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Appendiceal Mucocele: A Rare but High-Risk Entity

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Abstract **Case Report**

Appendiceal mucocele is a rare condition resulting from mucin accumulation and appendiceal dilation. It may remain asymptomatic or present as right iliac fossa pain, often mimicking other pathologies. We report the case of a 54-year-old woman with no medical history who presented with localized abdominal pain. CT imaging revealed a well-encapsulated, non-enhancing oblong mass connected to the appendix, suggestive of a mucocele. Surgical exploration confirmed a low-grade mucinous neoplasm confined to the appendiceal epithelium without rupture or peritoneal spread. This case highlights the essential role of imaging in early diagnosis and surgical planning to avoid serious complications such as pseudomyxoma peritonei.

Keywords: Appendiceal Mucocele, CT Imaging, Low-Grade Mucinous Neoplasm, Pseudomyxoma Peritonei, Right Iliac Fossa Mass.

INTRODUCTION

Appendiceal mucocele is a rare condition, found in 0.2% to 0.6% of appendectomy specimens. It primarily affects women between 50 and 60 years of age. This pathology presents two major concerns: its potential malignancy and the risk of pseudomyxoma peritonei if rupture occurs, which happens in 10–15% of cases.

CASE REPORT

This is a 54-year-old female patient with no significant medical history, presenting to the emergency department with abdominal pain localized to the right iliac fossa. A previously performed CT scan revealed an oblong cystic mass located in the right parietocolic gutter, initially suggestive of a hydatid cyst.

A follow-up abdominal CT scan was subsequently performed at our institution, revealing an oblong formation connected to a structure resembling the appendix, with no enhancement or wall thickening. The diagnosis was consistent with an appendiceal mucocele. Figure 1

The patient subsequently underwent a laparotomy with appendectomy and closure of the cecal base. No metastases, peritoneal carcinomatosis, or ascites were observed. Histopathological analysis later confirmed a low-grade mucinous neoplasm confined to the appendiceal epithelium, without evidence of rupture. Figure 2

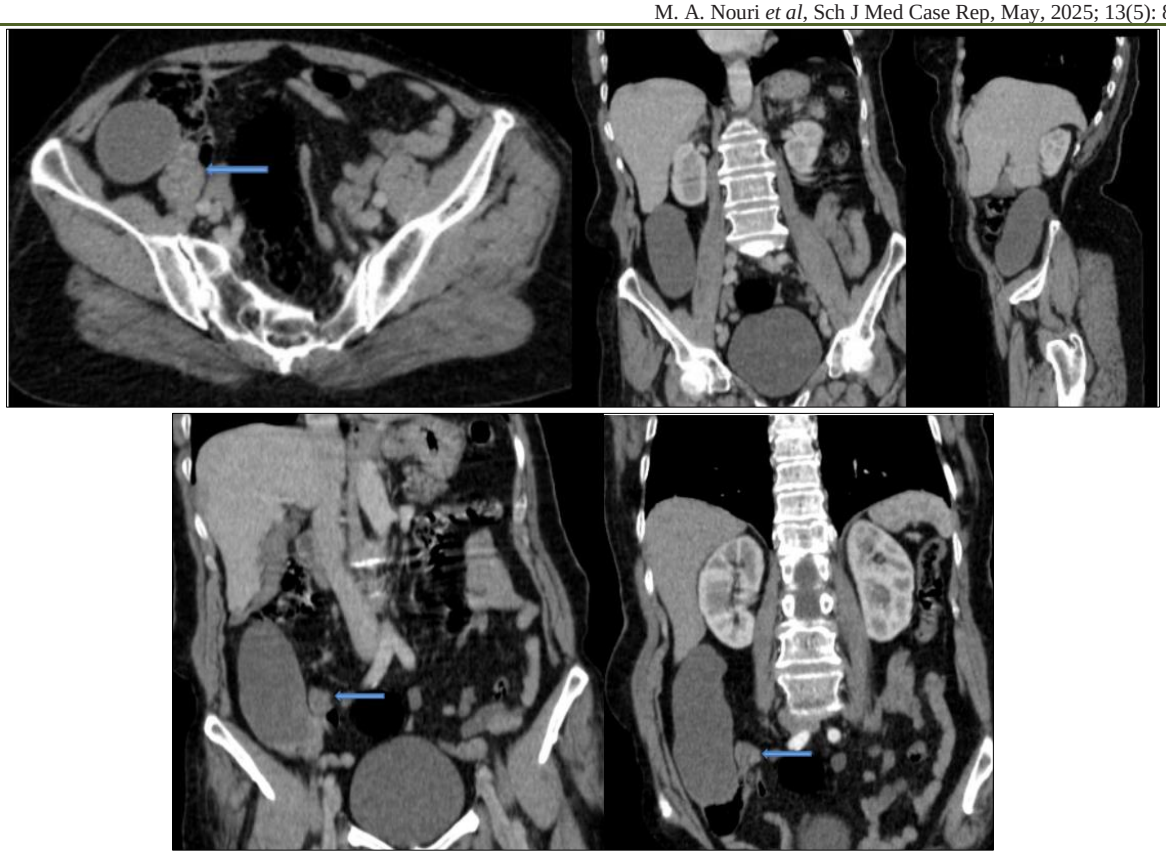

Figure 1: Axial, coronal, and sagittal CT images showing an oblong formation involving both ends of the appendix (blue arrow). The lesion shows no enhancement after contrast injection and demonstrates neither mural thickening nor wall enhancement

