

Low Grade Mucinous Appendicular Neoplasm: Incidental Finding Post Appendicectomy

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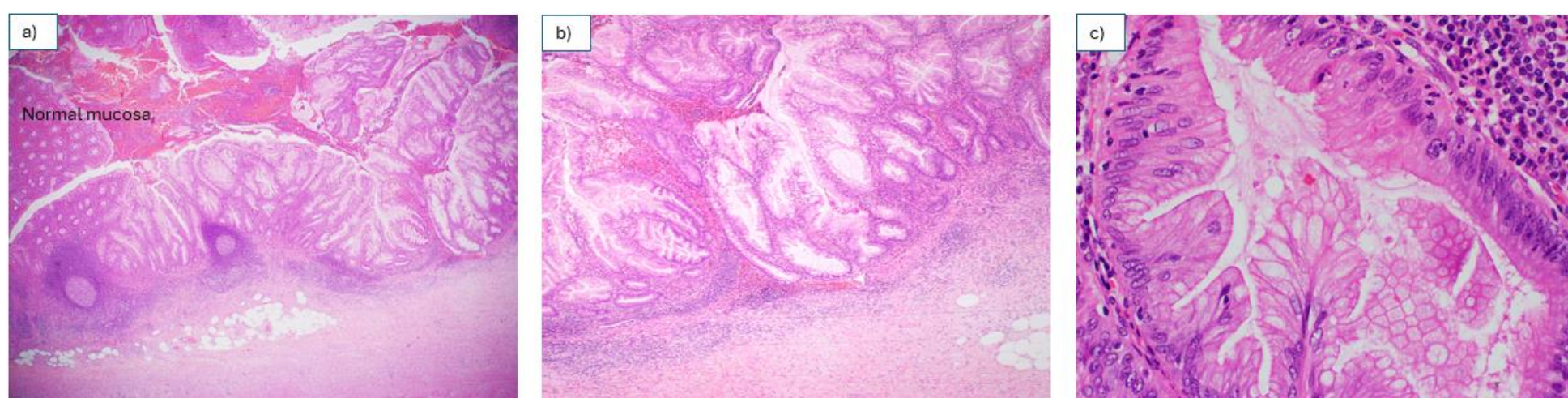
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Introduction:

Low-grade appendiceal mucinous neoplasms (LAMNs) are rare, non-invasive epithelial tumors of the appendix that produces abundant mucin. About 0.7-1.7% of appendicectomy specimens removed for suspicion of appendicitis, an epithelial appendiceal tumor is detected unexpectedly at histological assessment.⁽¹⁾ LAMNs in particular has an incidence of ~0.2-0.6 cases per 100 000 individuals per year.⁽⁵⁾ Although by definition they do not invade, their expansile growth may push and thin the appendiceal wall until the appendix ruptures, and the resultant spillage of mucin may lead to the potentially fatal pseudomyxoma peritonei (PMP) with 20-30% risk of progression.⁽⁴⁾ PMP is a clinical syndrome in which there is diffuse mucinous peritoneal collection which may lead to intestinal obstruction and eventually death if left untreated.

Case report:

We reported a case of a 21-year-old male who presented with short typical history of right iliac fossa pain and underwent laparoscopic appendicectomy for suspected acute appendicitis. Intraoperatively noted grossly inflamed appendix which was adhered to abdominal wall. He was well post-operatively and was discharged home on subsequent day. Histopathological diagnosis of appendix turned out to be LAMN and its characteristics as described below. Colonoscopy showed multiple small sessile polyps over terminal ileum which turned out to be lymphoid hyperplasia and tumor marker was normal. Computed Tomography (CT) Abdomen Pelvis showed ileocecal junction and distal ileum wall thickening. He was given options, either for surgical intervention due to unknown risk of spread or need for surveillance and opted for tumor marker and colonoscopy surveillance.



Histopathological examination of appendix:

- Focal villous proliferation of mucinous epithelial cells with pushing margin (right). Normal appendiceal mucosa is seen on the left side (H&E, x 2 HPF)
- There is lack of underlying lymphoid tissue and obliteration of muscularis mucosae layer beneath the epithelial proliferation (H&E, x 10 HPF)
- Mucinous cells with elongated nuclei and abundant apical mucin (H&E, x 40 HPF)

Discussion:

LAMNs are staged according to the ninth edition of the American Joint Committee on Cancer Staging Manual. The staging is depending on the depth of invasion through the appendiceal wall. However, assessing depth of invasion in LAMN is challenging, primarily owing to their lack of destructive invasion and the propensity for mucin extravasation. Studies have shown rare recurrences of in situ neoplasms (pTis) post appendicectomy supporting the uncertain malignant potential in LAMNs.⁽²⁾

Recommended guidelines suggest that appendicectomy with removal of entire mesoappendix is adequate for LAMNs that are confined to appendix with negative resection margin, provided that the appendix was handled with care during the operation. In this case, appendicectomy was performed for presumed suppurative appendicitis with adhesions to surrounding structures. Hence, mesoappendix may not be completely removed and there is risk of spread of mucin during the handling and extraction of appendix during a laparoscopic procedure as energy device used during ligation of appendix may not sealed the lumen of appendix completely. This may convert a localized to a disseminated disease.⁽⁴⁾

The extent of resection is predominantly based on involvement of base of the appendix and simple appendicectomy or partial caecectomy is adequate when a suspicious looking appendix is encountered intraoperatively. Retrieval bag is also helpful during extraction of specimen through laparoscopic port to avoid spilling of mucin.^(1,3,4)

While initial CT scan is recommended to evaluate the primary tumor and assess for metastatic disease, there is no specific guidelines for post-operative surveillance in LAMNs and should be catered individually as there is no proven benefit in frequent imaging for extended intervals.⁽⁴⁾ There is a known risk of 3-5% synchronous colorectal tumor and as appendix is embryologically derived from the colon there have been reports of colorectal neoplasm in patients with appendiceal neoplasm which necessitates the need for surveillance colonoscopy in all patients.⁽¹⁾

Conclusion:

Early appropriate surgical management of appendiceal tumors is critical to prevent tumor perforation and spread of mucin which may lead to life-threatening pseudomyxoma peritonei even in low-grade appendiceal neoplasm.

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