

Low-Grade Appendicular Mucinous Neoplasm Presenting with Obstructed Inguinal Hernia: A Rare Association

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ABSTRACT

Low grade appendicular mucinous neoplasm is a rare entity. It is grouped under appendicular adenocarcinoma. Though it presents commonly with pain abdomen, asymptomatic patients are not uncommon. It is a low grade neoplasm, but capable of producing progressive and fatal disease in the form of pseudomyxoma peritonei. Management in case of localised appendicular disease includes simple appendectomy. We came across a patient presenting with obstructed inguinal hernia. While performing emergency exploration, a dilated appendix was uncovered. Appendectomy was done as a result and the histopathological examination of the appendix revealed that it was a low grade appendicular mucinous neoplasm.

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Introduction

Low -grade Appendiceal Mucinous Neoplasm (LAMN) is a rare entity grouped under appendiceal adenocarcinoma which itself occurs in 0.1- 0.2 % of appendectomies. The median age of presentation of appendiceal adenocarcinoma is in the sixth or seventh decade of life and LAMN usually presents at more than 50 years of age. Patients commonly present with abdominal pain, though asymptomatic cases with incidental discovery of the lesion amount to one fourth of cases.^{1,2} We present here a case which presented as an obstructed inguinal hernia associated with a mucocele of appendix diagnosed as Low grade Appendiceal Mucinous neoplasm post operatively.

Case Report

Clinical findings: A 45-year-old man presented with a two day history of acute onset pain and tenderness in right inguino-scrotal swelling and got admitted through surgical emergency. The swelling was present for last two years without any medical advice. Examinations revealed an afebrile patient with a huge tender swelling in the inguino-scrotal region. Overlying skin was healthy looking. Taxis was tried gently but failed . Peristaltic sound was present over the swelling. It was provisionally diagnosed by the surgeons as obstructed irreducible inguinal hernia with small bowel content. As the patient presented as an emergency case of obstructed hernia, and time was crucial radiological imaging could not be availed.

Operative notes: After taking ileostomy and orchidectomy consent and performing the baseline investigations and serology, exploration was done with an inguino scrotal incision. It revealed a hugely distended viable caecum and appendix with mucoid material in surrounding



Fig. 1: Photograph showing intraoperative specimen of the appendix, separate from the hernia mass and showing distension.

area. A densely adherent hernial sac was identified separately(Fig.1). Appendectomy was done and specimen was sent for histopathology. Peritoneal cavity did not show any fluid collection. Content of the hernial sac was noticed to be viable small bowel. Caecum and small bowel were reduced after confirming their viability and mesh hernioplasty was done. Mesenteric lymph nodes were unremarkable. Ileostomy, colostomy and orchidectomy could be avoided. Post operative recovery was uneventful except for slight induration and pain over the scrotum during follow up.

Pathologic features: On gross histopathological examination, appendix was found to be hugely distended, measuring 5 cm X 2.5 cm; which on cutting open yielded mucoid material. No lymph nodes were sent with the specimen. On microscopy, the appendiceal mucosa showed serrated and undulating morphology and it rested on the fibrous tissue. (Fig.2a, Fig.2b) Some areas showed the denudation of mucosa. Cytologically, the mucosal cells had low grade dysplasia.(Fig.2c) There was invasion of the mucoid material into the muscular layer.(Fig.3). The diagnosis was given as low grade appendiceal mucinous neoplasm.

Discussion

LAMN is a rare tumor of the appendix which was termed variously earlier as mucinous cystadenoma, or mucocele when it is cystically dilated. LAMN was coined in 4th edition of WHO classification to omit “ adenoma” as this lesion has potential of producing widespread disease.² It is more common in females, usually observed in patients over 50 years age.³The most common presenting symptom associated with LAMN is abdominal pain. However, substantial numbers of patients are asymptomatic.

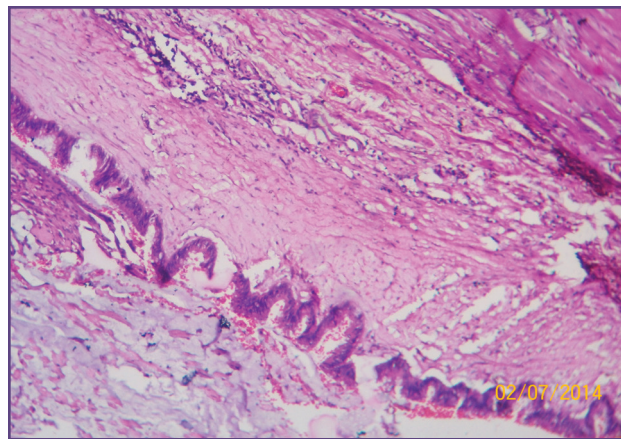


Fig. 2a: photomicrograph showing morphology of the appendiceal mucosa showing serrations and undulations (H&E, 100x)

