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## Incidental appendiceal mucinous neoplasm mimicking a left adnexal mass: A case report

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## ABSTRACT

**INTRODUCTION:** Appendiceal mucinous neoplasm is a rare type of appendiceal tumors which can present in a variety of symptoms and is difficult to diagnose. Preoperative diagnosis depends mainly on diagnostic imaging such as ultrasonography and computerized tomography (CT) scan. This uncommon case report discusses an appendiceal mucinous neoplasm mimicking a left adnexal mass on presentation, physical examination and diagnostic imaging findings.

**PRESENTATION OF CASE:** This is a 61-year-old female found to have a large left adnexal mass during follow up ultrasonography. The patient refused further imaging, and during laparotomy, she was found to have an appendicular mucocele with normal left and right ovaries.

**DISCUSSION:** Appendectomy was done and the final pathology came as appendiceal mucinous neoplasm. Her post-operative course and 3 years follow up were uneventful.

**CONCLUSIONS:** The equivocal signs and symptoms along with the anatomical position of appendiceal mucocele makes it difficult to diagnose and can mimic other types of tumors. Therefore, it should be considered in the differential diagnosis of lower abdominal and pelvic masses.

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## 1. Introduction

Appendiceal mucocele describes the progressive dilatation of the vermiform appendix due to abnormal mucus accumulation in the lumen [1]. Mucus is retained as a result of lumen obstruction or hyperproduction due to benign (fecaliths, post inflammatory fibrosis, hyperplastic polyps, serrated adenomas) or malignant (low or high-grade appendiceal mucinous neoplasm, carcinoid tumors, mucinous adenocarcinomas) pathologies [2]. Appendiceal mucoceles formed due to appendiceal mucinous neoplasms (AMNs) are exceptionally rare, with an incidence of 0.12 cases per 1 million individuals per year [3,4].

AMNs are occasionally found incidentally, during follow up or at the time of surgery for other causes, and frequently diagnosed in the late stages [5]. The symptoms of AMN vary significantly, and it is asymptomatic in 25% of cases [6]. Hence, a wide range of differential diagnoses should be considered, to include appendicitis, mesenteric or duplication cyst, or adnexal mass [7].

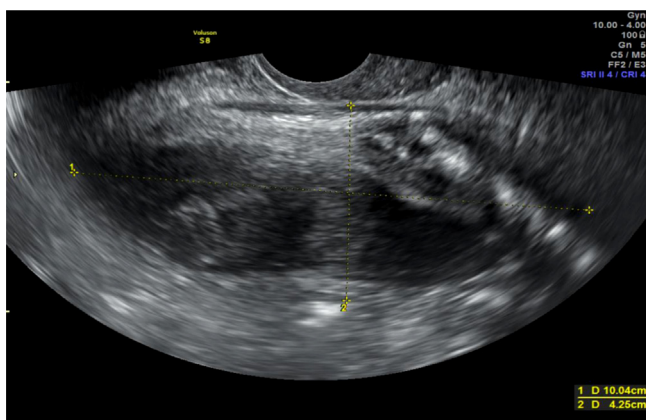
In the current case report, we present an incidental appendiceal mucinous neoplasm mimicking a left adnexal mass. We use the American Joint Committee on Cancer (AJCC) 8th edition [8] for the classification, prognosis and treatment of AMNs, and the case is reported in line with the updated consensus-based surgical case report (SCARE) guidelines [9].

## 2. Case presentation

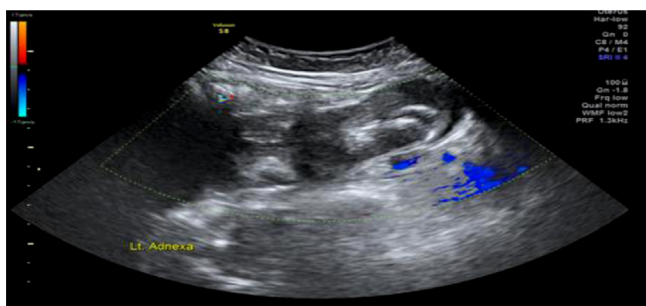
A 61 year old Jordanian female was referred to the obstetrics and gynecology clinic at our institution from health center due to suspected large left ovarian mass discovered by pelvic ultrasound (US). The patient was postmenopausal (gravida 7, para 6, 1 abortion). Her last childbirth was 25 years back and past history revealed that she underwent hysteroscopy and polypectomy of an endometrial polyp 3 years ago. The newly discovered mass was found during her regular follow up. Upon this incidental finding, the patient was not complaining of any gynecological symptoms. She was menopausal since 20 years, with no complaints of abdominal pain or mass, vaginal bleeding or discharge, or weight or appetite loss during the past months. There were no other gastrointestinal or urologic symptoms, and the rest of her systemic examination was unremarkable. Her family history was negative for gynecological or gastroenterology cancers, but the patient had comorbidities including type 2

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**Fig. 1.** Pelvic ultrasound showing large left adnexal mass: right ovary could not be seen at time of scan due to overlying bowel gas.