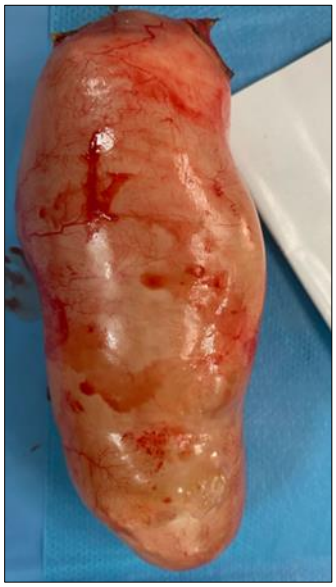

Figure 2 : Surgical specimen showing an oblong mass attached to the appendix

DISCUSSION

Etiologies and Classifications

Mucocele may be:

- Benign, such as retention cysts due to appendiceal obstruction by a fecalith or inflammatory stricture.
- Neoplastic, in mucin-secreting tumors, including:
- Villous hyperplasia (villous adenoma): mildly dilated appendix, thin mucosa, no atypia or mitoses.
- Mucinous cystadenoma: significant dilation, mucin-secreting epithelium, potential dysplasia.

- Mucinous cystadenocarcinoma: malignant form with muscular invasion and neoplastic cells in peritoneal mucin.

Histopathological examination is essential for definitive diagnosis.

Role of Imaging

Detailed Imaging Assessment:

- On **ultrasound**, mucoceles typically appear as anechoic or hypoechoic cystic masses in the right iliac fossa, adjacent to or behind the cecum. Internal echogenic layering—sometimes described as an "onion-skin" appearance—can be present, particularly when mucin is thick or mixed with debris. Mural nodularity, if detected, raises concern for neoplastic transformation.
- **CT scans** usually reveal a well-encapsulated cystic mass contiguous with the cecum, measuring low attenuation (10–30 HU). Wall calcifications, seen in up to 50% of cases, are often curvilinear and peripheral—considered highly suggestive of mucocele. Suspicious findings for malignancy include mural nodules, irregular wall thickening, or strong enhancement after contrast administration. CT also identifies complications such as rupture, abscess formation, or pseudomyxoma peritonei, visualized as mucinous ascites with scalloped liver margins.
- **MRI** provides further characterization, especially when iodinated contrast is contraindicated. Mucoceles usually show low signal on T1 and high signal on T2-weighted sequences. High-protein mucin may appear brighter on T1. Gadolinium enhancement of the wall, if nodular or irregular, is another red flag for malignancy.
- **PET-CT** is not standard for initial diagnosis but may be helpful in detecting malignant transformation or recurrence. Focal FDG uptake along the wall or in peritoneal implants can indicate cystadenocarcinoma.

Radiological red flags for malignancy include appendix diameter >15 mm, mural nodules, and heterogeneous enhancement patterns.

Differential Diagnosis

Differentiating benign (mucinous cystadenoma) from malignant (mucinous cystadenocarcinoma) appendiceal mucoceles remains difficult on imaging. The only suggestive radiological feature of malignant degeneration is nodular wall thickening with contrast enhancement. In our patient, the mass appeared encapsulated with fluid content and fine peripheral calcifications, without any significant wall thickening or contrast enhancement.

The differential diagnosis of appendiceal mucocele includes ovarian cystadenomas, lymphoceles, mesenteric cystadenomas, hematomas, and abscesses. Careful correlation with clinical findings and imaging features is essential for accurate diagnosis and appropriate surgical planning.

Pseudomyxoma Peritonei

This syndrome results from rupture of a mucinous appendiceal lesion, leading to mucin accumulation in the peritoneal cavity. It may be:

- Acellular: favorable prognosis.
- Cellular, with free-floating mucinous tumor cells: worse prognosis, associated with recurrent peritoneal disease (gelatinous disease).

Surgical Management

Surgery is always indicated:

- Laparoscopy for non-ruptured benign cases.
- Laparotomy for suspected or confirmed malignancy.

Benign mucoceles are treated with appendectomy. In malignant or suspicious cases, a right hemicolectomy may be needed. If peritoneal spread is found, intraperitoneal chemotherapy can be considered.

Prognosis

- Benign forms (retention cysts, mucosal hyperplasia, cystadenomas): excellent prognosis with complete resection, with 5-year survival near 100%.
- Malignant forms: prognosis depends on tumor spread; survival ranges from 30% to 80%.

CONCLUSION

Preoperative radiological diagnosis of mucocele is essential, guiding surgical approach and preventing rupture and peritoneal dissemination. In any case of an appendiceal mass with parietal calcifications, mucocele should be considered. Histopathological analysis of all appendectomy specimens is crucial for proper management.

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