CASE REPORT

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A rare case of esophageal mucoepidermoid carcinoma successfully treated via endoscopic submucosal dissection

So Eun Jeun[1,*], Kyung Bin Kim[2,*], Bong Eun Lee[1], Gwang Ha Kim[1], Moon Won Lee[1], Dong Chan Joo[1]

Departments of [1]Internal Medicine and [2]Pathology, Pusan National University School of Medicine and Biomedical Research Institute, Pusan National University Hospital, Busan, Korea

Esophageal mucoepidermoid carcinoma (EMEC) is a special subtype of esophageal malignancy, accounting for less than 1% of all cases of primary esophageal carcinoma. Pathologically, it consists of a mixture of adenocarcinoma and squamous cell carcinoma with mucin-secreting cells. Special staining for mucicarmine helps to diagnose EMEC. We present a rare case of EMEC successfully treated via endoscopic submucosal dissection (ESD). A 63-year-old man was referred to our tertiary hospital. On esophagogastroduodenoscopy, a 6-mm-sized subtle reddish depressed lesion was identified in the mid-esophagus. Diagnostic ESD was performed with a high suspicion of carcinoma. Histopathologic findings were consistent with EMEC which was confined to the lamina propria without lymphatic invasion. We plan to do a careful follow-up without administering adjuvant chemotherapy or radiotherapy. Due to the small volume of the lesion, establishing a diagnosis was difficult through forceps biopsy alone. However, by using ESD, we could confirm and successfully treat a rare case of early-stage EMEC.

Keywords: Carcinoma; Endoscopic mucosal resection; Esophagus

INTRODUCTION

Esophageal cancer is the 8th most common malignant tumor worldwide, with an incidence of 456,000 cases per year, and ranks as the 6th leading cause of cancer deaths globally.[1,2] Esophageal cancer shows variable histologic types, with squamous cell carcinoma (SCC) and adenocarcinoma comprising the majority.[1,3] Esophageal mucoepidermoid carcinoma (EMEC) is a very rare neoplasm, accounting for less than 1% of all cases of primary esophageal carcinoma.[4] Pathologically it consists of a mixture of adenocarcinoma and SCC with mucin-secreting cells, and special staining for mucicarmine helps to diagnose EMEC.[5] Since most previously reported cases were diagnosed at an advanced stage, surgery has been recommended as the treatment of choice. However, the clinical characteristics and prognosis of EMEC are still unclear. Thus, here we report a rare case of early-stage EMEC successfully treated via endoscopic submucosal dissection (ESD).

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Correspondence: Bong Eun Lee
Department of Pathology, Pusan National University School of Medicine and Biomedical Research Institute, Pusan National University Hospital, 179 Gudeok-ro, Seo-gu, Busan 49241, Korea
E-mail: bongsul@daum.net

*So Eun Jeun and Kyung Bin Kim contributed equally to the first author.

CASE REPORT

A 63-year-old man was referred to our gastroenterology clinic for further evaluation of an esophageal depressed lesion on screening esophagogastroduodenoscopy. Initial forceps biopsy specimen obtained at a local clinic showed a few atypical cells, raising suspicion of carcinoma but requiring confirmatory diagnosis (Fig. 1A). He was a heavy drinker with a smoking

