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MUCOCELE OF THE APPENDIX: CASE REPORT AND REVIEW OF LITERATURE

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Mucocele of the appendix is a descriptive term of a distended, mucus-filled appendix caused by various conditions, both benign and malignant. Computed tomography is the imaging modality of choice. Correct pre-operative diagnosis is important because of the possibility of peroperative rupture and subsequent development of pseudomyxoma peritonei. It is the task of the radiologist to alert the clinician and surgeon to the presence of this entity, the potential associated complications and possible signs of malignancy.

Key-word: Appendix, CT.

Case report

A 48-year-old man presented with abdominal pain and a small umbilical hernia. Because of the disproportional intensity of the associated abdominal complaints with regard to the size of the hernia, a computed tomography (CT) was performed.

The contrast-enhanced CT examination demonstrated a cystic, oval-shaped, thin-walled structure in the right lower quadrant (Fig. 1A) in continuity with the caecum. The wall contained punctate calcifications (Fig. 1B). There was no surrounding inflammation or abscess formation. Based on the radiological findings, an initial diagnosis of a mucocele was made. An elective appendectomy was preformed a few weeks later, during which the small umbilical hernia was also repaired. After resection, the diagnosis of a mucinous cystadenoma of the appendix was confirmed on pathological examination (Fig. 2).

Review of literature

Epidemiology and classification

A mucocele of the appendix is a rather rare entity, seen in only 0,2-0,3% of appendectomy specimens (1-3). Initially a female predominance was reported, although this has not been confirmed in more recent literature (4). It is commonly encountered in patients between 50 and 70 years-of-age with a mean age of 55 (3, 5).

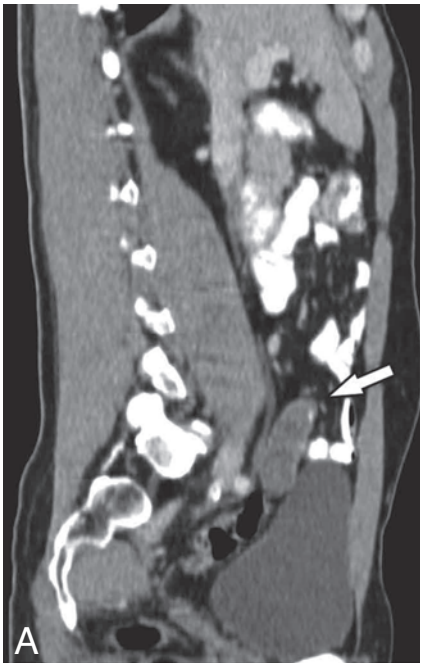
Mucocele of the appendix is not a true histopathological entity. It is a macroscopical descriptive term for a distended and mucus-filled appendix of variable etiology, and is generally divided into four histological groups (6) (Table I).

The first group consists of a simple retention cyst secondary to proximal occlusion of the appendix by e.g. a fecalith or scar tissue from previous inflammation, or in rare cases due to endometriosis (24). With rising pressure, degenerative changes in the appendiceal mucosa consecutively develop. This type of mucocele is usually smaller than 2cm in diameter (3, 7). The second group, called mucosal hyperplasia, has the same features as hyperplastic colon polyps. Benign mucinous cystadenomas form the third group. Finally, the fourth group encompasses the malignant mucinous cystadenocarcinomas, characterized by glandular stromal invasion and/or tumor cells in peritoneal implants i.e. pseudomyxoma peritonei.

In concordance with this classification, the course and prognosis of an appendiceal mucocele varies with the histological subtype (4-6, 8).

Clinical presentation and treatment options

The clinical manifestation is often non-specific, mostly presenting with vague pain and tenderness in the right lower quadrant. An abdominal mass is sometimes palpable. Nevertheless, an appendiceal mucocele is



