Int Canc Conf J (2017) 6:109–113
DOI 10.1007/s13691-017-0285-y

CASE REPORT

A case of low-grade appendiceal mucinous neoplasm with invagination resected laparoscopically

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Received: 2 December 2016 / Accepted: 12 March 2017 / Published online: 19 March 2017

Abstract Low-grade appendiceal mucinous neoplasm (LAMN) is rare disease, and the absence of characteristic clinical symptoms makes preoperative diagnosis difficult. The strategy of treatment for LAMN has not been established. Surgical approach and lymph node (LN) dissection are still controversial. We herein present a case of LAMN with difficulties in making the preoperative diagnosis, which exhibited invagination and was treated by laparoscopy-assisted ileocecal resection with LN dissection. When cystic mass is detected in the bowel, LAMN or mucinous adenocarcinoma should be considered as a different diagnosis. And the laparoscopy-assisted ileocecal resection is a feasible operation for LAMN with careful procedure.

Keywords Appendix · Appendiceal mucocele · Low-grade appendiceal mucinous neoplasm

Introduction

Low-grade appendiceal mucinous neoplasm (LAMN) is included in appendiceal mucinous neoplasm by definition of WHO classification. Pathologically, appendiceal mucinous neoplasm is designated if over 50% of the region consists of extracellular mucin, and classified as LAMN or as mucinous adenocarcinoma. LAMN is rare disease constituting about 1% of all colorectal malignancies [1], 8% of all appendiceal tumors and 0.3–0.7% of all appendectomy specimens [2]. Preoperative diagnosis of LAMN is difficult due to rarity of the disease and the absence of characteristic clinical symptoms [3]. And then, laboratory tests are not specific in patients with LAMN.

Surgical resection is the first choice for LAMN, and a chemotherapy has not yet been established [5]. Recently, the laparoscopic surgery is increasing; however, surgical approach and lymph node (LN) dissection are still controversial because of the difficulty of preoperative malignancy diagnosis and the risk of intraoperative injury of the mucinous tumor [6].

We present a case of LAMN with difficulties in making the preoperative diagnosis that exhibited invagination, and was treated by laparoscopy-assisted ileocecal resection.

Case report

The patient was a 40-year-old female, who had a gradually increasing pain in right lower quadrant with nausea and vomiting. Her body temperature was 37.3 °C and her abdomen was soft, but palpable mass was present in the hypogastric region with tenderness. In the laboratory tests, carbohydrate antigen 125 and carcinoembryonic antigen (CEA) levels were elevated to 54 U/ml and 7.7 ng/ml, respectively. Computed tomography (CT) showed a concentric circles containing cystic structure with calcification at the left side of the lesion (Fig. 1a, b). We diagnosed the invagination of intestine and an emergency laparoscopic examination was performed; nevertheless intestinal tract showed no abnormal finding.

After laparoscopic examination, the abdominal pain persisted. Total colonoscopy revealed a 5-cm

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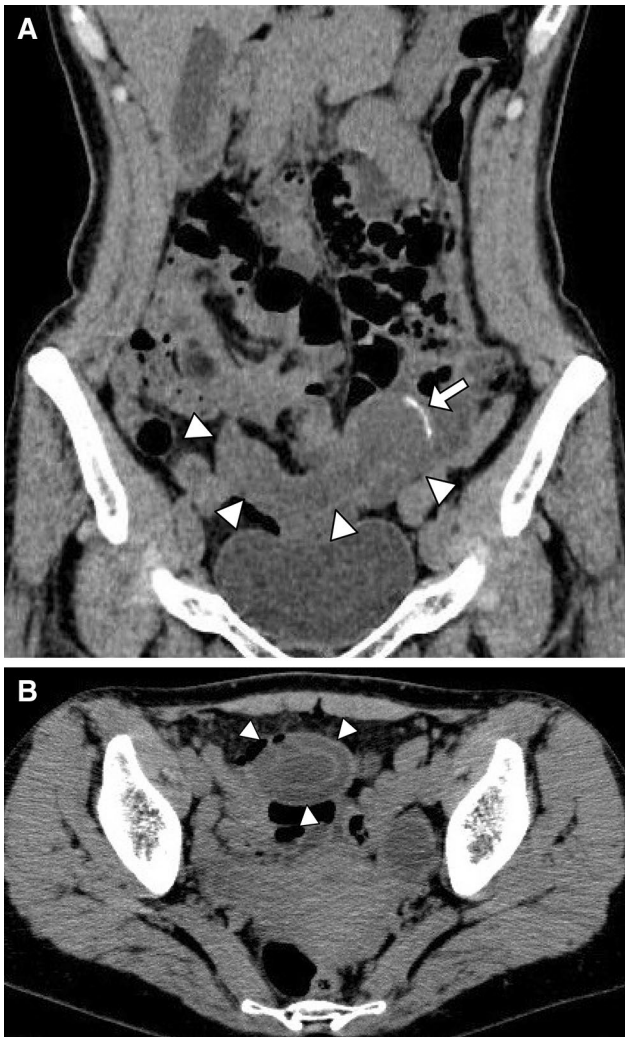

