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Case Report on Rare Appendiceal Mucinous Neoplasms – A Histopathologist's Role in Predicting the Consequence

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Abstract: *Appendiceal mucinous neoplasms are rare primary tumors of appendix which pose a major diagnostic challenge to histopathologists as categorizing these tumors is critical for further surgical management and predicting the disease-free survival rates. In the current case report, we put forth 2 cases of low grade appendiceal mucinous neoplasms (LAMNs), in middle aged women, with similar complaints of abdominal pain for a few weeks before seeking medical attention. On imaging studies, the lesion was suggestive of an appendiceal mucocele in one of the cases and as a metabolically inactive space occupying lesion in the right iliac fossa in the other. On histopathological examination, one of the tumors was entirely confined to the appendix and hence was categorized as stage Tis while the other was found to be producing copious amounts of mucin, dissecting through the appendiceal wall and involving the visceral peritoneum, manifesting in pseudomyxoma peritonei localized to the right iliac fossa and hence categorized as stage T4a.*

Keywords: *Histopathological examination, Low grade appendiceal mucinous neoplasm, Mucocele, Pseudomyxoma peritonei, Women.*

Key message: *Low grade appendiceal mucinous neoplasms – Histopathological features and their prognostic significance.*

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I. Introduction:

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy accounting for 1% of gastrointestinal neoplasms and is found in less than 0.3% of appendectomy specimens. [1-2] Since mucinous neoplasms are a rare cause of acute abdomen, they are often misdiagnosed as acute appendicitis or as retroperitoneal tumours initially. [3-4] An accurate preoperative diagnosis with the aid of imaging studies is essential in preventing accidental rupture of the mucinous neoplasm within the peritoneal cavity, which can lead to iatrogenic implantation of mucin and result in formation of pseudomyxoma peritonei (PMP).[5] These tumours also necessitate meticulous pathological assessment following surgery as their categorization and grading determine the overall outcome and disease-free survival rates.

II. Case Reports:

Case 1: A 57-year -old female patient presented with history of abdominal pain for 1 month on and off which was acute in onset and gradually increasing in intensity. There was no history of fever / vomiting or altered bowel and bladder habits. On sonography, a dilated non-inflammed appendix, with a possibility of appendicular mucocele was considered. The serum carcinoembryonic antigen (CEA) level was slightly elevated. With the above findings, an open appendicectomy with limited resection of terminal ileum and caecum was done and the specimen was subjected to histopathological examination. On gross examination, the appendix was enlarged, with a dilated lumen filled with mucinous material. The appendiceal wall was intact (Figure 1). Rest of the intestine was unremarkable. On microscopic examination the lesion was found to be a low grade appendiceal mucinous neoplasm – grade I based on histologic criteria, confined to the appendix (Tis) (Figure 2), classified according to WHO digestive system tumors (2019).[6]

Case 2: A 50-year-old female patient presented with abdominal pain on and off in the right lower quadrant for 3 months. On sonography, a large multiloculated cystic lesion was noted in the pelvis in relation to the right ovary. CECT showed a mass lesion in the right iliac fossa ventral to cecum and away from the ileocecal junction, suggestive of an appendiceal space occupying lesion. FDG-PET scan showed metabolically inactive non enhancing soft tissue density mass in the right iliac fossa. CA-125 and CEA were within normal limits. Laparotomy was performed which showed extensive collection of gelatinous material in the right iliac fossa, with bilateral normal ovaries. The patient subsequently underwent right hemicolectomy and the specimen was

sent for HPE along with about 500 ml of gelatinous material in the right iliac fossa. Gross examination showed a mutiloculated cystic mass replacing the entire appendix (Figure 3). Histopathologically, it was a low grade appendiceal mucinous neoplasm with mucin dissecting through the appendiceal wall and involving the visceral peritoneum, grade I based on histologic criteria (Figure 4) and stage T4a according to WHO digestive system tumors (2019).[6]

III. Discussion:

LAMNs are tumours predominantly confined to the appendiceal wall or rarely may involve the visceral peritoneum. These tumours characteristically cause cystic dilatation of the appendix due to the accumulation of copious amounts of gelatinous material within the lumen. They may dissect through the appendiceal wall and disseminate throughout the peritoneal cavity in the form of gelatinous deposits, termed pseudomyxoma peritonei (PMP).[7]

The first classification of mucinous neoplasms was made by Misdraji et al. as low-grade appendiceal mucinous neoplasm (LAMN) and mucinous adenocarcinoma based on the complexity of architecture and degree of cytological atypia in 2003,[8] which was adopted later by the World Health Organization classification in 2010.

