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clinical practice

# Pseudomixoma peritonei associated with appendiceal cistoadenoma rupture: case report

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SUMMARY: **Pseudomixoma peritonei associated with appendiceal cistoadenoma rupture: case report.**

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*Appendiceal mucocele represents specimen finding in 0.2-0.3% out of appendectomies. The rupture or perforation in peritoneal cavity might cause pseudomixoma peritonei (PMP), with multiple mucinous deposits in the abdominal cavity. We report a case of PMP caused by a perforated appendiceal cistoadeonoma.*

KEY WORDS: Pseudomixoma peritonei - Appendiceal mucocele - Appendiceal cistoadenoma - Rupture - Surgery.

## Introduction

Pseudomyxoma peritonei (PMP) is characterized by disseminated intraperitoneal mucinous ascites (1-5). Most PMP are originated by benign or malignant peritoneal neoplasia located in appendix and ovary. Histology reported mucinous neoplasm – whether adenomatous or carcinomatous – determining the prognosis (3-5). We report a case of PMP caused by a perforated appendiceal cistoadeonoma.

## Case report

A 75-year-old man was accepted to the emergency department of our Hospital with abdominal pain in the last seven days. Clinical conditions showed no peritoneal irritation, normal peristalsis and dyarrhea in the last three days. No vomiting or fever were reported. Blood exams showed a WBC count of 11,250/mm$^3$ with CRP value of 11 mg/l and CEA of 38,9 UI/ml. Abdominal ultrasound recorded ascites, cholelithiasis and prosthatic hypertrophy. CT scan showed an enlarged appendix with abnormal endoluminal distension, pericecal and diffuse peritoneal mucinous ascites (Figures 1, 2). Rupture of appendiceal mucocele was supposed (Figure 3).

Due to these findings patient underwent exploratory laparoscopy showing a pericecal collection with diffuse mucinous peritoneal deposits. These findings required laparotomic conversion. Pericecal drainage was performed and rupture of a large appendiceal mucocele was evidenced. We performed a blue charged endoGIA appendectomy (Figure 4) and extensive peritoneal irrigation due to the massive mucinous spread. Two tubular drainages were located in the pericecal space and Douglas cavity, respectively. Postoperative period was uneventful and patient was discharged five days after surgery.

Pathology showed an appendiceal mucinous cistoadenoma. No malignant cells in peritoneal washing were found on citology exam. One month after operation, patient underwent pancolonoscopy negative for residual disease. He is disease free after 18 months of follow-up.

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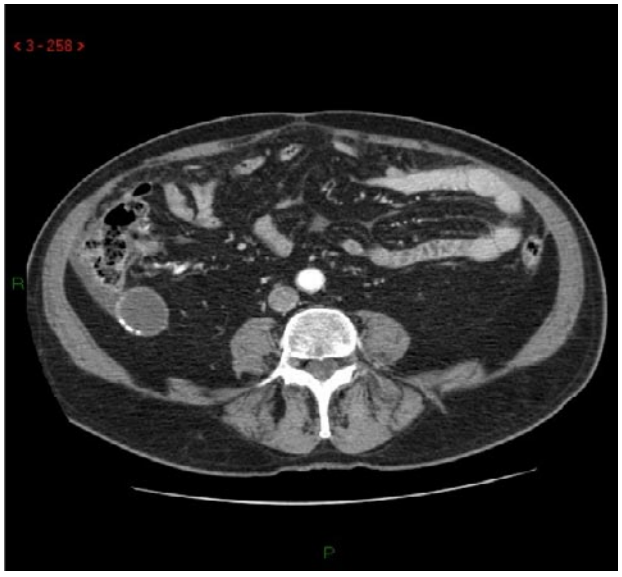

Figure 1 - Contrast-enanched CT scan showing enlarged appendix with abnormal distension and punctuated wall calcifications.

## Discussion

Rokitansky reported in 1842 the first case of gelatinous peritoneal masses related to an appendiceal benign mucocele (2). In 1884 Werth classified this clinical condition as "pseudomixoma peritonei" (PMP) (6). In the literature, PMP cases were related to colonic neoplasms, ulcerative rettocolitis and pancreatic cistic neoplasia. At present, isolated cases of PMP having an appendiceal or ovaric origin are described with an incidence of 2:10000 (4, 5, 7). Appendiceal mucocele represents specimen finding in 0.2-0.3% out of appendectomies, more frequently in female patients (3:1) (4, 5, 7). The rupture or perforation of appendiceal mucocele in peritoneal cavity causes PMP, with multiple mucinous deposits in the abdominal cavity. PMP caused by a ruptured cistoadenoma is very unusual.

