



Image of the Month

Pseudomyxoma peritonei of the appendix after ileocecal resection: Expect the unexpected

Melek Simsek^{a,*}, Annemiek J. Linn^b, Nanne K.H. de Boer^a^a Department of Gastroenterology and Hepatology, Amsterdam UMC, Vrije Universiteit Amsterdam, AG&M Research Institute, Amsterdam, the Netherlands^b Amsterdam School of Communication Research/ASCoR, University of Amsterdam, Amsterdam, the Netherlands

A 32-year-old female patient with Crohn's disease presented with vomiting, diarrhea, night sweating and pain in the lower right abdomen for over three months. She underwent an ileocecal resection ten years earlier and her Crohn's disease remained in remission without medication ever since. Initially, it was assumed that patient had a flare-up of the Crohn's disease. A computed tomography (CT) showed quiescent Crohn's disease, ascites and a cystic mass adjacent to the ascending colon without involvement of the right ovary (Fig. 1A and B).

Diagnostic paracentesis revealed mucinous ascites indicative of pseudomyxoma peritonei (PMP). Despite her historical ileocecal resection, the appendix was suspected as a source. Thereupon, the pathology report of the surgery was reiterated: the appendix was not removed completely. PMP of the appendix stump was confirmed during laparoscopy and pathology, classified as low-grade appendiceal mucinous neoplasm (LAMN) (Fig. 2) [1]. The peritoneal cancer index (PCI) score prior to removal was high: 31. In patients with low-grade PMP, however, complete surgical removal is still possible despite a high PCI score [2].

The patient underwent cytoreductive surgery (CRS), hyperthermic intraperitoneal chemotherapy (HIPEC), using Mitomycin C, and bilateral salpingo-oophorectomy (BSO) to minimize recurrence of disease. She recovered successfully and remained disease free during two years of follow-up. A year after surgery, she got pregnant after egg donation and gave birth to a healthy child. In hindsight, the alternative of a more conservative surgical approach to preserve fertility of our patient was discussed. In a couple of studies, it has been reported that ovarian preserving CRS in young women with low-grade PMP resulted in restored fertility and successful pregnancies whereas patients remained disease-free [3–5].

In conclusion, this case highlights that diagnosing PMP is a clinical challenge and physicians should be aware of the possible manifestation of the disease in young patients, even in those with (partial) removal of the appendix. In young women, adequate (in)fertility counseling is crucial

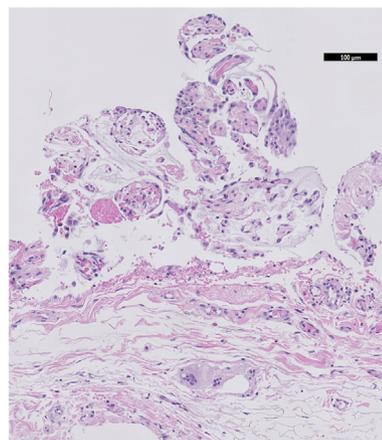


Fig. 2. Hematoxylin and eosin staining shows presence of low-grade atypical cells producing acellular mucin in the absence of infiltrative growth.

and ovarian preserving options should be considered in selected patients who wish to conceive.

Conflicts of interest

None declared.

Patient consent

Informed consent was obtained from the patient for the publication of their information and imaging.

Patient approval

Permission to publish patient information was provided by the patient. Our institution does not require IRB approval for case reports.

References

- [1] Misdraji J. Mucinous epithelial neoplasms of the appendix and pseudomyxoma peritonei. *Mod Pathol* 2015;28(Suppl 1):S67–79.
- [2] Gilly FN, Cotte E, Brigand C, et al. Quantitative prognostic indices in peritoneal carcinomatosis. *Eur J Surg Oncol* 2006;32(6):597–601.
- [3] Elias D, Duchalais E, David A, et al. Comprehensive study of ovarian metastases in young women with peritoneal pseudomyxoma: is a preservation of fertility possible? *Eur J Surg Oncol* 2013;39(7):748–53.
- [4] Sheehan LA, Mehta AM, Sawan S, et al. Preserving fertility in pseudomyxoma peritonei, a novel approach. *Pleura Peritoneum* 2017;2(1):33–6.
- [5] Ortega-Deballon P, Glehen O, Levine E, et al. Childbearing after hyperthermic intraperitoneal chemotherapy: results from an international survey. *Ann Surg Oncol* 2011;18(8):2297–301.

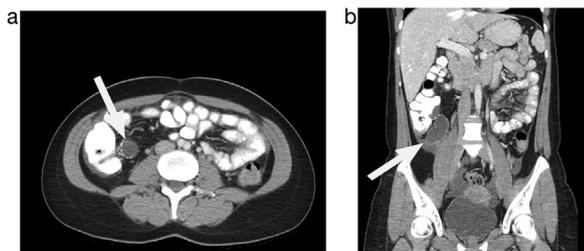


Fig. 1. (A) Axial and (B) coronal reformatted contrast-enhanced CT images of the abdomen. Arrow depicts the cystic mass adjacent to the ascending colon.

* Corresponding author at: Department of Gastroenterology and Hepatology, Amsterdam UMC, Vrije Universiteit, De Boelelaan 1118, 1081 HZ Amsterdam, the Netherlands.

E-mail address: m.simsek@amsterdamumc.nl (M. Simsek).