

Mucocele of the appendix and pseudomyxoma peritonei

A Fairise, C Barbary, AL Derelle, S Tissier, P Granger, F Marchal, V Laurent and D Regent

To cite the present paper, use exclusively the following reference. Fairise A, Barbary C, Derelle AL, Tissier S, Granger P, Marchal F, Laurent V, Regent D. Mucocele appendiculaire et pseudomyxome péritonéal (full text in English on www.masson.fr/revues/jr). J Radiol 2008;89:751-62.

A mucocele of the appendix corresponds to mucinous distension of the appendix. Rupture, the most severe complication, leads to pseudomyxoma peritonei (PMP) which frequently is associated with a poor prognosis.

The main role of medical imaging is to provide presurgical diagnosis so that all precautions may be taken to avoid peroperative rupture and peritoneal seeding. Early diagnosis of pseudomyxoma is also important to allow early radical surgery in patients with peritoneal involvement to reduce recurrences.

Radiologists must be familiar with the imaging features of this entity since imaging frequently is the first step in the diagnosis and management of this pathology and may contribute in avoiding errors with devastating impact on prognosis.

In this pictorial essay, the imaging features of 12 cases of mucocele of the appendix and 8 cases of PMP will be reviewed.

Anatomical and histological features

Knowledge of the anatomical and pathophysiological features as well as epidemiology of this pathology is mandatory for diagnosis.

Mucocele of the appendix

A mucocele of the appendix is defined by the distension of the appendiceal lumen by mucus (1).

Mucinous distension of the appendix lumen may be tumoral or non-tumoral,

benign or malignant in origin. Etiologies, by increasing order of severity, include (2):

- Retention cyst corresponding to the accumulation of mucus secondary to non-tumoral obstruction of the appendiceal lumen by an appendicolith, inflammatory stricture, extrinsic compression...
- Villous hyperplasia, diffuse or focal. Involvement is confined to the mucosa.
- Mucinous cystadenoma, a benign tumor of the mucosa, sometimes with areas of dysplasia.
- Mucinous cystadenocarcinoma, a malignant tumor characterized by malignant cells, desmoplastic reaction, and invasion of the muscularis mucosa. Well differentiated cystadenocarcinoma, defined by the presence of more than 50% of mucinous component, may be difficult to differentiate from cystadenoma in the absence of muscularis invasion; the tumor is often called mucinous tumor of uncertain malignant potential.

Pseudomyxoma peritonei

PMP corresponds to a clinico-pathological entity characterized by diffuse peritoneal involvement with mucinous ascites and multifocal mucinous epithelial implants; the diagnostic histological feature is the presence of extracellular mucin within the peritoneal cavity that may be associated with mucinous epithelial cells, more or less well differentiated (3).

It results from intraperitoneal rupture of a mucinous tumor, whose appendiceal or ovarian origin has been discussed for some time, and remains controversial.

Early reports on PMP described a female predominance with frequent ovarian involvement, often bilateral, hence the hypothesis for primary ovarian origin of this entity. However, recent studies, based on immunohistochemical analysis and molecular biology, have demonstrated the appendiceal origin of nearly all cases of

PMP, with eventual secondary ovarian involvement (4, 5).

These results were partly based on a study of MUC2-expression, a gene coding for a mucin protein. This protein was present in PMP and mucinous tumors of the appendix, but not in primary ovarian tumors. Primary ovarian tumors, initially considered as the origin of PMP, may cause peritoneal carcinomatosis with peritoneal tumor implants, but no true PMP. Peritoneal carcinomatosis from ovarian primary is characterized by a smaller amount of mucin relative to tumor cells compared to PMP (4).

Based on these data, the number of PMP from definite ovarian origin would correspond to a small percentage. The only primary ovarian tumors capable of true pseudomyxomatous dissemination would be cystic mature teratomas, maybe due to the presence of gastrointestinal tissue in these embryonal tumors (6).

Finally, a minority of PMP would result from rupture of non-appendiceal mucinous tumors, or gastric or colonic origin. A few cases of PMP secondary to urachal mucinous cystadenocarcinoma have been reported (7).

While prognosis of a non-ruptured mucocele is favorable, prognosis in cases of rupture with peritoneal dissemination is directly correlated to the cell type of the pseudomyxoma (8).

This classification includes two main categories (*table 1*):

- **Disseminated peritoneal adenomucinosis (DPAM)** corresponding to ascites mainly composed of extracellular mucin, with few epithelial cells, few atypical cells, and low mitotic activity; the underlying lesion is a mucinous adenoma of the appendix. The evolution is relatively benign with 10-year survival over 80%, and no metastatic potential.

Table I
Characteristics of the two main groups of PMP according to Ronnett, et al.

| | Disseminated peritoneal adenomucinosis DPAM | Peritoneal mucinous carcinomatosis PMC |
|-----------------------------|--|--|
| Initial appendiceal tumor | Mucinous adenoma | Mucinous adenocarcinoma |
| Macroscopic appearance | Mucinous ascites, spared small bowel | Carcinomatosis, with zones of infiltration |
| Cellularity | Low | Moderate to abundant |
| Morphology | Abundant extracellular mucin with bland to low-grade adenomatous mucinous epithelium | Moderate to abundant extracellular mucin with peritoneal lesions of proliferative mucinous carcinoma, glandular tissue, or malignant cells |
| Cellular atypia | Minimal | Moderate to marked |
| Mitoses | Rare | Few to frequent |
| Nodal invasion | Rare | Frequent |
| Invasion of adjacent organs | Rare (except ovaries) | Frequent |
| Survival at 5 years | 80 % | 10 % |

– **Peritoneal mucinous carcinomatosis (PMC)** corresponding to peritoneal collections of extracellular mucin with abundant epithelial cells, more or less structured, with frequent cellular atypia, and high mitotic activity; the underlying lesion is a mucinous adenocarcinoma of the appendix. The evolution is very poor, with 3-year survival inferior to 10%, and nodal, hepatic and pulmonary metastases. In addition to these two main categories of tumor, B. Ronnett has described two additional intermediate categories of malignant lesions, with intermediate prognosis (3-year survival inferior to 60%):

– **Peritoneal mucinous carcinomatosis with intermediate features (PMC-I):** disseminated peritoneal adenomucinosis with rare foci of well differentiated peritoneal mucinous adenocarcinoma; the underlying lesion is a well differentiated mucinous adenocarcinoma associated to a mucinous adenoma of the appendix.

– **Peritoneal mucinous carcinomatosis with discordant features (PMC-D):** foci of peritoneal mucinous adenocarcinoma, generally without low-grade components; the underlying lesion typically is a mucinous adenoma of the appendix with high-grade dysplasia or intramucosal carcinoma, without invasive carcinoma.

Even though more complex forms of PMP may sometimes occur, difficult to categorize using this classification, the system by Ronnett remains valuable because of its simplicity. In 1995, Sugarbaker proposed a “surgical” classifica-

tion of secondary peritoneal involvement by benign and malignant mucinous peritoneal lesions, that is valuable mainly for prognosis.

Pathophysiology: mechanism of peritoneal dissemination

Rupture of a mucocele of the appendix most frequently is intraperitoneal; it may be retroperitoneal in patients with retrocecal appendix.

After rupture, dissemination initially is loco-regional, with seeding in the vicinity of the ruptured appendix by extracellular mucin and mucinous epithelial cells, followed by diffuse seeding of the peritoneal cavity by mucin-producing cells.

Knowledge of preferential paths of distribution of tumor cells is helpful for surgical planning, characterized by localized resection of multiple sites of involved peritoneal.

