Endometriosis of the Appendix in the Presence of Low-Grade Appendiceal Mucinous Neoplasm

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Introduction

Endometriosis, a disorder predominantly attributed to the uterus and characterized by abnormal and painful growth of endometrial-like tissue outside the uterus, can present in the appendix. Obstruction due to appendiceal endometriosis tissue can, in rare cases, cause the appendix to swell and fill with mucus, known as an obstructive appendiceal mucocele. The simultaneous occurrence of **appendiceal endometriosis** and a **low-grade appendiceal mucocele neoplasms (LAMN)** has not previously been described.

Case Report

We present the case of a 63 year old woman evaluated in the ER with right flank abdominal pain for the past 24 hours admitted for possible appendicitis. CT scan revealed a distended appendix containing fecalith with possible appendicular neoplasm. She underwent a colonoscopy which revealed diverticular disease and benign polyps, and subsequently laparoscopic appendectomy. Pathology revealed a 4.5 cm low-grade appendiceal mucinous neoplasm (LAMN) with luminal calcification, as well as mural and subserosal appendiceal endometriosis. Margin was negative for neoplasia. She recovered without complications.

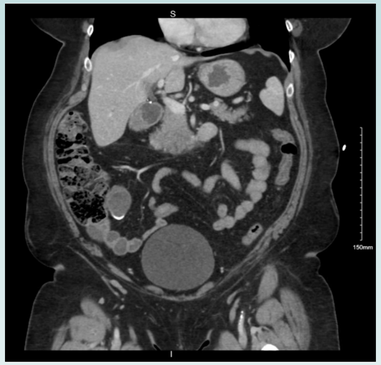


Figure 1. Abdominal CT scan view of distended appendix with mural calcification

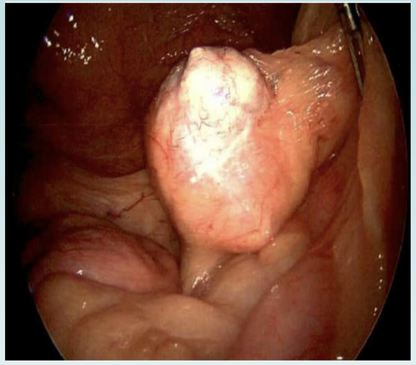


Figure 2. Laparoscopic view of the distended appendix with associated mass.

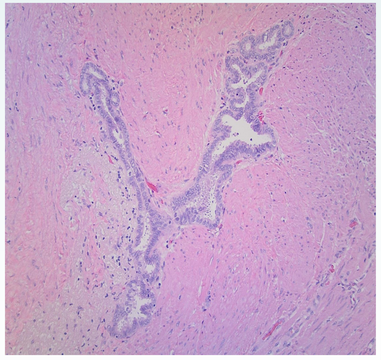


Figure 3. The expanded portion of the appendix is predominantly lined by atypical columnar epithelium with low-grade nuclear atypia and occasional complex tufting. No invasive carcinoma is identified.
(H&E, 100x original magnification).

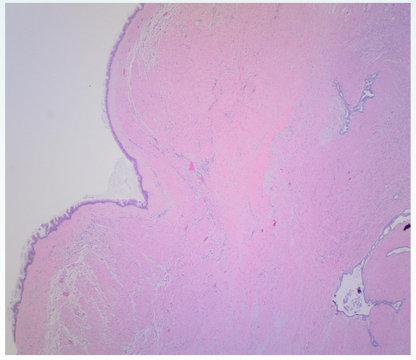


Figure 4. Next to the low-grade mucinous neoplasm, there are numerous foci of endometriosis within the muscularis propria and subserosal compartment of the appendiceal wall.
(H&E, 20x original magnification).

Results

Endometriosis affects 4%-50% of reproductive age women, commonly resulting in infertility and pain. 2.8% of endometriosis cases occur at the appendix. Endometriosis of the appendix can lead to an appendiceal mucocele. Mucoceles can be further categorized as neoplastic or non-neoplastic, with neoplastic mucoceles including **low-grade appendiceal mucocele neoplasms (LAMN)**. LAMN is a rare finding and accounts for only 1% of gastrointestinal neoplasms and is present in merely 0.3% of appendectomy specimens. We reported a rare discovery of LAMN in the presence of appendiceal endometriosis.

Conclusion

Endometriosis of the appendix is a very rare finding, with the definitive diagnosis usually coming from pathologic evaluation. **Appendiceal endometriosis in association with LAMN has not previously been described**. Further research of the relationship between appendiceal neoplasms and appendiceal endometriosis is required to improve operative treatment.