

Goblet Cell Carcinoid Tumor of the Appendix with Small Bowel Obstruction: A Case Report¹

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Goblet cell carcinoid tumor of the appendix (GCTA) is a tumor with histological features of both adenocarcinoma and carcinoid tumors. The most common clinical presentation of GCTA is acute appendicitis, although small bowel obstruction has been reported as a rare clinical symptom of GCTA. However, to the best of our knowledge, the CT feature of small bowel obstructions in patients with GCTA has not been reported to date. Here, we present a case of small bowel obstruction in a patient with GCTA caused by extensive tumor infiltration at the terminal ileum and distal ileum.

Index words : Carcinoid tumor

Appendix

Tomography, X-Ray computed

Intestine, small

Goblet cell carcinoid tumor is a rare neoplasm that shows histological features of both adenocarcinoma and carcinoid tumors. Goblet cell carcinoid tumor of the appendix (GCTA) primarily arises from the appendix, but can also occur in other parts of the gastrointestinal tract (1-5). The common clinical presentations in order of frequency are acute appendicitis (22.5%), asymptomatic (5.4%), non-localized abdominal pain (5.15%) and the presence of an appendiceal mass (3.09%) (5). Small bowel obstruction is one of the rare clinical presentations of GCTA (1-5). To the best of our knowledge, the CT finding of GCTA with small bowel obstruction has not yet been reported.

Here we present a case report of GCTA with small bowel obstruction in an 80-year-old man.

Case Report

An 80-year-old man was admitted to our hospital for an abdominal pain. He had been admitted to our hospital on two prior occasions and had been diagnosed with obstructive small bowel ileus two months earlier. The patient did not undergo abdominoplevic CT during the previous admissions. His medical history included hypertension, but he had no history of previous abdominal operations. Physical examination showed diffuse mild abdominal tenderness with hypoactive bowel sound. Laboratory tests were unremarkable except for the presence of an increased white blood cell count ($14.3 \times 10^3/\text{mm}^3$). Plain abdominal radiographs revealed dilatation of the small bowel loops with air-fluid levels compatible with a small bowel obstruction.

Abdominoplevic Multi Detector Row Computed Tomography (MDCT) was performed prior to the administration of intravenous contrast material, and a portal venous phase CT scan was taken after IV contrast administration. No oral or rectal endoluminal opacification