Fig. 1 Abdominal CT scan (**a** and **b**) showed smooth cystic bulge with calcification at the left side of the lesion (*arrow*). It showed a concentric circles structure (**b**)

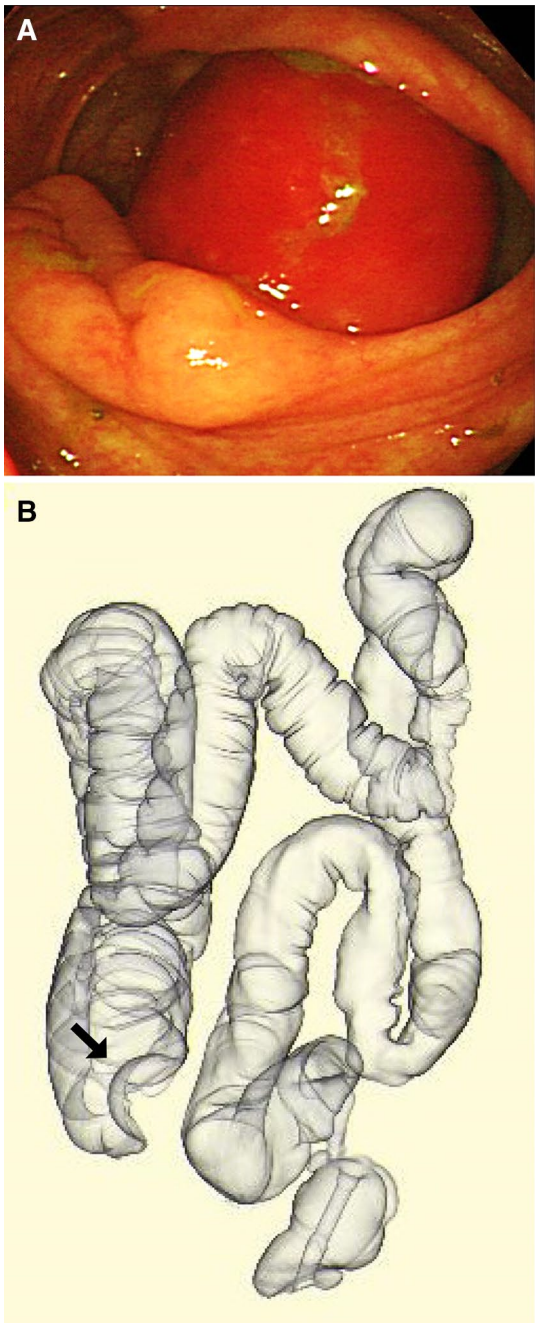

Fig. 2 Colonoscopy revealed a submucosal tumor in the transverse colon, and it was moved easily to cecum (**a**). CT colonography showed smooth hemispheric defect of the cecum (**b**)

submucosal-tumor-like lesion in transverse colon, and it was moved easily to cecum (Fig. 2a). CT colonography showed smooth hemispheric defect of the cecum (Fig. 2b). The mucinous adenocarcinoma of the appendix or LAMN was suspected, and laparoscopy-assisted ileocecal resection was performed. On entering the peritoneal cavity, swollen cecum and appendix with smooth serosal surface were found. Laparoscopy-assisted ileocecal resection with D2 LN dissection was performed. Ileum was excised at 5-cm oral side from terminal ileum, and ascending colon was excised at 8-cm anal side from tumor by electrocautery. Reconstruction of ileum and ascending colon was performed by end-to-end anastomosis using a two-layer anastomosis technique.

