



ORIGINAL RESEARCH PAPER

General Surgery

LOW-GRADE APPENDICEAL MUCINOUS NEOPLASM (LAMN) PRESENTING AS ILEO--COLIC INTUSSUSCEPTION.

KEY WORDS: Low-grade appendiceal mucinous neoplasm (LAMN), ileo-colic-intussusception, laparoscopic quarter colectomy, anastomosis.

Dr Rafique Umer Harvitkar

MBBS, MS (General Surgery), MRCS UK Clinical Associate (Dept of Surgery) Dr. L. H. Hiranandani Hospital Powai hillside avenue, Powai, Mumbai, India-400076.

Dr. Saiprasad Shetty

M.B.B.S, M.S(General Surgery) Clinical Assistant (Dept of Surgery), Dr. L. H. Hiranandani Hospital Powai S.T. Road Chembur, Mumbai 400071.

Dr. Abhijit Joshi

MBBS, MS (Gen Surgery), Fellowship in Laparoscopy, Diploma in Advanced Laparoscopy Consultant General, G I & Endo-Laparoscopic Surgeon Dr. L H Hiranandani Hospital, Powai, Mumbai, India.

ABSTRACT

Appendiceal neoplasms are rare, accounting for 0.6 percent of gastrointestinal malignancies and approximately 0.7-1 percent of appendectomies specimens (1,5). Most of the appendicular malignancies are detected incidentally. We herein report a case of 76 years old male with no comorbidities, presented to the emergency department with acute abdominal pain and vomiting. On physical examination the abdomen was distended without guarding or rigidity. An abdominal CT revealed an ileo-colic intussusception (fig1). Patient underwent laparoscopic quarter colectomy with ileo-colic anastomosis. Post-operatively patient recovery was uneventful. The histopathology report showed low-grade appendiceal mucinous neoplasm (LAMN) (fig 4).

INTRODUCTION:

Appendicular neoplasms are uncommon and rare tumors, accounting for less than 1.8 percent of the surgical specimen of the appendix (2). These tumors can mimic the symptoms of acute appendicitis. Broadly neoplasms of the appendix are divided as: epithelial tumors, mesenchymal tumors and lymphomas. Mucinous neoplasms of the appendix are of epithelial origin, and produces mucin. Appendiceal mucinous neoplasm accounts for 0.3-0.8 percent of appendectomies specimen (3).

Multiple efforts were taken in the past to categorise and classify mucinous tumors of the appendix. Various systems and classifications are existed for categorising the mucinous tumors of the appendix, the most consensus classification is of WHO 2010(2). However, in 2016 these controversial issues were discussed in Peritoneal Surface Oncology Group International (PSOGI) (4) conference. The consensus method for categorising tumors has been developed by the panel of experts. The panel categorised non-carcinoid epithelial neoplasia into multiple types as: Adenoma (low/high grade dysplasia), serrated poly, low-grade appendiceal mucinous neoplasm (LAMN), high grade appendiceal mucinous neoplasm (HAMN), mucinous and non-mucinous adenocarcinoma.

The mucinous neoplasms often cause accumulation of mucin along the inner part of the appendix, this lead to cystic dilatation of the appendix. However, perforation of these mucin containing appendix into the peritoneal cavity causes pseudomyxoma peritonei (PMP). Intussusception caused by LAMN is a rare complication. Here we present a case of ileo-colic intussusception secondary to low grade appendiceal mucinous neoplasm.

CASE REPORT:

A 76-year-old male presented with a one-month history of dull right lower quadrant abdominal pain and mild distention. He did not give any history of weight loss, change in bowel habits, bleeding per rectum or melena. Physical examination revealed mild abdominal distention with slight tenderness, and palpable lump of 5x4cm along right iliac fossa. The basic laboratorial analysis along with CEA levels were unremarkable.

An ultrasound scan of his abdomen revealed a 5 X 6 cm ill-defined lump related to bowel in the right lower quadrant was found. Abdominal computed tomography scan revealed a large ileo-colic intussusception. However, it did not reveal any mass at its lead point. (Fig 1).

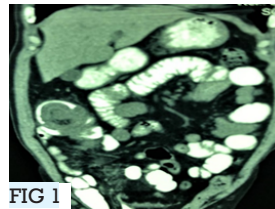


FIG 1 Fig 1. CT scan Coronal view, showing Ileo-colic intussusception along right side.



FIG 2 Fig 2. Intraoperative view where attempt to reduce intussusception is seen, with appendix and caecum.

The patient was planned for surgery. At Laparoscopy he was found to have a huge mass measuring 5 cm x 6 cm x 6 cm in the right iliac fossa. Gentle attempts to reduce the intussusception failed. Hence, laparoscopic quartercolectomy with totally intracorporeal ileo-ascending anastomosis performed. The ceecal mass and the appendix were removed intact without any intraoperative perforation (Figure 3). The mass was firm to touch and the appendix was markedly swollen (Fig2.)