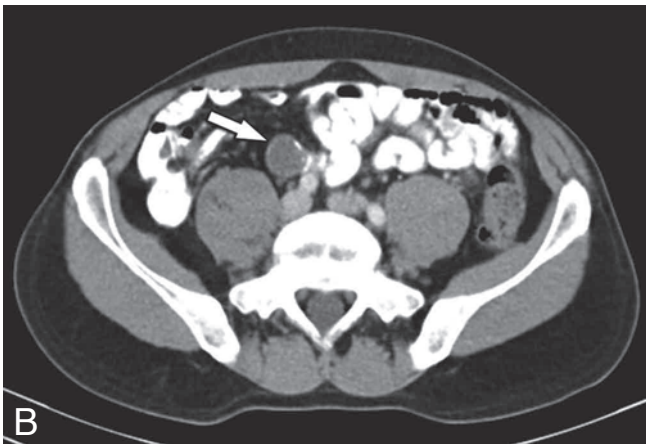


Fig. 1. — Sagittal (A) and axial (B) CT scan shows a cystic, oval-shaped mass in the right lower quadrant with punctuate calcification in the wall of the lesion.

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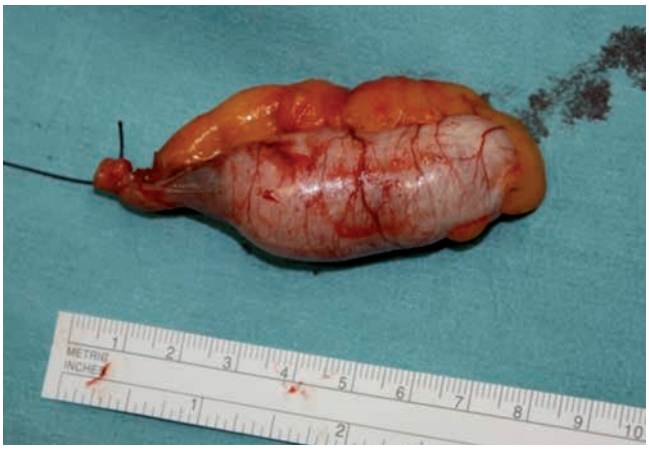

Fig. 2. — Photograph of the resected specimen.

Table I. — Histological division of mucocele of the appendix.

Simple retention cyst	18%
Mucosal hyperplasia	20%
Mucinous cystadenoma	52%
Mucinous cystadenocarcinoma	10%

often an incidental finding, with up to 25% of patients experiencing no related symptoms. When symptoms occur, they may be caused by complications like bowel obstruction, torsion or ureteral obstruction (3, 5, 6, 8).

Since imaging studies cannot reliably differentiate between benign and malignant mucoceles, surgery with complete resection is the treatment of choice. Furthermore, a neoplastic mucocele has the potential to rupture. Standard appendectomy is sufficient for retention cysts, mucosal hyperplasia and cystadenoma. In cystadenocarcinoma, choice of treatment depends on the extent of disease. If no mesenteric or adjacent organ involvement is present, standard appendectomy with resection of the appendiceal mesentery appears to be adequate (9). A right hemicolectomy is required in complicated mucoceles which involve the terminal ileum or caecum.

Imaging findings

The pre-operative radiological diagnosis of an appendiceal mucocele has important clinical consequences. The surgeon must be warned of the possibility of this pathological entity, since there is always a risk of rupture during surgery with subsequent evolution to a pseudomyxoma peritonei. Second, the risk of potential malignancy must always be assessed, especially in older people.

While different imaging techniques can visualize an appendiceal mucocele, it is particularly computed tomography (CT) and ultrasound which have the best diagnostic value (4, 10).

Currently, CT is the modality of choice for evaluating an appendiceal mucocele. The typical appearance is a well-circumscribed mass with a smooth thin wall, with or without mural calcifications located in the right lower quadrant (Fig. 3) (26). However, Kim et al (4) reported that the wall can have a variable thickness with many mucoceles having a thick wall, regardless of their etiology. The density of the mass may also vary, depending on the amount of contained mucin. Mostly it has a low, cystic (water) density, although a higher soft-tissue attenuation is also possible. An important feature is the usual lack of periappendiceal inflammation or abscess (10).

CT is a good technique for demonstrating the anatomic relationship between the mass and the adjacent structures. To adequately determine its anatomic localization, adequate opacification of the terminal ileum and caecum can be necessary.

Finally, CT is the best available modality for detecting mural calcifications, which may be punctuate or curvilinear. The presence of these calcifications is very typical for a mucocele. Nevertheless, calcifications may be absent in up to 50% of cases, making the diagnosis less straightforward (1, 7, 11). Furthermore, the diagnosis may be further compromised in large mucoceles where it may be difficult to determine the exact anatomic relationship with the caecum (12).

