Introduction

Primary appendiceal cancer is relatively rare. Appendiceal cancers are difficult to diagnose preoperatively, because the clinical features are non-specific and similar to appendicitis. Some cases are recognized by frozen sections obtained during aggressive surgical resection. Some appendiceal cancers are occasionally diagnosed by postoperative pathological findings. The prognosis of appendiceal adenocarcinoma is poor, partly because of the advanced stage of illness when it is ultimately identified\(^1\). It is considered that early diagnosis and appropriate surgery will improve the outcome of appendiceal cancer. Computed tomography (CT) and magnetic resonance imaging (MRI) may help to reach a diagnosis\(^2\). We report a case of appendi-
appendiceal adenocarcinoma that was suspected after positron emission tomography/computed tomography (PET/CT) and confirmed during exploratory surgery with single incisional laparoscopic surgery (SILS).

Case report

A 62-year-old woman was admitted to our department with the complaint of right lower quadrant abdominal pain. Her medical history was not remarkable. The body temperature was 36.6°C. Physical examination of the abdomen showed no significant abnormalities (no tenderness or palpable mass). Blood test showed normal haemoglobin, white cell counts (WBC) and C-reactive protein (CRP). Increased carcinoembryonic antigen (CEA, 40ng/dl) was detected; CA19-9 and CA125 were normal. CT revealed a 2.2cm mass located in the right side of the pelvis with remarkable calcification (Figure-1). MRI revealed the right pelvic mass, but was not sufficient to identify its origin and boundaries (Figure-2). In order to rule out metastatic tumor, further examinations were subsequently applied. Gastroscopy, enteroscopy, colonoscopy and MRCP provided no evidence of a tumor in the other digestive organs. Breast cancer was excluded by breast MRI. Cervical cancer was excluded by histological study. (18F-FDG) PET/CT showed a highly hypermetabolic lesion in the right lower abdomen (Figure-3). Considering the small right intrapelvic mass with high uptake signal in PET/CT, elevated CEA, and no elevation of CRP, and ruling out other malignancies, the preoperative differential diagnosis included appendiceal
cancer and right ovarian tumor. After obtaining informed consent from the patient, an exploratory operation employing SILS was performed. A SILS port (SILS™ Port, Covidien Inc.) containing two 5mm trocars and one 10mm trocar was inserted through a 2cm vertical trans-umbilical incision (Figure-4). After insufflation of the abdomen with CO₂, a 10mm 45° angle scope was inserted into the abdomen and two 5mm articulated clamps (Roticulator™, Covidien Inc.) were introduced. During the exploration at the SILS port, the lesion was proven to originate from the vermiform appendix, involving the right fallopian tube and invading to the distal mesoileum. The right ovary was normal (Figure-5 A, B). The lesion was highly suspected to be an advanced tumor of the appendix. Therefore, the SILS was converted to an open procedure with a lower midline abdominal incision. The tumor was resected en bloc for histopathological examination. Pathological examination of the frozen section indicated that the tumor was a primary appendiceal adenocarcinoma. In addition, radical ileocecal resection and oophorectomy were subsequently performed. The involved segments of the ileum and the mesentery were completely resected. A primary functional end-to-end ileocecal anastomosis was made.

The pathological diagnosis of the paraffin section specimen was well- and moderately-differentiated primary appendiceal tubular adenocarcinoma (Figure-6), infiltrating to the right fallopian duct wall, with no positive finding in the orifice or mesenteric lymph node metastasis (0/10). Pathological investigation found mild calcification inside the necrotic tumor tissue, but the calcification was not as remarkable as that indicated by CT. The patient recovered well without any complications and was discharged as expected.