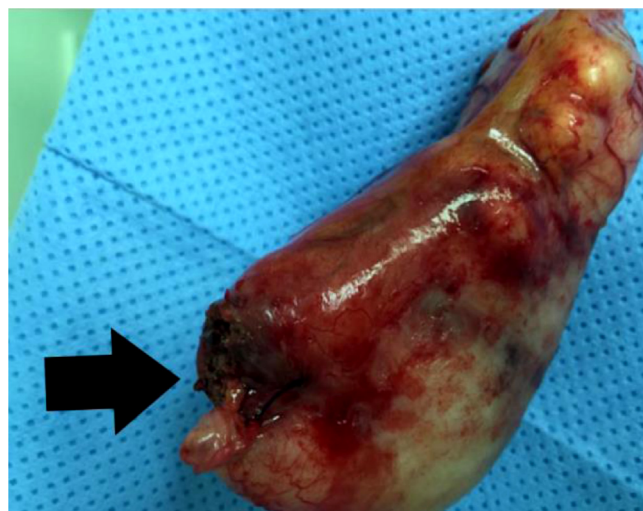


**Fig. 2.** Pelvic ultrasound showing large left adnexal mass (10.0 × 4.2 cm) with calcifications.

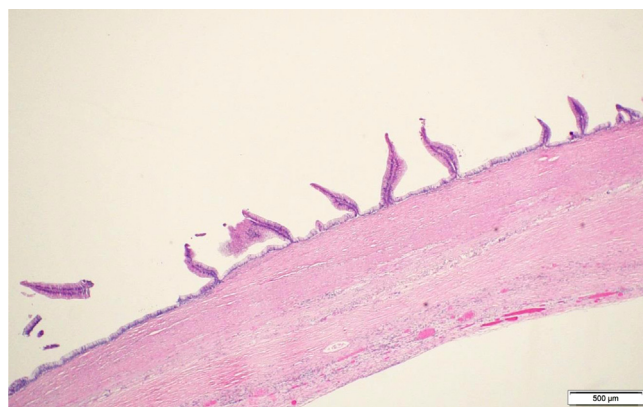
diabetes mellitus, hypertension and hyperlipidemia which were controlled by oral medications.

Examination of the patient revealed that she was vitally stable with unremarkable abdominal or gynecological findings, and no mass could be palpated in the lower abdomen. Investigations showed that her laboratory tests were unremarkable except for iron deficiency anemia and mildly elevated cancer antigen 19-9 (CA 19-9) (40 U/mL). Other tumor markers for ovarian and other solid intraperitoneal organs were within the normal range: cancer antigen 125 (CA 125), cancer antigen 15-3 (CA 15-3) and alpha-fetoprotein (AFP) were 9 U/mL, 13 U/mL and 2 Ng/mL respectively. Her pelvic US (Figs. 1 and 2) showed large left adnexal oval shaped heterogeneous solid mass lesion (10.4 × 4 cm), containing areas of calcification, and the left ovary could not be seen. Both the right ovary (2.9 × 2 cm) and the uterus were unremarkable. No free fluid was found in the pelvis. The patient was referred for MRI abdomen and pelvis, but she refused to proceed with any further imaging. The condition was discussed thoroughly with the patient and due to a high suspicion of ovarian malignancy, the decision was made to proceed with surgery. We did not start with laparoscopy due to large tumor size (10.4 × 4 cm), as exploring the pelvic region, resecting the tumor and delivering it would have been very difficult and could expose the patient to further complications. The patient was posted for exploratory laparotomy with or without total abdominal hysterectomy with salpingo-oophorectomy.

