The vermiform appendix: characterization with imaging of appendiceal mucocele

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Learning objectives

The aim of this paper is to review the radiological findings of appendiceal mucoceles and their complications using different imaging techniques.
Background

INTRODUCTION:

The vermiform appendix is a tubular structure that originates from the caecum approximately 2 cm below the ileocecal valve, at the point where the teniae coli converge [1].

Traditionally the appendix has been regarded as a vestigial organ with no clear function, but nowadays some consider that it may have a role in the immune system [1].

The appendix wall is formed by several layers. From outer to inner these are [2]:

- Serous layer.
- Muscular layer.
- Submucosa.
- Mucosa.

On ultrasound, the appendix is seen as a tubular aperistaltic blind-ending structure which originates from the cecum. It's compressible, with differentiation of layers (to more external to more internal layer are hyperechoic, hypoechoic, hyperechoic), with a diameter less than 0.6 cm and whose wall has a thickness less than 0.3 cm [2].

ENTITIES THAT MAY AFFECT THE APPENDIX.

There are several diseases that may affect the appendix, but by far the most common is acute appendicitis, which is one of the most frequent causes of the acute abdomen in the emergency department.

An inflamed appendix is thought to result from the obstruction of the lumen by different possible causes (such as appendicoliths, neoplasms or parasites) which leads to an increase in the intraluminal pressure secondary to the accumulation of secretions. This causes an alteration in the perfusion of the wall of the appendix [3], wall ischaemia, bacterial invasion, and if the process continues, necrosis of the wall with extension of the infection into the surrounding tissues.

The commonest symptoms are nausea, vomiting, fever and periumbilical pain which subsequently focuses in the right lower quadrant. The blood tests usually show raised white cell count with neutrophilia. Up to 20-30% of patients may have an atypical
manifestation, so imaging, especially ultrasound and CT, plays an important role on the diagnosis.

By neighbourhood, an inflamed appendix can also appear in processes affecting adjacent organs or structures (such as Crohn's disease with involvement the terminal ileum and cecum or with pelvic inflammatory disease), with similar signs in radiological imaging to those associated with acute appendicitis inflammatory changes of neighboring organs.

Furthermore, appendix can be affected by different benign and malignant types of tumors, identified only in the 0, 5 to 1% of appendectomies [4]. Between the tumors that can be found are [4]:

- **Carcinoid tumor:** It's the most frequent, but usually it has less clinical manifestation that epithelial tumors. It rarely presents as a mucocele (because it usually affects the tip of the appendix) or causes carcinoid syndrome. They are usually diagnosed after appendectomy for suspected acute appendicitis.
- **Epithelial tumors:** They are the following in frequency. They are divided into no mucinous and mucinous, belonging to the latter group the mucinous adenoma or mucinous cystoadenoma, which is responsible for most of the most appendiceal mucoceles.
- **Other:** Far less common, appendiceal lymphoma, mesenchymal tumors and other neuroendocrine tumors as paraganglioma, the latter rarer.

The appendiceal mucocele isn't a disease in itself, it's a form of presentation several entities.

**APPENDICEAL MUCOCELE:**

**Definition:**

Appendiceal mucocele is a rare entity that corresponds to a descriptive macroscopic term, referring to a focal or diffusely dilated appendix with accumulation of mucus in its lumen [4-8].

It doesn't correspond to itself a pathological diagnosis, because there are several causes that originate it.

**Pathology:**
Traditionally four causes of appendiceal mucocele have been described. The different findings that can be seen on the pathology are [5, 8-11, 13]:

- **Retention cyst.** It presents a squamous epithelium with atrophic aspect lining the lumen of the appendix.
- **Mucosal hyperplasia:** It’s a mucosal proliferation without atypia. It’s about 5 to 25% of cases. [7]
- **Mucinous cystoadenoma or mucinous appendiceal neoplasm of low grade:** It is a non invasive papillary or flat pattern mucin producing villous epithelium, with or without little atypia. Approximately 63-84% of cases. [7]
- **Mucinous cystoadenocarcinoma or mucinous appendiceal neoplasm of high grade:** It shows cellular atypia and invasion beyond the muscularis mucosa of the wall. It corresponds 11-20% of cases approximately. Fig. 2 on page 9

**Epidemiology:**

It has a low incidence, being an incidental finding in 0,2-0,3% of appendectomies [5-11, 13, 14].

