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Primary minute mucinous adenocarcinoma of vermiform appendix arising from appendiceal diverticulosis☆

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Abstract Primary mucinous adenocarcinoma (MA) of vermiform appendix is extremely rare; only three cases have been reported in the English literature. A 77-year-old man presented with abdominal pain, and was diagnosed with acute appendicitis. Appendectomy was performed. The resected appendix showed submucosal swelling measuring 0.7×0.6×0.6 cm in the tip of appendix. The appendix showed inflammation and numerous diverticuloses. Microscopically, the submucosal swelling was a mucin lake in which adenocarcinoma cells were floating. The adenocarcinoma cells were MA in 80% and signet-ring cell carcinoma in 20%. The carcinoma cells were located in the submucosa, muscular layer and subserosa, sparing the mucosa. No apparent lymphovascular permeation was seen. The surgical margins were negative for tumor cells. The non-tumorous appendix shows numerous diverticulosis, diverticulitis, and appendicitis. Immunohistochemically, the tumor cells were positive for CK CAM5.2, CK AE1/3, CK8, CK18, CK19, CK20, EMA, CEA, CA19-9, MUC1, MUC2, MUC5AC, MUC6, NCAM, p53 and Ki-67 (labeling index = 23%). The tumor cells were negative for CK34BE12, CD5, CK6, CK7, NSE, chromogranin, synaptophysin, CA125, KIT, and PDGFRA. No metastasis has been seen 2.5 years after the operation.

1. Introduction

Appendiceal cancer is rare; it accounts for only 0.5 % of all gastrointestinal neoplasms [1]. According to a nation-wide cancer database (SEER), the age-adjusted incidence of appendiceal malignancies was 0.12 case per 1,000,000 people per year [1]. Primary appendiceal cancer is diagnosed in only 0.9%–1.4% of appendectomy specimens [2]. Further, mucinous adenocarcinoma (MA) and signet-ring cell carcinoma (SRCC) of vermiform appendix are extremely rare, accounting only for 0.21% and 0.43% of all appendiceal malignancies, respectively [2]. To the best of the author's knowledge, there have been only three case reports of appendiceal primary MA [3–5] and also only three cases of primary SRCC [6–9].

2. Case report

A 77-year-old man presented with lower abdominal pain, and was admitted to a hospital. The blood data showed leukocytosis (12,000/μl) and increased C-reactive protein

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(6.2 mg/dl). Lanz's, and Kummel's, McBurney's tenderness, and Blunberg's sign were present. The CT imaging demonstrated a swelling of the appendix. The patient was diagnosed with acute appendicitis and appendectomy was performed.

The resected appendix showed submucosal swelling measuring 0.7×0.6×0.6 cm in the tip of appendix. The appendix also showed severe inflammation and numerous diverticuloses.

Microscopically, the submucosa swelling was a mucin lake in which adenocarcinoma cells were floating (Fig. 1A). The adenocarcinoma cells were MA in 80% (Fig. 1B) and signet-ring cell carcinoma (Sig) in 20% (Fig. 1C). The carcinoma cells were located in the submucosa, muscular layer and subserosa, sparing mucosa. No apparent lymphovascular permeation was seen. The surgical margins were negative for tumor cells. The non-tumorous appendix shows numerous diverticulosis (Fig. 1D), diverticulitis, and acute appendicitis.

Histochemical study was done as previously reported [10]. It showed neutral, carboxylated and sulfated mucins in the mucin pool and carcinoma cells (Fig. 2A and B).

An immunohistochemical study was performed with the use of Dako Envision method as previously reported [11,12]. Immunohistochemically, the tumor cells were positive for cytokeratin (CK) CAM5.2 (Fig. 3A), CK AE1/3, CK8, CK18, CK19, CK20 (Fig. 3B), EMA, CEA, CA19-9, MUC1, MUC2 (Fig. 3C), MUC5AC (Fig. 3D), MUC6, NCAM (Fig. 3E), p53, CDX-2 (Fig. 3F), and Ki-67 (labeling index (LI) = 23%). The tumor cells were negative for CK34BE12, CD5, CK6, CK7, NSE, chromogranin, synaptophysin, CA125, KIT, and PDGFRA. Since no carcinoma was seen in the mucosa and numerous diverticuloses were seen, the carcinoma seemed to arise from the diverticuloses. The location of the tumor is appendiceal tip, no lymphovascular permeation and the surgical margins were negative.

Post-pathological diagnosis whole body examinations including CT, MRI, PET, and endoscope revealed no tumors. The patient was not treated by chemotherapy, but strictly followed up. No metastasis has been seen 2.5 years after the operation. No pseudomyxoma peritonei was seen.

3. Discussion

The current case is the fourth case report of primary MA of the appendix [3–5], and the first report of primary MA clinically and pathologically manifesting as acute appendicitis. In the present study, the adenocarcinoma cells were composed of MA in 80% and signet-ring cell carcinoma (SRCC) in 20%. According to WHO, the only appendiceal adenocarcinoma containing more than 50% of SRCC elements is called appendiceal SRCC [9]. Therefore, the present case is not SRCC but MA. The primary SRCC of the current case was very small (0.7×0.6×0.6 cm) and the

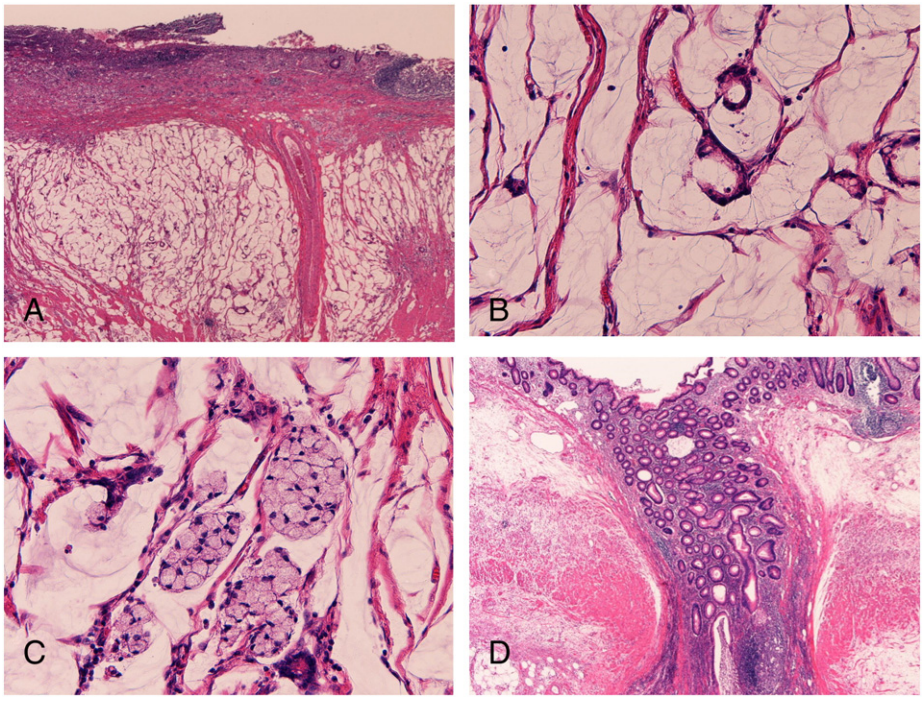

Fig. 1 Histologic findings of the appendix. A: A small mucinous tumor is seen. B: The most of carcinoma cells are tubular adenocarcinoma. HE, ×200. C: A minority is signet ring cell carcinoma. HE, ×100.