The paths of peritoneal redistribution are those for cells with low-adhesive potential (9, 10):

- zones of dependent stasis: cul-de-sac of Douglas, paracolic gutters;
- zones of GI structures with low mobility, zones of retroperitoneal attachment or hypoperistaltic regions: antro-pyloric region, duodenojejunal junction, ileocecal and rectosigmoid regions, with relative sparing of segments where peristalsis is more important: mainly jejunum and ileum;
- regions with posttraumatic or post-surgical scarring. Preferential tumor

adhesions on scars explains the need for early radical surgical resection, and poor prognosis related to multiple surgeries;

- regions of peritoneal fluid reabsorption: right hemidiaphragm, followed by left hemidiaphragm.

Clinical features

Mucocele of the appendix is most frequently asymptomatic, but may present with abdominal pain, chronic or acute, nausea, vomiting, and rarely a palpable mass (11). Malignant mucoceles would be symptomatic more frequently than mucoceles due to benign lesions.

Mucoceles may present in relation to a complication: acute intussusception, torsion, or compression of an adjacent organ, especially hydronephrosis due to ureteral obstruction from retroperitoneal mucocele. Acute appendicitis is more rare than with other appendiceal tumors, due to the chronic nature of luminal distension with mucocele.

Finally, the most severe complication from mucocele of the appendix is peritoneal rupture, spontaneous or preoperative (or from percutaneous puncture that should obviously be avoided...) with risk of tumor dissemination.

Clinical symptoms from PMP are non-specific, with heaviness, diffuse abdominal pain, variable gastrointestinal symptoms, resulting in frequent diagnostic delay (11).

Treatment

The treatment of non-ruptured mucocele of the appendix is surgical, preferably

from laparotomy as opposed to laparoscopy; in case of laparoscopy, a collection bag must absolutely be used to prevent peritoneal seeding (11). Appendectomy is performed, without appendiceal rupture, with complete resection of the meso-appendix and peritoneal fluid sampling for cytology.

Based on results from cytology, mucocele rupture, and involvement of the meso-appendix, additional treatment may be required: right hemicolectomy, tumor cytorreduction, intraperitoneal chemotherapy.

The treatment of PMP is similar for all types: radical tumor reduction surgery characterized by optimal gross total removal of mucus and tumor implants with omentectomy, localized peritoneal resection, right hemicolectomy, and bilateral oophorectomy in females, with peroperative intraperitoneal chemotherapy, with improved prognosis by treating residual microscopic disease (10, 12, 13).

Imaging features

Mucocele of the appendix

Abdomen radiographs

Radiographs of the abdomen are rarely helpful for diagnosis. Mucoceles are rarely visible on plain radiographs as a right flank or iliac fossa mass with curvilinear

mural calcifications (*fig. 1*). Calcifications are not always present (1). Abdomen radiographs are not part of the work-up of mucocele of the appendix.

Barium studies

Barium studies are no longer recommended in the initial work-up of abdominal pain.

The imaging features on barium enema described in association with mucocele of the appendix included extrinsic compression of the medial cecal wall, absence of appendiceal opacification, and presence of vertical folds on the cecal mucosa corresponding to a concentric distribution of mucosal folds around the obstructed appendiceal orifice (1, 14).

In rare cases of cecal intussusception (*fig. 2*), barium enema would demonstrate the presence of an endoluminal mass at the level of the cecum, right hemicolon, and even transverse colon, to be differentiated from intussusception due to carcinoma at the ileocecal valve or colonic lipoma.

The presence of thin mural calcifications would in this case be valuable to suggest a diagnosis of intussusception due to mucocele (1).

Ultrasound

Typical mucoceles of the appendix are hypoechoic masses of the right lower abdomen. They may have a layered appearance (onion skin sign) (*fig. 3 and 4*). They are well defined, cylindrical or lobulated

in shape (pear shaped), with increased through transmission, and frequent thin curvilinear or punctate echogenic mural calcifications (*fig. 5*). They are mobile but attached to the cecum (14, 15).

In the absence of these characteristic features, the sonographic appearance of mucoceles may be misleading. The US appearance of the mucocele content varies based on the presence of necrosis, cell debris, and especially the consistency of the mucus, which may cause the mucocele to have a spectrum of appearances from a cystic anechoic mass (*fig. 6*) to a "solid" mass with "soft tissue" echogenicity (1).

The wall may be of variable thickness, especially in the presence of mucosal hyperplasia, that may mimic acute appendicitis or a malignant tumor of the appendix.

Septations (*fig. 6*) and endoluminal polypoid nodules may be present. These features are not specific for diagnosis of the underlying lesion.

Mural calcifications may be present in less than 50% of cases (15).

The size is variable, up to 10 cm in some patients; the relationship to the cecum may be difficult to confirm for larger lesions (16).

Finally, myxoglobulosis, a rare variant of mucocele, is characterized by distension of the appendiceal lumen by small rim-calcified spherical globules, visible on radiographs, US and CT (1, 14).

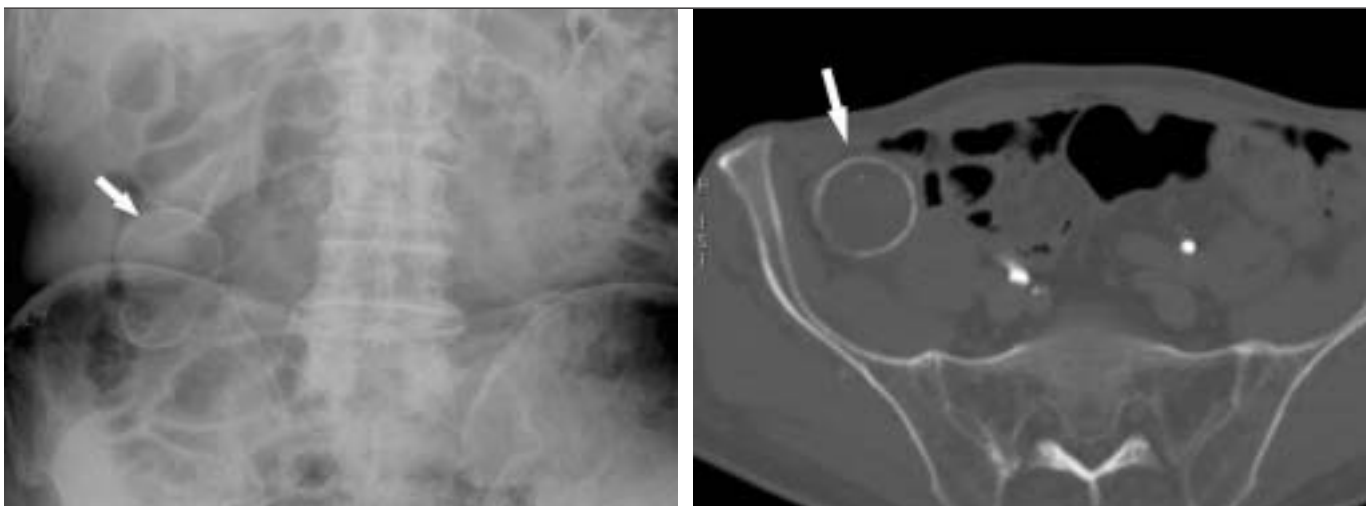


Fig. 1: Mucocele of the appendix (arrow) with calcified wall in the right lower abdomen.
 a Abdomen radiograph
 b Axial CT image

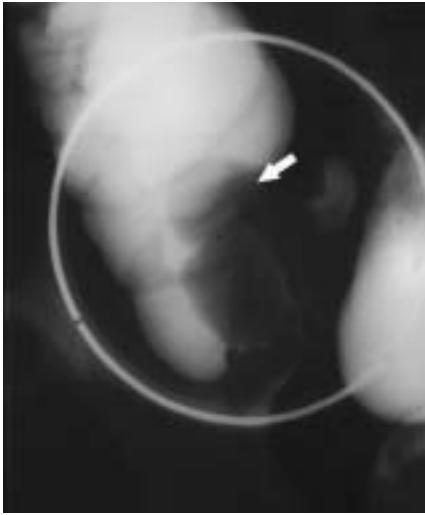


Fig. 2: *Mucocele of the appendix with cecal intussusception.*
Barium enema: absence of filling of the appendix and cecal pole with endoluminal mass (arrow).



