

Aspecific presentation of a cystadenocarcinoma originating from the appendix

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Section: Abdominal imaging

Imaging Technique: Ultrasound

Imaging Technique: CT

Imaging Technique: CT

Imaging Technique: CT

Case Type: Clinical Cases

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Patient: 82 years, female

Clinical History:

An 82-year-old female patient presented with fever, nausea, vomiting and pain in the right flank. Her medical history showed that she had undergone a total hysterectomy and bilateral adnexectomy 40 years ago. The laboratory findings showed an elevated sedimentation and white blood cell count. An ultrasonographic examination of the abdomen showed the presence of a 10 cm large thick-walled cystic mass with a slightly hyperechogenic and heterogeneous content. A discrete vascularization of the wall of the cystic mass was seen. A Computed Tomography (CT) examination of the abdomen was performed. It was found that the large cystic mass was located against the cecum, displaying discrete enhancement of the cystic wall with a small amount of air and oral contrast agent in its lumen. A right hemicolectomy was performed. The pathologic examination of the resected specimen was conclusive.

Imaging Findings:

An 82-year-old female patient presented with fever, nausea, vomiting and pain in the right flank. Her medical history showed that she had undergone a total hysterectomy and bilateral adnexectomy 40 years ago. The laboratory findings showed a sedimentation level of 89 mm/h and white blood cell count of $14.100/\text{mm}^3$. An ultrasonographic examination of the abdomen showed the presence of a 10 cm large thick-walled cystic mass with a slightly hyperechogenic and heterogeneous content (Fig. 1). A discrete vascularization of the wall of the cystic mass was seen. Differential diagnosis included an acute appendicitis with abscedation, a mucocele of the appendix (with suspicion of an underlying tumor regarding the age of this patient), a duplication cyst, a mesenteric cyst or a congenital cyst with internal hemorrhage. A tumoral process of the ovaries was not included in the differential diagnosis because of the medical history of this patient. A Computed Tomography (CT) examination of the abdomen was performed in the venous phase, after the administration of an intravenous contrast material. It was found that the large cystic mass was located against the cecum, displaying discrete enhancement of the cystic wall (Fig. 2). A small amount of oral contrast agent (Fig. 3) was seen together with a delicate amount of air (Fig. 4) in the lumen of the cystic mass. A limited inflammatory reaction in the surrounding mesenteric fatty tissue was noted. No ascites was present neither were lymphadenopathies visualized. A right hemicolectomy was performed. The pathologic examination of the resected specimen (Fig. 5) was conclusive.

Discussion:

Primary appendiceal neoplasms are uncommon, and comprise approximately 0.5%–1% of appendiceal neoplasms (3). The most important differential diagnoses include epithelial tumors, carcinoid tumors, lymphomas and very rare tumors such as ganglioneuroma, pheochromocytoma, Kaposi sarcoma and mesenchymal tumors (2). These tumors occur in the middle-aged or older patients, except the carcinoid tumors, which occur in young adults. Thirty to fifty percent of all appendiceal neoplasms will clinically present with signs and symptoms of acute appendicitis (3). Carcinoid neoplasms are the most common neoplasms found in the appendix (1). The incidence of carcinoid tumor is 0.32% and 0.054% in surgically removed appendices and in autopsy series respectively (1). This endocrine tumor originates from subepithelial neuroendocrine cells (2). Classic carcinoid tumors make up to 80% of all neoplasms of the appendix (2). Mucocele formation may occur, but is a rare finding (2). Epithelial neoplasms of the appendix are either benign or malignant (2). These neoplasms are less common than carcinoid tumors but are more often diagnosed by imaging due to their large size (2). A mucocele is a macroscopic descriptive term which means that the appendix is grossly distended by mucus. It can be caused by a variety of non-neoplastic, benign neoplastic, and malignant conditions (2). They are mostly found incidentally, because a mucocele results from a chronical luminal obstruction (2). The imaging diagnosis of neoplasms hinges primarily on detection of the resulting mucocele. An abdominal radiography may suggest the presence of a soft-tissue mass in the right lower quadrant, but is mostly aspecific. At ultrasound (US), an ovoid cystic mass, with or without acoustic shadowing from dystrophic mural calcification, is characteristic of mucoceles from mucinous neoplasms. The intraluminal ultrasonographic texture can have a variable appearance but hypo-echoic internal echoes are somewhat typical. The differential diagnosis might also include peri-appendiceal abscess, enteric duplication cyst, mesenteric cyst and hydrosalpinx (2). An additional CT scan can be useful for evaluating a mucocele of the appendix. The anatomic relationship between the elongated cystic mass and the cecum can be more apparent at CT than at US. Furthermore, CT is more sensitive than radiography in detecting mural calcifications. However, inadequate opacification of the ileocecal region with enteric contrast material is a well known cause of false-positive and –negative findings at CT (2). The difference in the frequency of a tumor manifesting as appendicitis, between mucinous neoplasms and all other appendiceal neoplasms, is significant ($P < 0.001$) (3). At an older age, in the case of appendiceal pathology, the possibility of an underlying malignancy must be excluded. The frequency of carcinoma 'masquerading' as appendicitis is likely to increase with age (3). Preoperative imaging is of importance as it can direct the surgeon in performing an appendectomy or right hemicolectomy, laparoscopically or by performing a laparotomy, if indicated. The CT findings strongly suggest the presence of an underlying neoplasm in the majority of patients (3). Our patient had a bilateral adnexectomy 40 years ago, rendering the possibility of a mucinous tumoral pathology of gynecological origin unlikely. In this case, the additional CT examination following the US examination was useful. The opacification of the lumen of the cystic mass by orally ingested contrast material allowed a better insight into the anatomic

