Case Report

DOI: 10.21276/APALM.3225

High Grade Appendiceal Mucinous Neoplasm Presenting as Intussusception In an Elderly Female - A Case Report

Manjari SKVSK*, Mark Ruth Prasanna, T Satya Prakash Venkatachalam

Pathology, GSL Medical College and General Hospital

Abstract

Introduction: Appendiceal mucinous neoplasms (AMNs) are rare and account for 1% of gastrointestinal neoplasms and the diagnosis is mostly incidental. In spite of advances in the understanding of AMNs and their association with peritoneal disease, AMNs are still confusing entities in respect to their diagnosis and treatment alike.

Case Report: We report a case of a 51year old female who presented with abdominal pain and vomiting and was diagnosed as ileo-colic intussusception on ultrasonography and CECT abdomen. Histopathological examination of right hemicolectomy specimen revealed a high grade appendiceal mucinous neoplasm (HAMN). The peri-appendiceal mucin deposition forming a mass in the caecal wall at the base of appendix was the lead point for intussusception in this case. No features of pseudomyxoma peritonei was seen in this case. This patient is well after 18 months of follow up period.

Conclusion: Appendiceal mucinous neoplasms are rare and diagnosis is often difficult as presentation is mostly non-specific. Hence, awareness and further classification of these tumours is essential for diagnosis, determining the risk of recurrence and malignancy and also prognostication.

Keywords: *Appendiceal mucinous tumors, intussusception, high grade, AMN, HAMN*

Introduction

Appendiceal mucinous neoplasms are rare and account for 1% of gastrointestinal neoplasms.[1] The diagnosis of these neoplasms is mostly incidental following a surgical intervention, radiological study or in pathological study of an appendectomy specimen.[2] Diagnosis at times is quite challenging as they can present with varied manifestations like abdominal pain, weight loss, nausea and vomiting, palpable mass, acute appendicitis or intussusception.[3] Intussusception is uncommon in adults and usually occurs secondary to a definable lesion. In AMNs appendix may be the lead point in case of intussusception.[4] AMNs are classified as either low grade appendiceal mucinous neoplasm (LAMN) or high grade appendiceal mucinous neoplasm (HAMN), the latter with areas of high grade cytological features. HAMN is a recently introduced term describing a rare epithelial neoplasm with pushing type invasion and high grade cytological atypia. They lack infiltrative pattern of growth and destructive invasion.[5] The most feared complication in these tumors is pseudomyxoma peritonei. The extra-appendiceal mucin may be cellular or acellular. It is unlikely for patients with AMNs with acellular extra-appendiceal mucin to develop recurrence unlike those with cellular mucin.[6] Treatment may include appendectomy and hemicolectomy depending on factors like histological type, tumor size and lymph node/organ involvement.[7]

Here, we report a rare case of high grade appendiceal mucinous neoplasm in an adult female patient who presented with intussusception. The lead point was the mass in the cecum near base of appendix which is organized peri-appendiceal extra cellular mucin with calcification and granulation tissue.

Case Report

A 51year old female presented with abdominal pain and watery stools of one month duration. There was history of loss of weight and loss of appetite. On examination there was tenderness in the right lumbar region. Ultrasonography showed thickened bowel loops in right para-umbilical region with bowel within bowel configuration suggesting intussusception. CECT abdomen also showed intussuscepted terminal ileal loop along with mesenteric vessels and fat into the ascending colon and hepatic flexure giving a target sign appearance consistent with the diagnosis of ileo-colic intussusception, however, no lead point could

be visualized. (Fig 1a) Lower gastrointestinal colonoscopy also revealed a polypoidal growth telescoping into the ascending colon suggesting ileo-colic intussusception. A hard growth was found near the appendix during exploratory laparotomy. The intussuscepted bowel loop was reduced followed by a right hemicolectomy.

Right hemicolectomy specimen was received for histopathological examination. The specimen measured 41.0cm in length with appendix measuring 4.2 cm in length. On cutting open, a firm nodular mass of size 2.5x2.0cm was seen in the cecum adjacent to the base of appendix. The overlying mucosa was ulcerated. The mass had grey white to grey brown cut surface with focal mucoid areas. Cut section of the appendix showed mild thickening of wall. (Fig 1b)

Figure 1: a: An axial CECT abdomen showing intussuscepted terminal ileal loop. b: Gross specimen showing appendix (orange arrow) and nodular mass at the base of appendix (black arrow)

Histopathological examination showed normal mucosa replaced by flat and short villous arrangement of columnar epithelial cells. These cells showed pseudo stratification, moderate nuclear pleomorphism with frequent mitoses, moderate amount of cytoplasm with areas showing intracytoplasmic mucin. There is atrophy of underlying muscularis mucosa, submucosa and lymphoid tissue. A small area of extra cellular mucin seen in the wall. (Fig 2) Nodular mass in the cecal wall showed large areas of extra cellular mucin with foci of calcification surrounded by proliferating capillaries and fibroblasts with lymphoplasmacytic infiltrate. (Fig 3) No epithelial component was seen in the mucin pools. A diagnosis of high grade appendicular mucinous neoplasm (HAMN) with low grade areas was made. No features of pseudomyxoma peritonei seen in this case. This patient is well after 18 months of follow up period.

