

# Mucinous Neoplasm of the Appendix as a Mimic of Cystic Adnexal Pathology

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## INTRODUCTION

Mucinous neoplasms of the appendix are an enigmatic and controversial entity with unpredictable biological potential. Their significance is their potential to spread to the peritoneum and viscera and form gelatinous deposits through mucin-secreting malignant epithelial cells. This is the most common cause of pseudomyxoma peritonei (PMP) which is a descriptive term that covers several histological entities.<sup>[1]</sup> In its most aggressive form, the gelatinous deposits coalesce to form large volume mucinous ascites. This can cause bowel obstruction and death. Appendix mucocele can represent a benign retention cyst caused by luminal obstruction by a fecalith.

Being alert to the possibility of a mucinous neoplasm of the appendix allows prompt surgical excision. Appendectomy before appendiceal rupture is critical to outcome. Certainly, the clinical presentation of mucinous neoplasms of the appendix is highly nonspecific. Chronic right lower quadrant pain is the most common presentation. PMP frequently presents with increasing abdominal girth or with a large mass. By this stage,

highly aggressive management is the patient's only hope for long term survival.<sup>[2]</sup>

We present a case where a young lady presented with minor chronic pelvic pain and who was otherwise asymptomatic with the imaging findings and selected images from her ultrasound, magnetic resonance imaging (MRI), and computed tomography (CT) to aid visualization. She was treated surgically and, at the time of writing, has not suffered from disease recurrence. We have reviewed the English language literature pertinent to mucinous tumors of the appendix and PMP and summarizes this complex topic in the discussion.

## CASE REPORT

A 47-year-old lady presented to her family doctor with pelvic pain and secondary amenorrhea. She was referred

### ABSTRACT

Pelvic pain presents a common diagnostic conundrum with a myriad of causes ranging from benign and trivial to malignant and emergent. We present a case where a mucinous neoplasm of the appendix acted as a mimic for tubular adnexal pathology on imaging. With the associated imaging findings on ultrasound, computed tomography, and magnetic resonance imaging, we wish to raise awareness of mucinous tumors of the appendix when tubular right adnexal pathology is present both in the presence of pelvic or abdominal pain or when noted incidentally. Tubular pathology such as uncomplicated paraovarian cysts or hydrosalpinx is frequently treated conservatively with long-interval follow-up imaging or left to clinical follow-up. Thus, if incorrectly diagnosed as tubular pathology, an appendix mucocele or mucinous neoplasm of the appendix is likely to be undertreated. We wish to clarify some of the confusion around nomenclature and classification of the multiple entities that are comprised by the terms mucocele and mucinous tumor of the appendix.

**KEYWORDS:** Adnexal, appendix neoplasm, mucocele, tubular

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for an ultrasound examination which showed complex fluid and a tubular lesion on the right side containing some sedimenting debris [Figures 1 and 2]. Overall impression was that of hydrosalpinx with adjacent loculated pelvic fluid. No vascular component was identified. The suggestion of layering of the echogenic material within the right-sided tube raised the possibility of previous hemorrhage or infection. Some fluid or low-density material appeared to undercut the thin wall of the structure, separating the layers. These somewhat atypical appearances led her to be referred for a pelvic MRI.

Our institutional routine pelvic MRI protocol was performed, and T2-weighted image (T2WI) HASTE sequences were obtained in three planes along the axis of the endometrial cavity. Axial T1WI and axial T1 fat saturated sequences were also obtained. Gadolinium was not administered [Figure 3a-d] MRI showed no evidence of hydrosalpinx or any ovarian abnormality. In the right lower quadrant, apparently contiguous with the base of the cecum and separate from the right ovary, a vertically orientated tubular cystic lesion with low-T1W and high-T2W signal intensity was noted. This measured 3.1 cm AP  $\times$  3.5 cm TR  $\times$  8.9 cm CC. It contained thin walls with mild wall irregularity and was unilocular with no solid components. The findings raised the suspicion of an appendix mucocele possibly caused by a mucinous tumor.

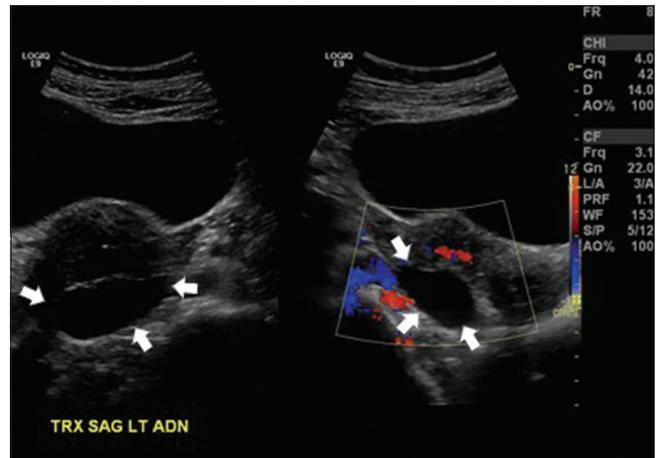
CT of the abdomen and pelvis with intravenous omnipaque in the portal venous phase was performed for confirmation and to allow evaluation of the remainder of the peritoneum and for staging.