It was considered more common in women than men with a variable ratio, generally about 4:1, although some authors consider the incidence is quite similar, even slightly higher in males [7,10, 12, 14].

The age of diagnosis is usually over 55 years, usually middle-aged or elderly patients [7,11, 12, 14].

**Clinical manifestation:**

The presentation can range from asymptomatic, as an incidental finding in a patient being studied by another cause, 25-50% of cases [7, 9, 12, 14], to the acute abdomen.

Signs and symptoms can be found are [4]:

- Colic pain in the right lower quadrant.
- Painful or painless mass in right lower abdominal quadrant.
- Gastrointestinal bleeding.
- Right ureteral obstruction.

Other: Clinical complications secondary to superinfection simulates an acute appendicitis, intussusception or pseudomyxoma peritonei with increased abdominal girth.

**Differential diagnosis.**
Differential diagnosis should be made with several entities such as [1, 4, 7, 8, 11, 15]:

1. Acute appendicitis.
2. Intestinal duplication cyst.
3. Mesenteric cyst.
4. Adnexal cyst.
5. Hydrosalpinx.
7. Appendiceal lymphoma.
8. Other: Abscess, hematoma.

The radiological findings of these entities are explained in the section "Imaging findings".

Complications:

Complications that can be found are: superinfection, intussusception and pseudomyxoma peritonei.

**Superinfection** may mimic acute appendicitis and the superinfected mucocele may be diagnosed in surgery. It has to be suspected when the blood tests and symptoms suggest infection.

**Intussusception** is a rare complication of appendiceal mucocele (incidence about 0.01 %) [11, 13] that can manifest as colic pain in right lower quadrant, obstruction and gastrointestinal bleeding.

**Pseudomyxoma peritonei** means "false peritoneal tumor". It’s a complication can be seen in up to 6 % of appendiceal mucoceles [7, 11]. It appears mainly in tumoral cases, and particularly in mucinous cystadenocarcinomas. It is a rare entity and according to some authors corresponds to a descriptive term and a syndrome rather than to a specific disease.

It’s characterized by accumulation of mucin or gelatinous material in the peritoneal cavity [4].

The most common causes are appendiceal mucoceles or ovarian neoplasm, also be associated with other malignancies, including colon or gastric tumors [8, 11, 17].

When it finds pseudomyxoma peritonei, it is important to do a detailed analysis to determine if it is only formed by mucus or if there are epithelial cells, because this influences the management and prognosis.
Histologically two conditions that can involve the peritoneum and produce mucin have been described [16,17]:

- **Diffused peritoneal adenomucinosis.** Epithelial cells without atypia can be seen and mitosis are rare. The cells can be floating in the mucus content in the peritoneal cavity or localised in the dependant areas like non invasive mucinous implants.
- **Peritoneal mucinous carcinomatosis.** It usually occurs in high grade tumors and is characterised by invasive implant that make excision almost impossible.

The difference between these two entities also results in a different outcome, so in diffused peritoneal adenomucinosis survival at 5 years is approximately 50 %, and for the peritoneal mucinous carcinomatosis the 5 year survival is about 10% [16].

In pseudomyxoma peritonei is required to review appendix and ovaries in imaging test, it has been observed appendiceal mucoceles and synchronous or metachronous ovarian tumors in the same patient.

Pseudomyxoma peritonei may be focal or diffuse. Clinically, the most striking finding is an insidious increase in the abdominal girth (with a 5 year survival of about 65 %) [4].

