Pseudomyxoma peritonei

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ABSTRACT

Pseudomyxoma peritonei is very rare, and its exact pathogenesis is unknown. It is characterized by intra-abdominal extracellular gelatinous fluid collections. We report a case of pseudomyxoma peritonei in a 38-year-old Saudi male who presented with right iliac fossa mass and weight loss. He was treated initially as an appendicular mass and computed tomography was helpful in making the diagnosis. He was treated by laparotomy, right hemicolectomy and omentectomy, but no perioperative intraperitoneal chemotherapy was instilled. He received postoperative chemotherapy and remained alive with no recurrence at 18-month follow-up.

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Pseudomyxoma peritonei (PP) is a slowly progressive condition, characterized by copious amounts of mucoid fluid and tumor that, over time, fills the peritoneal cavity. It literally means 'false mucinous tumor of the peritoneum'.[1] The exact pathogenesis is unknown, but it is believed to originate from a mucinous adenoma of the appendix, a mucus-producing gastrointestinal adenocarcinoma, and a primary ovarian mucinous tumor, or a mucinous peritoneal carcinomatosis of an unknown primary tumor.[2] The incidence is very rare; it occurs in one per million per year.[3] We report a case of PP to highlight its surgical presentation, preoperative diagnosis and surgical management.

Case Report. A 38-year-old Saudi male school headmaster was admitted through the emergency room with right iliac fossa (RIF) pain and fullness of a year-duration, but has increased in severity 4 weeks prior to admission. The pain was colicky in nature with no radiation and was aggravated by fatty meals and associated with flatulence and weight loss of approximately 14 kg. On examination, there was no pallor; jaundice or lymphadenopathy and the vital signs were stable. The abdomen was soft, lax with 8 × 10 cm RIF mass, which was slightly fixed and was tender on deep palpation. His blood investigations were normal with no leucocytosis. Ultrasonography and CT scan of the abdomen (**Figure 1**) revealed an irregular walled-off RIF collection with the enhancing wall. He was treated initially as an appendicular mass, but as his clinical response was very slow percutaneous aspiration of the cystic part of the mass was carried out. Cytology of the aspirated gelatinous fluid showed malignant cells. Colonoscopy showed a malignant looking lesion at the appendiceal orifice (**Figure 2**), biopsies revealed adenocarcinoma. Carcinoembryonic antigen (CEA) was elevated. He underwent laparotomy that showed large number of mucinous deposits throughout the greater omentum and the small bowel mesentery. An appendiceal mass which was fixed and adherent to the posterior abdominal wall was noted. The diagnosis of PP secondary to mucinous adenocarcinoma of the appendix was made. Right hemicolectomy, greater omentectomy and excision of all the mucinous deposits were carried out (**Figure 3**). No perioperative intraperitoneal chemotherapy was instilled as none

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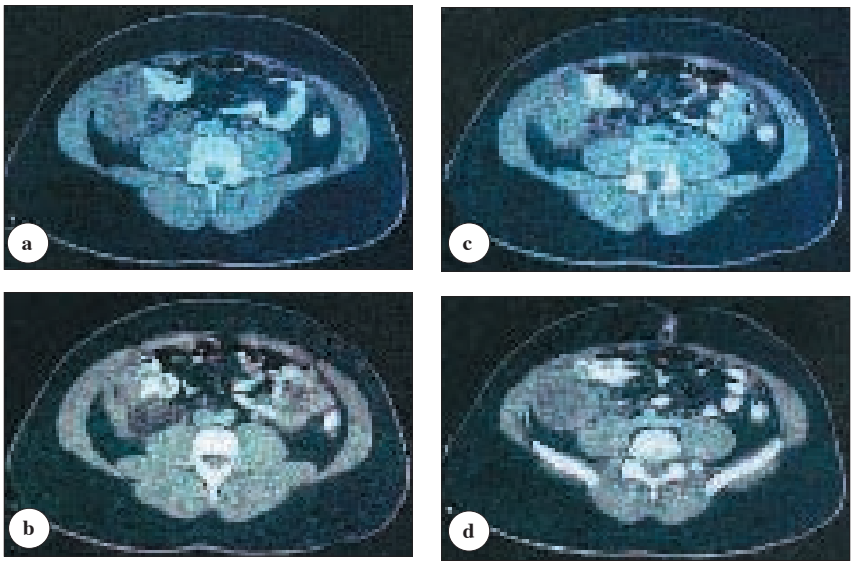

Figure 1 - Abdominal CT scan showing: **(a-d)** colonoscopic views of the cecum showing different appearances of the tumor in the base of the appendix.