A tubular fluid-filled lesion originating from the cecal pole with similar measurements to MRI was noted [Figure 4]. Coarse wall calcification was present with no obvious wall thickening or evidence of rupture. A small volume of free fluid was adjacent to its tip with no substantial ascites. No evidence of lymphadenopathy or other abnormalities were identified.

During the surgery, an abnormal appendix with paper thin walls and tanned smooth serosa was found. The lumen was dilated and contains yellow-tan gelatinous material.

Histopathology showed carcinoma *in situ* with the epithelial invasion of lamina propria. Acellular mucin had invaded through the muscularis propria into the subserosa but not to the serosal surface. No invasion by the epithelial cells was present.

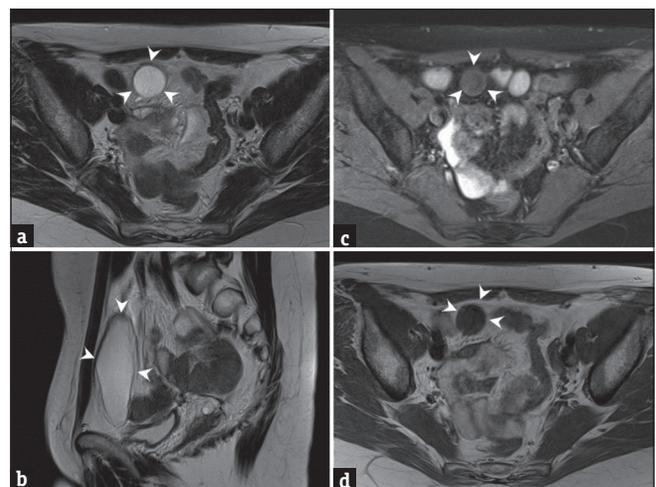
This is a controversial situation which can be classified by the latest AJCC classifications as pT3. The definition



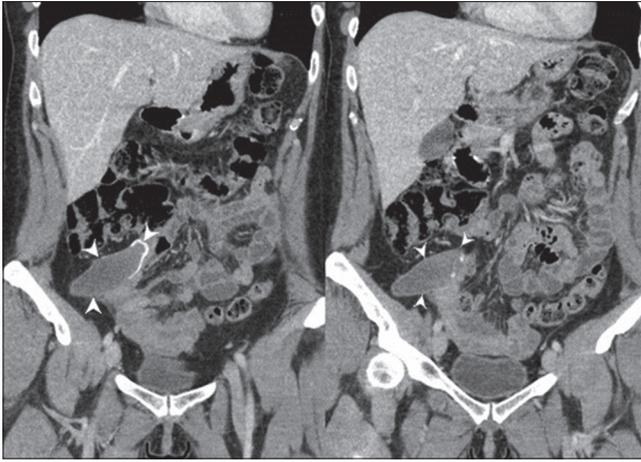
**Figure 1:** A 47-year-old lady presenting with pelvic pain and secondary amenorrhea. Transabdominal ultrasound of showing a dilated fluid-filled right adnexal lesion (arrows).



**Figure 2:** A 47-year-old lady presenting with pelvic pain and secondary amenorrhea. Transvaginal ultrasound showing the thin-walled tubular structure (arrows) with adjacent pocket of free fluid (arrowheads).



**Figure 3:** A 47-year-old lady presenting with pelvic pain and secondary amenorrhea. (a and b) Axial and sagittal T2 turbo spin echo showing a vertically orientated tubular fluid-filled structured with thin wall, pointing toward the right lower quadrant (arrowheads). (c and d) Axial T1 fat sat and axial T1 show the structure to be low in T1-weighted signal (arrowheads).



**Figure 4:** A 47-year-old lady presenting with pelvic pain and secondary amenorrhea. Coronal reformats from portal venous phase computed tomography abdomen and pelvis demonstrating the distended, mucous-filled appendix with thin walls and coarse wall calcification (arrowheads).

given currently for pT3 is that “Tumor extends through the muscularis propria into the subserosa or mesoappendix;” however, it has not been clarified as to whether the term “tumor” includes acellular mucin such as in this case or if it necessitates epithelial invasion, which is not present in this case.

The patient made an uneventful recovery from her appendectomy with no evidence of disease recurrence at this time, 6-month postprocedure.

