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Asymptomatic Primary Mucinous Cystadenocarcinoma of the Appendix with a Large Abdominal Mass: Report of a Case

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Abstract: A case of cystadenocarcinoma of the appendix with a large cystic lesion is reported. A 49-year-old man undergoing a routine ultrasonic scan was incidentally found to have an abdominal mass measuring some 30 cm in size. The clinical presentation was asymptomatic, and the patient underwent a laparotomy without ascertaining a diagnosis preoperatively. The lesion, which derived from the appendix, was removed and was found to be cystic and contained huge amounts of mucin. The histological findings revealed a well-differentiated cystadenocarcinoma of the appendix, and immunohistochemical staining of the epithelium and mucinous implants in the mass demonstrated a positive reaction for carcinogenic antigens, including carcinoembryonic antigen and carbohydrate antigen.

Key Words: Adenocarcinoma of the appendix, CEA, CA19-9

Introduction

Adenocarcinoma of the appendix is very uncommon and is also rarely diagnosed preoperatively.[1–4] The most commonly demonstrated presentations of malignant tumors of the appendix are acute appendicitis, a right quadrant abdominal mass, or an acute intestinal obstruction.[1,4] Asymptomatic adenocarcinoma of the appendix has seldom been reported except for incidental discoveries at the time of laparotomy for unrelated conditions. We herein report a case of cystadenocarcinoma of the appendix which presented as a large abdominal cavity mass with an increased serum level of CEA. This appears to be the first reported case of asymptomatic cystadenocarcinoma of the appendix in which a huge cystic lesion was initially demonstrated.

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Case Report

A 49-year-old man was examined ultrasonographically as part of a mass screening when a cystic mass measuring 30 cm in size was incidentally found in the left abdominal cavity. Computed tomography (CT) also revealed a 30 × 20 × 10-cm mass which was well-encapsulated, noninvasive, and cystic (Fig. 1). A barium meal and follow-through examination showed that the abdominal mass compressed the small intestine. A barium enema also revealed a massive compression of the transverse, descending, and sigmoid colon, while the appendix was not enhanced. An investigation of tumor markers in the serum revealed an elevated carcinoembryonic antigen (CEA) level at 12.3 ng/ml (normal <2.5 ng/ml) while carbohydrate antigen 19-9 (CA19-9) was 23 IU/ml (normal <37 IU/ml), which suggested the possibility of epithelial malignancy.

The patient underwent a laparotomy, and a fibrous adherent mass involving the tip of the appendix was dissected free from the small intestine, colon, and bladder wall; thereafter an appendectomy was carried out. The mass revealed an expansive growth into the left abdominal cavity without any neoplastic invasion (Fig. 2). It was totally cystic, and contained 1,050 g of gelatinous mucin (Fig. 3). There was no mucus dissemination or ascites in the abdominal cavity. On observing a cut section, the epithelium of the appendix tip focally filled the lumen. The tumor marker levels of CEA and CA19-9 of the involved mucinous implants revealed substantial increases to 28,000 ng/ml and 11,000 IU/ml, respectively.

Histologically, the tumor formed a papillary growth of the atypical epithelium showing a remarkable

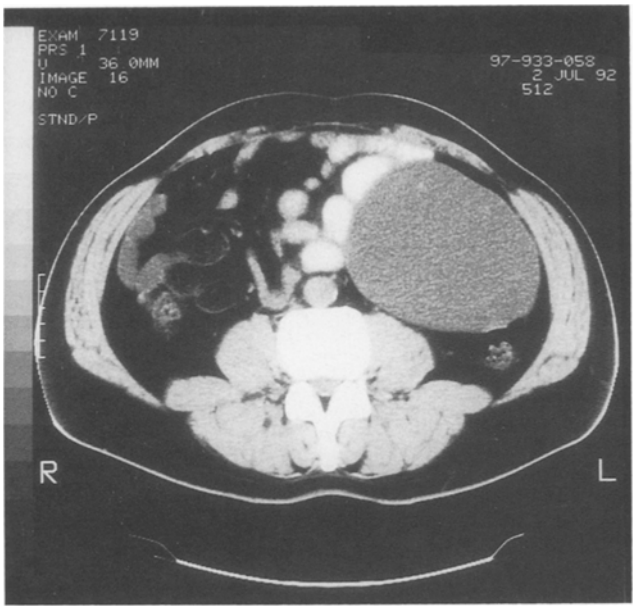

Fig. 1. Computed tomography shows a large cystic mass in the left abdominal cavity

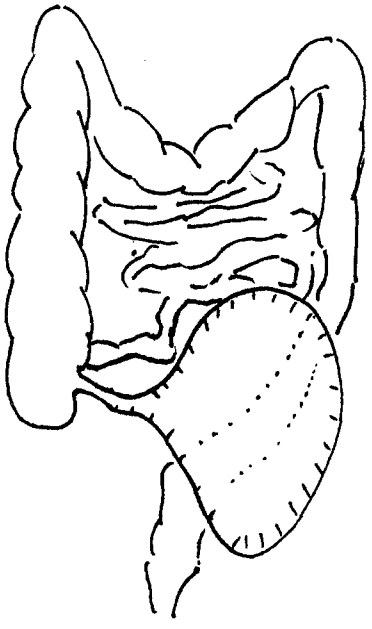

Fig. 2. Schema of the location of the cystic mass in the abdominal cavity

amount of mucin production. The tumor cells invaded to the muscular layer of the appendix. Immunohistochemical staining revealed positive reactions for CEA and CA19-9 in all tumor cells. Numerous crystals and minute calcifications associated with foreign reaction were also noted in the cystic wall.