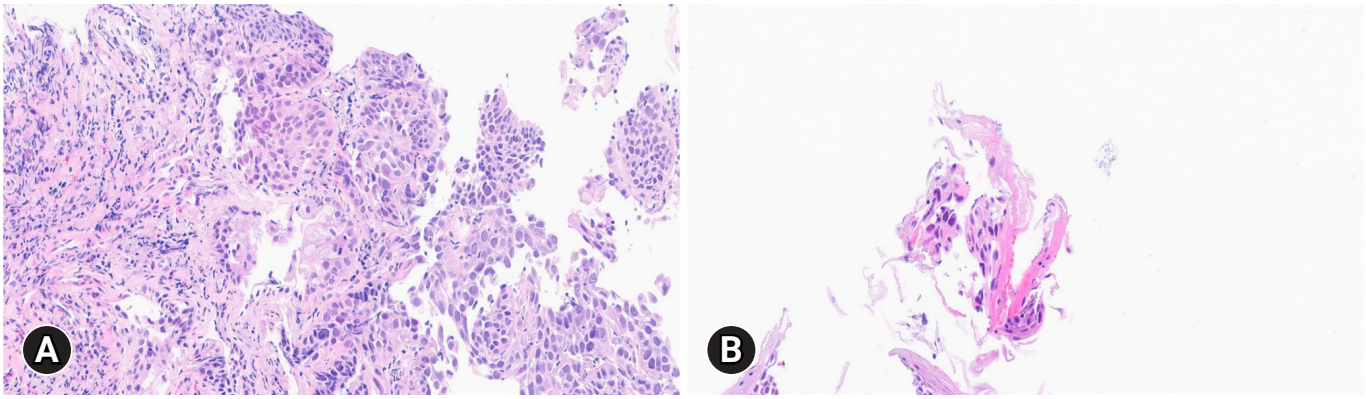

Fig. 1. (A) The initial biopsy specimen obtained at a local clinic shows atypical cells in an erosive background, which are suspicious of carcinoma (hematoxylin & eosin [H&E] stain, ×200). (B) The second biopsy specimen also shows a few atypical cells of indeterminate malignant potential (H&E stain, ×200). The patient provided written informed consent for the publication and use of his images.

history of 30 pack-years. He had no gastrointestinal symptoms, and physical examination and laboratory tests were normal. On esophagogastroduodenoscopy at our clinic, there was a 6-mm-sized reddish depressed lesion in the mid-esophagus, 30 cm below the upper incisor teeth (Fig. 2A). The lesion was unstained by Lugol's iodine solution (Fig. 2B), and narrow band imaging showed tortuous, irregular intrapapillary capillary loops (Fig. 2C, D). Repeat forceps biopsy was performed, but the obtained sample also showed a few atypical cells that were indefinite for carcinoma (Fig. 1B). Since carcinoma was suspected based on the endoscopic features, we decided to perform diagnostic ESD. ESD was successfully completed without complications such as bleeding or perforation (Fig. 3). On microscopic examination, the tumor showed a mixed expanding and infiltrative growth pattern consisting of a variable admixture of dominantly squamoid and mucous cells with scattered intermediate-type cells, and aggregates of mucous cells with intermixed squamoid cells (Fig. 4A, B). Immunohistochemically, the squamous component of the tumor was positive for p40 (Fig. 4C), while the mucinous component tumor was focally positive for CK7 (Fig. 4D). In addition, mucicarmine staining revealed mucin-producing cells with intracytoplasmic mucin (Fig. 4E). Based on these findings, the tumor was diagnosed as an EMEC. The tumor measured 6 mm×3 mm in size with clear lateral and deep resection margins, and was confined to the lamina propria without any lymphovascular invasion. Chest computed tomography was performed to confirm the final staging, and no significant lymphadenopathy was observed. Since the lesion was completely resected and fulfilled the generally accepted curability criteria for esophageal ESD, we planned to do a careful follow-up without administering adjuvant chemotherapy or radiotherapy. This study was approved by the Institutional Review Board of Pusan National University Hospital (No. 2402-015-136).

DISCUSSION

Mucoepidermoid carcinoma (MEC) represents a distinct type of tumor consisting of a mixture of a varying degree of squamous, mucinous, and intermediate cells. It commonly occurs in the salivary gland, lacrimal gland, thyroid gland, and trachea, but rarely in the esophagus. Esophageal adenosquamous carcinoma (ASC) was first reported in 1947 as an adenoacanthoma by McPeak and Arons,[6] and EMEC and ASC were initially considered as a single disease entity.[7] Subsequently, the World Health Organization distinguished EMEC from ASC in the World Health Organization classification of tumors of the esophagus.[7] However, due to the small volume of the biopsied specimens, it is difficult to obtain an accurate pathological diagnosis before endoscopic or surgical resection. A few case reports and case series have been published; however, most patients were diagnosed at an advanced stage and treated surgically. With regard to the reported cases that underwent endoscopic resection (ER), submucosal invasion was observed in all cases, indicating that ER alone did not achieve curability.[8,9] As far as we know, this is the first reported case in which ESD was successfully performed for an early stage mucosa-confined EMEC.