relationship between the cystic mass and the cecum. Although in this case all radiological examinations were unable to visualize the tumoral mass itself, the additional CT examination was important in the decision-making to operate on the patient. Suggestion of an underlying tumor was made because of the age of this patient. Therefore, the decision was made to perform a right hemicolectomy. The pathological examination revealed the presence of a mucinous adenocarcinoma of the appendix, causing the mucocele.

Differential Diagnosis List: Mucinous cystadenocarcinoma of the appendix.

Final Diagnosis: Mucinous cystadenocarcinoma of the appendix.

References:

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- Pickhardt P, Levy A, Rohrmann C, Kende A Jr, Kende A. Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. Radiographics. 2003; 23:645-62. (PMID: [12740466](#))
- Pickhardt P, Levy A, Rohrmann C, Kende A Jr. Primary neoplasms of the appendix manifesting as acute appendicitis: CT findings with pathologic comparison. Radiology. 2002; 224:775-81. (PMID: [12202713](#))

Figure 1

a



Description: An ultrasonographic image showing the presence of a large cystic mass with a slightly hyperechogenic and heterogeneous content. This cystic mass is delineated by a thick wall. **Origin:**

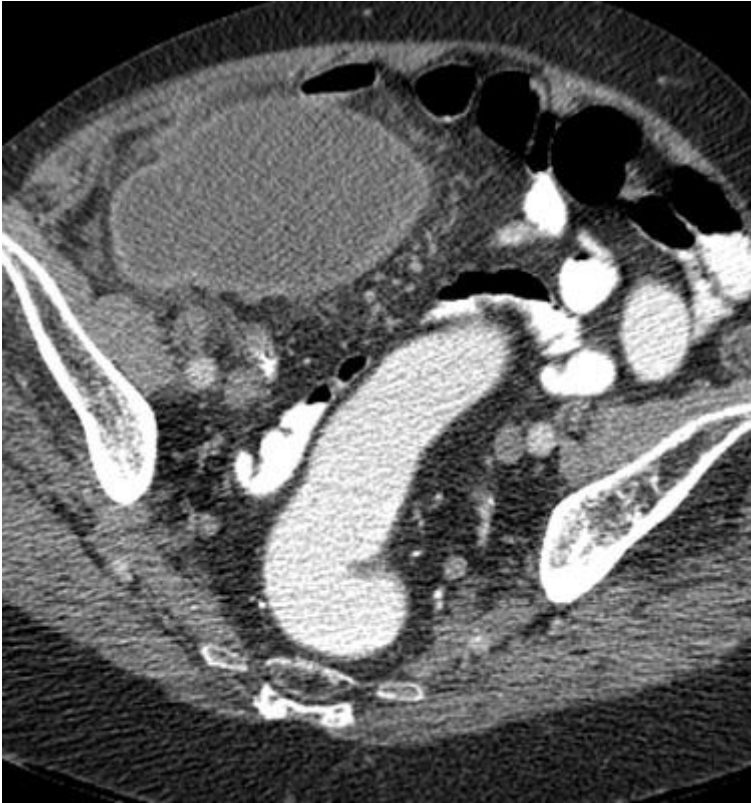
b



Description: An ultrasonographic image showing the presence of a large cystic mass with a slightly hyperechogenic and heterogeneous content. This cystic mass is delineated by a thick wall. **Origin:**