**Approach to treatment:**

Once mucocele is diagnosed and the remainder abdominal organs and peritoneal cavity, are assessed we must determine which treatment is the most appropriate for each case [9].

Generally nearly all appendiceal mucoceles are resected, given the potential although not proven risk of transformation of a benign lesion into a malignant lesion, or the risk of rupture [12, 18, 19]. Moreover, an underlying tumor cannot be excluded even in the absence of radiological signs suggestive of malignancy. These signs include:

- Large mucocele [5, 7, 9, 13, 17].
- Mural enhancements in abdominal CT with intravenous contrast [9, 13, 17].
- Pseudomyxoma peritonei [5, 13].

Surgical treatment and management will depend on the findings and histology of the lesion.

Simple appendectomy is the treatment of choice when lesion is small (lower risk of malignancy) and the base of appendix is free [7, 8, 10, 14]. Sometimes when lesion affects the base a small caecectomy may be performed.
Right hemicolecctomy is usually the treatment of choice when associated appendix malignancy, or is required to remove the tumour completely, or pathologic lymph nodes were found in previous surgery. It is very important manipulate the appendix carefully during surgery because its rupture may cause tumoral spread into the peritoneal cavity, which can worsen the prognosis [4,9, 13, 14].

The presence of small amount of periappendiceal liquid does not mean that there is pseudomyxoma peritonei, and similarly, the presence of a small localized focal pseudomyxoma peritonei does not necessarily associate a worse prognosis [11].

The treatment can be performed in a diffuse pseudomyxoma peritonei are resection of peritoneum or peritoneectomy appears to have better results than surgery cytoreductive or "debulking" (which according to the center and the cases can also include appendectomy, omentectomy and bilateral oophorectomy) [4, 14]. Intracavitary hyperthermic chemotherapy can also be used in association with cytoreductive surgery [4]. Systemic chemotherapy can be performed and is preferred in patients with extensive peritoneal involvement and high grade tumor [9].
**Fig. 1:** Cross section of appendiceal mucocele stained with hematoxylin-eosin (1.25X magnification). The anatomopathological study showed a retention cyst. Note on the magnified image on the right bottom corner the mucus filling the lumen of the appendix (black arrow) and the flattened atrophic epithelium (white arrow). (Department of Pathology, Hospital Virgen de la Luz, Cuenca).

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Fig. 2: Section of mucinous carcinoma of appendix stained with hematoxylin-eosin (1.25X magnification). Image A shows a detail of the mucosa without atrophic appearance, unlike the retention cyst (red arrow) and neoplastic cells with loss of differentiation invading the appendiceal wall. Picture B shows invasion of the wall by tumoral cells and mucin lakes within it (black arrow). (Department of Pathology, Hospital Virgen de La Luz, Cuenca).

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Imaging findings OR Procedure details

APPENDICEAL MUCOCELE

Semiology in imaging:

Some radiological techniques, such as the plain abdominal film or barium enema, only allow a diagnostic suspicion. Others allow not only the diagnosis of this entity but also the detection of its possible complications, as well as the evaluation of the rest of the peritoneal cavity. These include ultrasound, magnetic resonance imaging and particularly CT, the modality of choice [7].

On abdominal films, signs that can raise the suspicion of a mucocele are increased density in the right lower quadrant with possible mass effect on adjacent intestinal loops and curvilinear calcifications in the area of the lesion (these are seen in less than 50% of the cases) [4]. Fig. 3 on page 16

The barium enema may show a filling defect in the caudal and medial aspect of the cecum with estrinsic or submucosal mass effect [4], smooth edge and no contrast in the lumen of the appendix.