Because of the rarity of EMEC, the origin, clinical characteristics, and prognosis are still unclear. Regarding the origin, Stout and Latters[10] suggested that EMEC originates from the esophageal gland or ductal cells, based on its usual submucosal

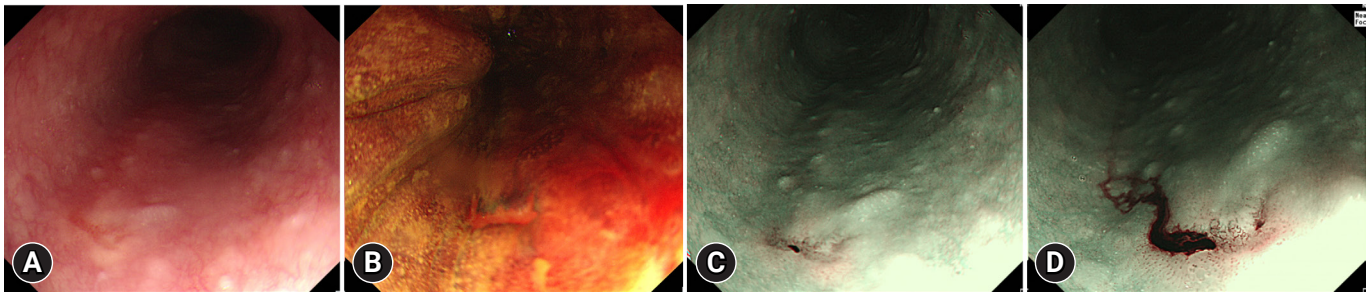

Fig. 2. (A) Conventional endoscopy show a 5-mm-sized reddish depressed lesion in the mid-esophagus, 30 cm below the upper incisor teeth. (B) The lesion is unstained by Lugol's iodine solution. (C, D) Narrow band imaging shows tortuous, irregular intrapapillary capillary loops, and these features are apparently visualized with a near focus mode. The patient provided written informed consent for the publication and use of his images.

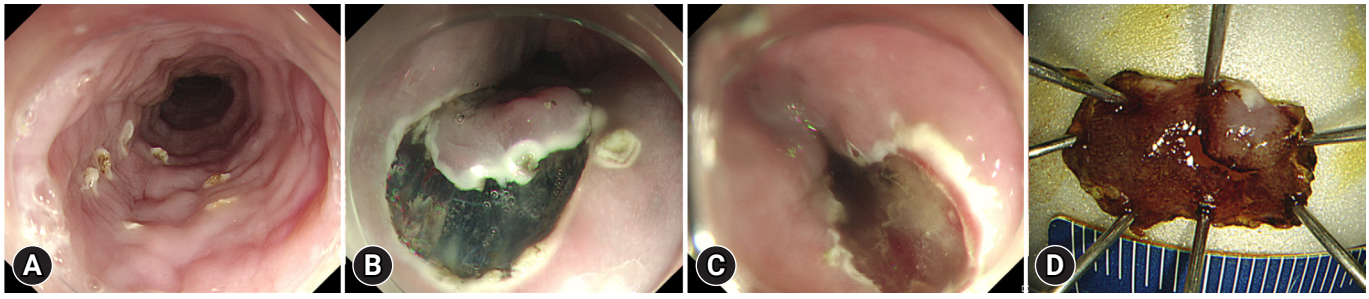

Fig. 3. (A) Markings around the lesion. (B, C) The tumor was completely resected by endoscopic submucosal dissection. (D) Gross finding show a 5-mm-sized Lugol-unstained lesion with clear margins. The patient provided written informed consent for the publication and use of his images.

