Case Report

A Case of Primary Carcinoma of the Vermiform Appendix in an Early Stage

Nobuaki SAKAMOTO, Keizoh YONEDA, Jiroh OGATA, Munetaka MORI, Tatehiko WADA, Kiyoharu UMEZU, Susumu SATOH, Kazuhiko KASUYA, Akira MAJIMA, Kohichiroh KATOH, Tatsuya AOKI and Yasuhisa KOYANAGI

Department of Surgery, Tokyo Medical University

A 76-year-old woman, presented with positive fecal occult blood. She underwent a resection of the pancreatic body and tail under a diagnosis of serous cystadenoma of the pancreas on July 21, 1999. The patient had no particular subjective symptoms, but laboratory examination during a follow-up visit revealed occult blood in the feces. Based on the findings of a barium study and colorectal endoscopy, she was diagnosed as having cancer of the vermiform appendix. Resection of the ileocecum and regional lymph node dissection (D2) were performed. The tumor filled the lumen of the appendix and resembled a polypoid Is tumor, protruding into the cecum. The tumor was a well-differentiated adenocarcinoma arising from the mucosa, with a papillary to tubular structure. The pathological stage of the tumor was rated as 0 (well-differentiated adenocarcinoma in a tubulo-villous adenoma). The depth of tumor invasion was m, ly(0), v(0), and no signs of lymph node metastasis were found. Primary cancer of the vermiform appendix is rare, and early detection is very uncommon. We report a case of primary carcinoma of the vermiform appendix that was detected at an early stage and discuss the relevant literature.

Key Words : Primary carcinoma of the vermiform appendix, Early appendiceal cancer

Introduction

The vermiform appendix can be affected by a wide variety of diseases, such as malignant tumors (cancer, malignant lymphoma, carcinoid, etc.), benign tumors (mucous cyst, etc.), and inflammatory diseases (appendicitis, etc.), but unlike other organs of the digestive tract, it has a blind sac structure, and only its root can be visualized by endoscopy. Lesions from the center to the tip of the vermiform appendix can only be visualized by endoscopy as extramural compressive lesions, and only if the lesion is a mass lesion. Primary cancer of the vermiform appendix is rare, and detection at an early stage is seldom possible for the reasons mentioned above. We recently encountered a case of early primary cancer of the appendix that arose at the root of the organ, grew towards the cecum, and presented a characteristic endoscopic sign of the disease.

Case report

The patient was a 76-year-old woman, whose chief presenting feature was positive fecal occult blood. She underwent resection of the pancreatic body and tail for a diagnosis of serous cystadenoma of the organ on July 21, 1999. She was taking oral medication for diabetes mellitus. The patient was being foll-
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owed up at an outpatient clinic after resection of the pancreatic body and tail. She had no particular symptoms, but a laboratory examination during a follow-up visit revealed fecal occult blood. Based on the results of a subsequent barium study and colorectal endoscopy, she was diagnosed as having cancer of the vermiform appendix. On admission, the patient's height was 155 cm, and she weighed 45 kg. Her abdomen was flat and soft, and there was no tenderness. There was no palpable mass in the abdomen.

Laboratory test results on admission:
WBC, 5,200/μl; RBC, 523 × 10^4/mm³; and Plt, 27.5 × 10⁴/mm³. The serum levels of Hb (13.7 g/dl), TP (8.1 g/dl), CRP (0.3 mg/dl), and tumor markers (CEA and CA-19-9) were all within their normal ranges. Other hematological parameters examined were also normal.

Diagnostic imaging findings on
1) Plain abdominal X-rays: A small collection of gas was noted in the upper small intestine, but no other abnormalities were found.
2) Barium study: A compressive lesion, about 20 mm in size, was visible in the ileocecum. The appendix did not fill. Multiple diverticula were seen in the ascending colon (Fig. 1).
3) Colorectal endoscopy: An elevated lesion, about 20 × 20 mm, was visible at the opening of the appendix. A depression, probably representing a lumen, was seen at the center. Vertical folds were visible at the base. Biopsy revealed a diagnosis of well-differentiated adenocarcinoma (Fig. 2).

Based on these findings, the patient was diagnosed as having primary cancer of the vermiform appendix in an early stage.
vermiform appendix. In view of the likelihood of adhesions having formed as a result of the previous operation, open surgery with celiotomy was performed.