Discussion

Primary appendiceal adenocarcinoma is a rare neoplasm that constitutes less than 0.5% of all gastrointestinal neoplasms\textsuperscript{1}. The incidence of primary appendiceal adenocarcinoma in appendectomy specimens is 0.1%~0.39%, with an increasing ten-
The clinical manifestations of primary appendiceal carcinoma are not specific. Abdominal pain and mass are common symptoms. Preoperative diagnosis of primary carcinoma of the appendix is rarely made. Most patients are not diagnosed until the disease has become advanced. Nitecki reported a series of 94 appendiceal adenocarcinoma patients, of which none were objectively diagnosed before operation, and about 50% of the patients were presumed to be acute appendicitis[1]. A mistaken or delayed diagnosis may compromise the outcome of patients with primary appendiceal cancer. In a report of 532 patients with mucinous epithelial cancers of the appendix, CEA was elevated in 56% and CA 19–9 was elevated in 67.1% of these patients[6]. This study also showed that serum CEA and CA19–9 levels had diagnostic and prognostic significance in primary appendiceal carcinoma.

In our case, the main complaint was right lower abdominal pain. No positive signs were found on abdominal physical examination. WBC and CRP were normal in blood test, but CEA was elevated. The presentation of this patient could not be explained by common appendicitis or adnexitis. Subsequent abdominopelvic CT and MRI showed an impalpable mass situated in the right side of the intrapelvic cavity, and a high uptake signal in the lesion was detected with PET/CT. FDG PET can show increased glycolysis in cancer cells, as well as in activated white blood cells. However in this patient, without features indicating infection, the remarkable hypermetabolic mass was more likely a malignance rather than an infectious disease.

A precise diagnosis of a primary appendiceal cancer is rarely made by imaging studies alone. Neither abdominal ultrasound nor CT can differentiate appendiceal cancer from ovarian carcinoma in most female patients[7]. In the present case, CT and MRI imaging studies were not able to identify the origin of the tumor. A study of the CT features in primary appendiceal tumor found that calcification was presented in 32% (7/22) of patients[2]. Punctate mural calcification was seen in one case of adenocarcinoma in that series. In our case, CT showed remarkable calcification in the lesion, but it was still hard to distinguish calcifications seen in either the appendix or the ovary.

The purpose of surgical exploration with SILS was to identify the origin of the lesion. After confirmation of the tumor’s origin, SILS was converted to open surgery. A surgical specimen was acquired for frozen-section pathological examination. The appropriate surgical procedure was subsequently determined based upon these pathological findings. Single-port laparoscopic surgery has the potential ability to meet these requirements. We selected SILS because it was adequate for exploration and less invasive, and it could easily be converted to open surgery or full laparoscopic surgery. The SILS procedure has been successfully implemented in the fields of general, gynecological and urological surgery[8]. SILS appendectomy is technically feasible and safe[9]. Single-port right hemicolectomy for a patient with persisting polyp was reported by Remzi[10] in 2008. SILS can be completed through a minor incision across the umbilicus. The SILS port is flexible in that it can accommodate up to 3 instruments without significant interference. In our case, we applied a SILS port and 2 forceps. As a result of this procedure, a tumor originating from the appendix was ultimately confirmed. The tumor invaded extensively to the surrounding organs. For the patient’s safety, we converted the procedure directly to open surgery instead of standard laparoscopic surgery. We enlarged the 2cm trans-umbilicus incision to an appropriate direction and length according the minimum requirements. SILS showed its advantage in...
identifying a confusing intra-abdominal situation in the present case. We reported an uneasy case of appendiceal cancer that was ultimately diagnosed with a new tool and procedure, and had interesting findings.

References

症例は62歳，女性で主訴は右下腹部痛。血液検査でCEAとCA19-9の上昇を認め，CTとMRI検査では骨盤の右側に2.2cm大の腫瘤をみとめた。またPET/CT検査では同部位に強い集積を認め虫垂原発の悪性疾患を疑った。手術はSingle incisonal laparoscopic surgery（SILS）で開始したが虫垂腫瘍が右付属器と回腸末端部に癒着しており，開腹移行となった。術中迅速病理検査で虫垂癌の右卵管浸潤と診断され，回盲部切除術を施行した。虫垂原発の悪性疾患の鑑別診断にPET/CTは有用であると考えられた。

キーワード：虫垂癌，PET/CT，SILS