Fig. 3: *Ultrasound.*
Typical mucocele (arrow), with layered appearance and thin walls, posterior to the bladder.

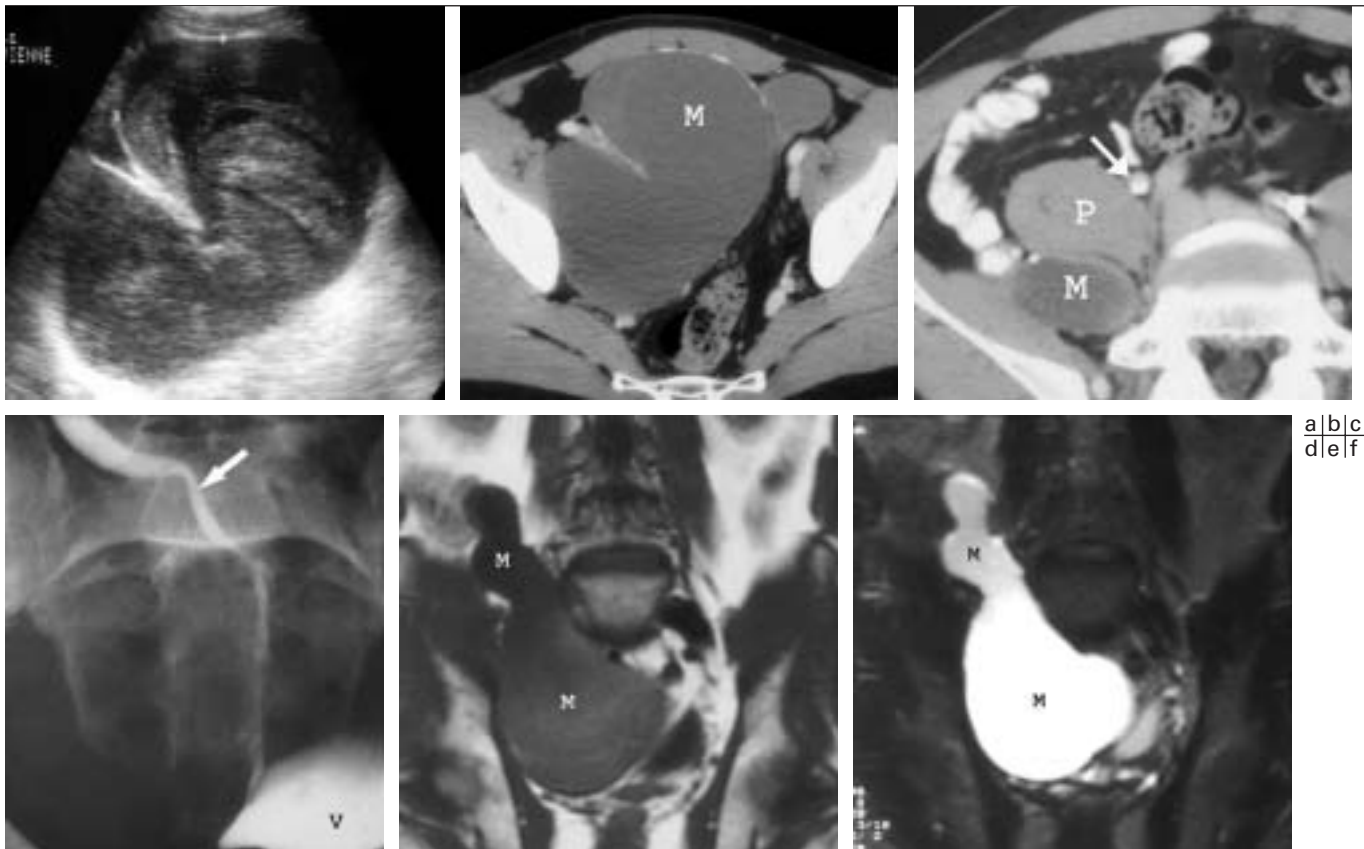


Fig. 4: *Large retroperitoneal mucocele with underlying villous hyperplasia.*
a *Ultrasound:* pelvic mass with layered appearance.
b *Corresponding CT image:* hypodense pelvic mass (M) with mural calcifications.
c *Axial CT image (bowel opacification):* retroperitoneal location of the mucocele, posterior to the right psoas muscle (P), displacing the right ureter (arrow).
d *Intravenous urogram* showing mass effect upon the right ureter and bladder (V) from the mass.
e *Coronal noncontrast T1W image*
f *Coronal T2W image:* T2W hyperintense pelvic mucocele (M) with retroperitoneal extension superiorly.

Computed-Tomography

Similar to US, mucoceles present as well defined rounded masses near the cecum, with thin wall and mural calcifications

(fig. 5 and 7); the attenuation is variable, from fluid to soft tissue density. An appendicolith is sometimes identified at the base of the appendix.

The wall may be thickened, irregular, with enhancing nodules, suggesting a diagnosis of cystadenocarcinoma (fig. 8); however, no imaging finding can be used to confidently infirm or confirm a diagnosis of underlying malignancy (2, 15).

Non-specific peri-appendiceal stranding may be present, inflammatory or tumoral in origin (fig. 9).

Mural calcifications, when present, are helpful to differentiate from acute appendicitis in patients with acute pain (fig. 9 and 10) (14).

Calcifications are usually thin and curvilinear, rarely chunk like (fig. 5).

Mucocele may become quite large, and have unusual locations relative to the cecal pole; mucocele of a retrocecal appendix may develop and rupture in the retroperitoneal space (fig. 4); a pelvic mucocele may be confused for an adnexal mass; accurate diagnosis is based on the detection of the mass attachment to the cecum, a feature that should be carefully assessed in all patients.

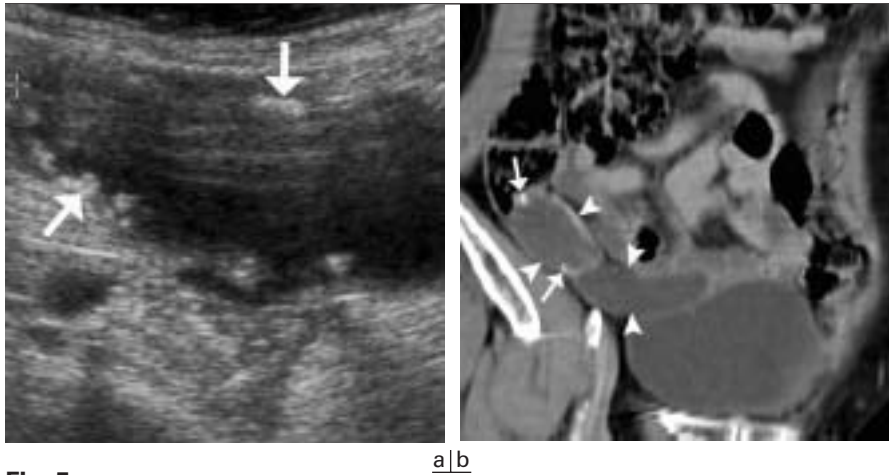


Fig. 5: *Mucocele of the appendix.*

a *Ultrasound:* large anechoic mucocele with echogenic mural calcifications (arrows).
b *CT, oblique coronal reformatted image:* mucocele (arrowheads), mural calcifications (arrows).