The appendiceal tumor measured 10 cm in length and 4 cm in diameter, and it continued into the cecum. The tumor was filled with jelly-like mucus (Fig. 3a, b). Histopathological examination revealed the papillary growing glandular epithelium with tall columnar mucinous cells. It showed elongated swollen nuclei at the base and low-grade dysplasia (Fig. 3c). Final pathologic diagnosis of the mass was LAMN.

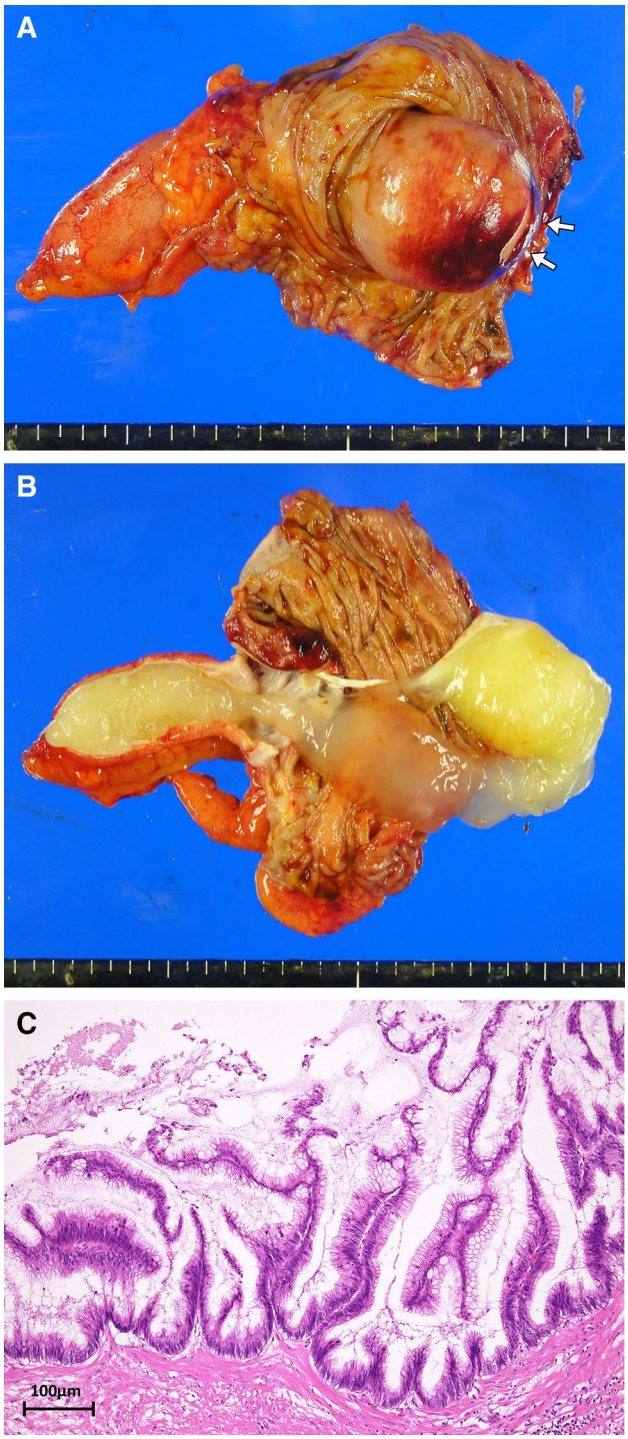

Fig. 3 Macroscopic view of the resected specimen showed the submucosal tumor originated from appendix and the appendiceal orifice (*arrows*). The tumor was filled with jelly-like mucus (**a** and **b**). Histopathologic image demonstrating the papillary growing glandular epithelium covered by a single layer of columnar mucinous cells, and elongate swelling nuclei at the base (**c**)

The postoperative course was uneventful; patient was discharged 8 days after surgery and remained well with no recurrence for 16 months.