In the current report, we evaluated 2 cases of low grade appendiceal mucinous neoplasms, wherein the first case was radiologically suggestive of mucocele of the appendix. On post-surgical pathologic examination, the appendix was found to be enlarged in size, with an attenuated wall and lumen filled with gelatinous material. The mucinous lesion measuring 4x3x2.5cm was confined to the appendiceal lumen and the serosal aspect of appendix was unremarkable. Microscopic examination showed a well circumscribed cystic neoplasm lined by mucin secreting columnar epithelium showing focal nuclear stratification and tufting. The epithelium showed low grade dysplasia. A pool of acellular mucin was seen within the cavity. The appendiceal wall was fibrotic with no evidence of lymphoid tissue. With these features, the lesion was classified as LAMN, grade I and stage Tis.

The second case was also observed in middle aged post-menopausal woman, with similar clinical history and on imaging studies, was found to be a metabolically inactive space occupying lesion in the right iliac fossa. On laparotomy, the lesion was found to be enmeshed in a pool of gelatinous material, confined to the right iliac fossa. The patient subsequently underwent right hemicolectomy with en-masse removal of appendiceal lesion. Around 500ml of gelatinous material was cleared from the right iliac fossa. On pathological examination, the appendix was grossly enlarged, measuring 7x4x4cm, lumen filled with gelatinous material, involving the serosal aspect of the appendix. On microscopy, the cystic lesion was lined by a single layer of mucin secreting columnar cells with basally located nuclei demonstrating low grade dysplasia and apically located intracellular mucin. There was focal tufting and stratification of the nuclei. The acellular mucin was dissecting through the appendiceal wall and involving the serosa. Also seen was a single glandular structure on the serosal aspect, lined by columnar epithelium with bland nuclear features. With these features the tumour was categorized as LAMN, grade I, stage T4a.

The histological grading and TNM staging of these tumours are essential in planning further management and follow-up of these patients. LAMNs in stage Tis have excellent prognosis following surgical excision while those with peritoneal dissemination require frequent follow up at 6-month intervals until 5years [9] due to the risk of pseudomyxoma peritonei or recurrence of malignancy. The addition of hyperthermic intraperitoneal chemotherapy (HIPEC) along with complete cytoreduction could increase the survival rates in patients with disseminated disease.[6]

IV. Conclusion:

Low grade appendiceal mucinous neoplasms deserve clinical and pathological attention not only due to their rarity but also for their broad spectrum of presentation and their varied clinical implications. However, with meticulous examination, state of the art management and subsequent clinical follow up could offer promising results in the current scenario.

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Images with Legends:

Figure 1: Right hemicolectomy resection of terminal ileum showing an enlarged appendix, lumen filled with gelatinous material. Serosa appears unremarkable.