FIG 3 Fig 3 Specimen with "Volcano" sign with intussusception which could not be reduced.

Gross examination of the specimen revealed a protruding smooth mass in the luminal aspect of the caecum with a 'crater' like opening at its peak (Fig 3). This mass was found to be located luminally exactly at the point of origin of the appendix

in the caecum. Base of the appendix is merged with caecum with presence of polypoidal mucosal elevation.

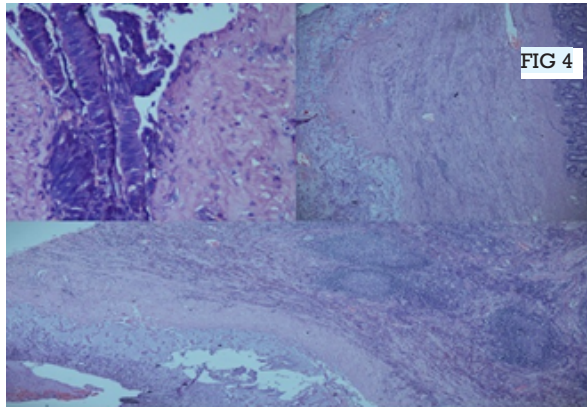


FIG 4

Fig 4:
 No.1: Photomicrograph shows flat mucinous epithelium of low-grade cytology lining appendiceal mucosa and absence of muscularis mucosae, (H&E x 40)
 No2: Photomicrograph showing acellular mucinous areas in the wall of caecum. (H&E x 4)
 No3: Photomicrograph showing acellular mucinous areas in the wall of Appendix (H&E x 4)

The histopathology(fig4) report revealed the appendicular mucosa at the base is lined by single layer of mucinous columnar epithelium with nuclear atypia, atrophy of the lymphoid follicles. LAMN confined to the appendix and without extrusion of mucin or mucinous neoplasia beyond the appendiceal serosa, or without involvement of the specimen margin by either neoplastic epithelium or acellular mucin. The specimen was retrieved in a plastic bag and hence a redo surgery was not required. The patient was stable postoperatively with no surgical complications. He was discharged from the hospital on post-operative day seven.

DISCUSSION:

Low-grade appendiceal mucinous neoplasms are uncommon and rare tumor of the appendix, accounting for 0.4 per cent of the appendicular specimen. The disease is more common in females and features most commonly in 5th or 6th decade of life, though they could be detected at any age. (1,5).

Low-grade appendiceal neoplasm (LAMN) can be define as presence of low-grade cytology and any of the following features:(PSOGI 2016) (4). Loss of muscularis mucosae, fibrosis of submucosa, undulating or flattened epithelial growth, “pushing invasion” (Expansile or diverticulum like growth), dissection of acellular mucin in the wall, mucin or neoplastic cells outside the appendix.

LAMNs are usually asymptomatic and diagnosed incidentally during abdominal imaging or on diagnostic laparoscopy. When the tumour is large, LAMN patient may present with vomiting, pain in abdomen or palpable mass in the right iliac fossa. However, LAMNs can be accompanied with diverticula, herniations, dissections, and rupture leading to intestinal obstruction or intussusception or pseudomyxoma peritonei. (6)

Laboratorial analysis for LAMNs are nonspecific. However, patients are also noted with low levels of hemoglobulin and increased levels of tumor markers (e.g. CA19-9, CEA, CA 125) (7). Radiological modalities for diagnosis include ultrasound (US) and CT scan of the abdomen. It is difficult to distinguish mucinous from no-mucinous neoplasm on these imaging techniques. However, there are certain CT characteristic features of the tumor suggestive of neoplasm includes:

irregular and calcified wall with thickening of the soft tissue. Appearance of ascites or peritoneal thickening may suggest intraabdominal spread. (8)

There are different treatment modalities and controversies around the management of appendicular mucocele. The primary surgery for appendiceal mucinous lesions should be appendicectomy with complete clearance of meso appendicular tissue and any mucus found during surgery should be sent for cytological evaluation. (9) If on the frozen section meso appendicular lymph nodes are negative then no further treatment is required. However, if the frozen section is positive then quartercolectomy is sufficient.(9) However, If the base of the appendix is involved in the disease process so that a clear margin cannot be achieved by stapling, a partial caeectomy , ileocecectomy, or right colectomy can be performed on the basis of surgeon's experience and knowledge, with the ultimate goal of achieving a clear margin.

Right hemicolectomy should be considered to clear the tumor margin in case of involvement after appendectomy. It is usually indicated when:-

- Tumor involve peri-appendiceal region
- Tumor size of 2 cm or greater
- High grade Histology
- Tumors that invades across muscularis-propria. (9)

LAMN with peritoneal metastasis can be treated with Optimal cytoreductive surgery (CRS) and hyper thermic intraperitoneal chemotherapy (HIPEC). Neoadjuvant systemic chemotherapy is not recommended. High-grade appendiceal adenocarcinomas should be considered for neoadjuvant systemic chemotherapy and if there is positive response to chemotherapy then surgical resection with HIPEC is to be considered (10).