Ultrasound shows a structure compatible with the appendix increased diameter and pathological aspect. Features can be found:

- **MORPHOLOGY**: Usually elongated with tubular shape (sometimes described like "chicken drumstick") [4], although it can also have rounded morphology. Fig. 4 on page 16
- **SIZE**: Diameter equal or greater 0,6 cm, but it's variable. It should be suspected if the diameter of the appendix exceeds 1,5 cm. Generally when the process is benign, such as retention cyst, they rarely exceed 2 cm [4, 7]. When the diameter is larger than this, probable underlying malignancy must be suspected, and the risk of rupture increases with the diameter.
- **WALL**: It's usually thin. A thin hypoechoic outer layer corresponding to the intestinal wall, and a thin inner echogenic ring have been described [5]. Curvilinear or punctate hyperechogenic images corresponding to calcifications can be seen. Although rare, polypoid projections into the lumen can also be present. Fig. 4 on page 16 Fig. 5 on page 17
- **ECHOGENICITY**: It's variable depending on the ratio between the mucinous and water content. It can be anechoic structure, hypoechoic with non mobile low intensity echos, usually in dependant zone (it allows differentiate them...
from gas bubble) have echogenic contents or show "onion-skin" appearance [9]. Fig. 6 on page 18

- **ACCOUSTIC ENHANCEMENT:** The mucous may attenuate the ultrasound beam [10], so posterior acoustic enhancement is variable and may appear to a greater or lesser degree. Fig. 5 on page 17

- **OTHER FINDINGS:** The Doppler study does not show intralesional flow. The mucocele may be or may be not accompanied of small amount of regional intraperitoneal free liquid, which does not necessarily correspond to localised pseudomyxoma peritonei. The findings related to the complications are discussed in another section. Fig. 7 on page 19

Findings on CT studies are:

- **MORPHOLOGY:** It can be seen as a tubular or rounded cystic-like structure in the theoretical location of the appendix Fig. 8 on page 20

- **WALL:** It can show curvilinear or punctuate hyperdense areas corresponding to calcifications. Nodular areas of parietal enhancement may also be seen, in which case we must suspect a possible cystadenocarcinoma [12]; this a finding that should be highlighted on the radiological report to guide proper surgical management. . Fig. 9 on page 21 Fig. 10 on page 22 Fig. 11 on page 23

- **ENHANCEMENT:** Normally there is not significant enhancement of the wall. If there is wall enhancement or a nodular area is shown, a cystadenocarcinoma area must be suspect [12].

- **CONTENT:** Usually it’s hypodense, with attenuation values among water density and soft tissue density. It depends on the amount of mucous, water, protein... [6].

- **OTHER FINDINGS:** The manifestations of complications will be described in other section.

On MR, it can be seen as a cystic lesion of intermediate or hypodense signal on T1-weighted sequences (depending on the amount of water and proteins) and high signal intensity on T2-weighted sequences presented in the latter a hypointense ring in the wall [7]. The evaluation of mural calcifications is worse on this imaging modality [4], but it provides a better assessment of gynaecological anatomy in women, which are very important in these cases as sometimes appendiceal mucoceles may be associated with ovarian mucinous tumors [4, 11, 16, 17].

**DIFFERENTIAL DIAGNOSIS AND IMAGING FINDINGS.**

Differential diagnosis should be made with several entities [1, 4, 7, 8, 11, 15]:
Acute appendicitis

- **Sonographically** is characterized by an appendix greater than 0.6 cm in diameter, with internal content and non compressible. It can present flow in its wall in the Doppler study. In addition, it may be accompanied by other findings: positive sonographic McBurney sign, abnormal echogenicity of the surrounding fat, free intraperitoneal fluid and small reactive regional lymph nodes. When it progresses, sometimes it's impossible to identify the appendix, substituted by a phlegmon or an abscess in the right lower quadrant seen as an heterogeneous collection. Fig. 12 on page 24  Fig. 13 on page 25

- **On CT** the appendix shows increased in diameter, enhancement of its wall in early stages, and frequently stranding and increased attenuation of the fat, as well free fluid and regional lymphadenopathy. If it progresses, phlegmons and abscesses can form, with or without an identifiable appendix and signs of peritonitis can appear, with intraperitoneal free fluid and peritoneal enhancement.