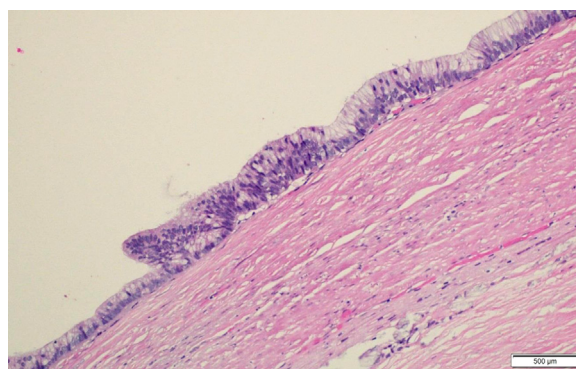
During lower midline laparotomy by an experienced surgeon and after full inspection of the pelvic organs including right and left adnexa along with the uterus and sigmoid colon, the patient was unexpectedly found to have a large appendiceal mass at the terminal portion of her appendix approximately 15 cm (long) × 5 cm (wide). The appendix was intact with no spillage or perforation, and no peritoneal nodules were found (Fig. 3). The uterus and both ovaries were normal. Appendectomy with excisional biopsy of the



**Fig. 3.** Appendicular mucocoele post resection: appendix was removed intact without spillage. Arrow points to surgical knot near the proximal resection margin which was found to be free of malignancy.



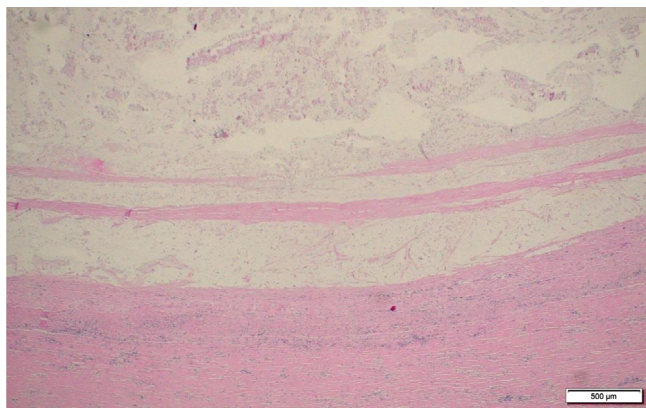
**Fig. 4.** Villous and flat proliferation of mucinous epithelial cells lining the appendiceal mucosa in LAMN, HX&E, x 4.



**Fig. 5.** Flat proliferation of mucinous epithelial cells with low grade nuclear atypia in LAMN, HX&E, x 20.

omentum that was attached to the appendix was hence performed. The post-operative period was unremarkable, with no complaints or complications, the patient was stable vitally and started diet on day 1. She was followed for the next 2 days then discharged.

Her histopathology results indicated a diagnosis of low grade appendiceal mucinous neoplasm (LAMN, 17 × 3 cm). Figs. 4–6 show villous and flat proliferation of mucinous epithelial cells lining the appendiceal mucosa with low grade nuclear atypia. The acellular



**Fig. 6.** Acellular mucin dissect the muscularis propria of the appendix in LAMN, HX&E, x 10.

mucinous cells had invaded into the muscularis propria, the proximal and mesenteric margins were uninvolved by tumor, and there were no identifiable tumor deposits, lymphovascular or perineural invasion. No lymph nodes were found in the submitted specimen and the pathological classification was pT2, pNx [10]. Staging computerized tomography (CT) scan of the chest and abdomen in order to detect any distant metastasis were negative. The patient was referred and discussed at our gastro-intestinal multidisciplinary (MDT) meeting, and the plan was to follow her up in the General surgery outpatient clinic with no need for further intervention or completion surgery. The patient was followed for the next 3 years with no signs of recurrence. She was satisfied with the outcome.

### 3. Discussion

AMN is a very rare disease with heterogeneous clinical features [11]. To the best of our knowledge, the current case could be the second case reported describing an AMN mimicking a left ovarian tumor, the first being from United States [12], despite that many case reports described AMN mimicking a right ovarian tumor [13,14]. Accurate preoperative diagnosis of AMN is usually difficult due to its diverse clinical features, lack of specific tumor biomarkers, and susceptibility of appendiceal neoplasms to involve/ metastasize to the ovaries hence mimicking ovarian tumors [11,15].

In terms of demographics, the current patient was a female aged 61 years, in agreement with other reports suggesting that AMN tends to have a slight female prevalence and is typically found in patients in their 5th and 6th decades; although, it may occur at any age [6,16].

As for presentation, the possible symptoms of appendiceal mucocele range from lower abdominal or pelvic pain, fever, nausea, and vomiting to asymptomatic presentation [16,17]. Our case represents a classic example of the lack of symptoms and difficulty in diagnosis as the patient was actually asymptomatic and the mass was found incidentally during routine US follow up.

