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Primary mucinous cystadenocarcinoma of the appendix: An unusual presentation of a rare tumor

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A primary mucinous cystadenocarcinoma of the appendix is a rare tumor, accounting for approximately 25% of all appendiceal adenocarcinomas.[1] Preoperative diagnosis is very difficult and is mainly based on computed tomography (CT) scan findings. Furthermore, in many cases difficulties in establishing an accurate intraoperative diagnosis have resulted in a two-stage surgical intervention. We present here a rare case of an appendiceal mucinous cystadenocarcinoma (AMC) in a patient who presented with an unusually large cystic mass in the right lower abdomen.

A 67-year-old man with a past medical history remarkable for diabetes mellitus, presented with a 1-month history of lower abdominal discomfort and dull pain localized mainly to the right lower abdomen. The pain became progressively worse over the two days prior to his admission and was associated with nausea. No urinary dysfunction or changes in bowel habits were reported.

On admission his blood pressure and body temperature were normal. A physical examination revealed a firm tender mass measuring approximately 15 cm with muscle rigidity in the right iliac fossa. There was no abdominal distension and his bowel sounds were normal, but his liver and spleen were not palpable. A rectal examination revealed no abnormalities. On laboratory investigation, his peripheral blood counts and biochemical markers were normal. The serum concentrations of carcinoembryonic antigen, carbohydrate antigen and alpha fetoprotein were within normal range as well.

Contrast-enhanced CT scans of the abdomen revealed the presence of a thick-walled cystic mass with enhanced borders near the cecum. The mass, measuring 15 cm at maximum diameter, was associated with inflammatory changes at its periphery. A giant appendiceal mucocele and a mesenteric cyst were considered in the differential diagnosis.

At an exploratory laparotomy performed through a midline incision a large cystic mass measuring 15 × 5 cm was found, involving the appendix. The mass was in contact with the anterior abdominal wall and extended into the urinary bladder, and loops of the small bowel adhered to its surface. The mass was resected and found to contain a thick gelatinous fluid and a number of papillary structures on its inner surface.

A frozen section examination revealed malignancy. A right hemicolectomy with side-to-side ileocolic anastomosis was then performed. A histopathological examination of the resected specimen revealed a well-differentiated AMC (Figs 1,2). No venous or lymphatic

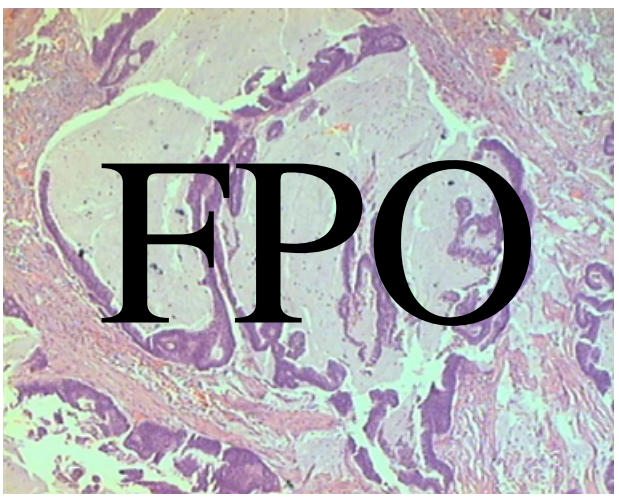

Figure 1. Cystically dilated carcinomatous structures with extracellular production of mucus (HE stain, original magnification 25×).

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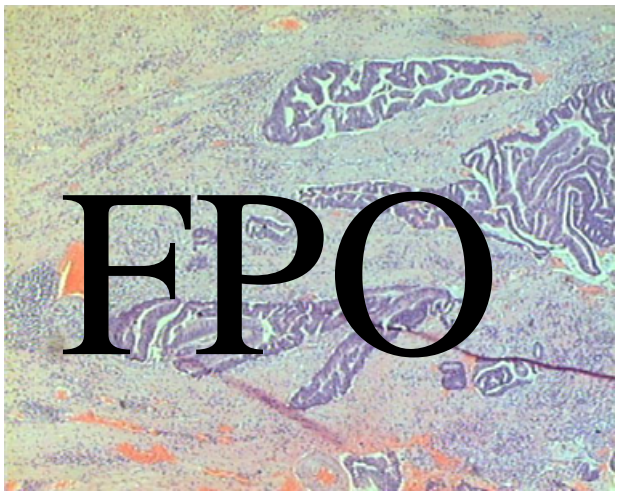

Figure 2. Carcinomatous infiltration of the appendiceal wall. (HE stain, original magnification 25×).

