Case Report

A Case of Mucinous Cystadenocarcinoma of the Appendix : Difficult Preoperative Differentiation from an Ovarian Tumor

Nobuaki SAKAMOTO, Motohiro SAIGUSA, Jeong Sik LEE,
Yoich WAKAMA, Yasuharu MORI, Kenichiro HASUE,
Tatehiko WADA, Akira MAJIMA, Akihiko TSCHIDA,
Koichiro KATO, Tatsuya AOKI and Yasuhisa KOYANAGI
Department of Surgery, Tokyo Medical University

We report a case of primary mucinous cystadenocarcinoma of the vermiform appendix. A 54-year-old woman presented to a local clinic with a chief complaint of right lower abdominal pain. A right ovarian tumor was suspected, and the patient was referred to our gynecology department. Surgery was performed for a preoperative diagnosis of right ovarian tumor, but laparotomy revealed a primary tumor of the appendix, and appendectomy was performed instead. The intraoperative pathological examination did not reveal any evidence of malignancy, however, the final histological diagnosis was primary mucinous cystoadenocarcinoma of the appendix, and right hemicolectomy with lymph node dissection (D3) was performed two weeks after the initial operation. The tumor was histological stage II (p0, h0, n0, and ss). Primary mucinous cystadenocarcinoma of the appendix is rare and difficult to differentiate from appendicitis and other diseases preoperatively.

Key Words : primary carcinoma of the vermiform appendix, mucinous cystadenocarcinoma

Introduction

Primary carcinoma of the appendix is a rare disease. It tends to be found after laparotomy for diagnosis of appendicitis or discovered incidentally during abdominal surgery for an unrelated condition (especially gynecologic or urologic disease). When carcinoma of the appendix is associated with inflammation, differentiation from appendicitis is extremely difficult by abdominal examination alone. Among the primary carcinomas that occur in the appendix, mucinous cystadenocarcinoma has been reported to pursue a different course from intestinal-type lesions, however, there are many uncertainties about their pathology. We report a case of a patient who underwent lapar-

otomy for a suspected ovarian tumor in whom the final pathological diagnosis was primary mucinous cystoadenocarcinoma of the appendix.

Case Report

The patient was a 54-year-old woman with a chief complaint of right lower abdominal pain. There was no relevant family history or past history. She visited a local clinic where she was referred to our gynecology department due to suspicion of a right ovarian tumor. When abdominal ultrasonography also suggested a right ovarian tumor, she was admitted to our hospital for treatment. On admission, her height was 155cm, and her weight was 45kg. The abdomen was flat, soft, and nontender, and

Fig. 1 Transvaginal ultrasonography showed the uterus to be of normal size, although a small subserosal myoma was observed. A mass was noted to the right of the uterus, and its interior consisted of a mixture of high and low echoic areas.

Fig. 2 Pelvic CT scan.

A roundish cystic mass lesion was seen in the right pelvis, and calcification of its wall and a slight enhancement effect were observed. Small nodular shadows were noted in portions of it.

Fig. 3 Pelvic magnetic resonance imaging (MRI).

a) A roundish cystic mass with a thick wall and containing some nodular areas is seen occupying the right half of the pelvis. b) An annular structure connected the mass to the ileocecal region.

no masses were palpable. Laboratory tests on admission revealed the following: leukocyte count, $5,200/\mu\text{L}$; erythrocyte count, $523 \times 10^4/\text{mm}^3$; hemoglobin, 13.7g/dl ; platelet count, $27.5 \times 10^4/\text{mm}^3$; total protein, 8.1g/dl ; and C-reactive protein, 0.3mg/dl . The tumor markers CEA and CA19-9 were within the normal range. Other laboratory parameters examined were also normal. Plain abdominal radiography yielded no significant findings. Transvaginal ultrasonography (US) showed the uterus to be of normal size, although a small subserosal myoma was observed to the right of the uterus, a mass was noted and its interior consisted of a mixture of high and low echoic areas (Fig. 1). A pelvic computed tomography (CT) scan showed a roundish cystic mass lesion in the

right pelvis, and calcification of its wall and slight enhancement effect were observed. Small nodular shadows were also noted in portions of it (Fig. 2). Pelvic magnetic resonance imaging showed a roundish cystic mass located in the right half of the pelvis, with a

Fig. 4 Macroscopic view of the resected specimen, which measured $6.5 \times 4.5 \times 2.0$ cm. The appendix was enlarged and had a rough surface.