**Intestinal duplication cyst:** It's a cystic lesion usually seen in young patients. Mural calcifications are rare. Its walls double layered, with an echogenic inner layer corresponding to the mucosa, and a hypoechoic halo related to the muscular layer.

**Mesenteric cyst:** Multilocular cystic lesion. Rarely it shows mural calcifications.

**Adnexal cyst:** Transvaginal ultrasound has an important role to determine the origin of the lesion. Its wall is usually thin and it normally does not show internal mucin echoes or septa. Fig. 14 on page 26

**Hydrosalpinx:** A serpiginous tubular cystic structure in right lower quadrant, with or without internal contents (these are usually present when there is associated). Determining its relation to the uterus is essential for diagnosis.

  - **Ultrasound:** Dilatation of the Fallopian tubes can show content and increased flow in the wall when there is associated infection. Fig. 15 on page 26
  - **CT:** It shows a tubular structure in the adnexal region in relation to the uterine horn, with or without enhancement of the wall with intravenous contrast (usually present when there's superadded infection). Fig. 16 on page 27

**Lymphocele:** Cystic lesion with different sizes and generally with internal septa.
Appendiceal lymphoma: The gastrointestinal tract is the site where extranodal non-Hodgking lymphomas more often appear. Despite this, appendiceal lymphoma is rare, and even rarer that it is associated to a mucocele. They usually present clinically as acute appendicitis [4].

- **Ultrasound:** It can show an enlarged appendix, sausage shaped, but with preserved vermiform appearance, with marked hypoechoic thickening of the wall simulating a mucocele.
- **CT:** It shows increased size appendix, even aneurysmal dilatation, but with soft tissue attenuation.

**Other:** Abscess, hematoma.

**COMPLICATIONS AND IMAGING FINDINGS.**

**Superinfection:** On imaging, we can find inflammatory changes of the periappendiceal fat (increased echogenicity or attenuation). The presence of gas bubbles is a finding that strongly suggests infection [4, 7]. Fig. 3 on page 16 Fig. 17 on page 28

**Intussusception:** On imaging we can see:

- **Ultrasound:** An image of "cup-and ball" [11] with a hypoechoic lesion corresponding to the appendiceal mucocle entering a bowel loop (normally the colon) and dragging with it mesenterium and hyperechogenic mesenteric fat.
- **CT:** It can be seen a filling defect within the right colon secondary a cystic lesion related to the appendix.

**Pseudomyxoma peritonei** can be characterized by:

- **Plain film:** The findings are similar to those of ascites: medialization of bowel loops, blurring of fat planes of the paracolic gutters, properitoneal fat and psoas line, and increased abdominal density.
- **Ultrasound:** The peritoneal fluid may be anechoic or hypoechoic with echos (non-mobile, which helps to differentiate it from loculated ascites) and internal septa. Sometimes it may cause scalloping surface of liver and spleen, which can also help to differentiate from ascites. Fig. 18 on page 29
- **CT:** It's the test of choice, to assess the mucocele and its complications. In pseudomyxoma peritonei, bowel loops are medializated as in other types of ascites, and the content of the peritoneal cavity can have a density similar to water of slightly higher. It can show tin septa and implants with or without curvilinear or punctuate calcifications (the calcifications usually appear after treatment, but occasionally can be seen above). Typically, there is scalloping of the surface of the liver, spleen and bowel loops caused by
the hypodense implants [7]. It's calling "scalloping" [17]. The omentum may show increased density (which can be caused by fibrosis with no tumoral infiltration). There are no liver metastases, lymphadenopathy or chest involvement as a difference with peritoneal carcinomatosis.

Fig. 19 on page 30 Fig. 20 on page 31
Fig. 3: Plain abdominal film. Note the presence of an area of increased density in the right lower quadrant (red arrow) with a pocket of gas in this location. This gas has an atypical morphology in comparison with the bowel gas visualized in the rest of the abdomen. An infected appendiceal mucocele was subsequently demonstrated.