Some authors have suggested that ultrasound (US) can be more useful in making the diagnosis of mucocele (12, 13). US typically shows an oval-shaped cystic mass with or without acoustic shadowing, depending on the presence of mural calcifications. The internal echotexture may be variable, with internal consistency varying from water-like to gelatinous (1, 4). This, content may be layered, causing the 'onion skin' sign that represents the concentric pattern of mucoid material in the lesion (11, 12, 13). Similar to CT, an important feature is the lack of inflammation, with no wall thickening (> 6 mm) as seen in acute appendicitis. The wall typically has a layered appearance with an echogenic inner layer and a hypoechogenic outer layer (4, 10).

While only rarely requested, magnetic resonance imaging (MRI) can also demonstrate a cystic mass like CT or US. Given its nature, it is less suited than CT for detecting calcifications. The signal intensity of the mass depends on its content. A high fluid content will lead to a high signal intensity on T2-weighted images (T2WI), with a low signal intensity on T1-weighted images (T1WI). However, if the mass has a high mucin content it will present with a high signal on both T2WI and T1WI (7, 14).

Barium enema can show indirect signs of a mucocele such as an impression on the caecum, but it can not demonstrate the mucocele itself nor its extent (1, 10). It is in practice never used for this indication.

Although there is incidental mentioning of the use of virtual colonoscopy in diagnosis of mucocele, at this time there is no large scale evidence to warrant the routine use of this technique in the evaluation of mucocele (27). There are only a few references in the literature regarding the potential use of ^{18}F-flurodeoxyglucose positron emission tomography/computerized tomography (FDG PET/CT) to detect underlying malignancy in mucoceles. These show that PET has a rather low sensitivity in detecting underlying malignancy and has limited use in the evaluation of appendiceal carcinomas (28, 29).

While there are no pathognomonic signs that can differentiate between a cystadenoma and a cystadenocarcinoma, some radiological features may suggest malignancy. Solid mural wall nodules which enhance after intravenous contrast administration and presence of internal papillary vegetations are suspected signs for a

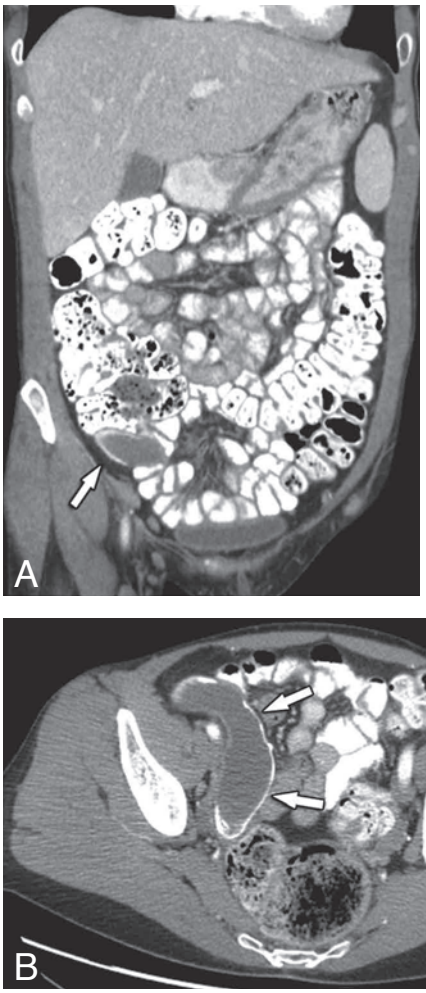

Fig. 3. — Coronal (A) and axial (B) CT scan shows the typical appearance of an appendiceal mucocele with cystic content and mural calcifications.