Discussion

Appendiceal mucinous neoplasms are a poorly understood heterogenous pathology with unusual presentation and unpredictable biologic behavior.[7]

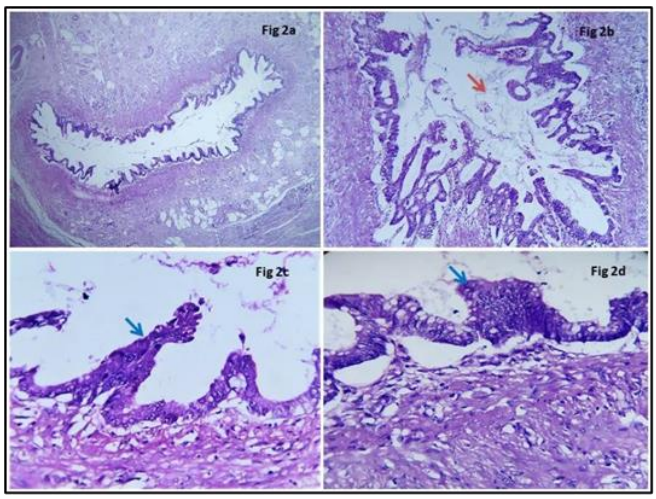

Figure 2: a: Flat and short villous arrangement of columnar epithelial cells, atrophy of underlying muscularis mucosa, submucosa and lymphoid tissue (H&E 4X). b: Intraluminal mucin (orange arrow) (H&E 10X). 2c, 2d -Pseudo stratification, nuclear pleomorphism and increased mitoses(blue arrow) (H&E 40X) .

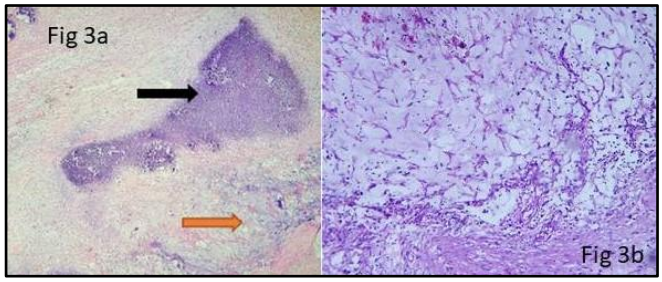

Figure 3: a and b(H&E 10X,40X): Nodular mass showing areas of extracellular mucin (yellow arrow) with foci of calcification (black arrow).

Intussusception is a quite unusual presentation in these cases with appendix being the lead point. In this case mucin surrounded by tissue reaction resulted in the formation of a mass in the cecal wall at the base of appendix leading to intussusception. The terms low grade appendiceal mucinous neoplasm (LAMN) and high grade appendiceal mucinous neoplasm (HAMN) refer to non-invasive lesions with varying degrees of cytological atypia.[5] HAMN diagnosis is uncommon and clinically poorly understood due to its rarity.

Though grading and staging of mucinous neoplasms of appendix is challenging and fraught with terminology problems, it has critical prognostic and therapeutic implications. Complications depend on the tumor size and histological type. [8] These neoplasms if confined to the

mucosa have a benign course but those with disseminated peritoneal deposits often follow an indolent but malignant course. Peri-appendiceal or extra-appendiceal mucin deposits may be cellular or acellular. [6]

Conclusion

Ileocecal intussusception secondary to extra-appendiceal mucin deposition in a case of high grade appendiceal mucinous neoplasm is extremely rare. Awareness of such presentation and thorough histopathological evaluation is needed for diagnosis and further treatment of such patient. A close follow up was recommended for our patient.

Acknowledgements: Nil

Funding: None

Competing Interests: None declared

Reference

1. Ramaswamy V. Pathology of Mucinous Appendiceal Tumors and Pseudomyxoma Peritonei. Indian J Surg Oncol. 2016;7:258-267.
2. Nutu OA, Marcacuzco Quinto AA, Manrique Municio A, et al. Mucinous appendiceal neoplasms: Incidence, diagnosis and surgical treatment. Cir Esp. 2017;95(6):321-327.
3. S.H. Tirumani. M. Fraser-Hill, R. Auer et al., "Mucinous neoplasms of the appendix : a current comprehensive clinicopathologic and imaging review," Cancer Imaging.2013;13(1):14-25.
4. Siddiqui G, Lipka S, Freedman L, Takeshige U, Rizvon K, Mustacchia P. Mucinous adenocarcinoma of the appendix presenting as intussusception in 27 year old Female. J Gastrointestinal Oncol. 2012;3(4):369-372.
5. Gonzalez RS, Carr NJ, Liao H, Pai RK, Vulaj D A, et al. High grade appendiceal mucinous neoplasm. Clinicopathologic findings in 35 cases. Arch Pathol Lab Med.2022;4(30)1-8.
6. Rhonda Y, Jinru S, David S, Hejin H, Robert OD. Prognostic significance of localised extra-appendiceal mucin deposition in appendiceal mucinous neoplasms. Am J of Surgical Path. 2019;33(2):248-255.
7. Chirca A, Negreanu L,Iliesiu A, Costea R. Mucinous appendiceal neoplasm: A case report. World J Clin Cases.2021;9(7):1728-33.
8. Umetsu SE, Shafizadeh N, Kakar S. Grading and staging mucinous neoplasms of the appendix: a case series and review of the literature. Hum Pathol.2017;69:81-89.

Corresponding Author:

Dr Manjari SKVSK

Plot No 48, Srinivasa Nagar, Kommadi Jn, Madhurawada,

Visakhapatnam-530048

drmanjariskvsk@gmail.com

Date of Submission	**14 October 2022**
Date of Final Revision	**1 December 2022**
Date of Acceptance	**10 December 2022**
Date of Publication	**31 December 2022**