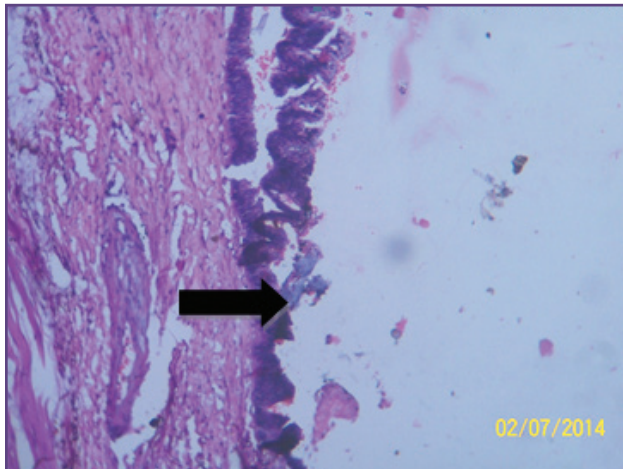


Fig. 2b: photomicrograph showing mucosa directly resting on fibrous tissue (H&E, 100x)

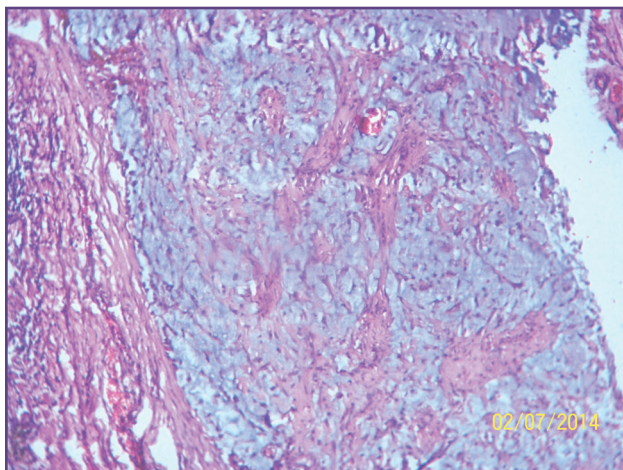


Fig. 3: photomicrograph showing invasion of muscle layer by mucin (H&E, 400X)

Presentation as obstructed inguinal hernia along with separate LAMN is a very rare finding, and to our best knowledge, yet to be reported. However, presentations with bloody stool, intussusception have been reported.⁴ Imaging studies like ultrasonography, computed tomography are required for differential diagnoses as symptoms are non specific. The imaging studies also help to uncover incidental cases. However, the diagnosis is usually made intraoperatively or postoperatively on histopathological examination.^{3,5,6}

LAMN generally grow slowly, and tend to produce the clinical picture of low-grade pseudomyxoma peritonei in which spread beyond the peritoneum or nodal metastasis is unusual. Histologically, LAMN may have villous, serrated or undulating morphology, but unlike adenomas, they rest on fibrous tissue rather than lamina propria. Nuclei

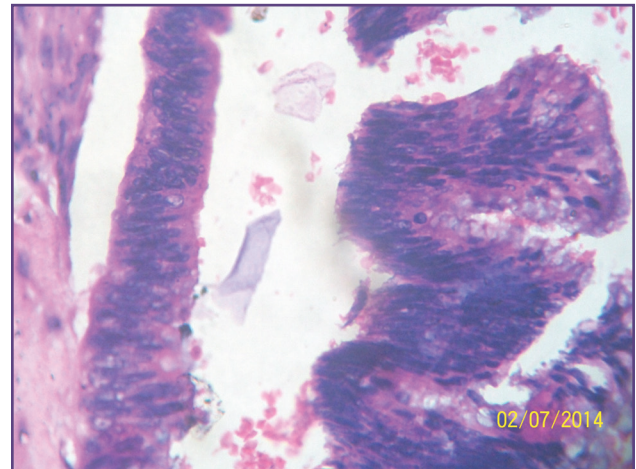


Fig. 2c: photomicrograph showing appendiceal mucosa with low grade dysplasia, (H&E, 400x)

are small and regular with low grade dysplasia. Often there is atrophy and fibrosis of underlying submucosa and muscularis propria, but desmoplasia is absent. They tend to produce atrophy of the underlying lymphoid tissue.¹

The morbidity/mortality associated with LAMN results from rupture and intraperitoneal spread of mucin-producing epithelium, which may cause pseudomyxoma peritonei, which warrants careful intraoperative tissue handling.⁷ LAMN is found to be associated with colonic and ovarian malignancy.⁷⁻⁹

Since there are no evidences in regard to lymphatic or hematogenous spreading of LAMN, if the mass confines in the appendix body without local invasion or caecal involvement, simple appendectomy and mesoappendix excision is considered sufficient treatment. For mass involving the caecum or adjacent organs, right hemicolectomy is often required.⁷⁻⁹

Conclusion

Low grade appendiceal mucinous neoplasm, though a low grade neoplastic entity, can result in fatalities owing to development of pseudomyxoma peritonei. When the lesion is confined to the appendix only, simple appendectomy suffices. So, the aim of the diagnosis should focus at proper management and avoid confusions regarding nomenclature.

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Declaration

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Competing interests

none declared

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