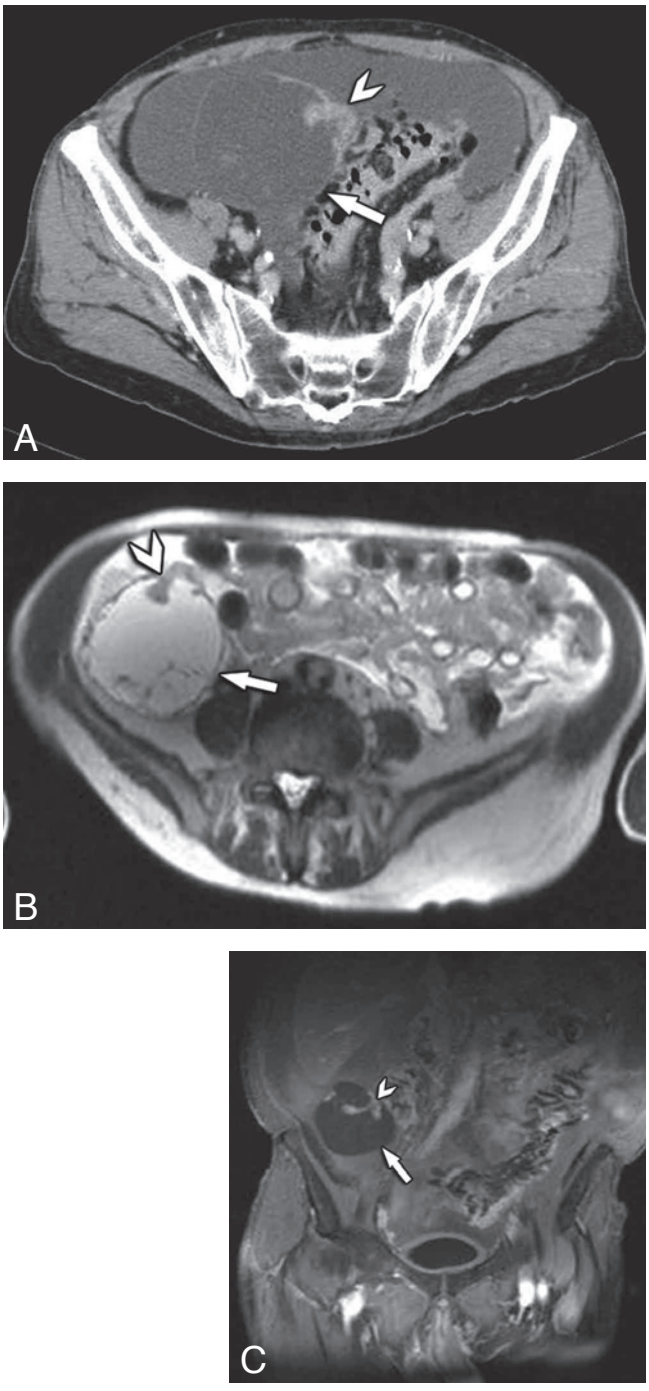

Fig. 4. — Axial CT scan (A), axial MR T2 HASTE (B) and coronal MR T1 GRE (C) with gadolinium from a 75-year-old man with a mucinous cystadenocarcinoma (white arrows) show internal septations, a papillary projection and irregular mural wall thickening (white arrowheads).

mucinous cystadenocarcinoma (4, 15, 16). Furthermore, mesenterial infiltration, peritoneal implants with or without omental cakes, and ascites are also signs that suggest a malignant origin (Fig. 4).

Myxoglobulosis is a rare variant of mucocele where the appendix is filled with translucent spheres. These spheres may calcify, which makes them visible on plain films and CT scans. It is believed that they are derivatives of granulation tissue from the wall of the mucocele that loosen, necrotize and may calcify (1, 17).

Complications of mucocele

It is important to recognize that complications may be the first manifestation of an appendiceal mucocele. It is an infrequent cause of bowel obstruction in adults due to an intussusception or a volvulus. This gives typical imaging findings with a cystic mass as leading point, with associated mesenteric and obstructive signs (Fig. 5). Furthermore, it can also present as an acute appendicitis due to torsion or cause genito-urinary symptoms as a result of ureteral obstruction or bladder compression. Mucoceles, both benign and malignant, can be complicated by a superimposed infection, which causes gas bubbles or an air-fluid level within the mass (5, 6, 7, 18, 25).