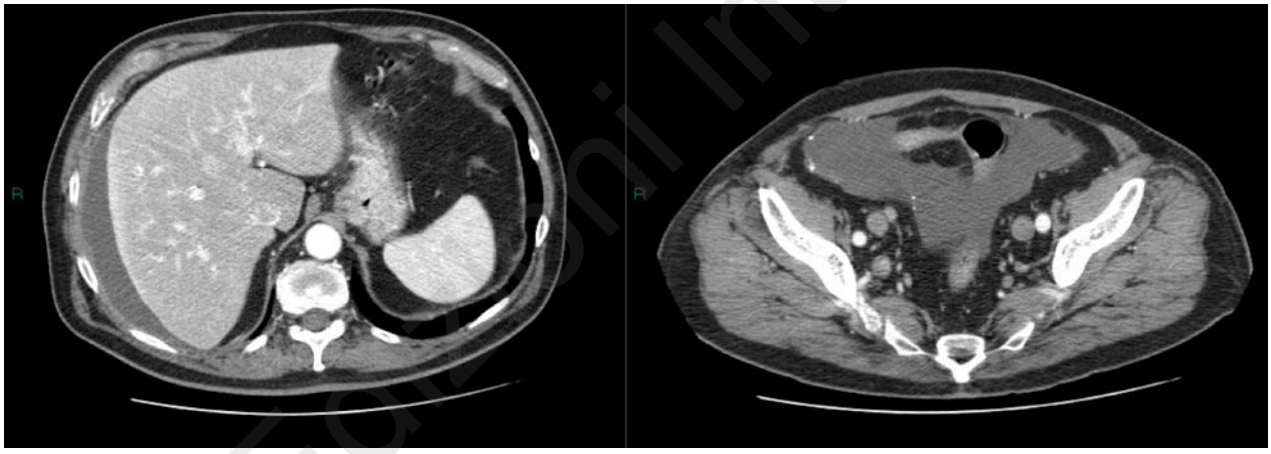

Figure 2 - Contrast-enanched CT scans showing perihepatic (left side) and pelvic ascites (right side) as sign of rupture.

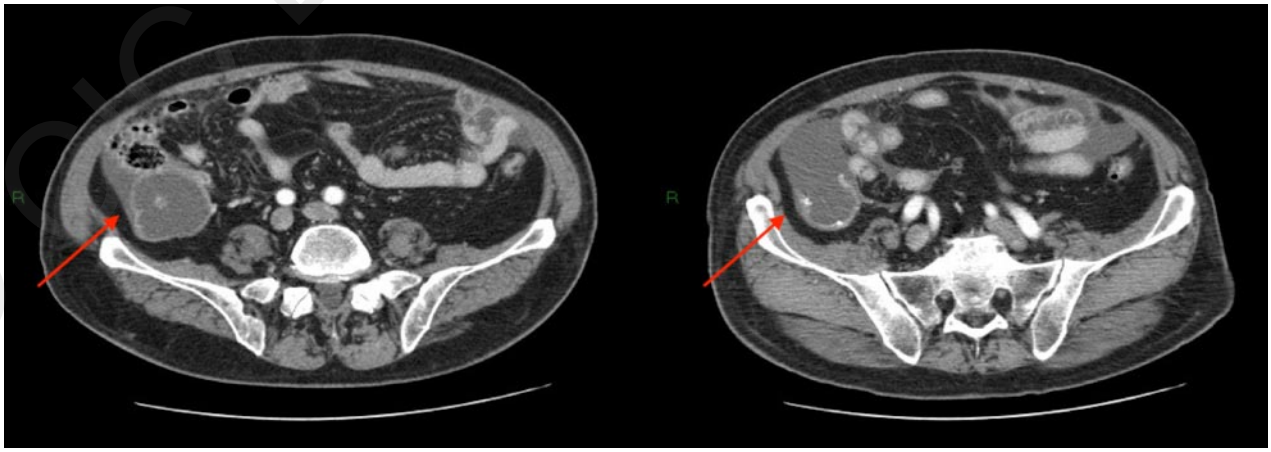

Figure 3 - Contrast-enanched CT scans showing the enlarged appendix with pericecal fluid (left side) and the rupture of appendiceal mucocele (right side).