## DISCUSSION

Appendiceal tumors are very uncommon, if not exceedingly rare, with a large review of appendectomies finding primary appendiceal malignancies in 0.1% of specimens.<sup>[3]</sup> Malignancies can be epithelial, neuroendocrine tumours (NET) or lymphomatous. NETs were previously thought to be more common, but a large review has now demonstrated that epithelial tumors are more prevalent, with NETs only comprising 11% of appendiceal tumors.<sup>[4]</sup>

Epithelial tumors range from benign retention cysts to cystadenocarcinoma. Mucocele is a descriptive term to describe a distended appendix filled with mucoid fluid and covers a wide range of entities. Typically mural thickness in a mucocele is <6 mm and they can demonstrate an echogenic submucosal layer sandwiched by echogenic edematous layers which can give a characteristic targetoid appearance which is used to differentiate them from acute appendicitis.<sup>[5]</sup> Should a benign mucocele rupture, there is no risk of disease spread.

Mucoceles can also be caused by malignant epithelial tumors and imaging cannot reliably differentiate these from retention cysts. NETs most commonly arise at the tip of the appendix and hence rarely cause luminal

obstruction. Lymphoma tends to be non-Hodgkin’s type and is rarer than the other malignancies. It most frequently presents with perforation acutely and rarely causes mucocele formation.<sup>[6]</sup>

Mucinous tumors of the appendix underwent specialist review in 2016 by the Peritoneal Surface Oncology Group International to resolve some of the controversy surrounding their classification.<sup>[1]</sup> Various systems were previously in place including the WHO classification of 2010.<sup>[7]</sup> These were in consensus in defining a benign neoplastic adenoma as disease confined to the mucosa, without mucin or cells penetrating the muscularis mucosa or evidence of perforation. They are also concordant that adenocarcinoma is a frankly invasive neoplastic process with cellular invasion beyond the muscularis mucosa. The previous classifications differed in how they defined adenomatous growths with intermediate cellular atypia or with mucin dissection beyond the muscularis mucosa in the absence of cellular invasion.

Appendiceal adenomas are low grade, confined to the mucosa of the appendix with no evidence of invasion beyond the muscularis mucosa. They should be considered similar to traditional colorectal adenomatous polyps.

Low-grade appendiceal mucinous neoplasm (LAMN) is a term that should be reserved for histologically LAMNs with any of the following features: loss of muscularis mucosae, fibrosis of submucosa, dissection of acellular mucin into the appendix wall, rupture of the appendix, or presence of mucin or cells outside of the appendix. They must not include any of the features used to describe high-grade lesions or adenocarcinoma.<sup>[1]</sup>

The term high-grade appendiceal mucinous neoplasm (HAMN) was devised to describe lesions that had high-grade cellular atypia that could not be included in the LAMN but that did not have the infiltrative invasion of cystadenocarcinoma.<sup>[1]</sup>

Calcification, internal septations, and periappendiceal fat stranding can be present in LAMN or HAMN. The distinction between the two is histopathological and important for patient prognostication. Soft-tissue thickening inside the appendix, wall irregularity, or presence of PMP have been shown to be the most reliable markers for malignancy.<sup>[8]</sup>

When associated with LAMN, the presence of intraperitoneal extracellular mucin deposits (PMP) is classified as disseminated peritoneal adenomucinosis (DPAM). When associated with HAMN, PMP is classified as peritoneal mucinous carcinomatosis (PMCA). In one review undertaken as part of classification, 5-year survival rates were 84% for

patients with DPAM and 6.7% for patients with PMCA, demonstrating the importance of histological grading.<sup>[9]</sup>

If PMP is present in the presence of cellular atypia (DPAM or PMCA), aggressive management involves debulking, peritonectomy, and hyperthermic intraperitoneal chemotherapy.<sup>[10,11]</sup>

A distinctive feature of PMP is its redistribution phenomenon. The mucus follows the distribution of peritoneal fluid within the peritoneal cavity to sites of fluid absorption where the cells concentrate. Consequently, mobile loops of small intestine are spared, but the pelvis, paracolic gutters, omentum, and liver capsule are affected. The capsular involvement causes the almost pathognomonic “scalloped” appearance. These deposits can demonstrate coarse calcification on CT, a further specific finding.

MRI has been shown to be more sensitive in detecting peritoneal deposits than CT, usually with low T1WI, high T2WI, and variable enhancement. Cellular deposits frequently exhibit diffusion restriction.<sup>[10]</sup>

## CONCLUSIONS

In a female patient, a blind-ending, dilated and fluid-filled structure could reasonably be considered to represent a hydrosalpinx, paratubal, or paraovarian cyst on ultrasound.<sup>[12]</sup> Thus, if the reporting radiologist does not consider the possibility of an appendiceal mucinous neoplasm as a mimic for gynecological disease, the abnormality may be undermanaged, leading to long-term surveillance or no routine imaging follow-up at all.

Delay in diagnosis may allow for tumor progression to high grade or perforation with the subsequent dramatic increase in morbidity and mortality. Features to be aware of that may indicate appendiceal neoplasm rather than gynecological pathology and include lamellated appearance to the wall of the tubular structure, sedimenting debris, and an origin in the region of the cecal pole. We urge reporting radiologists to give consideration to the possibility of appendiceal tumors in these cases.

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## Conflicts of interest

There are no conflicts of interest.

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