Fig. 5 Microscopic findings in the appendix (H&E stain, $\times 40$ in each).

a) The cells contain an extremely large amount of mucin, and their nuclei are relatively regularly aligned at their bottoms (rapid intraoperative pathological examination). They consist of epithelium exhibiting a mild increase in nuclear chromatin with a low degree of atypia. b) Near the tip of the appendix cancer cells were observed in an abundant mucous stroma.

thick wall and containing some nodular areas. An annular structure connected the mass to the ileocecal region. Since the cyst fluid was not

hyperintense on T1-weighted images, it was difficult to assess its characteristics (Fig. 3). Based on these findings, the patient was suspected of having a right ovarian tumor, and laparotomy was performed in our gynecology department. At operation, the appendix was found to be enlarged and partly adherent to the greater omentum. The appendix and part of the greater omentum were resected, but the intraoperative pathological examination did not reveal any evidence of malignancy, and the abdomen was closed without further intervention (Fig. 4). The postoperative pathological examination showed that the resected appendix measured $6.5 \times 4.5 \times 2.0$ cm and that the attached fat was prominent at the cecal border. The cut surface showed severe inflammation of the appendix that appeared to involve the cecal stump. On histological examination, the epithelial component in the inflamed region was mainly non-neoplastic, and the occasional neoplastic cells seemed to be due to contamination. Near the tip of the appendix, however, tumor cells were observed in an abundant mucous stroma, and a diagnosis of mucinous cystadenocarcinoma of the appendix was made (Fig. 5). Based on this pathology report, right hemicolectomy with lymph node dissection (D3) was performed two weeks after the initial operation. The tumor was judged to be histological stage II (p0, h0, n0, and ss). The patient made good progress postoperatively, and was discharged from hospital after receiving intravenous 5-FU and CDDP adjuvant chemotherapy. She is now being treated with oral 5'-DFUR on an outpatient basis.

Discussion

Primary carcinoma of the appendix is a rare disease, accounting for only 0.3-0.6% of all colorectal cancers¹⁾. Ogawa et al.²⁾ reported that tumors of the appendix could be diagnosed

preoperatively in 19.8% of patients, which is a rather low preoperative detection rate for malignancy. Carcinoma of the appendix is often found at laparotomy for a diagnosis of appendicitis or is incidentally detected during abdominal surgery for another conditions (particularly, gynecological or urological disease). Since tumors associated with inflammation are difficult to differentiate from appendicitis on the basis of an abdominal routine examination (without CT or US) alone. Our patient underwent laparotomy for a diagnosis of suspected ovarian tumor. It has been reported that cancer of the appendix is sometimes detected because of the presence of ovarian metastases³⁾ or when laparotomy is performed for a diagnosis of urinary tract tumor⁴⁾, showing that it is difficult to distinguish carcinoma of the appendix from primary tumors of adjacent organs. Since the muscularis propria of the appendix is poorly developed, cancer of the appendix tends to give rise to lymph node metastasis, and the prognosis is generally poor⁵⁾. However, mucinous cystadenocarcinoma of the appendix is thought to metastasize less often than adenocarcinoma. Carcinoma of the appendix is uncommon, and the presence of mixed lesions containing both mucinous cystadenocarcinoma and adenocarcinoma components has been reported⁶⁾. But since the pathology of carcinoma of the appendix is poorly understood, however, treatment methods have yet to be established, especially for mucinous cystadenocarcinoma. Nitecki et al.⁷⁾ reviewed 94 patients with various primary tumors of the appendix and reported finding that mucinous cystadenocarcinoma accounted for 8%, adenocarcinoma for 4%, and unusual adenocarcinoid tumors for 2% (the remainder were carcinoid tumors). Metastasis to mesenteric lymph nodes is detected with disease progression in 38% of patients initially treated by appendectomy alone, suggesting that

right hemicolectomy should be selected as the method of reoperation. In a study of 47 patients with mucinous cystadenocarcinoma, appendectomy alone was followed by a 5-year survival rate of approximately 50%, whereas right hemicolectomy improved the survival rate to approximately 80%. Rutledge et al.⁸⁾ stated that whenever carcinoma of the appendix is identified during surgery, they perform right hemicolectomy including lymph node dissection, even if the appendix is ruptured. They also stated that performing curative surgery within several weeks after making the diagnosis by pathologic examination following appendectomy should improve the outcome. Stephenson et al.⁹⁾ reviewed cases of mucinous cystadenocarcinoma with survival for up to 10 years and found that both appendectomy alone and right hemicolectomy were followed by a 5-year survival rate of approximately 70%, but that the 10-year survival rate was only approximately 37% after the former procedure, versus 65% after the latter. These data suggest that mucinous cystadenocarcinoma of the appendix is a slowly growing tumor, and based on these reports the appropriate method of surgery appears to be right hemicolectomy with lymph node dissection. However, the extent of lymph node dissection that should be performed is still unclear. There is no established regimen of postoperative adjuvant chemotherapy for carcinoma of the appendix, and in our patient it was administered according to the schedule for colorectal cancer. Investigation of more patients in the future may help identify adequate treatment. Finally, Nitecki et al.⁷⁾ reported that about 35% of patients with primary carcinoma of the appendix had other malignancies (tumors of the colon, rectum, pancreas, ovary, etc.), suggesting that careful consideration should be given to investigation of other organs during postoperative follow-up after

surgery for primary carcinoma of the appendix.

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