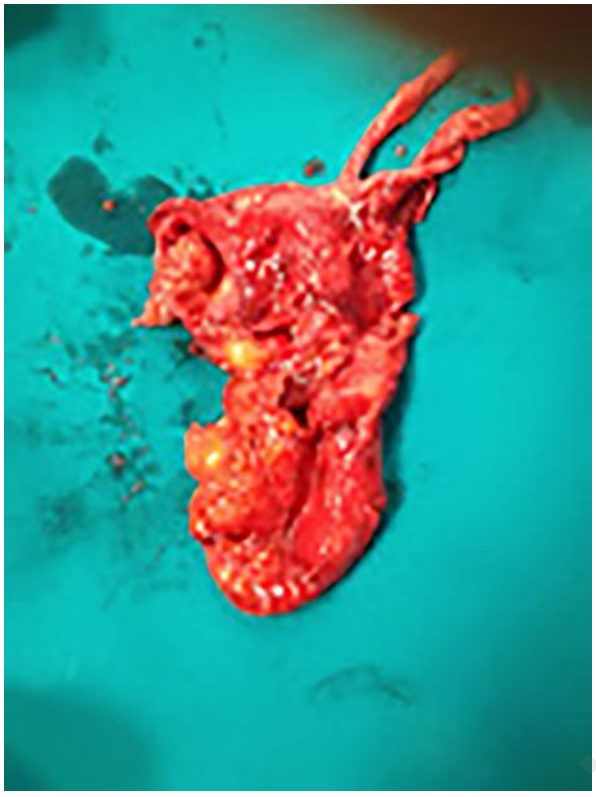

**Figure 4 - Appendectomy.**

Kim et al. described a rare case of a patient affected by appendiceal mucocele, who developed necrotizing fascitis caused by the formation of an enterocutaneous fistula (8). Chen et al. reported an unusual case of dedifferentiation of low-grade pseudomyxoma peritonei for mucinous appendicular neoplasm in high grade sarcomatoid carcinoma (9).

Clinical presentation of mucocele is often non-specific and up to 50% of patients could be asymptomatic (5). Therefore, CT scan findings are very important to confirm diagnosis. A low attenuated, well encapsulated mass with smooth regular wall in the right lower quadrant, suggests the diagnosis. The presence of curvilinear or punctuated wall calcification and displacement of the adjacent bowel without periappendiceal inflammation are important signs of mucocele (1, 4, 5, 10, 11). In our case, preoperative work up revealed the presence of mucinous ascites in the pelvis, right abdominal quadrants, between the liver and diaphragm, as a sign of rupture. The patient underwent appendectomy, ascites sampling and irrigation of surgical field. No perioperatory complications and no malignant tissues compromised a short and uneventful clinical course. When a dysplastic ruptured mucocele and mucinous carcinomatosis or PMP have been diagnosed, Dhage-Ivatury and Sugarbaker (4) recently recommended minimal surgery (including appendectomy with free margins, appendiceal lymphadenectomy, sampling of the ascites and irrigation of surgical incisions) with the goal of establishing a diagnosis and minimizing neoplastic seeding. On the basis of the pathological and cytological reports, patients should be referred to a specialised centre for the further treatment and follow-up as needed. Treatment may range from appendectomy to right colectomy and cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy and early postoperative intraperitoneal chemotherapy (4).

In 1995 Ronnet et al. described three types of PMP on the basis of prognosis: diffuse peritoneal adenomucinosis (DPMA), peritoneal mucinous carcinoma (PMCA) and intermedial/discordant group (PNCA-ID) (3). DPMA is originated by an appendiceal mucinous adenoma with favorable prognosis. PMCA is caused by an appendiceal mucinous adenocarcinoma with poor prognosis. The I/D group has variable prognosis due to appendiceal epithelial and glandular atypies (1, 3, 10-13). Prognosis of PMP originating by appendiceal mucinous cistoadenoma is favorable with a 91-100% rate survival. In PMP mucinous adenocarcinoma 5-years survival rate decreases down to 25% (1, 3, 10-13).

## Conclusions

Pseudomixoma peritonei caused by perforated appendiceal mucocele of cistoadenoma is unusual. In our case, sintomatology was unclear. The CT scan better clarified the diagnosis. Laparoscopic approach is useful to confirm diagnosis, recording intra-abdominal evidence. Laparotomy is then recommended to radically remove the pathological tissues and mucinous deposits in the peritoneal cavity. Thus, the prognosis is favourable even in case of rupture.

Finally, this unusual surgical pathology is important to take into consideration in urgency setting as well as in periferic hospitals.

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