Controversy remain on the optimal surgical technique (laparoscopic or exploratory). Laparoscopic management is still advocated by surgeons (11). Laparoscopic technique is advocated because it provides with zoomed and close vision, faster recovery, less hospital stay and early mobilization. Whatever the technique is use, the goal of the surgery should be to do the en-block resection and avoid tumor handling or tumor rupture. In our case, great care had been taken to avoid tumor spillage or rupture, and the specimen was retrieved intact in plastic bag (fig 3).

The final result of the recovery depends largely on the extent of disease spread. Intact removal without peritoneal spillage can increases the survival rate up to 100% for 5 years. However, 5 years survival rates drastically decrease up to 45% in cases of intraabdominal spillage or improper handling with ruptured specimen. Spillage can also put the patient at a risk of developing PMP.

CONCLUSION:

Ileo-colic intussusception caused by a low-grade mucinous neoplasm is very rare clinical presentation. CT scan of the abdomen and pelvis is the recommended modality for provisional diagnosis. However, the final diagnosis is made intraoperatively and on histopathological examination.

Quartercolectomy can be performed if LAMN is confined to the base of the appendix, un ruptured, without the need of extensive right hemicolectomy, as was the case of our patient. Right hemicolectomy is warranted for- tumor involving peri-appendiceal region, tumor size of 2 cm or greater, high grade Histology or tumor that invades across muscularis-propria (1). The primary surgery for appendiceal mucinous lesions should be appendicectomy with complete clearance of meso appendicular tissue and any mucus found during surgery should be sent for cytological evaluation. Hence, during emergency appendectomy it is important to consider every mucocele as malignant, in order to avoid iatrogenic

perforation causing pseudomyxoma peritonei or spillage or seeding.

REFERENCES:

1. Incidentally discovered low-grade appendiceal mucinous neoplasm: a precursor to pseudomyxoma peritonei. Padmanaban V, Morano WF, Gleeson E, et al. *Clin Case Rep.* 2016;4:1112–1116. [PMC free article] [PubMed] [Google Scholar].
2. Connor SJ, Hanna GB, Frizelle FA. Appendiceal tumors: retrospective clinicopathologic analysis of appendiceal tumors from 7,970 appendectomies. *Dis colon and rectum.* 1998; 41:75–80. [PubMed] [Google Scholar].
3. S.H. Tirumani, M. Fraserhill, R. Auer, W. Shabana, C. Walsh, F. Lee, J.G. Ryan. Mucinous neoplasms of the appendix: a current comprehensive clinicopathologic and imaging review. *Canc Image,* 13 (1) (2013), p. 14, 10.1102/1470-7330.2013.0003.CrossRefScopus Google.
4. Carr NJ, Cecil TD, Mohamed F, Sobin LH, Sugarbaker PH, González-Moreno S. A consensus for classification and pathologic reporting of pseudomyxoma peritonei and associated appendiceal neoplasia: The results of the peritoneal surface oncology group international (PSOGI) modified delphi process. *Am J Surg Pathol.* 2016;40:14-26.
5. Pathology of mucinous appendiceal tumors and pseudomyxoma peritonei. Ramaswamy V. *Indian J Surg Oncol.* 2016; 7:258–267. [PMC free article] [PubMed] [Google Scholar].
6. Primary epithelial neoplasms and other epithelial lesions of the appendix (excluding carcinoid tumors) Misdraji J, Young RH. *Semin Diagn Pathol.* 2004; 21:120–133. [PubMed] [Google Scholar].
7. Surgical approach to appendiceal mucocele mimicking an adnexal complex mass: case report. Scaffa C, Di Bella O, Tartaglia E, Rotondi M, Lup F, Messalli EM. *Eur J Gynaecology Oncol.* 2007;28(6):503.
8. PubMed. Predicting Underlying Neoplasms in Appendiceal Mucoceles at CT: Focal Versus Diffuse Luminal Dilatation. Marotta B, Chaudhry S, McNaught A, Quereshy F, Vajpeyi R, Chetty R, Ghai S. *AJR Am J Roentgenol.* 2019;213(2):343. Epub 2019 Apr 11.
9. J. G. D. A. Filho and E. F. D. Lira, "Mucocele of the appendix: appendectomy or colectomy?" *Journal of Coloproctology,* vol. 31, no. 3, pp. 276–284, 2011.
10. Sugarbaker PH, Chang D. Results of treatment of 385 patients with peritoneal surface spread of appendiceal malignancy. *Ann Surg Oncol* 1999; 6:727–731. [PubMed] [Google Scholar].
11. R. Miraliakbari and W. H. H. Chapman III, "Laparoscopic treatment of an appendiceal mucocele," *Journal of Laparoendoscopic & Advanced Surgical Techniques,* vol. 9, no. 2, pp. 159–163, 1999. [Google Scholar]