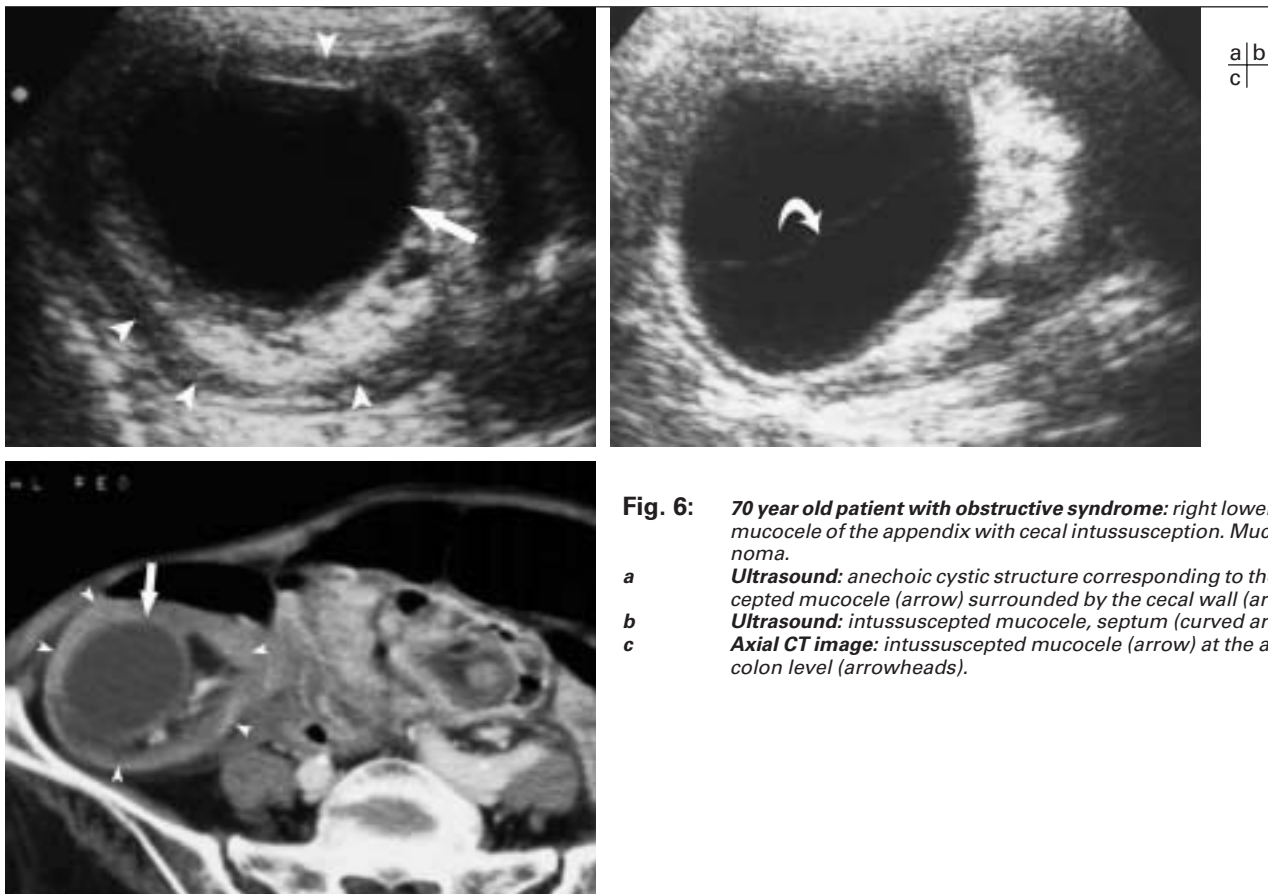


Fig. 6: *70 year old patient with obstructive syndrome: right lower quadrant mucocele of the appendix with cecal intussusception. Mucinous adenoma.*

a *Ultrasound:* anechoic cystic structure corresponding to the intussuscepted mucocele (arrow) surrounded by the cecal wall (arrowheads)
b *Ultrasound:* intussuscepted mucocele, septum (curved arrow)
c *Axial CT image:* intussuscepted mucocele (arrow) at the ascending colon level (arrowheads).

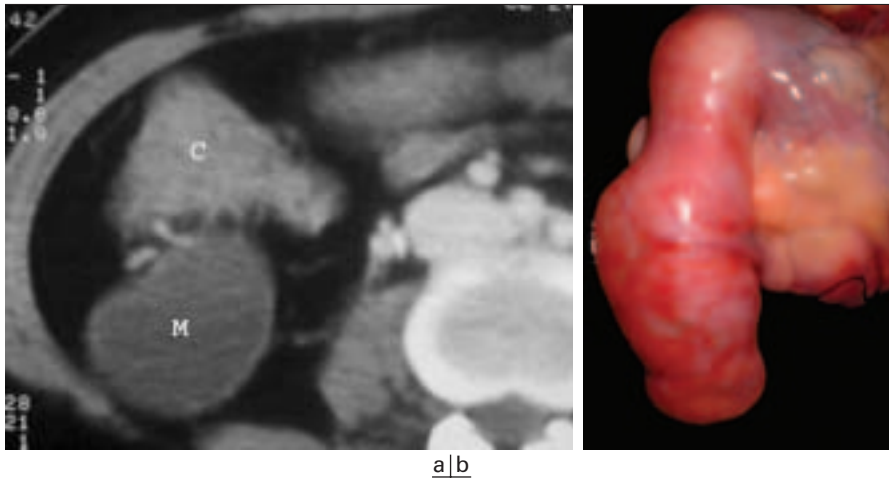


Fig. 7: *Mucocele of the appendix with underlying diffuse hyperplasia.*
a *Axial CT image: hypodense mass (M) behind the cecum (C), with thin calcified wall. The presence of wall calcifications differentiates the mass from a retrocecal abscess.*
b *Macroscopy.*



Fig. 8: *Mucocele of the appendix with underlying adenocarcinoma; pericecal mass with enhancing irregular nodular walls (arrows), septations.*

Complications may be detected both on US and CT: intussusception (*fig. 6*), torsion, ureteral compression (*fig. 4*).

Magnetic Resonance Imaging

Lesion location and morphological features are identical to CT; the mucocele is T1W hypointense and T2W hyperintense, but signal is variable based on mucin content (*fig. 4*). Calcifications may be less conspicuous (2, 3).

Pseudomyxoma Peritonei

On US and CT, the diagnosis of PMP is based on the identification of three lesions: characteristic mucinous ascites, nodular peritoneal implants, and the primary tumor; the latter is only rarely visualized.

Ultrasound

Mucinous ascites typically is heterogeneous, hypoechoic, poorly mobile, with

a more or less layered appearance. It may be loculated, with multiple septations, and thin septal calcifications (*fig. 11*).

It is classically more heterogeneous, less anechoic, and less mobile than simple ascitis. In some cases, it may even appear as an echogenic soft tissue mass.

Mass effect with extrinsic compression and scalloping of the liver and spleen and displacement of hollow structures may be seen both on US and CT (17, 18).

Computed-Tomography

On CT, mucinous ascites typically is hypodense, but usually slightly denser than a simple transudate (*fig. 10, 12 and 13*); it may be loculated (*fig. 11 and 14*) and contain thin curvilinear calcifications (17).

Thick gelatinous ascites and solid nodular peritoneal implants may cause extrinsic compression with typical scalloping of upper abdominal solid organs (*fig. 15*).

A characteristic feature of liver scalloping is the discordance between the presence of multiple or large hypodense peripheral lesions and the absence of intraparenchymal lesions, away from the liver capsule (*fig. 15*).

Gelatinous ascites may also cause deformity and displacement of lower abdominal structures and vessels, with fixation of small bowel loops (*fig. 14 and 16*) (9, 19).

Peritoneal implants correspond to heterogeneous nodules that may enhance following intravenous administration of contrast. Their presence should be assessed at the level of the greater omentum, cul-de-sac of Douglas, ovaries, paracolic gutters, and subphrenic regions (*fig. 11, 12 and 15*) (9).

They may be disseminated and involve multiple peritoneal and retroperitoneal structures.

In patients with ascites and peritoneal implants, differentiation between PMP and peritoneal carcinomatosis may be difficult; the presence of calcifications and/or septations, the degree of scalloping and the hypodense nature of the gelatinous masses all favor a diagnosis of PMP (20).