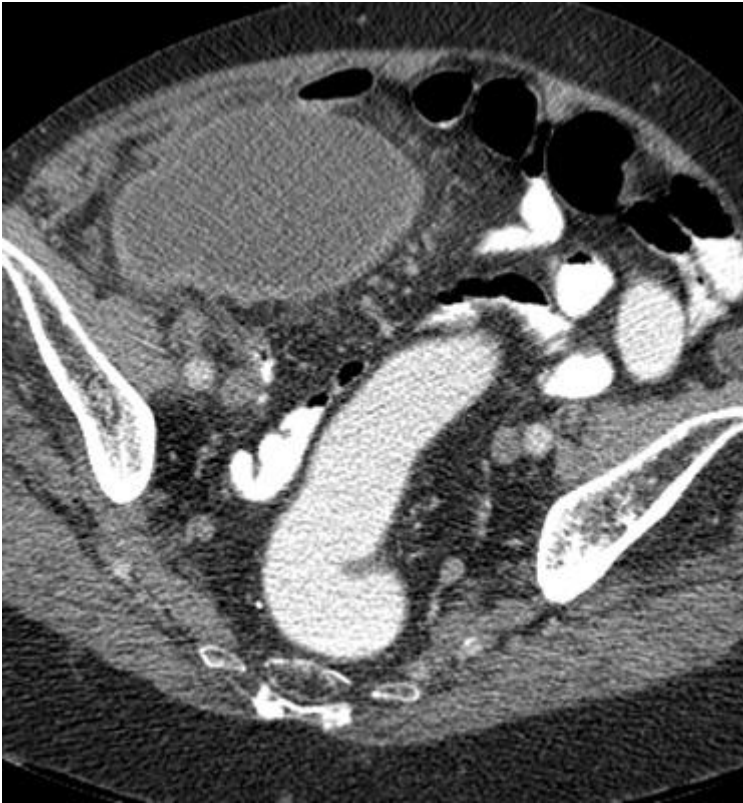
Figure 2

a



Description: A CT image showing the presence of a large cystic mass with discrete enhancement of its wall. A limited infiltration of the surrounding mesenteric fatty tissue is noted. **Origin:**

b



Description: A CT image showing the presence of a large cystic mass with discrete enhancement of its wall. A limited infiltration of the surrounding mesenteric fatty tissue is noted. **Origin:**