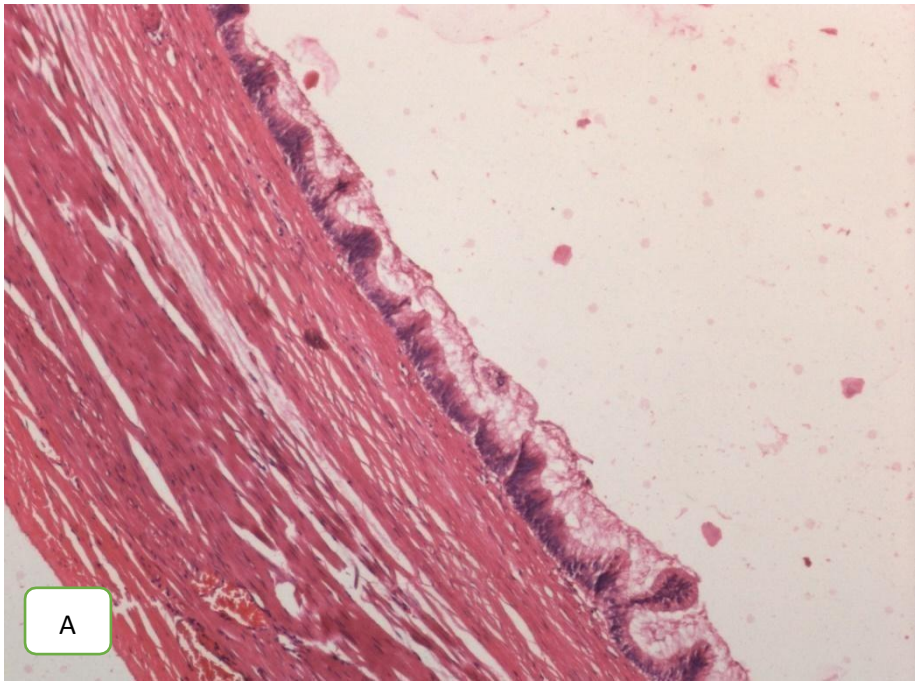
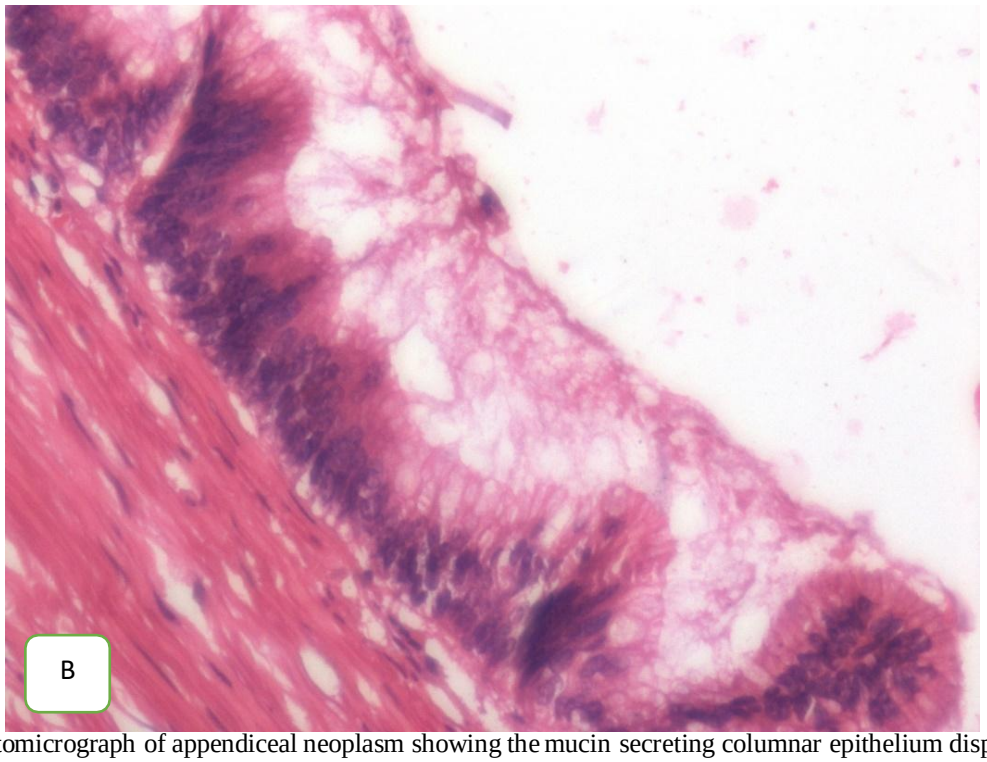


Figure 2: (A) Photomicrograph of appendiceal neoplasm showing a fibrotic wall lined by mucin secreting columnar epithelium showing focal tufting and nuclear stratification. (H and E stain, x100 magnification)



(B) Photomicrograph of appendiceal neoplasm showing the mucin secreting columnar epithelium displaying nuclear stratification and low-grade dysplasia. (H and E stain, x400 magnification)

Figure 3: Right hemicolectomy specimen with limited resection of terminal ileum showing an irregularly enlarged appendix, with multiple cystic locules filled with gelatinous material located on the serosal surface of appendix.

