumor demonstrated an almost normal esophageal mucosa, but it was reddish and with capillary dilatation (Fig. 4).

Biopsy specimens from the tumor and the oral side epithelium of the tumor revealed adenocarcinoma and a columnar epithelium which were positive for alcian blue pH 2.5 (AB pH 2.5). Based on these findings, we suspected esophageal adenocarcinoma arising from

Barrett's epithelium, and acquired eventration of the diaphragm.

On August 8, a proximal gastrectomy with reconstruction of the jejunal interposition was performed. At laparotomy, the left diaphragm was thin, relaxed, and elevated up to the thorax. The relaxed and deformed stomach extended up to the high left diaphragm, but no gastric volvulus was observed.

Fig. 1. Chest X-ray showing elevated left diaphragm; the mediastinum is shifted toward the right

Pathology findings

Macroscopically, the cauliflower-type tumor extended over the esophagogastric junction, and measured 5.7×5.2 cm (Fig. 5).

Microscopically, the oral side of the surgical specimen showed normal squamous epithelium, columnar epithelium with goblet cells, dysplastic epithelium, and well differentiated adenocarcinoma. All these tissues, except for the squamous epithelium, were positive for AB pH 2.5 (Fig. 6). The anal side mucosa of the tumor was covered with a typical gastric mucosa without intestinal metaplasia. The gastric mucosa was not positive for AB pH 2.5 but was positive only for periodic acid-Schiff (PAS). The depth of cancer invasion reached to the adventitia. There was no evidence of tumor spread to the adjacent structures or metastases to the lymph nodes and distant organs.

Discussion

There are two general theories regarding the etiology of eventration of the diaphragm: the congenital and acquired theories. It is presumed that acquired

Fig. 2a,b. Esophagography shows barium meal **a** stagnating in the esophagus while only slightly streaming into the stomach and **b** mass with a ragged surface obstructing the lower esophagus

Fig. 3. T2-weighted coronal magnetic resonance image shows thin diaphragm (*arrow*), and thickened wall of the abdominal esophagus and the cardia (*arrowhead*)

Fig. 4. Endoscopic examination showing cauliflower-type tumor and capillary dilatation of the mucosa in the lower esophagus

Fig. 5a,b. Photomacrographs of the tumor showing **a** relaxed stomach, and **b** cauliflower-type tumor extending over the esophagogastric junction

eventration of the diaphragm is induced by either an idiopathic atrophic change in the diaphragm or by secondary atrophic changes caused by various factors related to neurological dysfunction [3]. The most typical symptom is respiratory insufficiency. Other symptoms may simulate those of large diaphragmatic hernias, with dysphagia, belching, heartburn, and epigastric pain. However, most patients with eventration of the dia-

Fig. 6a–c. Photomicrographs of the surgical specimen showing **a** the sequence of squamous epithelium, columnar epithelium with barrel-shaped cells, dysplasia, and carcinoma (H&E, $\times 25$). **b** High-power view of columnar epithelium with barrel-shaped cells and dysplastic epithelium (H&E, $\times 250$) **c** with well differentiated adenocarcinoma (H&E, $\times 250$)

phragm are asymptomatic [4]. In our patient, no abnormality of the diaphragm had been observed until she was 44 years of age, and no contributory events had been identified during that period. Based on her clinical course, we inferred that the diaphragmatic eventration was acquired.

Volvulus of the stomach is a well known digestive condition associated with eventration of the diaphragm [5]. In this our patient, however, no volvulus of the stomach was seen, but Barrett's adenocarcinoma was observed. To the best of our knowledge, no previous case of Barrett's adenocarcinoma associated with eventration of the diaphragm has been reported in the literature (MEDLINE search 1966–1997).

In recent years, the incidence of adenocarcinomas of the esophagus and gastric cardia has increased in western countries. Barrett's esophagus has also been noted

as a risk factor for these malignancies [6–8]. Barrett's esophagus has generally been defined as a columnar epithelium at least 3 cm in length above the esophago-gastric junction [9,10]. However, short-segment Barrett's esophagus (SSBE), which has a columnar epithelium measuring less than 3 cm, is also recognized as Barrett's esophagus [11,12]. SSBE is also considered to be a premalignant condition and there is a wide consensus that most adenocarcinomas of the esophagus and gastric cardia evolve from SSBE [13–15]. On the other hand, it is widely accepted that the genesis of adenocarcinoma arising from Barrett's esophagus is related to the metaplasia-dysplasia-carcinoma sequence [16].

In our patient, we diagnosed the lesion as Barrett's adenocarcinoma arising from SSBE, because: (1) the biopsy specimen from lower esophagus revealed a co-

lumbar epithelium, (2) the surgical specimen showed a gradual change from squamous epithelium and columnar epithelium to severe dysplasia, and adenocarcinoma was seen on the oral side of the tumor; (3) the gastric mucosa was normal and no intestinal metaplasia was seen; and (4) the columnar epithelium, with goblet cells, dysplastic epithelium, and tumor were all similarly positive for AB pH 2.5 [6,17,18].

Barrett's esophagus is considered to be an acquired condition caused by hiatal hernia, gastroesophageal reflux, and esophagitis. In patients with SSBE, the typical symptoms are those of gastro-esophageal reflux disease, such as heartburn, regurgitation, and dysphagia. However, about half the patients with SSBE do not have these symptoms [12,19].

In our patient, we assumed that the abnormal relaxation and elevation of the stomach may have induced a pooling of the gastric juice and localized regurgitation at the esophagogastric junction. Subsequently, both esophagitis and SSBE occurred and thereafter Barrett's adenocarcinoma also developed. Interestingly, our patient's surgical specimen showed serial changes from normal squamous epithelium to SSBE, dysplasia, and adenocarcinoma during the course of the disease, with the same sequelae as those seen in the metaplasia-dysplasia-carcinoma sequence.

In conclusion, to the best of our knowledge, no case of Barrett's adenocarcinoma associated with acquired eventration of the diaphragm has previously been reported in the literature (according to Medline search, 1966–1997). We have reported one such rare case, with a brief literature review. It is possible that the eventration of the diaphragm observed in this patient was complicated not only with volvulus of the stomach but also with SSBE and Barrett's adenocarcinoma.

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