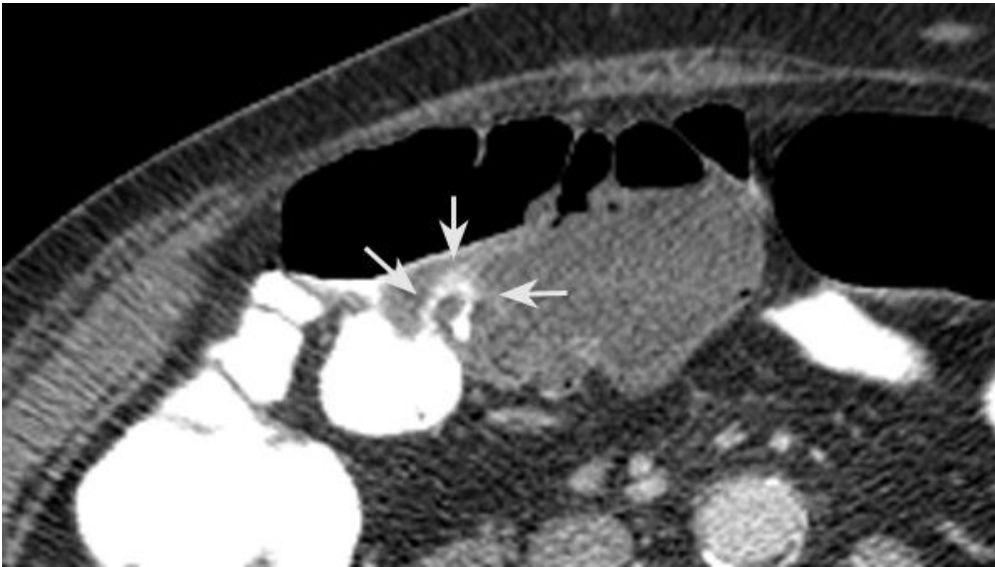
Figure 3

a



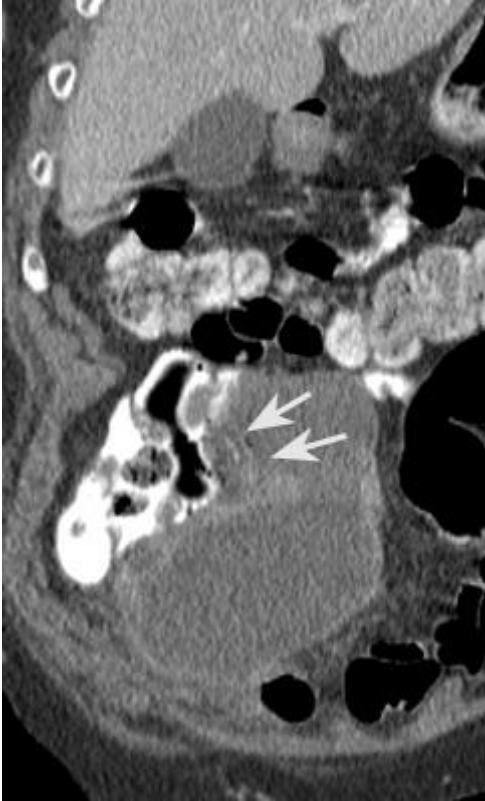
Description: A CT image showing the presence of a delicate amount of an oral contrast agent (indicated by white arrows) in the lumen of the cystic mass. **Origin:**

b



Description: A CT image showing the presence of a delicate amount of an oral contrast agent (indicated by white arrows) in the lumen of the cystic mass. **Origin:**

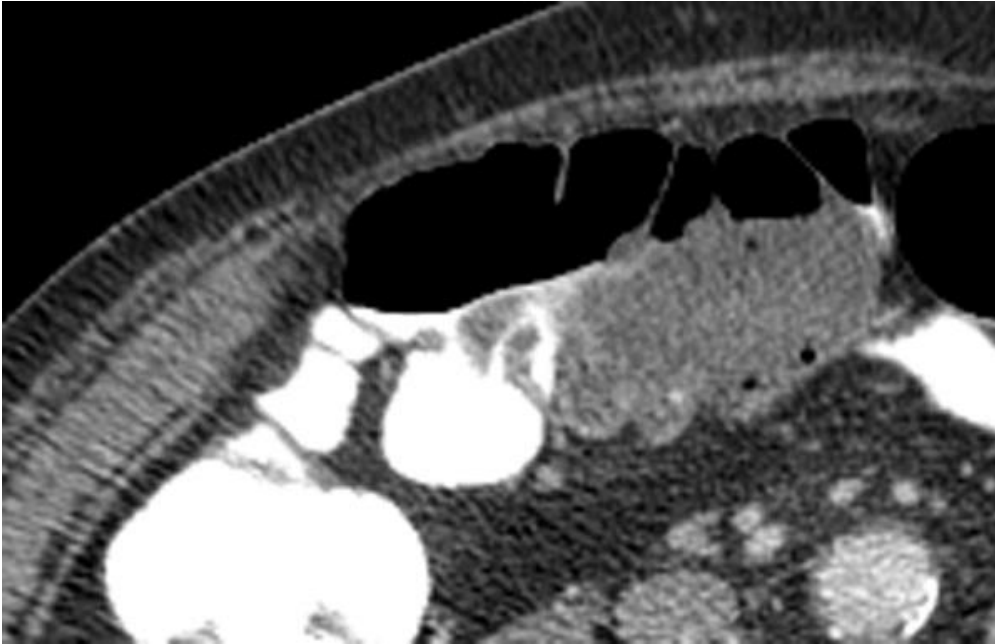
c



Description: A CT image showing the presence of a delicate amount of an oral contrast agent (indicated by white arrows) in the lumen of the cystic mass. **Origin:**