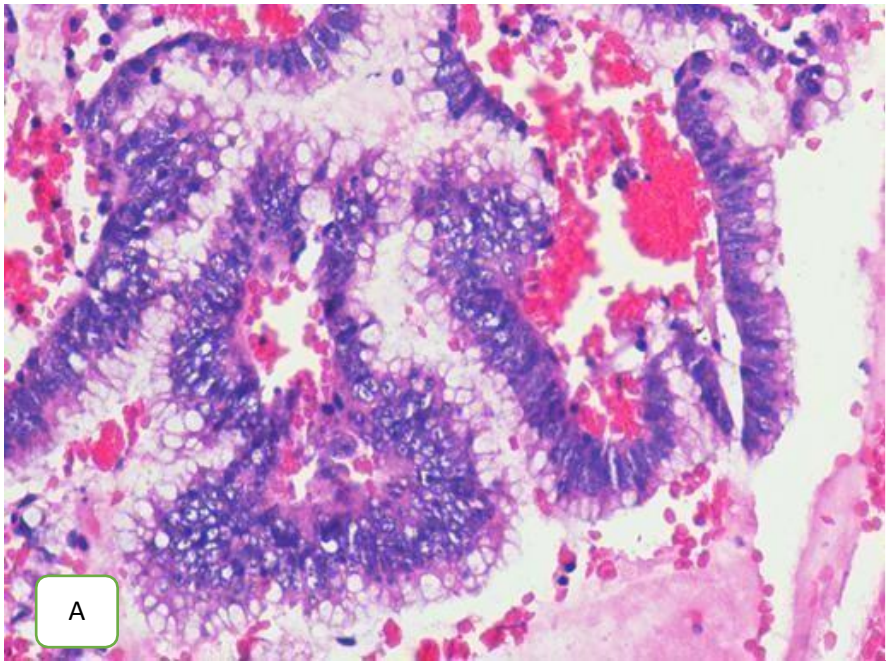
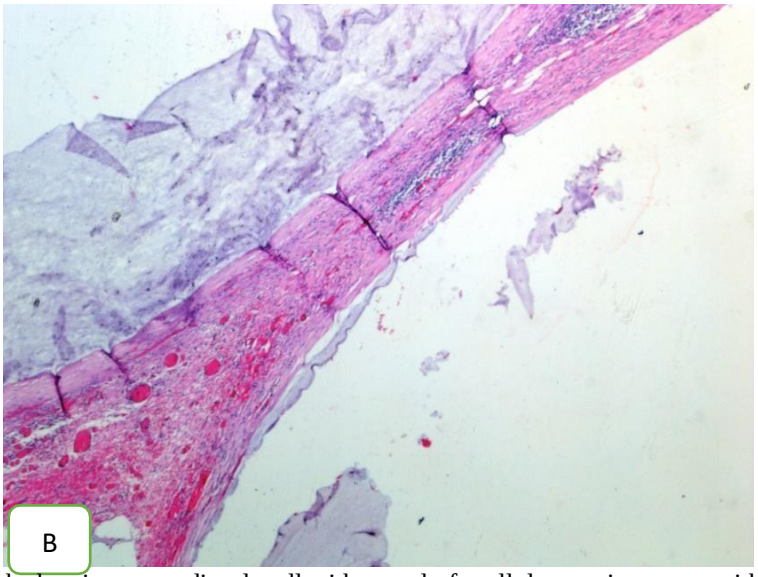
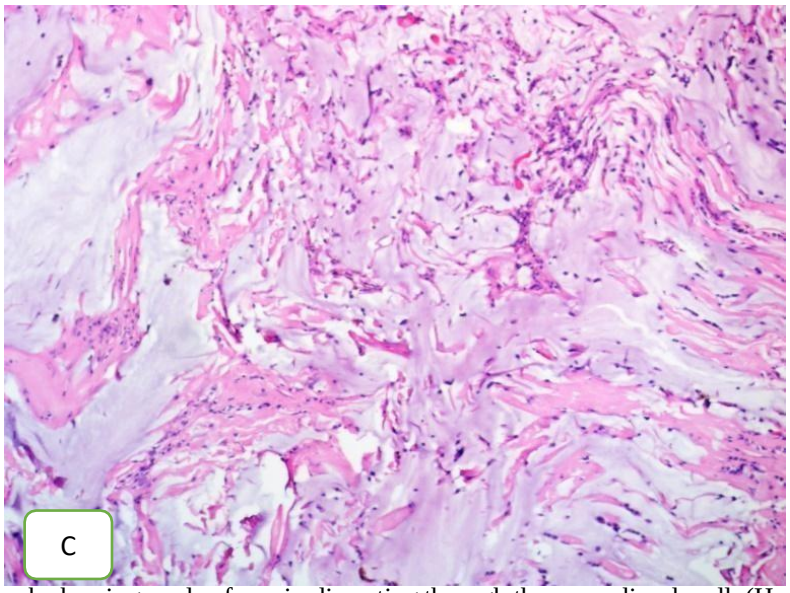


Figure 4: (A) Photomicrograph of low grade appendiceal neoplasm showing mucin secreting columnar epithelium with low grade dysplasia, resting on a fibrotic appendiceal wall. (H and E stain, x400 magnification)



(B) Photomicrograph showing appendiceal wall with a pool of acellular mucin seen on either side of the wall, indicating the extension of the mucin beyond the serosa of the appendix. (H and E stain, x100 magnification)



(C) Photomicrograph showing pools of mucin dissecting through the appendiceal wall. (H and E stain, x100 magnification).

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