Colonoscopic Feature of Primary Adenocarcinoma of the Appendix:

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Abstract

We herein report a case of primary adenocarcinoma of the appendix, a very rare disease that is seldom diagnosed before surgical intervention. This case was first suspected for its unique colonoscopic presentation as a cecal submucosal tumor with an overlying mucin-coat at the appendiceal orifice. The diagnosis was later confirmed after the operation. The imaging features of this exceptional disease are presented in detail.

Key words: adenocarcinoma of appendix, appendiceal carcinoma, appendiceal tumor, colonoscopy

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Introduction

Primary adenocarcinoma of the appendix is an extremely rare disease with an incidence of approximately 0.08%~0.1% of all previously reported appendectomy specimens (1, 2). Appropriate preoperative diagnosis of appendiceal adenocarcinoma is rare and difficult by either radiology (3) or endoscopic findings. Here, we present a patient with primary adenocarcinoma of the appendix with unique colonoscopic features mimicking cecal submucosal tumor.

Case Report

A 29-year-old Chinese man with a medical history of chronic duodenal ulcer visited our hospital in March 2003. He complained intermittent vague, non-radiating abdominal pain and postprandial bloating for half a year. According to his statement, the pain was limited to the periumbilical area, and was associated with nausea, decreased appetite and tenesmus. Several episodes of black stool passages before admission were also mentioned. Physical examination revealed right lower quadrante abdominal tenderness and slightly hyperactive bowel movement. His body temperature was 36°C. Laboratory profiles including complete blood count and biochemical parameters were all within normal limits, except for a slightly elevated carcinoembryonic antigen (6.96 ng/mL).

Colonoscopy study showed one bulging submucosal tumor about 3 cm in diameter over the anterior wall of cecum near the ileocecal valve (Fig. 1). The overlying mucosa was congestive, and was covered by a thin layer of mucin. An obliterated appendiceal orifice was present on the top of the tumor. Biopsy was taken around the orifice of appendix, but only nonspecific inflammatory changes were revealed. Double contrast barium study of the colon demonstrated an appendiceal tumor about 2 cm in diameter over the anterior wall of the cecum with an umbilicus at the top (Fig. 2). Abdominal computed tomography demonstrated a cecal mass without significant lymph nodes enlargement in the abdominal cavity (Fig. 3).

After completing the above examinations, a mucin-secreting appendiceal tumor was diagnosed. During the operation, a 2×2 centimeter whitish firm tumor mass was found at the appendiceal orifice, which obliterated the lumen of the appendix (Fig. 4). The other portion of the appendiceal mucosa was grossly intact, and the tumor was not adhesive to the adjacent organ. Frozen sectional pathology showed adenocarcinoma with mucoid material and thus right hemicolectomy and ileo-colostomy were performed. No intra-abdominal tumor metastasis was found grossly. The final pathological report revealed a well-differentiated adenocarcinoma originating from the appendiceal mucosa with focal invasion to submucosa, but no perineural, lymphatic or vascular invasion was found (Fig. 5). The post-operative course was smooth and he was discharged one week later.

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Figure 1. One bulging submucosal tumor about 3 cm in diameter was found in the cecum near the ileocecal valve with a congestive surface and covered by a thin layer of mucin. An obliterated appendiceal orifice (arrowheads) was present on the top of the tumor.

Figure 2. Double contrast barium study of the colon demonstrated an appendiceal tumor (black solid arrow) about 2 cm in diameter over the anterior wall of the cecum with an umbilicus at the top.

Figure 3. Abdominal computed tomography demonstrated a cecal mass (white solid arrow) without significant lymph node enlargement in the abdominal cavity.

Figure 4. The cross-section showed a tumor (white solid arrow) with mucoid material obliterating the appendiceal orifice.

Figure 5. Pathology revealed a well-differentiated adenocarcinoma arising from the appendiceal mucosa with focal invasion to the submucosa.

