Mucocele of the Appendiceal Stump due to Benign Mucinous Cystadenoma

D.P. KORKOLIS1, K. APOSTOLAKI2, G.D. PLATANITIS1, J. TZORBATZOGLOU1, I.G. KARAITIANOS1 and P.P. VASSILOPOULOS1

1First Department of Surgery and 2Department of Surgical Pathology, Hellenic Anticancer Institute, “St. Savvas” Hospital, Athens, Greece

Case Report

A 49-year-old Caucasian female presented with a 6-month history of vague right lower quadrant pain radiating through to the back. The patient had undergone an appendectomy 25 years previously. Total abdominal hysterectomy with bilateral salpingo-oophorectomy for bleeding myomatous disease of the uterus had been also performed 5 years earlier, in our institution.

The patient described no fever, malaise, nausea or vomiting, change in bowel habits, rectal bleeding or weight loss. Physical examination of the abdomen revealed slight tenderness in the vicinity of the old appendectomy scar. Laboratory analysis, including serum levels of CEA and CA-125, were found to be within normal limits. Colonoscopy showed no pathological findings. An abdominal CT scan demonstrated a well-defined, elliptical, 7x5 cm cystic mass, lying at the inferior aspect of the cecum, just above the iliac fossa. The lesion had smooth walls, scattered mural calcifications and no surrounding inflammation (Figure 1).

The patient underwent exploratory laparotomy through a midline incision. A firm, well-encapsulated and calcified tumor, 8x5.5x4 cm in size, was discovered at the base of the cecum. It was easily mobilized from adjacent structures and found to originate from the 1-cm, unburied, appendiceal stump. The lesion was removed intact, together with the appendiceal remnant and a safe rim of the cecal wall (Figure 2). Frozen section examination of the lesion and the margin of resection were both negative for malignancy.

The resected mucocele contained about 75 ml of mucin. Histopathological evaluation revealed crowded, irregular, villotubular structures, with mild to moderate epithelial atypia together with acellular mucin pooling. No evidence of malignancy was found (Figure 3). The final diagnosis ascertained the presence of an appendiceal stump mucocele associated with a benign mucinous cystadenoma.

Correspondence to: Dimitris P. Korkolis, MD, 22 Socratous Street, 1st Floor, 14561 Kifissia, Athens, Greece. Tel: +30 210 8083743, Fax: +30 210 8012689

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The patient had an uneventful recovery and was discharged home on the fifth postoperative day. Eighteen months after surgery, she is free of symptoms with no evidence of recurrence.

Discussion

Although widely used, the term mucocele is inherently imprecise and inclusive of both benign and malignant lesions. Mucoceles are found in only 0.07% to 0.3% of appendectomies and are histologically subdivided on the basis of the World Health Organization classification (5, 6). Specifically, simple mucocele is appendiceal dilation with accumulation of mucus due to obstruction of the lumen. Mucinous cystadenoma is defined as the dilated, mucus-filled appendix containing hyperplastic adenomatous mucosa, and cystadenocarcinoma as the presence of adenocarcinoma associated with a dilated, mucus-filled appendix. The last two types comprise the majority of cases.

Yeong et al. (3) reported, for the first time, a unique case of a mucocele of the appendiceal stump related to a papillary cystadenocarcinoma arising at the base and causing pseudomyxoma peritonei. More recently, Lien et al. (4) described a mucinous cystadenoma of the appendiceal stump as the cause of a mucocele developed 30 years after appendectomy. Our report, in which a specimen was available, represents the second ever described case of an appendiceal stump mucocele associated with a benign condition.

Clinical manifestations of appendiceal mucoceles are non-specific, thus diminishing the rate of accurate preoperative diagnosis. These include abdominal pain, palpable mass, weight loss, nausea and vomiting, change in bowel habits, appendicitis or lower gastrointestinal bleeding (7). One-quarter of mucoceles are asymptomatic. However, when symptoms are present a cystadenocarcinoma is more likely to be encountered. In general, only one-third of them are malignant, whereas the risk of malignancy increases with size (8). These data may be influenced by the fact that routine incidental appendectomy is generally performed during gynecological or other surgical procedures, increasing the rate of incidental benign appendiceal mucoceles, as they would not be detected otherwise. In
Figure 2. The resected specimen included an intact, firm, well-encapsulated and calcified tumor, measuring 8x5.5x4cm.

Figure 3. Microscopic evaluation revealed crowded, irregular, villotubular structures, with mild epithelial atypia and acellular mucin pooling, suggesting the presence of a benign mucinous cystadenoma.
addition, there might be a referral bias in favor of tertiary referral centers for symptomatic patients with a large abdominal mass, rectal bleeding and weight loss, who are more likely to harbor an appendiceal cystadenocarcinoma.

Of note is the elevated incidence of associated neoplasms which occur in almost one-third of cases. These include neoplasms of the colon and rectum and, less frequently, ovarian, endometrial, bladder, prostate or other gastrointestinal tract tumors (9). It would, therefore, seem prudent to recommend surveillance colonoscopy in, at least, those patients with an established diagnosis of an appendiceal mucinous cystadenoma.

The initial diagnostic modalities include ultrasound, barium enema, colonoscopy and CT scan. The latter has been invaluable in the diagnosis of appendiceal mucocele. It usually reveals a low-attenuation, well-encapsulating, cystic mass with smooth walls located at the base of the cecum, sometimes having mural calcifications (10).

All mucoceles should probably be removed in order to eliminate the chance of progression to malignancy. Although an open approach is usually recommended, laparoscopic appendectomy for mucocele removal has been described. Caution is advisable because of the risk of port-site recurrences, as well as accidental intraoperative rupture, which increases the likelihood of peritoneal spread (11, 12).

At the time of surgery, a spontaneous appendiceal perforation or any mucus extravasation from the appendiceal lumen is strongly suggestive of an underlying malignancy and should prompt an oncologically-performed right hemicolecystectomy. If the lesion is confined to the appendix, has no surrounding infiltration and is followed by negative frozen-section examination and clear margins of resection, wide local resection is usually adequate (2, 13).

Conclusion

Mucocele of the appendiceal stump is an extremely rare oncological entity. It should, however be, included in the differential diagnosis of a cystic mass detected in the area of a previous appendectomy, necessitating thorough investigation and prompt surgical intervention.

References


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