Mucinous Cystadenoma of the Appendix: Diagnosis, Surgical Management, and Follow-up

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PURPOSE: To review the diagnostic examination and clinical presentation of mucinous cystadenoma of the appendix.

METHODS: Case report from experience at an Air Force tertiary care hospital in a 66-year-old woman with chronic right lower quadrant pain.

RESULTS: After extensive preoperative evaluation and subsequent diagnostic laparoscopy, a right hemicolectomy was performed for a mucinous cystadenoma of the appendix.

CONCLUSIONS: Appendiceal mucinous cystadenoma is a rare entity found in only 0.3% of appendiceal specimens. Preoperative evaluation with radiologic and endoscopic methods is helpful but not always diagnostic. Although a benign disease process, complications from rupture, invasion into adjacent organs, or recurrence warrant adherence to strict oncologic principles for resection. (Curr Surg 60:341–343. © 2003 by the Association of Program Directors in Surgery.)

KEY WORDS: mucinous cystadenoma, appendix, appendiceal tumors, mucinous cystadenocarcinoma

INTRODUCTION

Mucinous cystadenoma of the appendix is an uncommon clinical finding. It is the most common classification of what has been generally termed "mucocele" of the appendix. Overall, appendiceal mucoceles make up about 0.3% of appendix specimens.[1] The average general surgeon can anticipate encountering several in one's career. In fact, based on retrospective case series, appendiceal mucoceles are second only to carcinoid tumors.[2] The world literature highlights many case reports of unusual findings in association with appendiceal mucoceles. A common theme throughout is that preoperative and often intraoperative diagnosis remains elusive.

The term mucocele simply refers to a cystic mass filled with mucin, as in a dilated appendix. Simple mucoceles secondary to inflammatory obstruction do occur with or without atrophic epithelial changes. These are benign entities without pathologic significance. However, a more descriptive classification system based on the changes to the underlying appendix epithelium has been proposed to try and define biologic behavior. These histopathologic lesions and percentage of overall mucoceles are then defined as mucosal hyperplasia (25%), mucinous cystadenoma (63%), and mucinous cystadenocarcinoma (12%).[3]

CASE REPORT

Our patient was a 66-year-old woman with a history of stage II breast carcinoma treated with modified radical mastectomy and chemotherapy in 1994. She presented to her gynecologist complaining of a vague right lower quadrant pain for several months. She denied any weight loss, fevers, flushing symptoms, or change in bowel habits. On pelvic examination, she was noted to have a palpable right adnexal mass, but pelvic ultrasound was normal. A computed tomography (CT) scan was obtained and revealed a nonobstructing 2-cm by 3-cm ileocecal mass (Fig. 1). A barium enema was also obtained that showed only mild diverticulosis and no intraluminal or extraluminal lesion. She was referred to our surgical service; at which time, she underwent colonoscopy. A prominent mass in the region of her ileocecal valve was noted with normal overlying mucosa (Fig. 2). Biopsies were benign.

At this point, surgical options were discussed and the patient was taken for diagnostic laparoscopy. At the operation, a smooth mass was noted at the base of the appendix that continued into the cecum at the ileocecal junction. A formal low midline celiotomy was performed with 4-quadrant exploration. The ovaries appeared normal. No other lesions in the liver or mesentery were found. The mass extended to the ileocecal junction; so a right hemicolectomy was performed, including the right mesocolon and nodes. The patient recovered unevent-

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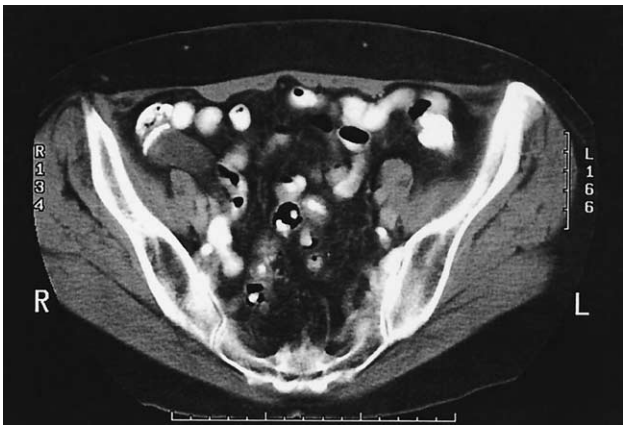