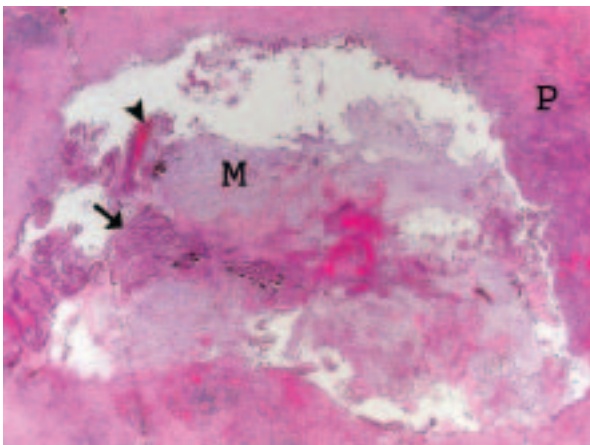
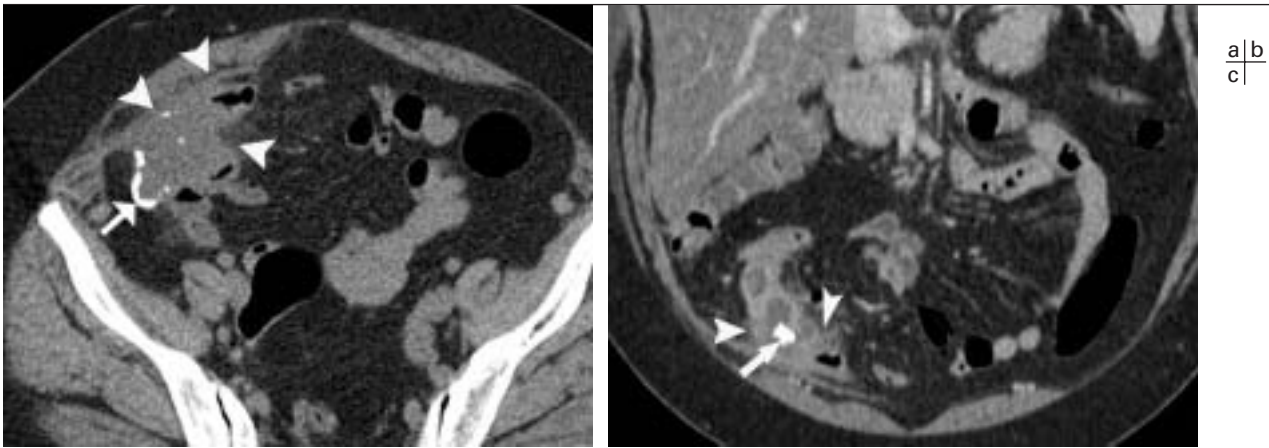


Fig. 9: *Acute appendicitis and perforated mucocele. Mucinous adenocarcinoma and PMP, PMC-D type.*

a *Axial CT image.*

b *Coronal reformatted image: perforated mucocele with wall calcifications (arrow); infiltration of the meso-appendix and periappendiceal peritoneum (arrowheads)*

c *Microscopy: transverse section of the appendix (Hemalun Eosin Safran, X25): appendicitis with inflammatory infiltrate of the wall (P); mucus present in the appendix lumen (M), with calcifications (arrowhead); desquamated dysplastic pseudo-stratified epithelial cells within the appendix lumen (arrow).*

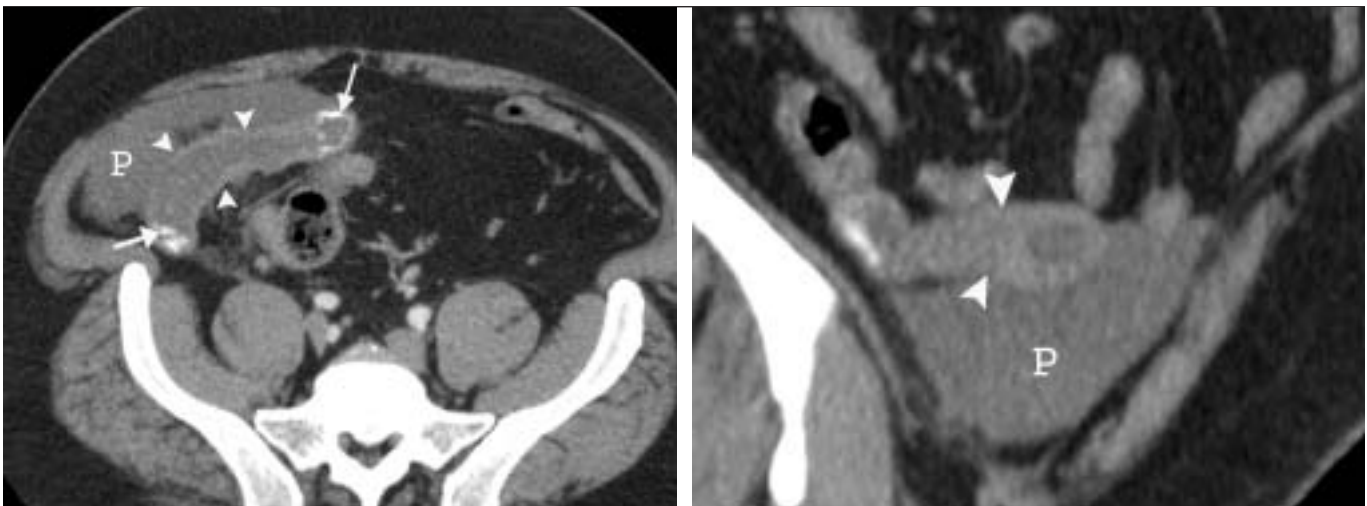
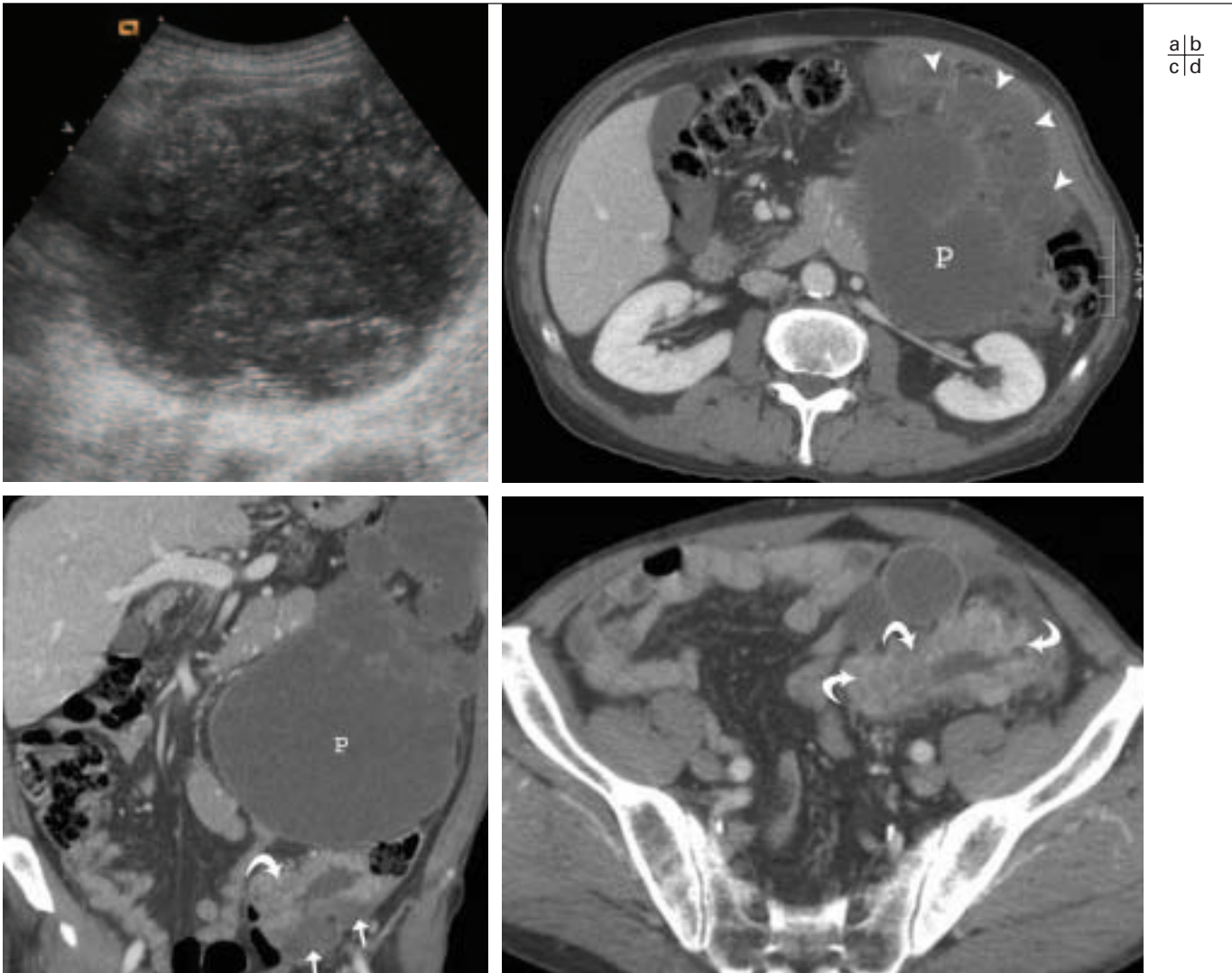


