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Primary signet-ring cell carcinoma of vermiform appendix clinically and pathologically presenting as acute appendicitis

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Abstract Primary signet-ring cell carcinoma (SRCC) of vermiform appendix is extremely rare; only three cases have been reported in the English literature. An 89-year-old man suddenly presented right lower abdominal pain, and transferred to a hospital, where he was diagnosed with acute appendicitis by physical data, blood data, and CT. He was further transferred to our hospital for emergency operation. Physical examination showed positive abdominal pain, Blunberg sign, and Rosenstein sign. Blood test showed leukocytosis and increased C-reactive protein. An appendectomy was performed. Gross examination during operation showed inflamed appendix, appendiceal adhesion, and acute peritonitis. Gross pathological examination showed no apparent tumor, but the proximal appendix showed wall thickening and luminal occlusion. The appendix was cut into three sections, and was observed under microscopically. Nests of carcinoma cells were seen in the proximal appendix. The carcinoma was composed of SRCC (70%) and mucinous carcinoma (30%). The size of carcinoma was 6 × 7 mm. The carcinoma cells invaded into muscular layer. No lymphovascular permeation was seen. The cut margins were negative for carcinoma cells. Immunohistochemically, SRCC cells were positive for cytokeratin (CK) AE1/3, CK CAM5.2, CK8, CK18, CK19, CK20, EMA, CEA, CA19-9, p53, Ki-67 (labeling = 30%), CDX2, MUC2, and MUC5AC. They were negative for CK34PE1, CK5/6, CK7, CK14, p63, vimentin, TTF-1, MUC1, MUC 5AC, NSE, synaptophysin, chromogranin, and CD56. No further treatments were performed, because the appendiceal carcinoma was small, the surgical margins were negative and the patient was very old. He was followed up by various imaging modalities. No recurrence or metastasis is found 17 months after the operation.

1. Introduction

Appendiceal cancer is very rare; it accounts for only 0.5% of all gastrointestinal neoplasms [1]. According to a nationwide cancer database (SEER), the age-adjusted incidence of appendiceal malignancies was 0.12 cases per 1,000,000 people per year [1]. Primary appendiceal cancer is diagnosed in only 0.9%–1.4% of appendectomy specimens [2]. Further, signet-ring cell carcinoma (SRCC) of vermiform appendix is extremely rare, accounting for 0.43% of all appendiceal malignancies [2]. To the best of the author's knowledge, there have been only three case reports of appendiceal SRCC [3–5]. Herein, reported is a

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very rare case of primary appendiceal SRCC clinically and pathologically presenting as typical acute appendicitis.

2. Case report

An 89-year-old Japanese man suddenly presented with right lower abdominal pain, and transferred to a hospital, where he was diagnosed with acute appendicitis by physical test, blood data, and CT. CT demonstrated appendiceal swelling. He was further transferred to our hospital for emergency operation. Physical examination showed positive abdominal pain, Blunberg sign, and Rosenstein sing. Blood test showed leukocytosis (13,200/gl: normal 3500–9000) of predominant segmented neutrophils (94%, normal 43%–59%), and increased C-reactive protein (2.24 mg/dl: normal 0–0.3). An appendectomy using laparoscopy was performed. Gross examination during the operation showed inflamed appendix, appendiceal adhesion, and acute peritonitis. Gross pathological examination showed no apparent tumor, but the proximal appendix showed wall thickening and luminal narrowing (Fig. 1). After fixation in formalin, the appendix was cut into three sections, and was observed under microscopically. Nests of carcinoma cells were seen in the proximal appendix (Fig. 2A). The carcinoma was composed of SRCC element (70%) (Fig. 2B) and mucinous carcinoma element (30%). The size of carcinoma was 0.6 × 0.7 cm. The carcinoma cells invaded into the muscular layer (pT1). No lymphovascular permeation was seen. The cut margins were negative for carcinoma cells. No distant metastasis, lymph node metastasis or peritoneal dissemination was found by imaging techniques as well as by clinical findings. The TNM classification and stage of the appendiceal carcinoma were rated pT2N0M0P0H0 and Stage I or Dukes A.