Pseudomyxoma peritonei is a complex and confusing entity. It is defined by the presence of mucine and debris in the peritoneal cavity (1, 5, 10, 15, 19, 20). First it was thought only to be present in malignant mucocele (21), but more recently two different categories were defined (1, 19). The first group refers to disseminated peritoneal adenomucinosis

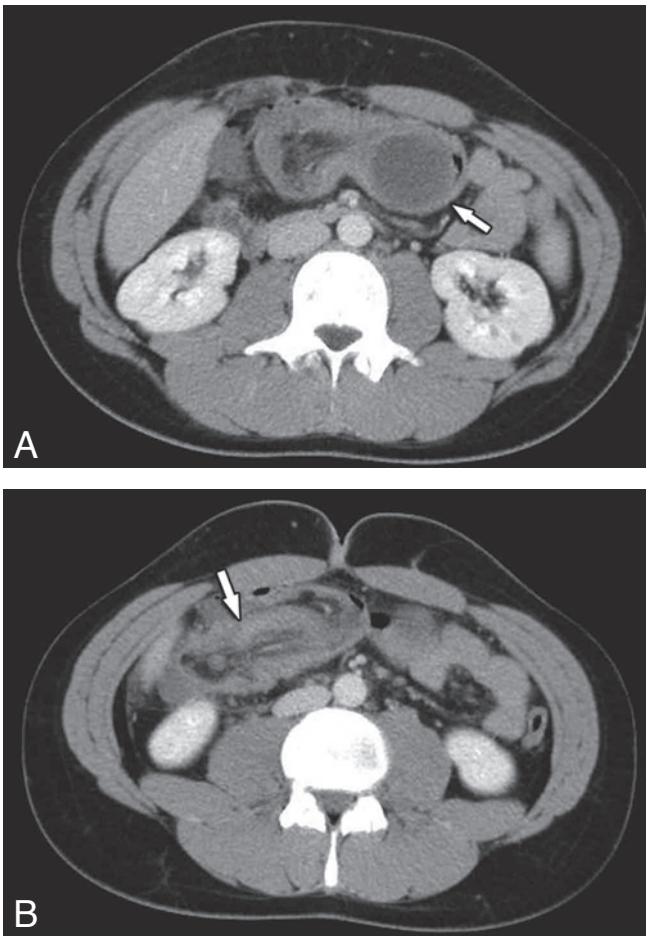

Fig. 5. — Axial CT scan in a 26-year-old male with appendical mucocele (A) presenting as an intussusception, note the mesenteric vessels and fat in the lumen of the colon (B).