Fig. 10: *PMP in a patient with sigmoid adenocarcinoma. Histological diagnosis of mucinous peritoneal carcinomatosis.*
a *Axial US image through the left mid abdomen: multi-layered heterogeneous echogenic mass corresponding to loculated PMP.*
b *Axial CT image: loculated mucinous ascites of the left mid abdomen (P), with epiploic implants (arrowheads).*
c *Coronal reformatted image: loculated ascites (P), sigmoid mass (curved arrow), and peri-sigmoid implants (arrows).*
d *Axial CT image: primary sigmoid tumor (curved arrows).*



| |
|-----|
| a b |
| c d |

Fig. 11: *Acute appendicitis. Acute appendicitis and perforated mucocele with PMP, and cecal, omental and right ovarian implants. Mucinous adenocarcinoma and PMC.*

a *Axial CT image.*

b *Coronal reformatted image: perforated mucocele (arrowheads), PMP (P).*

c *Mucocele (arrowheads), omental (white arrow) and right ovarian (black arrow) implants*

d *Histology, appendix wall, (HES, X100): appendiceal mucosa with large areas of mucus (M), with hypersecreting cells (arrow), and zones of hyposecreting cells with hyperchromatic nuclei (arrowheads)*

e *Histology (HES, X400): atypical dysplastic elements with prominent nucleoles and nuclei (arrow) in the peritoneal cavity, within collection of mucus (M): mucinous carcinomatosis.*

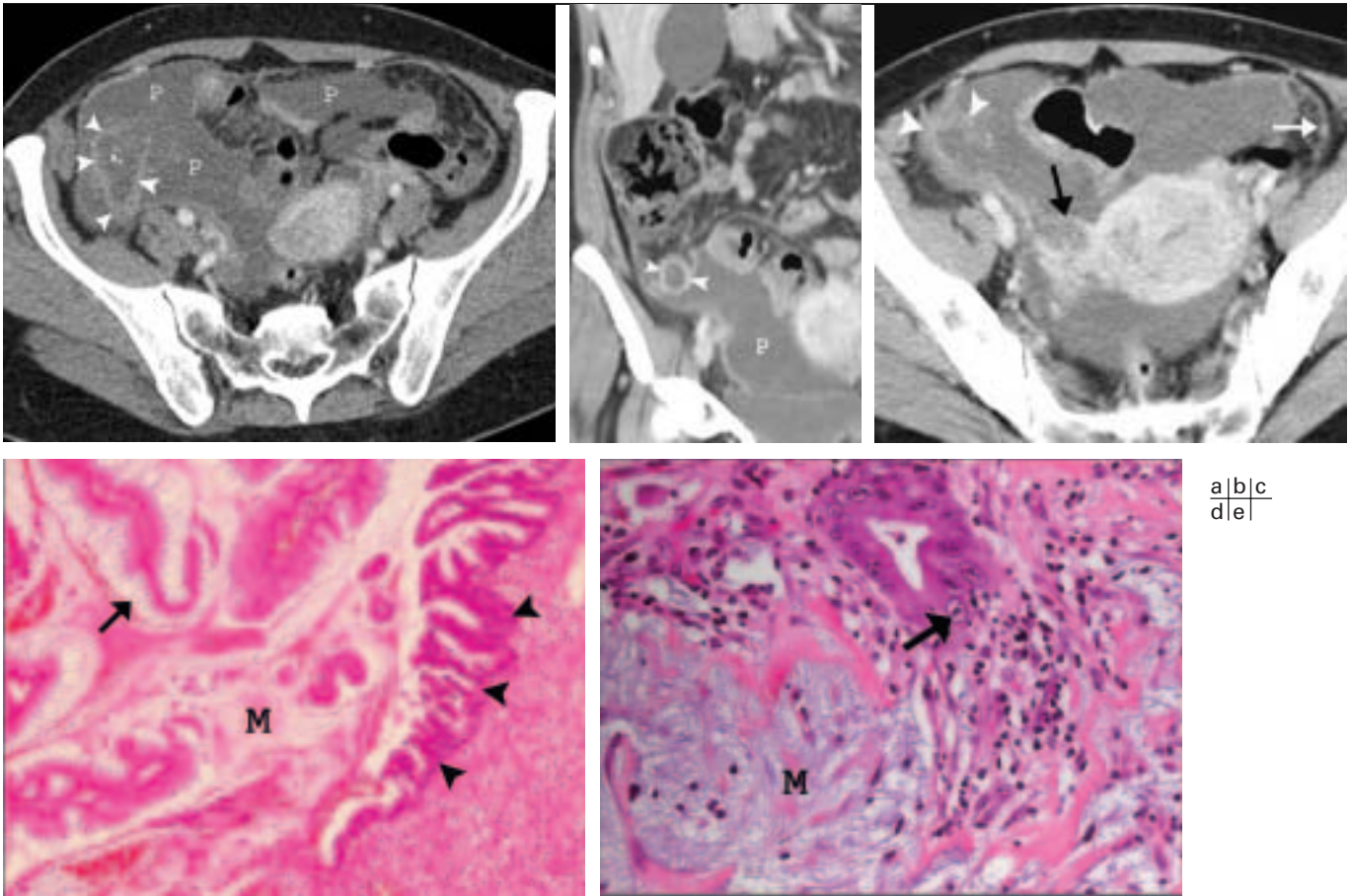
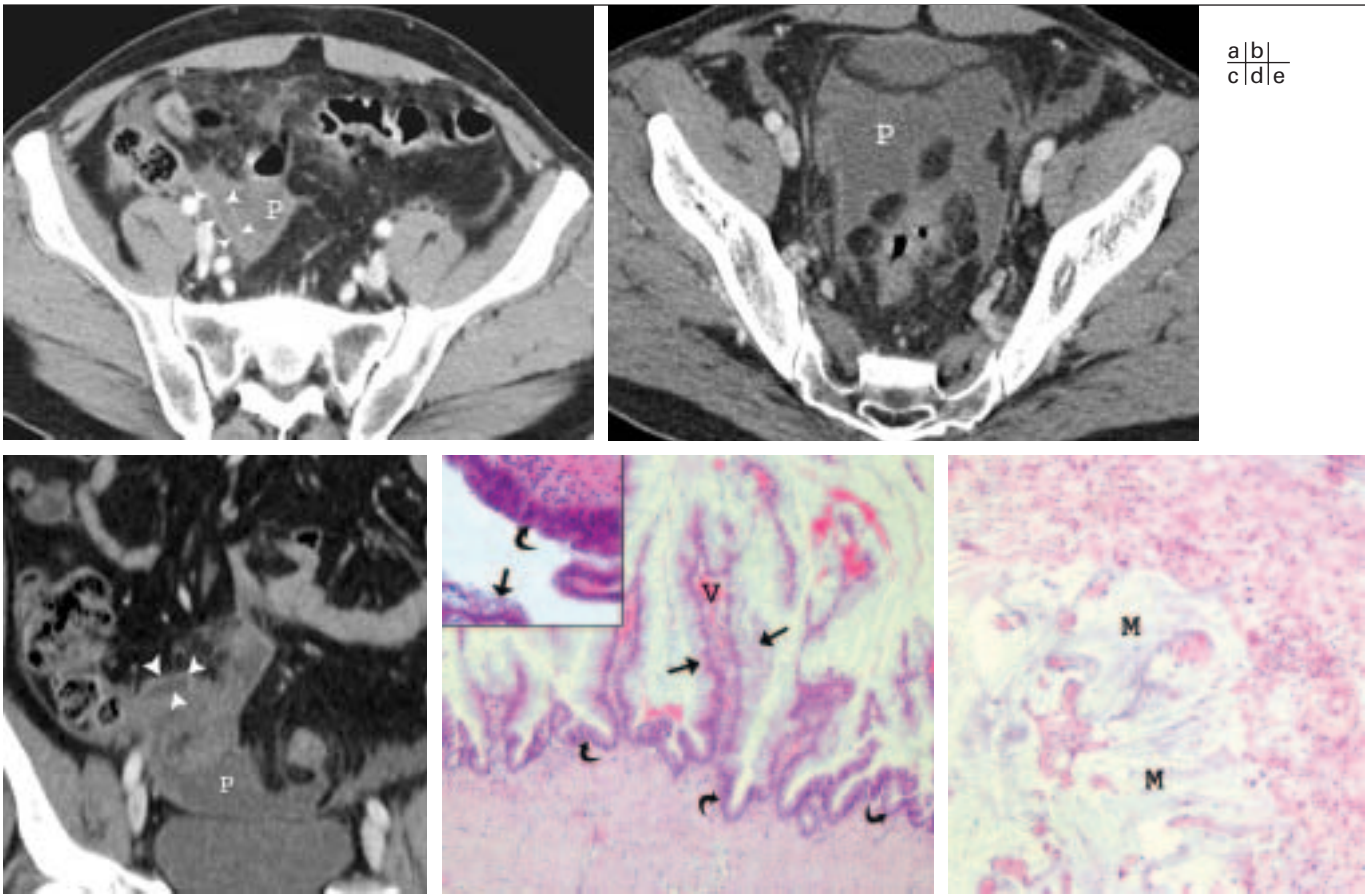