The postoperative convalescence was uneventful and the patient was discharged 15 days after surgery.

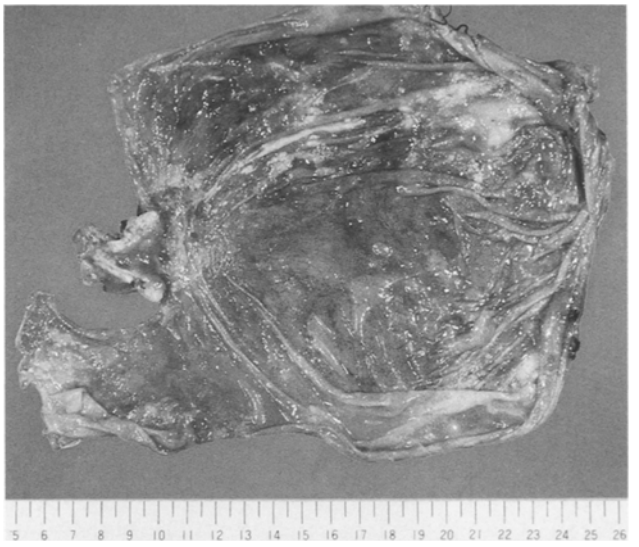

Fig. 3. Resected tumor showing the cyst and appendix

Discussion

The incidence of adenocarcinoma of the appendix is rare, ranging from 0.025%–0.2% of appendectomy specimens.[5,6] In addition, an accurate preoperative diagnosis of malignancy of the appendix has seldom been made[7] since most cases either underwent laparotomy for acute appendicitis, or it was found unexpectedly in the course of other pelvic or abdominal operations.[1–4] On the other hand, a huge-sized mucinous cystadenocarcinoma could be found to favor symptoms caused by its cutaneous invasion, vesical fistula, or bowel obstruction.[8,9]

The most common presentation of adenocarcinoma of the appendix is appendicitis, appendicular mass, or intestinal obstruction.[1,4] Our case was unusual since the patient had not complained of any symptoms such as abdominal pain, diarrhea, or constipation. Furthermore, as our patient had a cystic mass which expanded into the left abdominal cavity, it was difficult to accurately ascertain the origin of this tumor. In addition, this patient had never noticed any abnormality of his abdomen before the ultrasonic scanning demonstrated the mass. Hence, in this case, the lack of such symptoms and the laterality of the tumor location led us to perform an exploratory extirpation of the abdominal mass without first securing an accurate preoperative diagnosis.

In the absence of an inflammatory episode, as in this case, the tumor may remain asymptomatic until it presents with the spread to other abdominal or pelvic organs or to the skin.[4] Another possibility is that the

disseminated mucinous implants derived from the ruptured mass demonstrate severe abdominal distention. Pseudomyxoma peritonei, which might originate from ruptured mucinous tumors of the appendix or ovary, has been reported to have a high incidence of recurrence.[10–12] In our case, the cystic mass was completely encapsulated, and contained a huge number of gelatinous implants. Since the cystic wall had an extremely thin membrane, we were fortunate in being able to resect the mucocele in an unruptured condition.

Virtually the only positive laboratory finding in this patient was the elevated serum CEA level, which is frequently associated with malignancy especially in the digestive system. Since cystadenocarcinoma of the ovary or pseudomyxoma peritonei has been reported with an increased CEA level of serum or mucinous implants,[13–15] we investigated the immunohistochemical findings of CEA and CA19-9 in cystadenocarcinoma of the appendix. A marked increase in these antigens in the mucinous products along with substantial immunohistochemical staining of the epithelium in this case also helped in determining neoplastic behavior.

A comparative study between colonic adenocarcinoma and cystadenocarcinoma of the appendix suggests that the colonic adenocarcinoma of the appendix has the same prognosis as other colon adenocarcinomas and thus requires a right hemicolectomy, while mucinous cystadenocarcinomas are slowly progressive.[16] In this case, however, although it was a cystadenocarcinoma, a higher serum level of CA19-9 in colorectal adenocarcinomas is also correlated with its clinical staging status;[17] therefore an additional radical resection and strict clinical follow-up should be completed. In fact, we are planning to perform a second-look right-hemicolectomy in this patient. In addition, the serum levels of CEA and CA19-9 were downregulated to 1.0 ng/ml and 18 IU/ml, respectively, at 4 weeks after operation, suggesting that these markers could reflect the clinical status of this patient.

In summary, we herein reported a case of cystadenocarcinoma of the appendix in a 49-year-old man. Unlike common adenocarcinoma of the appendix, this patient was asymptomatic, and the cystadenocarcinoma was demonstrated to be a huge-size cystic mass. The cystic lesion was successfully resected, and there was no mucinous dissemination to the abdominal cavity. Furthermore, an immunohistochemical investigation of CEA and CA19-9 was performed, suggesting that the CEA serum level might be a useful biomarker for either the diagnosis or clinical follow-up in cystadenocarcinoma of the appendix.

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