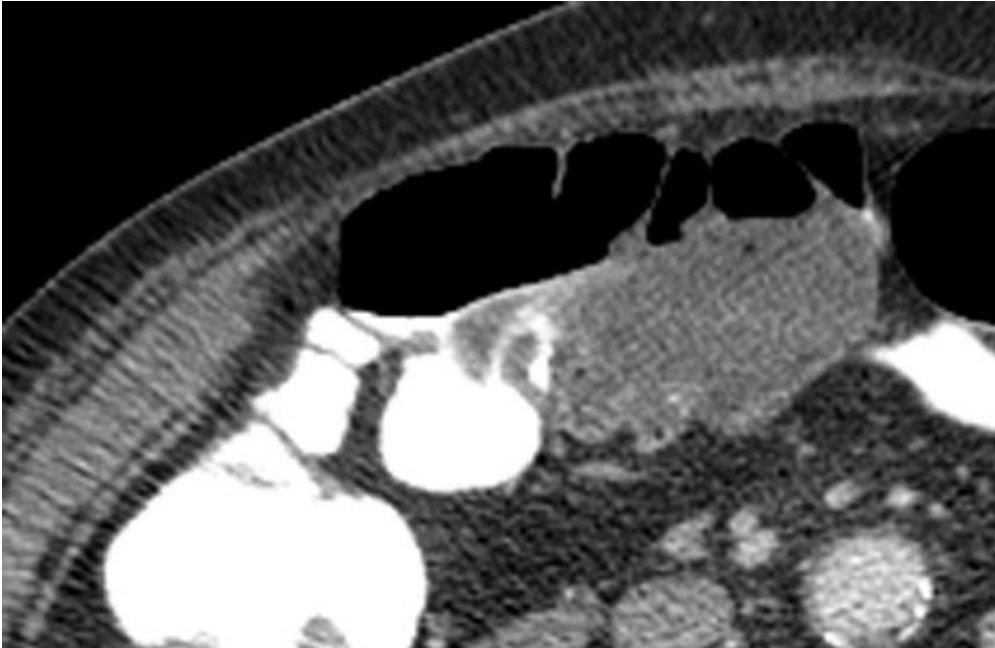
Figure 4

a



Description: A CT image showing the presence of a small amount of air in the lumen of the cystic mass. **Origin:**

b



Description: A CT image showing the presence of a small amount of air in the lumen of the cystic mass. **Origin:**

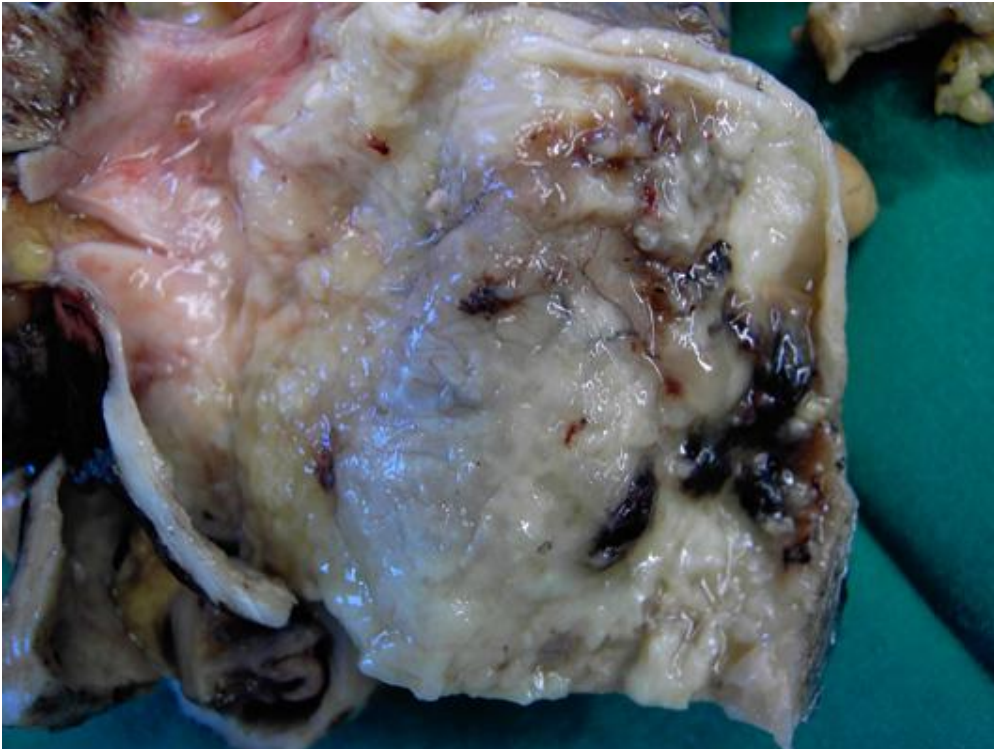
Figure 5

a



Description: A gross specimen of the resected appendix. **Origin:**

b



Description: A gross specimen of the resected appendix. **Origin:**