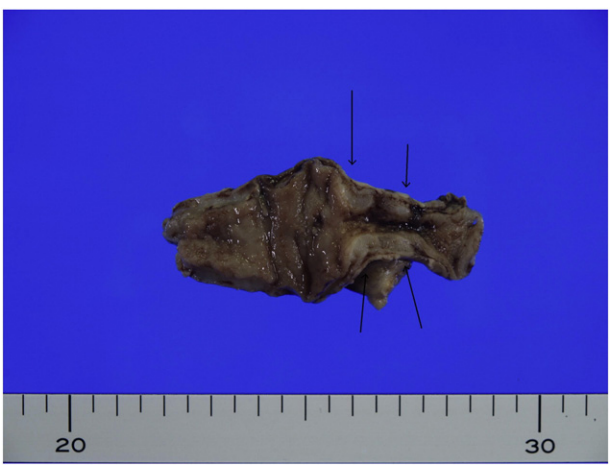

Fig. 1 Gross findings of the appendix. The lumen is open, but the walls of the proximal appendix (right) are thickened (arrows). The appendix distal appendix (left) shows severe appendicitis.

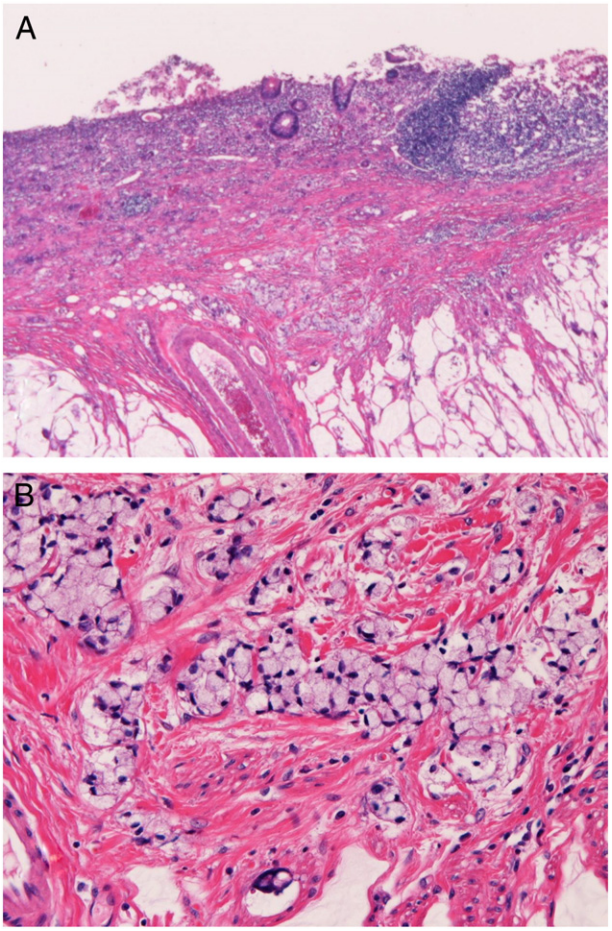

Fig. 2 Histological findings of the appendix. A: The proximal appendix shows proliferation of signet bring cell carcinoma and mucinous adenocarinoma. HE, ×40. B: Higher power view of the signet ring cell carcinoma element. Features of signet ring cell carcinoma are apparent. HE, × 200.

An immunohistochemical study was performed by the use of Dako Envision system and its variation methods (Dako, Glostrup, Denmark), as previously described [6–14]. Immunohistochemically, the SRCC cells were positive for cytokeratin (CK) AE1/3, CK CAM5.2, CK8, CK18 (Fig. 3A), CK19, CK20, EMA, CEA, CA19-9, p53, Ki-67 (labeling = 30%), CDX2 (Fig. 3B), MUC2 (Fig. 3C), and MUC5AC. They were negative for CK34PE1, CK5/6, CK7, CK14, p63, vimentin, TTF-1, MUC1, MUC 5AC, neuron specific enolase (NSE), synaptophysin, chromogranin, and CD56.

The other parts of the appendix showed typical severe acute phlegmonous appendicitis (Fig. 4). No further therapeutic treatments were performed, because the appendiceal carcinoma is small, the surgical margins were negative, and the patient was very old. He was followed up by various imaging modalities. No recurrence or metastasis is found 17 months after the operation.