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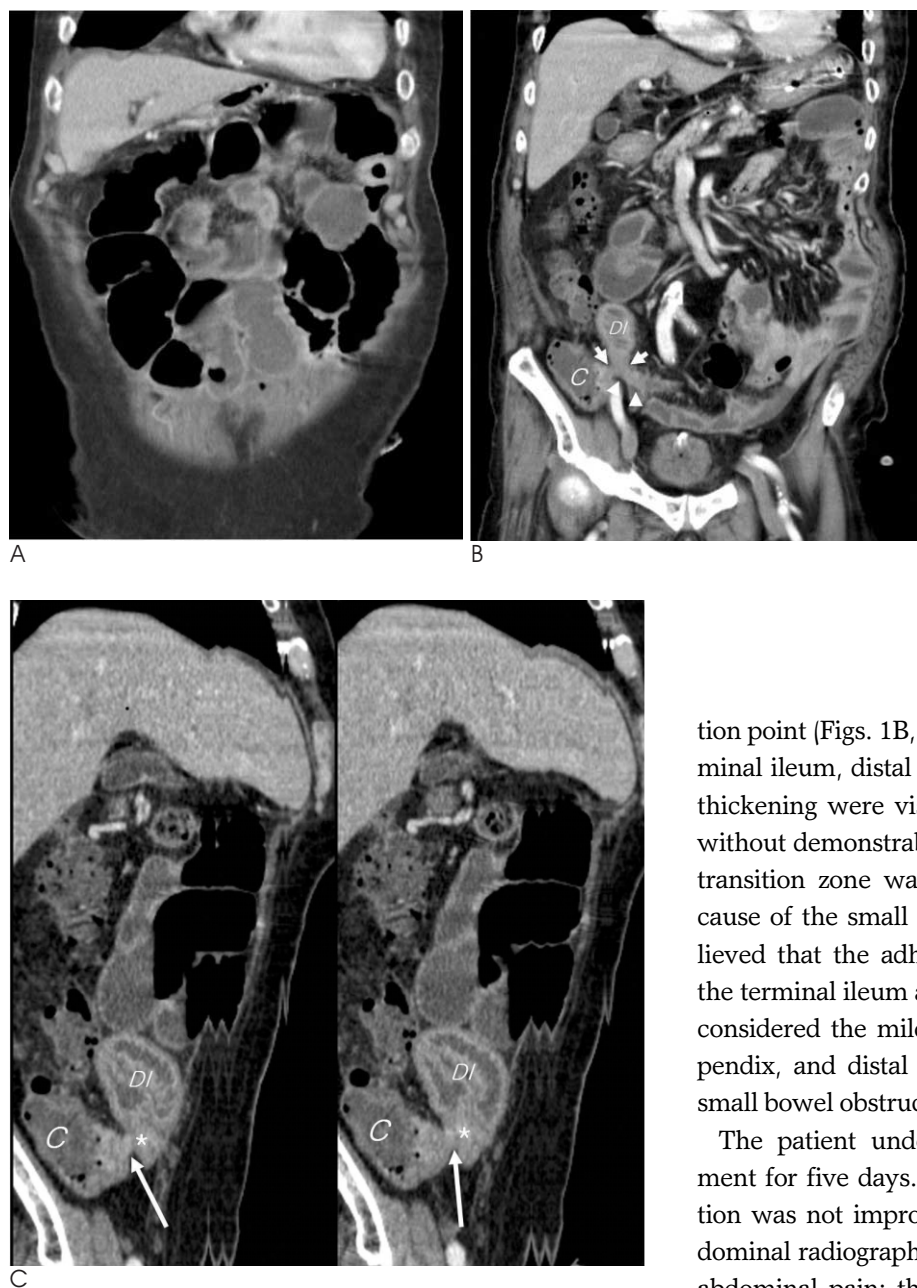


Fig. 1. Goblet cell carcinoid tumor of the appendix in an 80-year-old man.

A, B. Reformatted coronal contrast-enhanced CT images show dilated small bowel loops with a transition point (short arrows) at the medial aspect of the cecum (C) and narrowed terminal ileum (arrowheads). At the transition point, distal ileum (DI) is adhered to the terminal ileum (arrowheads).

C. Two serial curved planar reformation images of a transition point show the partially visible appendix (long arrow) placed at the inferior aspect of the transition point and adhered to the terminal ileum (asterisk). There is mild diffuse wall thickening with enhancement at the appendix and cecum (C) near the orifice of the appendix.

tion point (Figs. 1B, C). Mutual adhesions among the terminal ileum, distal ileum, and appendix with mild wall thickening were visualized around the transition point without demonstrable pathologic process, and a smooth transition zone was noted. We could not predict the cause of the small bowel obstruction precisely and believed that the adhesion itself provoked narrowing of the terminal ileum and the small bowel obstruction. We considered the mild wall thickening at the cecum, appendix, and distal ileum as secondary findings of the small bowel obstruction.

The patient underwent conservative medical treatment for five days. However, the small bowel obstruction was not improved upon serial follow-up plain abdominal radiographs, and he complained of aggravating abdominal pain; therefore, he underwent laparotomy. During surgery, severe inflammation at the appendix that was adhered to the terminal ileum was observed. Multiple subcentimeter peritoneal seeding nodules were also observed. Because these findings suggested appendiceal carcinoma, the intraoperative frozen biopsy of the peritoneal seeding nodule was performed. Pathological examination of the frozen section revealed poorly differentiated metastatic adenocarcinoma. Based on the pathological results, an extended right hemicolectomy was performed.