FIGURE 1. Computed tomography with paracecal mass.

fully. Final pathologic analysis of the mass was mucinous cystadenoma of the appendix (Fig. 3). Histologically, there was mucinous epithelium with small papillary projections and areas of low-grade dysplasia. There was also extravasation of mucin into the submucosa, but no epithelial cells within the mucin pools.

DISCUSSION

The differential diagnosis of a cystic right lower quadrant mass is extensive and includes primary adenocarcinoma, carcinoid tumor, mucinous cystadenoma, mucinous cystadenocarcinoma, lymphoid hyperplasia, lymphoma, peri-appendiceal abscess, and in females, ovarian tumors. When symptoms do not suggest an inflammatory, hemorrhagic, or acute obstructive process, ancillary studies may help to refine the diagnosis. The most common presenting symptom associated with mucinous cystadenoma has been abdominal pain; however, one-fourth of patients are asymptomatic and are found incidentally.[4] Case reports of bleeding, intussusception, and local invasion into surrounding structures are described with associated symptomatology.[5-7]

We relied on ultrasound, CT, barium enema, and colonoscopy to assist in preoperative diagnosis. Retrospective series

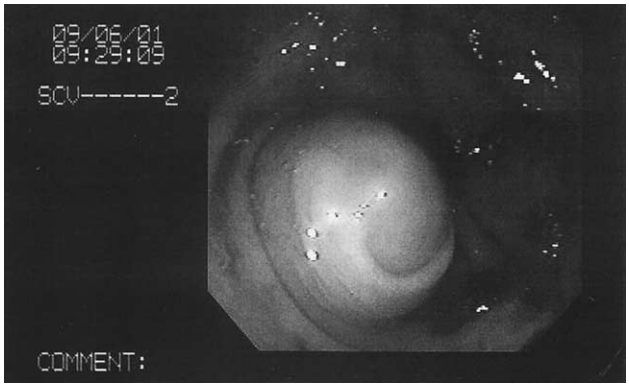

FIGURE 2. Colonscopic view of mass adjacent to ileocecal valve.

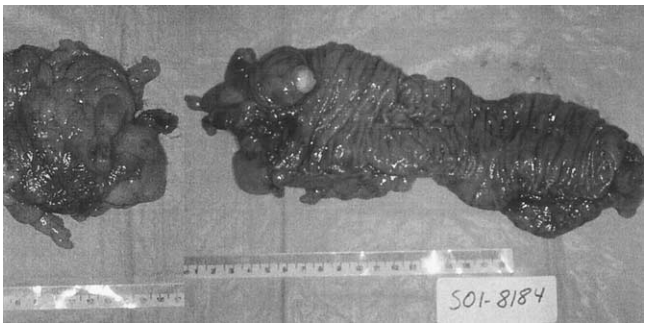

FIGURE 3. Gross specimen: (left) external view and (right) internal view shows mucinous contents upon opening of cyst.

have looked at radiologic findings of pathologically confirmed mucinous cystadenoma and mucinous cystadenocarcinoma to determine if any findings were suggestive of malignancy.[8] By ultrasound, cystic masses with varying internal echogenicity and a layered wall with calcification may be seen. Computed tomography findings show well-encapsulated cystic masses with low attenuation, usually without associated appendiceal inflammation. Varying thickness of the mass wall did not seem to correlate with malignancy, although contrast-enhancing nodules may suggest cystadenocarcinoma. Computed tomography does help in evaluating other pathologic processes. Colonoscopy is usually nondiagnostic, as mucosal biopsies will often be normal. Suggestion of extrinsic compression or mass protrusion of the appendiceal orifice, as in our case, can be helpful.