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Fig. 4: Tubular blind-ending lesion in the right iliac fossa, with echogenic content and posterior acoustic enhancement, appearances suggestive of appendiceal mucocele. Note the presence of some small echogenic foci in the wall which could correspond to small calcifications.

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Fig. 5: Appendiceal mucocele with echogenic and hypoechoic content and posterior acoustic enhancement. Note a thin hyperechoic inner ring (white arrow) and a peripheral hypoechoic ring (red arrow) in the wall of the mucocele, corresponding to the intestinal wall.

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Fig. 6: These are examples of mucoceles with different types of appendicular content: hypoechoic (A), echogenic (B) and "onion skin" (C).

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**Fig. 7:** Color Doppler imaging of a lesion suggestive of appendiceal mucocele. No flow is detected.

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Fig. 8: Appendiceal mucoceles seen on axial CT images (A and B) as well as coronal (image C) and sagittal reconstructions (image D). They are well defined hypodense lesions, with tubular or "chicken drumstick" morphology, (C) protruding into the cecum (images A and C, white arrows).

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Fig. 9: Abdominopelvic CT with intravenous contrast, coronal reconstruction. Small calcification in the superointernal aspect of the wall of an appendiceal mucocele.

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**Fig. 10:** Abdominopelvic CT with intravenous contrast. Note the presence of a curvilinear calcification in the tip of the appendix, in a retrocaecal location. The patient had a known appendiceal mucocele.

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Fig. 11: Abdominopelvic CT with intravenous contrast, coronal reconstruction. Note the presence of a small punctate calcification in the caudal wall of an apparently cystic lesion in the right lower quadrant with ill defined margins. This lesion was proven to be a perforated appendiceal cystadenocarcinoma.

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**Fig. 12:** Right iliac fossa US with linear transducer shows a tubular structure in keeping with an enlarged appendix, with flow in the appendiceal wall on Power Doppler. There are inflammatory changes in the surrounding fat and a trace of free liquid. These findings were consistent with acute appendicitis.

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**Fig. 13:** Right iliac fossa US shows an enlarged appendix with hypoechoic content, simulating a mucocele. The structure of the wall layers is lost and the appendix
contains an appendicolith in its lumen (right). These findings are compatible with acute appendicitis.

Fig. 14: Abdominal ultrasound. Cystic lesion with thin smooth wall in the hypogastrium suggestive of adnexal cyst.

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Fig. 15: Abdominal ultrasound with color Doppler shows a tubular cystic lesion related to the uterus, with no detectable flow, appearances compatible with hydrosalpinx.
Fig. 16: CT abdomen with oral and intravenous contrast, axial image. Note the presence of a hypodense lesion adjacent to the right lateral margin of the uterus in keeping with hydrosalpinx.

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**Fig. 17:** Infected appendiceal mucocele. Image A: Abdominal ultrasound showing a tubular lesion with heterogeneous content and internal foci suggestive of gas (white arrow). Image B: Axial CT image which confirms the presence of gas within a mucocele in the appendix. Image C: Abdominal CT, coronal reconstruction, showing gas within the appendicular mucocele and adjacent fat stranding suggesting inflammatory changes (detailed image, black arrow).

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Fig. 18: Abdominal ultrasound showing peritoneal fluid with heterogeneous content and septa suggestive of pseudomyxoma peritonei.

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**Fig. 19:** CT abdomen showing pseudomyxoma peritonei. Image A: There is medialization of the bowel loops (white arrows) and "scalloping" of the splenic contour (black arrow). Image B: "Scalloping" of the liver contour (black arrow).

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**Fig. 20:** CT abdomen. Note the loss of the normal smooth liver contour ("scalloping" liver) in a patient with pseudomyxoma peritonei.

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Conclusion

The appendiceal mucocele is an entity with a low incidence and rare cause of acute abdominal pain, but which should be familiar to perform appropriate clinical management.
References


Personal Information

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