Upon pathological examination, the mucosal surface of the resected specimen showed a small nodular lesion at the appendiceal orifice and mild to moderate narrowing of the terminal ileum (Figs. 2A, B). The serosal sur-

was administered. The CT scan showed dilated small bowel loops with a transition point at the medial aspect of the cecum and a narrowed terminal ileum (Fig. 1). At the transition point, the distal ileum was collapsed and adhered to the terminal ileum (Fig. 1B). The proximal portion of the appendix was also adhered to the terminal ileum at the inferior aspect of the transition point (Fig. 1C). The appendix was not thoroughly traced on the CT scan. The cecum near the orifice of the appendix and the portion of the appendix that was visible showed mild diffuse wall thickening with enhancement (Figs. 1B, C). The CT also showed mild diffuse wall thickening with enhancement at the distal ileum around the transi-

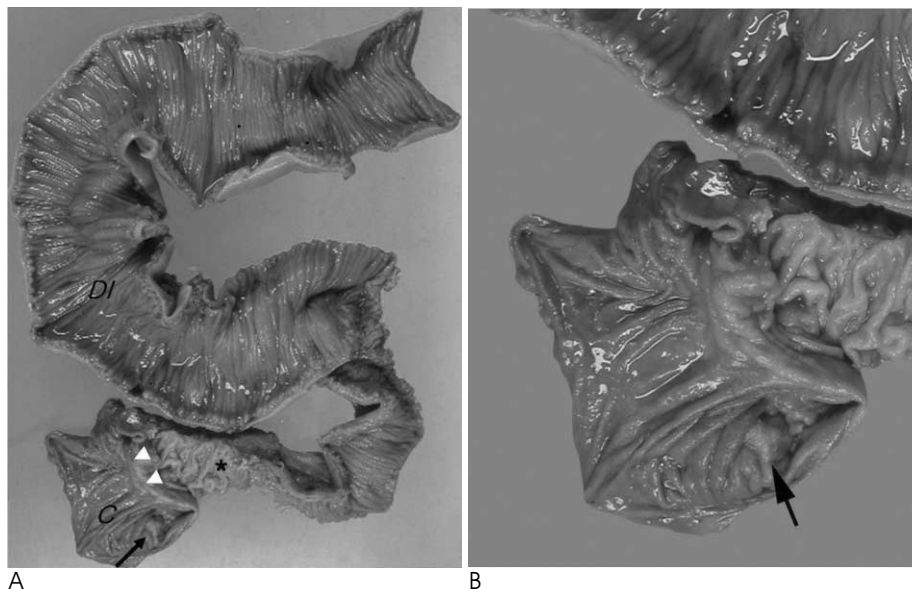
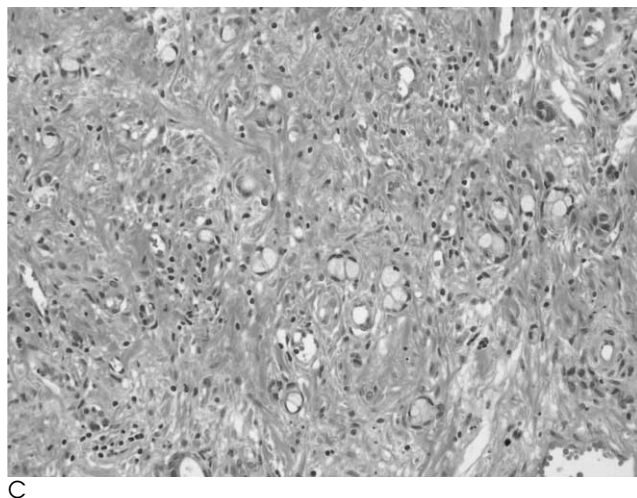


Fig. 2. Pathologic findings of the resected specimen following right hemicolectomy.

A, B. The resected gross specimen of the right hemicolectomy shows mild to moderate narrowing of the terminal ileum (asterisk) and dilated distal ileum (DI). The appendiceal orifice of the resected cecum (C) shows a small elevated submucosal lesion (long arrow). Arrowheads = Ileocecal valve
C. On the photomicrograph (original magnification, $\times 200$; H-E stain) there are scattered islands of tumor cells showing abundant intracytoplasmic mucin in the appendix.



face of the small intestine and cecum was diffusely fibrotic. Except for the appendiceal orifice, the appendix was not identified. The pericecal soft tissue showed diffuse fibrotic changes without discrete mass formation. Upon microscopic examination, there were diffusely infiltrated islands of tumor cells distended by mucus that resembled signet-ring cells in the appendix (Fig. 2C). The tumor involved the serosa, subserosal soft tissue, proper muscle, and submucosa of the cecum, and there were scattered islands of tumor cells in the subserosal soft tissue of the small intestine. Immunohistochemical studies revealed that the tumor cells were reactive for chromogranin, synaptophysin, and CD56, and the final pathologic diagnosis was goblet cell carcinoid of the appendiceal orifice.