There was no tumor recurrence after 44 months of follow-up.

Discussion

Primary adenocarcinoma of the appendix is a rare disease, representing less than 0.5% of all gastrointestinal malignancy (4, 5). Even with the technological advancement of examination techniques, it is still difficult to make a precise diagnosis before surgical intervention possibly because of the small size of the lumen of the appendix.

When a tumor mass is discovered over the right lower quadrant of the abdomen, the following differential diagnoses should first be ruled out: adenocarcinoma, carcinoid, mucinous cystadenoma, mucinous cystadenocarcinoma, lipoma, lymphoma, and pericolonic abscess. Until recently, no typical signs or symptoms could be regarded as particularly representative of appendiceal adenocarcinoma. The most common clinical presentation reported (5, 6) is acute appendicitis. The narrow appendiceal lumen may be occluded by the tumor early during the course, and predisposes it to appendicitis and rupture. Other presentations in-

cluded palpable abdominal mass, ascites, peritonitis resulting from perforated appendix, intussusception and a variety of non-specific gastrointestinal or genitourinary symptoms. Some of the above presentations were incidental findings during intra-abdominal surgery for other medical conditions (5, 7). In the present case, the clinical presentation was non-specific abdominal symptoms, e.g. vague abdominal pain and bloating. The only positive physical finding was right lower quadrant abdominal tenderness, but no definite mass was palpated. Therefore, clinicians should be alert and remember to take this diagnosis into consideration if a right lower abdominal lesion is suspected.

Before the operation was performed, computed tomography, double-contrast barium study and colonoscopy were done for evaluation. During colonoscopic examination there was an appendiceal tumor that obliterated the orifice of the appendix, resembling a submucosal tumor of the cecum. Sakamoto et al (8) reported an experience of an attempt to pull the tumor out from the orifice of appendix with biopsy forceps, but this technique was difficult to perform upon this patient due to his already tightly obliterated orifice. What came to our notice was that a thin layer of mucin-coat, which was present on the mucosal surface of the tumor, although biopsy specimen taken from the mucosa revealed a negative finding for malignancy. It brings the idea that when mucin was present over the tumor at the appendix orifice, an appendiceal mucinous adenocarcinoma is highly suspected. It is also suspicious a sign of appendiceal tumor if there is negative demonstration of the appendix and/or extrinsic compression in the cecum during a barium study (3, 6, 8).

Whether to manage appendiceal adenocarcinoma by right

hemicolectomy or appendectomy is controversial. Some authors suggest simple appendectomy for adenocarcinoma of any differentiation confined to mucosa or well-differentiated adenocarcinoma with submucosal invasion (9). Moreover, right hemicolectomy is suggested for invasive adenocarcinoma or tumor involving the cecum or adjacent organ (10). Some investigators recommend right hemicolectomy with the presence of malignancy to take care of the risk of possible nodal metastasis (5, 11). The 5-year survival rate reported by Nitecki et al (5) is superior in the patient receiving right hemicolectomy to those receiving appendectomy alone (68% versus 20%). Nonetheless, 38% of patients that underwent right hemicolectomy as a secondary procedure were upstaged after the operation because of the involvement of the mesenteric lymph nodes. Adjuvant chemotherapy might be beneficial for more advanced diseases. A high incidence (35%) of secondary malignancy is also noted, and it is mainly located at colorectal region (5). Carefully colonoscopic examination is needed pre-operatively to reveal synchronous malignancies (18%) and post-operatively for metachronous malignancies (17%). This patient underwent right hemicolectomy and ileo-colostomy with a good prognosis.

Based on the experience of this case report, we learned that the cecum and appendiceal orifice should be carefully inspected during colonoscopic examination in order to detect appendiceal tumors. If a mucin coating over a submucosal tumor with obliterated appendiceal orifice is present, an appendiceal tumor should be suspected. The other parts of the colon should also be examined to detect secondary malignancies.

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