**Intraoperative and histological findings**

Adhesions as a result of the previous operation were visible, and the appendix was slightly swollen. Because no marked lymph node enlargement was noted, resection of the ileocecum and regional lymph node excision (D2) were performed. The tumor filled the lumen of the appendix and assumed the form of a polypoid Is type tumor that protruded into the cecum (Fig. 3).

**Histopathological findings**

The tumor was a well-differentiated adenocarcinoma arising from the mucosa, and it possessed a papillary to tubular structure (Fig. 4a–d). The pathological stage of the...
tumor was rated as 0 (well-differentiated adenocarcinoma in a tubulo-villous adenoma). The depth of tumor invasion was m, ly(0), v(0), and lymph node metastasis was negative.

**Discussion**

Primary cancer of the vermiform appendix is a rare condition, having been reported to account for about 0.3-0.6% of all cancers of the large intestine. Ogawa et al. reported that only 19.8% of all cases of cancer of the appendix could be diagnosed preoperatively. The preoperative diagnosis rate for cancer of the appendix is thus significantly lower than for malignant tumors arising from other sites in the large intestine. There have been many reports of cases in which cancer of the appendix was detected during surgery for a preoperative diagnosis of appendicitis and cases in which it was incidentally detected during surgery for cancer of other organs (in particular, cancers of the genitourinary system). When cancer of the appendix is associated with signs of inflammation, it may be quite difficult to distinguish it from appendicitis by abdominal palpation. Because colon preparation using purgatives is usually not performed in the preoperative examination of cases suspected of appendicitis, the diagnosis is often only made on the basis of the intraoperative findings or pathological examination of the resected specimens.

In the present case, the positive fecal occult blood test provided the clue that led to the rare detection of an appendiceal cancer at an early stage. In our patient the cancer was associated with diverticulosis, and it remained unknown whether the cancer or a diverticulum was the source of the fecal blood.

Cancer developing at the root of the vermiform appendix is often associated with a characteristic endoscopic sign called the “vertical fold”, and a similar sign detected in barium studies is called “coiled spring appearance” or “vertical fold”. This sign is produced by full-circumferential elevation of the intact cecal mucosa by gas and a tumor developing in the root of the appendix and extending towards the cecum. It is also seen in cases of mucous cyst or cystadenocarcinoma. Therefore, when performing colorectal endoscopy, the physician must bear in mind that, although not pathognomonic, vertical folds suggest cancer of the appendix. The incidence of appendiceal cancer is very low compared to cancers of other sites of the large intestine, and in most cases, it is detected in an advanced stage. This is primarily because of the blind sac structure of the appendix, which differs from the structure of other organs of the digestive tract, and only permits endoscopic visualization of lesions that develop at the root of the appendage. Ogawa et al. reviewed cases of appendiceal cancer reported over the last 10 years and reported that preoperative diagnosis was impossible by a barium study in 46% of the cases and impossible by a barium study + colorectal endoscopy in 25% of the cases. They therefore concluded that preoperative diagnosis of cancer of the vermiform appendix is difficult. Whenever an elevated lesion with vertical folds is found at the opening of the appendix during colorectal endoscopy, attention must be directed toward excluding the possibility of cancer of the vermiform appendix. Reports of early cancer of the vermiform appendix are rare. In Japan, only 21 cases of cancer of the appendix confined to the mucosa, including the present case, have been reported. However, reports of cancer of the appendix diagnosed at an early stage have increased in recent years, and they are likely to increase further, if more careful examinations are conducted during colorectal endoscopy to detect this type of cancer.
Since the muscle layer is not well-developed in the appendix, cancers tend metastasize early, and the prognosis is usually poor\(^1\),\(^2\). When advanced cancer of the appendix is diagnosed, right hemicolecetomy is the standard operative procedure\(^6\),\(^7\). No reports on operative procedures or the prognosis of cancer (mucosal or submucosal cancer) of the appendix diagnosed at an early stage have been published. Preoperative evaluation of the depth of tumor invasion is also difficult for this type of cancer. A case has been reported in which perforation developed after endoscopic surgery, necessitating re-operation with celiotomy\(^10\). Endoscopic surgery is therefore not indicated, even in cases of early cancer of the appendix, and open surgery is performed as a rule. It will be possible to establish a standard operative procedure for early cancer of the vermiform appendix, if a large number of reports of treatment are accumulated in the future.

**Conclusion**

A rare case of early primary cancer of the vermiform appendix that was diagnosed preoperatively as a result of investigation of a positive fecal occult blood test has been reported. Careful observation of the opening of the appendix during colorectal endoscopy would seem essential for early detection of this cancer.

**References**

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