Discussion

GCTA accounts for less than 5% of primary tumors of the appendix, whereas the classic carcinoid tumor of the appendix accounts for 32% to 57% of all appendiceal tumors (5, 6). The classic carcinoid tumor of the appendix demonstrates a uniquely indolent clinical course and has a 5-year survival rate of greater than 90%. In addition, carcinoid tumors metastasize in 2 to 5% of all cases (5, 6). In contrast, GCTA is a low-grade malignancy with a 5-year survival rate of between 60 and 84%. Moreover, GCTA metastasizes in 15 to 30% of all cases (5, 6). Therefore, right hemicolectomy is usually performed to reduce the risk of metastatic disease (2). However, the vast majority of goblet cell carcinoid tumors are rarely diagnosed preoperatively and are usually diagnosed during operation or the pathologic examination of the appendiceal specimen (2). GCTA is an infiltrative tumor that typically involves the entire appendix. These characteristics are manifested as a diffuse mural thickening of the appendix with or without peritoneal seeding lesions or ovarian metastasis upon CT (7). The infiltrative nature of GCTA was demonstrated as a partially visible appendix with mild diffuse enhancing wall thickening in this case (Figs. 1B, C). Moreover, the extraappendiceal tumor infiltration into the terminal ileum caused a small bowel obstruction in this case.

In the evaluation of small bowel obstructions on CT, it is important to define the cause of the obstruction. If the cause is not identified, a diagnosis of adhesions is usually inferred. Adhesive ileus is common in patients who

have undergone previous abdominal operations (8). In this case, the patient had no history of previous abdominal operations. However, we suggested adhesive ileus preoperatively because we thought that the small bowel obstruction had no cause and we overlooked the possibility of an infiltrative appendiceal lesion. In addition to GCTA, the classic carcinoid tumor, lymphoma, and nonmucinous adenocarcinoma of the appendix can show an infiltrative growth pattern (7, 9, 10). Classic carcinoid tumors of the appendix are not typically detected by direct imaging studies due to their small size and location in the distal appendix. Lymphoma manifests as marked enlargement of the appendix with diffuse wall thickening but relative maintenance of its vermiform shape. Thus, in this case, the possibilities of lymphoma and classic carcinoid tumor are low. However, nonmucinous adenocarcinoma of the appendix can manifest as an infiltrative appendiceal lesion with extraappendiceal infiltration and direct invasion of the adjacent organs (7). Nonmucinous adenocarcinomas rarely occur in the appendix and tend not to form mucoceles. Its reported CT finding is a subtle infiltrative appendiceal lesion with surrounding periappendiceal infiltration with or without direct invasion of the adjacent organs (5).

In summary, GCTA in this case presented as a small bowel obstruction. On pathological examination, GCTA was found to be associated with diffuse extraappendiceal infiltration of the terminal ileum and distal ileum, which caused a small bowel obstruction. However, we did not notice the malignant appendiceal cause of the small bowel obstruction preoperatively. When small bowel obstructions with a transition point at the right lower quadrant abdomen are encountered, it is impor-

tant to evaluate the appendix thoroughly. If the appendix is partially visible at this transition point, the possibility of an infiltrative appendiceal lesion such as a goblet cell carcinoid tumor should be considered, especially in patients with no previous abdominopelvic operation.

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소장폐쇄를 일으킨 충수돌기의 술잔세포 유암종:

증례 보고¹

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충수의 술잔세포 유암종은 유암종과 선암종 두 종양의 조직학적 특징을 모두 가지는 종양이며, 충수염이 가장 흔한 합병증이다. 드물게 충수의 술잔세포 유암종에 의해 장폐색이 유발될 수 있으며 보고된 바 있다. 그러나, 현재까지 충수의 술잔세포 유암종에 의한 장폐색의 전산화단층촬영 소견에 대한 보고 및 고찰은 없었다. 이에 저자는 충수의 술잔세포 유암종이 말단회장 및 원위부 회장 침윤으로 인해 소장의 폐색을 초래한 사례를 보고하고자 한다.