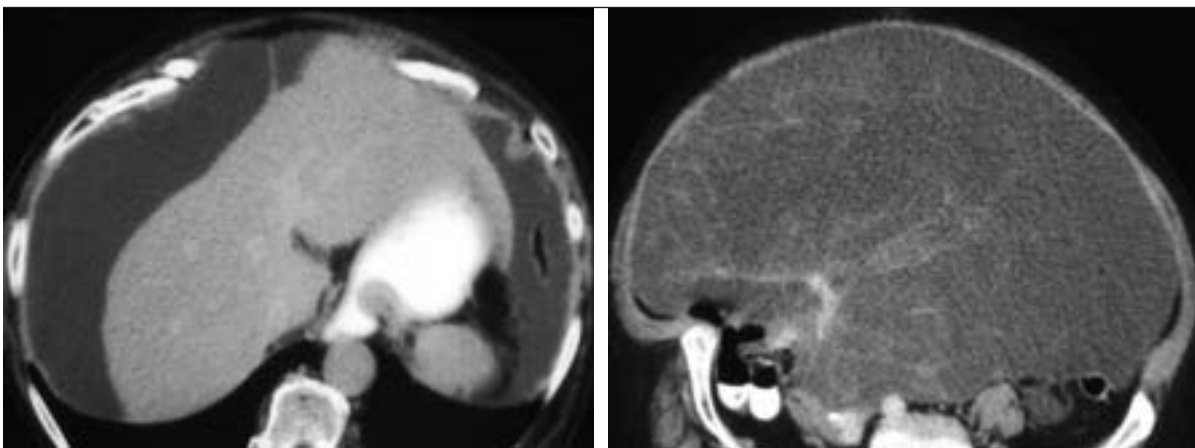
Fig. 12: *Acute appendicitis. Perforated mucocele of the appendix and PMP; adenoma and well-differentiated adenocarcinoma, PMC-I. Despite the presence of mural calcifications (arrows), the diagnosis was not made preoperatively.*

- a** *Axial CT image: RLQ mucocele (arrowheads), regional hypodense peritoneal infiltration: PMP (P). Note the mural calcifications (arrows).*
- b** *Oblique reformatted image.*



a|b|
c|d|e

Fig. 13: *Acute appendicitis. Perforated mucocele of the appendix and PMP. Difficult differential diagnosis with peri-appendiceal abscess in the absence of mural calcifications. Perioperative diagnosis. Villous adenoma with focus of severe dysplasia: mucinous tumor of uncertain malignant potential and PMC-D.*
a *Axial CT image:* mucocele of the appendix (arrowheads) and regional hypodense peritoneal infiltration corresponding to PMP (P).
b *Axial CT image.*
c *Coronal reformatted image:* hypodense PMP (P), mucocele (arrowheads).
d *Microscopy (HES, X100, insert: HES, X400):* villous adenoma of the appendix; multiple villosities (V) composed of hypersecreting cells with regular nuclei (arrows); the villosities are partially superimposed and thickened at their base, bordered by pseudo-stratified columnar epithelium with basophilic cytoplasm, and irregular hyperchromatic nuclei (curved arrows)
e *Microscopy (HES, X100):* distal third of the appendix: large areas of mucin (M) dissecting into the sub-serosal region. No neoplastic cell.



a|b

Fig. 14: *Diffuse PMP, axial CT images.*
a *Hypodense perihepatic fluid causing scalloping.*
b *Loculated ascites, causing posterior displacement of bowel loops.*