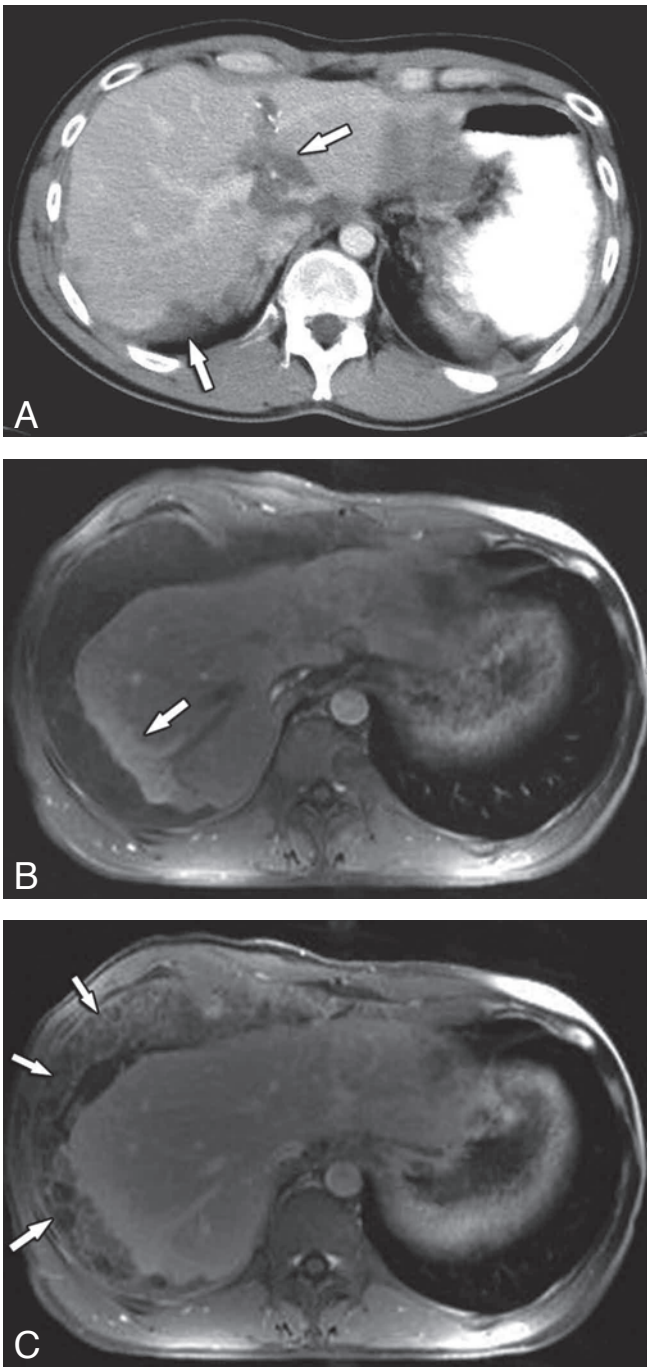

Fig. 6. — Axial contrast-enhanced CT scan (A) and MR GRE T1 +Gd early (B) and delayed (C) phase from a 40-year-old male with pseudomyxoma peritonei: scalloping of the liver with secondary perfusion defects and calcified implants at the falciform ligament.

implants caused by a localized rupture of a benign mucocele. It has an uneventful clinical course, and is thought to occur in approximately 20% of benign mucinous cystadenomas.

The second group, called peritoneal mucinous carcinomatosis, is characterized by diffuse mucinous implants on the peritoneal surfaces and mucus accumulation within the peritioneal cavity. This is caused by mucinous adenocarcinoma, with a poor prognosis and no reported 5-year survival rate (5). Recurrent mucinous ascites with intestinal obstruction is the major cause of morbidity (1).

CT-scan shows scalloping of liver, spleen and mesentery, corresponding to the peritoneal implants (1, 10). Ascites is of low-attenuation or slightly higher in density than a transudative (5-20 HU) ascites due to mucinous material. Mucinous nodules may be seen and may calcify, usually in a rim-like fashion. MRI can also demonstrate pseudomyxoma peritonei. T2WI give optimal contrast differentiation with normal tissue. T1WI after intravenous contrast administration is useful for the evaluation of visceral invasion (Fig. 6).

Pseudomyxoma peritonei, with similar findings, can also be caused by primary ovarian processes (15, 19). In patients with pseudomyxoma peritonei from ovarian cystadenocarcinoma, a mucinous neoplasm of the appendix is nearly always present. Whether this are two primary processes or whether the ovarian tumour is secondary to the appendiceal one remains controversial, with some studies (3, 15) reporting concomitant presence of appendiceal adenoma-type mucocele and ovarian mucinous cystadenoma. Careful examination of the ovaries in women with appendiceal mucocele is therefore recommended.

Table II. – Differential diagnosis.

Intraperitoneal lesions	Retroperitoneal lesions
ovarian cysts and tumours	inflammation, tumours and haemorrhages
enteric duplication cysts	lymphocele
mesenteric and omental cysts	renal cysts
mesenteric hematoma or tumours	pancreatic pseudocysts
abdominal abscesses	

Treatment consists of surgical debulking. Complete surgical tumor removal combined with intraoperative heated chemotherapy during surgery, followed by postoperative intraperitoneal chemotherapy (Sugarbaker technique) may improve symptom-free survival (23).

Finally, while some studies mention an association of roughly 20% between colonic adenocarcinoma and appendiceal mucocele due to adenoma (8, 22)], this is not universally accepted (15).

Differential diagnosis

The differential diagnosis of an appendiceal mucocele is broad and includes both intra- and retroperitoneal lesions (2) (Table II).

Conclusion

An appendiceal mucocele is a descriptive term of a distended, mucus-filled appendix caused by various conditions, both benign and malignant. Correct pre-operative diagnosis is important because among others the possibility of peroperative rupture and subsequent development of pseudomyxoma peritonei. It is the task of the radiologist to alert the clinician and surgeon to the presence of this entity, the potential associated complications and possible signs of malignancy.

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