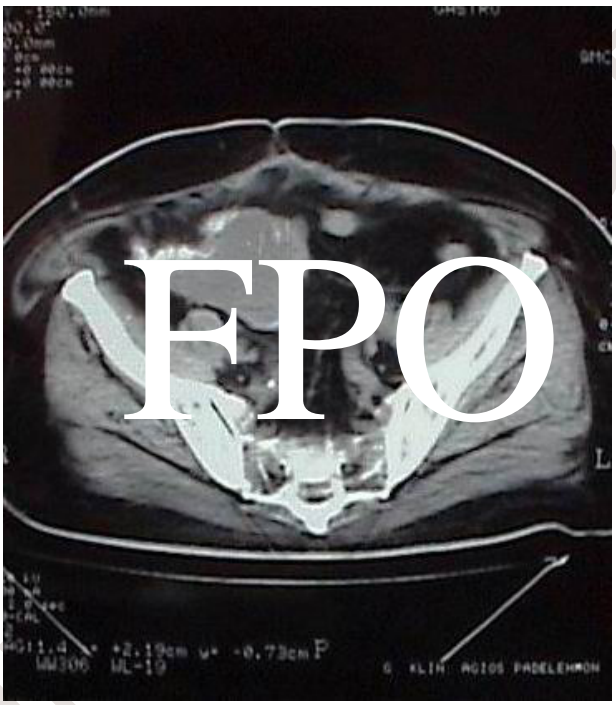

Figure 3. Computed tomography scans demonstrate a 5 × 7 cm cystic collection at the area of the primary appendiceal tumor following an anastomotic leak.

invasion was observed in the resected specimen, and the surgical margins and all 16 resected lymph nodes were free of tumor cells. The patient's postoperative course was complicated by an anastomotic leak that occurred 6 days after surgery (Fig. 3). The leak was diagnosed after a CT scan was obtained, following a localized tenderness in the right iliac fossa and mild leucocytosis. The patient was closely monitored and treated conservatively with total parenteral nutrition. No surgical intervention was required and he was discharged 18 days after the operation.

Differential diagnoses of such large cystic lesions include omental and mesenteric cysts, tumors of the cecum and ovarian tumors in women. Some of these cystic masses are considered to be appendiceal mucoceles (a descriptive term for any condition in the appendix in which the lumen is grossly distended with mucus that can be the result of a mucosal hyperplasia, a mucinous cystadenoma and a mucinous cystadenocarcinoma).[1]

An AMC is a rare neoplasm comprising about 0.5% of all intestinal tumors.[2] It is more prevalent among women than men (3:1) and occurs mainly in the fifth and sixth decades of life.[3] Presenting symptoms include right abdominal pain that is suggestive of acute appendicitis, abdominal distention and pelvic mass, whereas a possible rupture of the tumor may result in the clinical condition of pseudomyxoma peritonei.[3] The AMC is usually a well-differentiated, slowly progressive neoplasm that has a mass effect on adjacent organs rather than infiltrating them. A round, low-density, thin-walled, encapsulated mass communicating with the cecum is the most typical finding at a CT scan.[1]

Although there are not any diagnostic imaging findings pathognomonic for an AMC, some authors however, suggest that the presence of enhancing nodular lesions on the mass wall may indicate malignancy. Furthermore, findings on CT, like size, wall thickness, septations and wall calcifications are not helpful in differentiating a benign mucocele from a mucinous cystadenocarcinoma.[1]

In fact, most reported patients had the diagnosis made either several days after a simple appendectomy at the time of histological examination or by frozen section examination during surgery. At histopathology, invasion of the appendiceal wall by acellular mucin, cytologic features of epithelial cells, and the presence of epithelial cells in any intraperitoneal mucinous collection are the criteria that suggest the diagnosis of a malignancy.[4] The diagnosis is made intraoperatively in approximately 30% of patients.[3]

Perforation of the appendix is found in approximately 45% of patients with mucinous cystadenocarcinoma. A perforated tumor does not necessarily lead to diffuse pseudomyxoma peritonei, most probably due to a localized sealing process by the greater omentum.[3]

Once a diagnosis of an AMC has been achieved, a right hemicolectomy with lymph node dissection and an ileocolic anastomosis has been the accepted form of definitive therapy.[5] Aggressive debulking may improve survival when associated with pseudomyxoma peritonei, whereas intraperitoneal chemotherapy may be beneficial but requires further evaluation.[5] An accurate exploration of the abdomen during laparotomy is advised, because of the association between AMC and other tumors (such as carcinomas of the colon and ovarian tumors).[6] The 5-year survival of patients who undergo a right hemicolectomy is 73% in comparison with only 44% of patients who undergo an appendectomy alone.[3] In addition, there appears to be no significant difference in survival between patients with perforated and non-perforated neoplasms.[3]

In conclusion, we present a rare case of mucinous cystadenocarcinoma of the appendix, with an unusual appearance on preoperative CT evaluation. We would like to point out that although this clinical entity is very rare, it should however, be considered especially in elderly patients presenting with right lower abdominal pain associated with a cystic mass, regardless of size, with enhancing borders on CT scans.

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