In terms of imaging, diagnosis of appendiceal mucocele often depends on diagnostic imaging, and a main feature that distinguishes appendiceal mucocele from uncomplicated appendicitis by US is the lack of appendiceal wall thickening of > 6 mm [18,19]. We were unable to confirm the presence or absence of appendiceal wall thickening as the appendix could not be separately identified in right iliac fossa due to obscuring bowel gas and large pelvic mass.

Several US appearances specific for appendiceal mucocele might be encountered e.g. a bottle-like shaped solid appendicular mass sliding over the uterus and ovaries [19,20] or the highly specific “onion skin sign” (concentric echogenic layers with septa and fine

echoes) [21,22] can help to distinguish appendiceal mucocele from an ovarian cyst. Unfortunately, such diagnostic appearances were not evident in the US we undertook. Others suggested that the presence of the onion skin sign within a cystic mass in the right lower abdominal quadrant, with a normal right ovary, could be specific for the diagnosis of appendiceal mucocele [22].

Colonoscopy, MRI, and computed tomography (CT) are all valuable investigations that can aid in reaching an accurate diagnosis [23]. Colonoscopic findings e.g. “volcano sign” (appendiceal orifice seen in the center of a firm mound covered by normal mucosa) and a bulbous submucosal lesion of the cecum, establish precise diagnosis and are useful for the management [24]. In MRI, appendiceal mucocele lesions are well encapsulated cystic masses, hyperintense on T2-weighted sequences, and hypo- or isointense on T1-weighted sequences [25]. The typical CT appearance of an appendiceal mucocele is a large and well-encapsulated cystic mass in the expected region of the appendix; calcifications of the cyst wall are very specific to appendiceal mucocele and are valuable to characterize the mucocele from an abscess collection [25,26]. Although CT is considered the most informative imaging technique, the diagnosis is more difficult in the absence of cystic calcifications and failure to identify the organ of origin [27]. Unfortunately, the current patient refused to provide consent to proceed with any further imaging (either CT abdomen or MRI abdomen) as a second modality of diagnosis.

Although all the imaging techniques and related signs highlighted above can help to differentiate an appendiceal mucocele from primary ovarian tumors, a primary AMN is rarely diagnosed before operation and histopathological examination when compared with other more frequent types of appendiceal or colonic tumors [15]. Hence, recent views advocate considering AMN in the differential diagnosis of any pelvic mass in elderly female patients, and not to rely mainly on preoperative imaging tools [13–15].

In our case, the pre-operative misdiagnosis was possibly due to: a) the specific ‘onion skin’ sign was not evident on US; and, b) although calcifications within the mass and the wall were present, the right ovary was not recognized due to obscuring bowel gas, and the mass was mimicking a left adnexal mass (Fig. 2). Finding of a large left adnexal mass occupying the pelvic region usually prompts additional diagnostic imaging to further outline the anatomy before intervention, but the patient refused further MRI imaging. Others have reported a misdiagnosed ovarian mass during preoperative evaluation which turned out to be appendiceal mucocele during surgical exploration, and attribute such misdiagnosis to the mimicking symptoms, nearby surgical anatomy and invasion of the neoplasm to the surrounding ovaries and uterus [13].

As for the procedure, upon direct visualization of the lesion at the time of surgery, the surgeon recognized this pathological entity and performed a simple appendectomy that was needed, and the patient was referred for long term follow-up after discussion at our gastrointestinal MDT meeting. The patient was recently seen at the clinic after 3 years of follow up and was recurrence-free, using tumor markers (no elevation), colonoscopy, and imaging.

### 4. Conclusions

The symptoms of appendiceal mucocele vary significantly and are relatively similar to ovarian tumors. These similar signs and symptoms, along with the anatomical position of the appendiceal mucocele, renders it difficult to diagnose appendiceal mucocele as it can mimic other types of tumors, particularly ovarian tumors. Although multiple diagnostic imaging techniques can aid in reaching an accurate diagnosis, a primary AMN is rarely diagnosed before operation and histopathological examination. Hence, high suspi-

cion is required, and recent views advocate considering AMN in the differential diagnosis for any pelvic mass in elderly female patients.

### Declaration of Competing Interest

The authors report no declarations of interest.

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Nothing to declare.

### Ethical approval

Approved by the Medical Research Center (IRB), Hamad Medical Corporation, reference number (MRC-04-20-133).

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Author's contribution

Ammar Aleter: data collection, interpretation, writing the paper, editing the paper. Walid El Ansari: study concept, data interpretation, writing the paper, editing the paper. All authors reviewed and agreed on the final version of the paper.

### Registration of research studies

Not first in Man.

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