Peritoneal Metastases from Appendiceal Cancer



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KEYWORDS

- Appendiceal cancer Pseudomyxoma peritonei Low-grade mucinous tumors
- Low-grade lesions

KEY POINTS

- The early symptoms of appendiceal cancer may mimic the clinical picture of appendicitis.
- Most patients are diagnosed incidentally during surgical exploration or late when peritoneal or systemic dissemination has already occurred, as colonoscopy rarely will diagnose an appendiceal cancer.
- Systemic/extraperitoneal metastases are distinctly unusual for appendiceal mucinous lesions.

INTRODUCTION

Appendiceal neoplasms are diagnosed in approximately 1% of all appendectomy specimens. The early symptoms of appendiceal cancer may be nonspecific, or they may mimic the clinical picture of appendicitis. Not surprisingly, most patients are diagnosed incidentally during surgical exploration or late when peritoneal or systemic dissemination has already occurred, as colonoscopy rarely will diagnose an appendiceal cancer.

The common pathway of all appendiceal tumors regardless of grade and cell of origin involves invasion of the appendiceal wall, luminal obstruction, and perforation with subsequent dissemination of malignant epithelial cells throughout the peritoneal cavity (Figs. 1 and 2). Systemic/extraperitoneal metastases are distinctly unusual for appendiceal mucinous lesions. The subsequent course of disease for a mucinous lesion depends on the grade of appendiceal primary as defined by Bradley and colleagues.²

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Fig. 1. Ruptured appendiceal mucocele.

Patients with low-grade mucinous tumors (LGA) will typically progress slowly over years and can possibly develop pseudomyxoma peritonei, which describes accumulation of mucinous ascites within the peritoneal cavity. Approximately 7% of the low-grade lesions have lymph node involvement, and up to 16% will dedifferentiate into higher-grade lesions during the course of the disease.^{3–5}

High-grade lesions (HGA) are more likely to metastasize systemically, resulting in a poorer prognosis. The variability of HGA clinical presentation stems from the variability in biologic behavior and grade of the primary lesions: moderately differentiated, poorly differentiated, as well as signet ring cell histologies that are all included in the HGA group.²

Pseudomyxoma peritonei (PMP) is a descriptive term, referring to the presence of muscinous ascites. Mucinous ascites is by for most commonly associated with mucinous appendiceal neoplasms. However, pseudomyxoma can be produced by several primary tumor types, including appendiceal, colon, ovarian, mucinous pancreatic, and low-grade urachal primaries among others. The prognosis of patients with PMP of nonappendiceal origin depends on the primary tumor type, while the surgical selection for cytoreductive surgery (CRS)/hyperthermic intraperitoneal chemotherapy (HIPEC) for non-LGA PMP patients follows different selection criteria.



Fig. 2. Eviscerated omental metastatic deposits form a low-grade appendiceal primary.

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