

Appendiceal Mucinous Neoplasm

Nurse education about a rare and complex disease

Alaine M. Kamm, MSN, APRN, ACNP-BC, and Kiran K. Turaga, MD, MPH

BACKGROUND: Appendiceal mucinous neoplasms (AMNs) are a rare and complex spectrum of disease involving a mucinous tumor within the appendix, which can range from benign to malignant. If not limited to the appendix, the mucinous tumor can spread diffusely throughout the peritoneum.

OBJECTIVES: Because of the low incidence of AMN, this study describes the diagnosis and treatment process for this disease, which is not well known to most care providers.

METHODS: An extensive analysis of the current literature, including incidence, diagnosis, and surgical treatment, was performed. A review of the pre- and postoperative needs for patients undergoing surgery was also conducted.

FINDINGS: There is minimal information in the nursing literature about AMN and the complexity of surgical management. Nurses play an important role in caring for these patients and their unique needs both before and after surgery.

KEYWORDS

appendix; mucinous neoplasm; HIPEC; pseudomyxoma peritonei; jelly belly

DIGITAL OBJECT IDENTIFIER

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A 61-YEAR-OLD MAN PRESENTED TO HIS PRIMARY CARE PROVIDER with abdominal discomfort on his right side. He was initially treated with oral antibiotics for presumed early appendicitis. His symptoms did not improve and a computed tomography (CT) scan was ordered, which revealed a hepatic cyst and a mucocoele of the appendix. He was taken to surgery for a laparoscopic appendectomy; however, the surgery was aborted because of peritoneal implants and the concern for hepatic metastasis. Serum tumor markers demonstrated an elevated CA 19-9 of 130 and a CEA of 4.6. He had a colonoscopy within a year of presentation, which was significant for a benign polyp.

The patient was referred to a tertiary medical center for management of his appendiceal mucinous neoplasm (AMN). After additional testing, he underwent cytoreductive surgery (CRS), which involved removal of the anterior and pelvic peritoneum, greater omentum, and en bloc resection of the appendiceal mass with the base of the cecum. The appendiceal mass measured greater than 12 cm. In addition, there were over 100 mesenteric and serosal implants on the small bowel serosa. Following the cytoreduction, the patient received 90 minutes of hyperthermic intraperitoneal chemotherapy (HIPEC) with mitomycin-C chemoperfusion.

Pathology showed mucinous adenocarcinoma in the cecum and appendix with adjacent tissue specimens, indicating no cancer. He recovered from surgery and is followed every three months with tumor markers (CA 19-9, CEA, and CA 125) and magnetic resonance imaging (MRI) of the abdomen and pelvis.

Background

Significant improvements have been made in defining and classifying AMNs. In 2016, a clinical working group convened to help standardize the classification of appendiceal tumors. As part of the group's consensus recommendations, several definitions were agreed on (Carr et al., 2016) (see Table 1). AMNs have variable malignant potential. They may occur as a slow-growing benign tumor, as is the case with low-grade AMNs, or they may exhibit intermediate malignant potential, such as with peritoneal mucinous