Histologically, the classification scheme for appendiceal mucoceles outlines a spectrum of epithelial changes that are analogous to changes in the colon. Mucosal hyperplasia and mucinous cystadenomas are akin to colonic hyperplastic polyps and adenomatous polyps.[9] Concomitant cases of mucinous cystadenomas and colon cancer have been described with an incidence of up to 20%.[3,10] The progression to malignancy has not been proven for mucinous cystadenomas, but it is suggested. The intraoperative distinction between benign mucinous cystadenoma and mucinous cystadenocarcinoma remains difficult without the clinical picture of wall invasion, local spread, or widespread peritoneal implants (pseudomyxoma peritonei). This can be made more difficult in the face of acute inflammation. Histologic analysis is not always straightforward and relies on evidence of stromal invasion and marked nuclear atypia of the appendix epithelium to diagnose mucinous cystadenocarcinoma. Initial reported series seemed to suggest that only mucinous cystadenocarcinoma with rupture had the ability for intraperitoneal spread. In these series, pseudomyxoma peritonei was found in all cases at diagnosis. These had dismal 25% 5-year survival.[3] However, Prayson et al. reported a series of 19 patients with pseudomyxoma peritonei, confirmed by malignant epithelium in peritoneal implants, where greater than 75% had only mucinous cystadenomas of the appendix as the primary lesion. Although these patients were not immune from complications of bowel obstruction and recurrence, overall they fared better than did those with cystadenocarcinoma. They also

noted that patients with absence of neoplastic epithelial cells in their peritoneal implants had no evidence of disease on follow-up out to 7 years.[11]

The morbidity/mortality associated with both cystadenoma and cystadenocarcionoma stems from rupture and intraperitoneal spread of mucin-producing epithelium. No reports of lymphatic or hematogenous spread are found in the literature. It is apparent that both mucinous cystadenocarcinomas and cystadenomas have the capacity to produce pseudomyxoma peritonei, although the clinical burden is likely dependent on the degree of dysplasia.

Regardless of the ultimate histology, extreme care must be taken in tissue handling. Masses in the appendix body without local invasion or cecal involvement can be treated with simple appendectomy and mesoappendix excision. Most seem to advocate completion right hemicolectomy if final histology proves cystadenocarcinoma. Any mass involving the cecum or adjacent organs should be resected by a right hemicolectomy. Intraoperative exploration of the entire gastrointestinal tract and ovaries in females is warranted. All gross peritoneal implants should undergo a biopsy and be removed with grading of the degree of epithelial atypia for prognostic purposes.[12]

In our case, we chose initial diagnostic laparoscopy to assist with operative planning. It allowed assessment of the mass and evaluation of the remainder of the abdominal cavity. Ultimately, a mass freely involving the appendix body could be treated by a standard open approach as for appendectomy. When we determined that it encroached on the ileocecal junction, we felt a right hemicolectomy was warranted and we proceeded through a midline incision. Successful removal of appendiceal mucoceles laparoscopically, even those requiring ileocecal resection, has been reported. The ability to safely handle tissues and adequately assess the cecum for the lesion extent is a clinical decision left up to the operating surgeon. Reports of missed lesions and widespread peritoneal implants after laparoscopic removal would tend to support open conversion in most circumstances.[13,14]

Cystic lesions of the appendix especially mucinous cystadenoma and mucinous cystadenocarcinoma are difficult to diagnose despite extensive preoperative evaluation. Although the surgical treatment is straightforward, proper management of the incidentally found lesion requires understanding of the potential complications of widespread peritoneal disease. Thorough intraoperative gastrointestinal, ovarian, and peritoneal examination is required. Follow-up colonoscopy and pelvic examination is also warranted for the high association with other colon and ovarian malignancies.

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