Clinical Images

Constance Houlzé-Laroye and Clarisse Eveno*

Low-grade appendiceal mucinous neoplasms with bowel obstruction

<https://doi.org/10.1515/pp-2019-0020>

Received July 04, 2019; accepted August 27, 2019;

previously published online October 11, 2019

Abstract

Background: Perforation of low-grade appendiceal mucinous neoplasms (LAMN) is characterized by its potential to spread mucin into peritoneal cavity, giving rise to pseudomyxoma peritonei, commonly treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy.

Symptoms of intestinal obstruction and appendiceal infection are rare at early stages of the disease, while abdominal distension are observed in the later stages due to mucin spread.

Methods: We report herein a case of LAMN with atypical symptoms in a 35-year-old woman with abdominal symptoms evoking an intestinal obstruction.

Results: An abdominal CT scan revealed an ileo-caecal intussusception. An exploratory laparotomy and examination of the peritoneal cavity ruled out an exteriorization of mucin and the bowel was resected.

Conclusions: The pathology analysis confirmed the diagnostic of LAMN. This report aims to raise awareness among surgeons, of rare clinical presentations that LAMN may show, to adapt the surgical treatment to these patients and assign them to referral centers.

Keywords: ileo-caecal intussusception, low-grade appendiceal mucinous neoplasma, mucin

We report a case of 35-year-old female patient with no medical history presented to the emergency department with acute vomiting and abdominal pain. The abdomen was distended with significant guarding in the left upper quadrant. An abdominal CT scan revealed an ileo-caecal intussusception through the left transverse colon (Figure 1, panels A, B with a target sign, arrow) caused by an appendicular mucocele filled with mucin, presenting parietal calcification (Figure 1, panel C, arrow). An exploratory laparotomy and examination of the peritoneal cavity ruled out an exteriorization of mucin and the bowel was resected. The pathology analysis confirmed the low-grade appendiceal mucinous neoplasms (LAMN) diagnosis with necrosis of appendiceal basis [1]. A perforation of LAMN is characterized by its potential to spread mucin into peritoneal cavity, giving rise to pseudomyxoma peritonei, commonly treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy [2, 3]. Symptoms of obstruction and appendiceal infection at early stages of the disease, while abdominal distension is observed in the later stages due to mucin spread.

Ethical considerations: Informed consent given by the patient (Registration number 24009 in RENAPE network, organization for the treatment of rare tumors of the peritoneum).

Author contributions: All the authors have accepted responsibility for the entire content of this submitted manuscript and approved submission.

Research funding: None declared.

Employment or leadership: None declared.

Honorarium: None declared.

Competing interests: The funding organization(s) played no role in the study design; in the collection, analysis, and interpretation of data; in the writing of the report; or in the decision to submit the report for publication.

*Corresponding author: Clarisse Eveno, Department of Digestive and Oncologic Surgery, Claude Huriez University Hospital, Centre Hospitalier Universitaire (CHU) Lille, Université de Lille, Lille, France; INSERM Unité Mixte de Recherche 1172-JPARC Jean-Pierre Aubert Research Center, Team “Mucins, epithelial differentiation, and carcinogenesis”, Lille, France, E-mail: clarisse.eveno@chru-lille.fr

Constance Houlzé-Laroye, Department of Digestive and Oncologic Surgery, Claude Huriez University Hospital, Centre Hospitalier Universitaire (CHU) Lille, Université de Lille, Lille, France, E-mail: constance.laroye@gmail.com

Figure 1: Abdominal CT scan revealed an ileo-caecal intussusception through the left transverse colon (panels A, B with a target sign, arrow) caused by an appendicular mucocele filled with mucin, presenting parietal calcification (panel C, arrow).

References

1. Carr NJ, Bibeau F, Bradley RF, Dartigues P, Feakins RM, Geisinger KR, et al. The histopathological classification, diagnosis and differential diagnosis of mucinous appendiceal neoplasms, appendiceal adenocarcinomas and pseudomyxoma peritonei. *Histopathology* 2017;71:847–58.
2. Benhaim L, Faron M, Gelli M, Sourrouille I, Delhorme JB, Elias D, et al. Survival after complete cytoreductive surgery and HIPEC for extensive pseudomyxoma peritonei. *Surg Oncol* 2019;29:78–83.
3. Ansari N, Chandrakumaran K, Dayal S, Mohamed F, Cecil TD, Moran BJ. Cytoreductive surgery and hyperthermic intraperitoneal chemotherapy in 1000 patients with perforated appendiceal epithelial tumours. *Eur J Surg Oncol* 2016;42:1035–41.