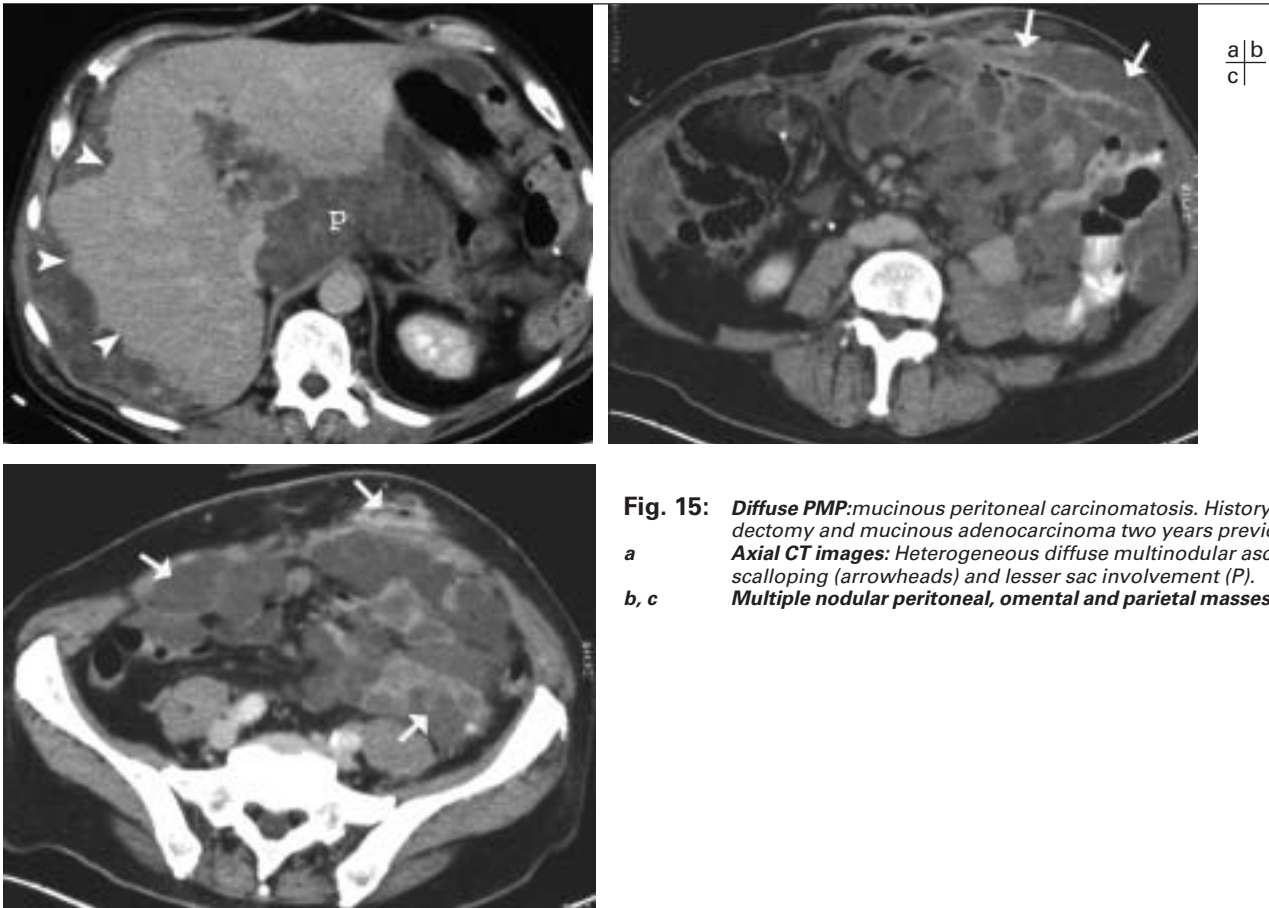


Fig. 15: *Diffuse PMP: mucinous peritoneal carcinomatosis. History of appendectomy and mucinous adenocarcinoma two years previously.*
a *Axial CT images: Heterogeneous diffuse multinodular ascites; liver scalloping (arrowheads) and lesser sac involvement (P).*
b, c *Multiple nodular peritoneal, omental and parietal masses (arrows).*

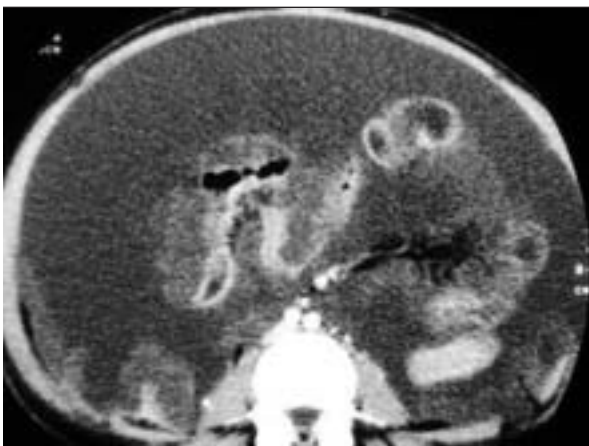


Fig. 16: *Axial CT image: diffuse PMP with displacement and grouping of bowel loops at the center of the abdomen.*

Conclusion

The detection and diagnosis of a non-ruptured mucocele of the appendix on imaging studies is a significant prognostic factor, allowing surgeons to take the necessary precautions to avoid peroperative rupture with peritoneal seeding. While diagnosis on imaging studies may not

always be straightforward, the presence of a few imaging features including appendiceal distension, mural calcifications and septations, should raise the possibility of mucocele, irrespective of the presenting clinical symptoms.

Early detection of a ruptured mucocele of the appendix with PMP plays a significant role by enabling tailored and radical

surgical intervention in order to reduce the number of subsequent surgeries with worsened prognosis.

References

1. Dachman AH, Lichtenstein JE, Friedman AC. Mucocele of the appendix and pseudomyxoma peritonei. *AJR Am J Roentgenol* 1985;144:923-9.
2. Pickhardt PJ, Levy AD, Rohmann CA Jr, Kende AI. Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. *Radiographics* 2003;23:645-62.
3. Derelle AL, Tissier S, Granger P et al. Diagnostic précoce de pseudomyxome péritonéal localisé à la zone de rupture d'une mucocèle appendiculaire: imagerie et aspects anatomopathologiques. *J Radiol* 2007;88:289-95.
4. O'Connell JT, Tomlinson JS, Roberts AA et al. Pseudomyxoma peritonei is a disease of MUC2-expressing goblet cells. *Am J Pathol* 2002;161:551-64.
5. Szych C, Staebler A, Connolly DC et al. Molecular genetic evidence supporting the clonality and appendiceal origin of

- pseudomyxoma peritonei in women. *Am J Pathol* 1999;154:1849-55.
6. Stewart CJ, Tsukamoto T, Cooke B et al. Ovarian mucinous tumour arising in mature cystic teratoma and associated with pseudomyxoma peritonei: report of two cases and comparison with ovarian involvement by low-grade appendiceal mucinous tumour. *Pathology* 2006;38:534-8.
 7. Takeuchi M, Matsuzaki K, Yoshida S et al. Imaging findings of urachal mucinous cystadenocarcinoma associated with pseudomyxoma peritonei. *Acta Radiol* 2004;45:348-50.
 8. Ronnett BM, Zahn CM, Kurman RJ et al. Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to 'pseudomyxoma peritonei'. *Am J Surg Pathol* 1995;19:1390-408.
 9. Régent D, Laurent V, Cannard L et al. Le péritoine « témoin » de la pathologie abdomino-pelvienne. *J Radiol* 2004;85:555-71.
 10. Sugarbaker PH. Pseudomyxoma peritonei. A cancer whose biology is characterized by a redistribution phenomenon. *Ann Surg* 1994;219:109-11.
 11. Dixit A, Robertson JHP, Mudan SS et al. Appendiceal mucocoeles and pseudomyxoma peritonei. *World J Gastroenterol* 2007;13:2381-4.
 12. Li C, Kanthan R, Kanthan SC. Pseudomyxoma peritonei – a revisit : report of 2 cases and literature review. *World J Surg Oncol* 2006;4:60.
 13. Qu ZB, Liu LX. Management of pseudomyxoma peritonei. *World J Gastroenterol* 2006;12:6124-7.
 14. Madwed D, Mindelzun R, Jeffrey RB et al. Mucocele of the appendix : Imaging findings. *AJR Am J Roentgenol* 1992;159:69-72.
 15. Kim SH, Lim HK, Lee WJ et al. Mucocele of the appendix : ultrasonographic and CT findings. *Abdom Imaging* 1998;23:292-6.
 16. Francica G, Lapicciarella G, Giardiello C, et al. Giant mucocele of the appendix. *J Ultrasound Med* 2006;25:643-8.
 17. Walensky RP, Venbrux AC, Prescott CA et al. Pseudomyxoma peritonei. *AJR Am J Roentgenol* 1996;167:471-4.
 18. Hanbidge AE, Lynch D, Wilson SR. US of the Peritoneum. *Radiographics* 2003;23:663-85.
 19. Seshul MB, Coulam CM. Pseudomyxoma Peritonei : Computed tomography and sonography. *AJR Am J Roentgenol* 1981;136:803-6.
 20. Taourel P, Baud C, Lesnik A et al. Le péritoine acteur de la pathologie abdominale. *J Radiol* 2004;85:574-90.