Discussion

WHO classification of tumors 2010 divided appendiceal mucinous neoplasm into mucinous adenocarcinoma and LAMN, previously called "mucinous cystadenoma." Pathologically, LAMN exhibits mostly epithelial villous adenomatous changes without malignancy and the prognosis is good. And mucinous adenocarcinoma demonstrates glandular stromal invasion, desmoplastic reaction, and/or presence of epithelial cells in the peritoneal implants and associated with a very poor survival rate and a high rate of lymph nodes or liver metastases [5]. While the nomenclature is controversial because LAMN can lead to diffuse peritoneal dissemination of the cells within the peritoneal cavity, known as pseudomyxoma peritonei, even distant metastases [7]. Like our case, LAMN is incidentally discovered at surgery or during imaging for unrelated symptoms, in patients with chronic abdominal pain, palpable mass in the right lower abdomen, or in patients with an intestinal tract invagination [4, 8–11]. Laboratory tests are not specific but some case reports showed that high serum CEA levels could be detected in patients with LAMN [12].

The diagnosis of LAMN is established basically by abdominal CT scan; its appearances include a well-encapsulated, round, and thin-walled cystic mass. Calcification is found in 50% of cases [13] and mucoceles less than 2 cm are rarely malignant but larger mucoceles (6 cm or more) are usually associated with adenocarcinoma and a higher perforation rate (20%) [5]. The enhancing nodules in the mucocele wall are suggestive of mucinous adenocarcinoma [5] but preoperative diagnosis of malignancy was difficult [3]. Colonoscopy may show a pathognomonic image called "volcano sign" [14]; the appendiceal orifice may be seen in the center of a mound-like elevation of the cecal wall [15]. In our case, the cecum was loosely fixed to retroperitoneum, and appendix was in an abnormal position by invagination, so we could not suppose appendiceal mucocele. However, LAMN or mucinous adenocarcinoma should be considered as a different diagnosis by primary CT findings. And we should have inspected the cecum and appendix more carefully at laparoscopic examination.

Surgical resection is considered as the only curative treatment for LAMN and appendectomy is performed when the appendiceal root is intact [16]. Even in the case of appendiceal mucinous adenocarcinoma, González et al. reported

that overall survival rate of appendectomy was similar to that of right hemicolectomy and suggested hence appendectomy or cecectomy is preferable [17]. The incidence of LN metastasis has been reported in 1.7% cases of LAMN [18]; however, that of mutinous adenocarcinoma ranging from 25 to 50% [19] and preoperative definitive diagnosis may be difficult [20]. Therefore, ileocecal resection or right hemicolectomy with LN dissection is often performed [4]. In our case, laparoscopy-assisted ileocecal resection with D2 LN dissection was performed since perioperative diagnosis of progression was difficult.

Recently, the laparoscopic resection for LAMN has been increasing, and this approach is minimally invasive as demonstrated by minimal postoperative pain and quick recovery. The rupture of LAMN leads to pseudomyxoma peritonei that worsens the outcome of the disease [22] and previous report showed cases of pseudomyxoma peritonei due to perioperative perforation in both open and laparoscopic surgery [23]. Some authors recommended the open surgery to avoid the rupture of the mucocele [6, 24]. But there is no comparative study between open and laparoscopic surgery. To prevent the injury of the mucocele, surgeon should not grasp a tumor directly and ensure the margin especially in the invagination case [11], and sometimes it needs mobilizing the entire right hemi-colon [25]. In the accredited facilities, laparoscopic surgery including mobilization of the colon and LN dissection is standard procedure, And in this case laparoscopy-assisted ileocecal resection was safely performed without grasping the appendix, and D2 LN dissection was successfully completed.

In conclusion, we reported a case of LAMN with difficulties in making the preoperative diagnosis that exhibit invagination. And the laparoscopy-assisted ileocecal resection and D2 LN dissection were performed safely for LAMN.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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