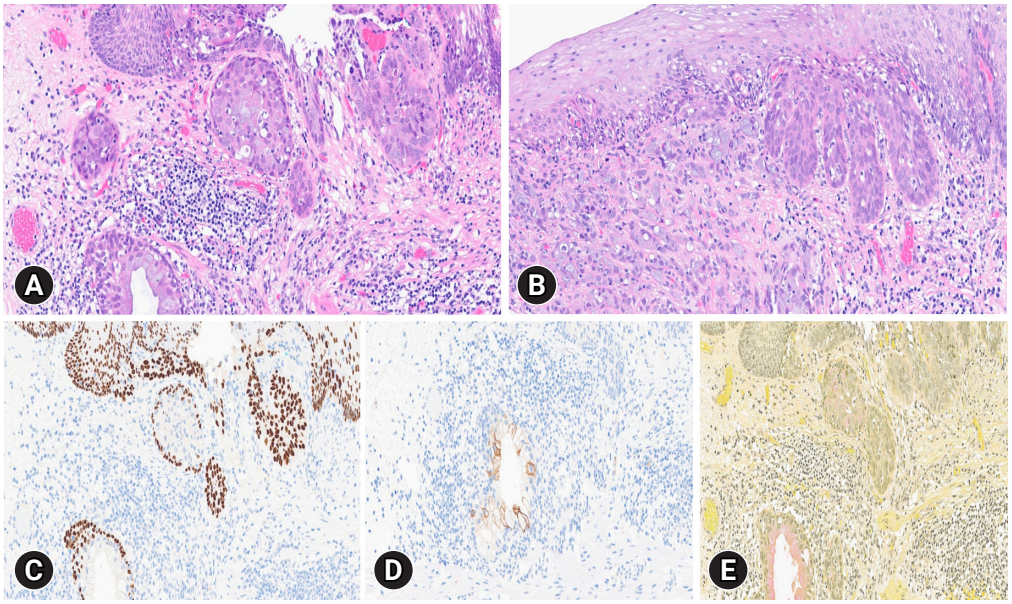

Fig. 4. (A) Tumor cells demonstrate a mixed expanding and infiltrative pattern consisting of a variable admixture of squamoid and mucous cells with scattered intermediate-type cells (hematoxylin & eosin [H&E] stain, ×200). (B) In some area, aggregates of mucin-producing cells with an unusual pattern can be observed (H&E stain, ×200). (C) p40 staining highlights the squamous component (×200). (D) CK7 staining focally highlights the mucinous cells (×200). (E) Mucicarmine staining demonstrates the mucin-producing cells with intracytoplasmic mucin (×200). The patient provided written informed consent for the publication and use of his images.

location and morphologic similarity with MEC of the salivary gland.[10,11] However, there is also a hypothesis that the mucus-secreting adenocarcinoma cells originate from the adjacent epithelial squamous cells through metaplastic change.[11] The existing evidence suggests that the behavior of this tumor differs from that of MEC of the salivary gland and SCC in situ coexists near the mucus-presenting cells. Our case supports the latter hypothesis in that the MEC cells were confined to the mucosa without a submucosal component. Kumagai et al. reviewed the macroscopic findings of superficial EMEC with mucosal or submucosal invasion.[11] Among 13 patients with superficial cancers, seven (53.8%) had type IIc lesions (slight depressed type), three (23.1%) had type IIa lesions (slight elevated type), and three (23.1%) showed mixed features of superficial and protruding types. In the present case, EMEC presented as a small 6-mm-sized reddish type IIc lesion with an abnormal microvasculature. Two sequential endoscopic forceps biopsy specimens could not confirm the diagnosis due to the small volume of the lesion. However, clinical suspicion based on the endoscopic findings and the decision to perform a diagnostic ESD led to an early diagnosis and successful outcome. Regarding the prognosis, most previous reports from the mid-1900s suggested that EMEC has a poorer prognosis than SCC.[4,12,13] In contrast, Wang et al. analyzed the relevant data since 1995 and found a similar 5-year survival rate between patients with EMEC and those with SCC.[14] However, the long-tern outcomes and prognosis after ER for EMEC have not been described yet. Suzuki et al. reported the long-term outcomes after ER was performed in patients with rare types of esophageal cancer.[15] They revealed that the curative resection rate was only 21.1% because of high rates of submucosal infiltration or lymphovascular invasion, and additional salvage surgery or adjuvant therapy were necessary in most cases. Furthermore, metastatic relapse and disease-specific mortality rates were higher in the special types of esophageal cancer compared to SCC, and the authors suggested that the special types of esophageal cancers have a high malignant potential. However, in this study, the number of enrolled patients with EMEC was only two, accounting for 10.5% of the special types of esophageal cancer. Although our case fulfilled the generally accepted curability criteria for esophageal ESD, there is no consensus for ER in EMEC. Therefore, we planned a meticulous follow-up examination.

In conclusion, we have described the first case of early stage mucosa-confined EMEC which was successfully treated via ESD. Due to the unknown clinical prognosis after ER, careful follow-up is necessary. More attention to this rare disease entity and accumulation of additional data are required.

Conflicts of Interest

Gwang Ha Kim is currently serving as a deputy editor in *Clinical Endoscopy*; however, he was not involved in the peer reviewer selection, evaluation, or decision process of this article. The other authors have no potential conflicts of interest.

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Author Contributions

Conceptualization: BEL; Resourses: SEJ, BEL; Supervision: BEL, KBK, GHK, MWL, DCJ; Writing–original draft: SEJ, Writing–review & editing: all authors.

ORCID

So Eun Jeun https://orcid.org/0000-0002-7541-6508
Kyung Bin Kim https://orcid.org/0000-0001-5430-4235
Bong Eun Lee https://orcid.org/0000-0003-2734-2134
Gwang Ha Kim https://orcid.org/0000-0001-9721-5734
Moon Won Lee https://orcid.org/0000-0002-8411-6398
Dong Chan Joo https://orcid.org/0000-0001-8734-4938

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