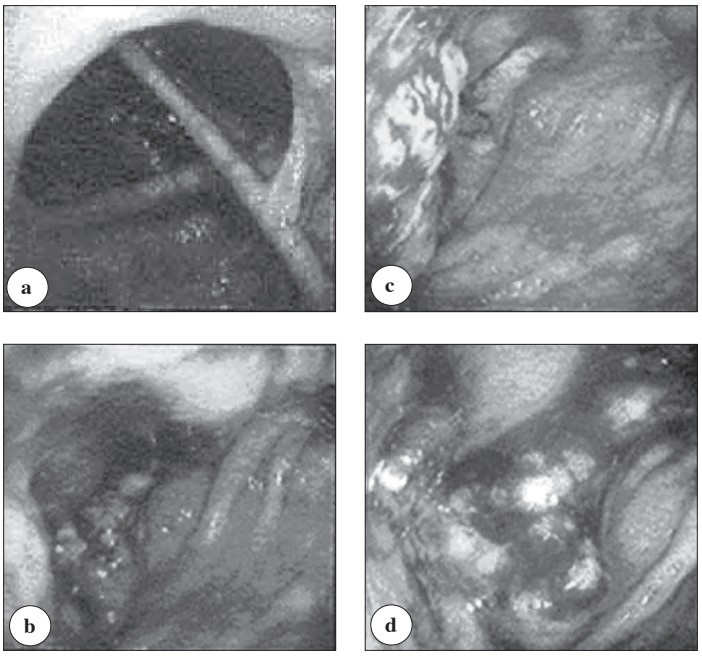

Figure 2 - Multiple sections of abdominal CT scan showing: **(a-d)** a heterogenous mass in the right iliac fossa closely adherent to the abdominal wall.

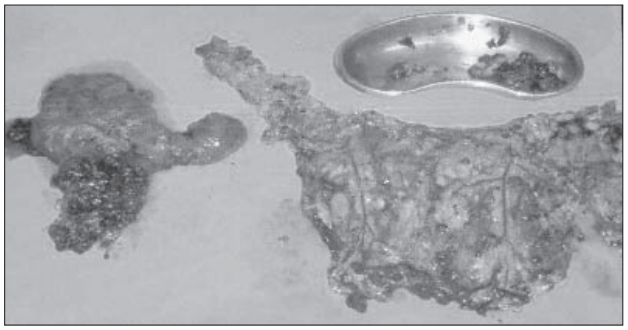

Figure 3 - Operative specimen showing the excised right colon together with the omentum.

were available. His postoperative recovery was uneventful except for a mild wound infection which was treated conservatively. He was discharged a week later in good general health and was referred to a tertiary oncology center for adjuvant chemotherapy in the form of 5-fluorouracil and leucovorin. He remained well with no recurrence at 18-month follow-up.

Discussion. Pseudomyxoma peritonei is a rare condition that is characterized by intra-abdominal extracellular gelatinous fluid collections and non-invasive mucinous implants on the peritoneum containing mucus-producing epithelium.[4] It is due to the neoplastic mucus-producing cysts within the peritoneal cavity that has a low grade and relatively benign cytological features. The cells are most commonly derived from an appendiceal cyst, benign adenoma and low-grade appendiceal adenocarcinoma.[4] It is usually diagnosed by combining the clinical findings of 'jelly belly' and the histological features. A differential diagnosis that is commonly confused with PP is carcinomatosis peritonei due to well differentiated mucinous carcinoma. When the mucinous tumor has a characteristic distribution and is accompanied by an appendiceal adenoma, it is referred to as PP syndrome.[5] Our patient might have had an adenoma-carcinoma sequence of the appendix and therefore, the term PPS may be applicable here. The presentation was that of vague abdominal symptoms suggestive of subacute appendicitis and later with what was believed to be an appendiceal mass. The most common presentation in patients with PPS is suspected acute appendicitis (27%).[2] The second most common presentation was an increasing abdominal girth (23%).[2] The new onset hernia was the presentation in 14% of cases.[2] The most common presentation in female is ovarian mass (39%).[2] Elevated tumor markers such as CEA, CA-19-9 and CA-125 indicates advanced and invasive disease and signify that complete cytoreduction is unlikely to be achievable. Monitoring levels in such patients predicts disease recurrence and progression. The CEA was elevated in our case at presentation.

Current management is by cytoreductive surgery with curative intent. This consists of peritonectomy procedure that excises all the involved parietal and visceral peritoneum combined with perioperative intraperitoneal chemotherapy (mitomycin C and 5-fluorouracil).[6] Multiple peritonectomy procedures are technically demanding, but can eradicate the disease especially if it is combined with some form of perioperative chemotherapy wash of the peritoneal cavity. Unfortunately, this modality was not readily available to us and therefore, our patient was deprived of this important step in treatment. Using both modalities has led to improvement in the outcome, and the 5-year survival rate of 87%.[5] A new treatment modality in the form of extensive surgical cytoreduction combined with intraoperative heated intraperitoneal chemotherapy has recently emerged with much improved long-term survival.[7] However, some patients may die as a direct result of this treatment. To facilitate complete cytoreduction, total gastrectomy and various colectomies may be performed.[8] Primary bowel anastomosis is usually deferred after adjuvant chemotherapy to avoid anastomotic site seeding.[7] Complete surgical removal of adenomucinosis is of paramount importance in determining long-term prognosis. At 8 years, the survival rate after complete cytoreduction was 72% vs. 0% after incomplete cytoreduction.[9] The long term survival of affected people remains poor; 5 years survival rates are 50% and 10 years survival rates are 10-30%.[4] Although, PP has an indolent course and is rarely aggressive, complete cure is seldom achieved despite repeated cytoreductions and the disease-free intervals between surgical interventions became progressively shortened. Eventually, further surgery becomes impossible due to extensive tumor entrapment of the bowel and other abdominal organs and patients eventually die of bowel obstruction and cachexia.[7] This case belongs to the aggressive category of PP which is associated with mucinous adenocarcinoma.

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