Appendiceal Mucinous Neoplasm: A rare clinical entity. A case report

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Case Report

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Background: Appendiceal mucinous neoplasms are uncommon, representing only 0.25% of all appendectomies and 8% of appendiceal tumors. They may be asymptomatic and are often discovered incidentally during radiologic or endoscopic evaluations, or during laparotomy or laparoscopy performed for other reasons. Approximately half of the cases present with right iliac fossa pain suggestive of acute appendicitis. These lesions may be benign or malignant, requiring individualized evaluation to determine their nature [6]. An estimated 10–15% of appendiceal mucinous neoplasms may evolve into pseudomyxoma peritonei, significantly affecting patient outcomes.
An appendiceal mucinous neoplasms is defined as a cystic dilation of the appendiceal lumen due to obstruction, leading to retrograde accumulation of mucinous material. This rare pathology is most frequently detected incidentally and can be classified into four histological subtypes, with mucinous cystadenoma being the most common.

Keywords: Appendix, Appendiceal mucinous neoplasms, Mucocele, Appendectomy, Mucinous Cystadenoma

Appendiceal mucinous neoplasms are rare lesions, accounting for 0.25% of all appendectomies and 8% of appendiceal tumors [1,2]. They may be asymptomatic and are often discovered incidentally during imaging or endoscopic examinations, or during laparotomy or laparoscopy performed for unrelated indications [4]. About half of the patients present with right iliac fossa pain that mimics acute appendicitis [1,3]. These lesions can be benign or malignant, making careful evaluation essential to establish their true nature [6].

First described by Rokitansky in 1842 and later named by Feren in 1876 [1], the appendiceal mucinous neoplasms is not a specific clinical or histological diagnosis but rather a descriptive macroscopic term for an obstructive cystic dilation of the appendiceal lumen with retrograde mucin accumulation [5]. Historical reports indicate that approximately half of the patients are asymptomatic, with the lesion discovered incidentally during surgery, imaging, or colonoscopy [4,6]. In a review of 135 patients by Stocchi et al., 51% were asymptomatic. Among symptomatic patients, abdominal pain occurred in 27%, a palpable abdominal mass in 16%, weight loss in 13%, and nausea or vomiting in 9%, with symptoms being more commonly associated with malignancy [2,5].

Surgical intervention is the cornerstone of treatment, but the optimal approach remains debated [3,6]. Accurate preoperative diagnosis is crucial to guide surgical strategy and prevent intraoperative rupture, as spillage of mucin into the peritoneal cavity can result in pseudomyxoma peritonei [7,8].

Case report

A 42-year-old man with no significant medical history presented to the emergency department with a three-day history of diffuse abdominal pain. He denied nausea and vomiting but reported altered bowel habits, with no bowel movement for five days. An abdominal ultrasound performed at a private clinic suggested sigmoid volvulus. On surgical evaluation, the patient was alert, oriented, and hemodynamically stable, with a soft, non-tender abdomen and no peritoneal signs. A palpable mass measuring approximately 10 × 5 cm was noted in the hypogastrium. A computed tomography scan revealed an intrapelvic mass consistent with a mesenteric cyst (Figure 1).

Due to the suspicion of intestinal obstruction, exploratory laparotomy was performed. Intraoperatively, a mass arising from the cecum at the base of the appendix was identified, consistent with an appendiceal mucinous neoplasm (Figure 2). A conventional appendectomy was carried out, yielding a specimen measuring approximately 15 × 8 cm with an intact base, using the Pouchet technique with Z-shaped Zuckermann stump inversion. (Figure 3).

The specimen was sent for histopathologic analysis, the result was obtained 10 days after the surgical procedure, where the pathologist reports the presence of mucinous cystadenoma.

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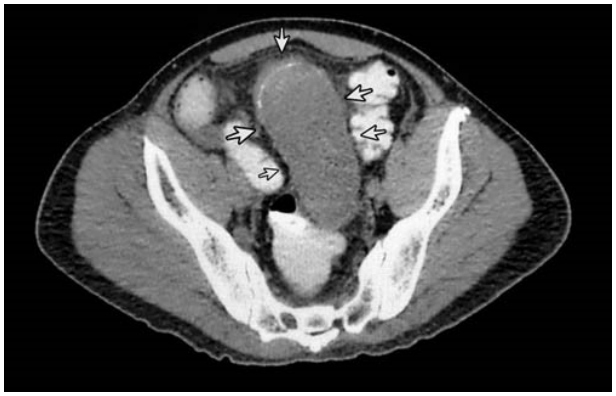

Figure 1. Abdominopelvic computed tomography revealing an abdominal mass located within the pelvic cavity. (Arrows)

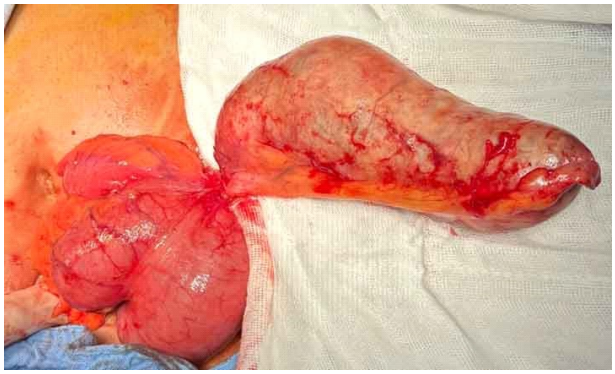

Figure 2. Cecum-associated mass consistent with appendiceal mucinous neoplasm.

Discussion

Appendiceal mucinous neoplasms is characterized by cystic dilation of the appendiceal lumen due to obstruction and retrograde mucin accumulation [1,2]. Its etiology can be benign or neoplastic, sometimes secondary to ovarian, breast, or liver malignancies [6,9]. Although rare, with an incidence of 0.2% of appendectomies, its presentation is often asymptomatic or mimics acute or chronic appendicitis [4,5]. Rokitansky first described the condition in 1842 under the term *Hydrops processus vermiformes* [1].

Histologically, four subtypes are recognized. Simple or retention mucoceles, accounting for 15–20% of cases, result from obstruction of mucus drainage, often by a fecalith, and feature a lumen usually less than 1 cm with atrophic, non-proliferative epithelium [1,3,9]. Mucosal hyperplasia mucoceles (5–25%) are benign lesions causing mild appendiceal dilation, typically diagnosed incidentally by pathologists [1,2]. Mucinous cystadenoma, or low-grade appendiceal mucinous neoplasm, represents approximately 50% of cases and is characterized by dysplastic epithelium with villous-papillary or tubular-glandular patterns producing abundant mucin [5,6]. Mucinous cystadenocarcinoma (11–20%) shows high-grade dysplasia with frequent invasion beyond the muscularis mucosae [6,10].

Appendiceal tumors are identified in 0.2–0.3% of appendectomies and account for 8% of all appendiceal tumors, with a mean presentation age of 55 years and a female predominance of 4:1 [3,4]. Clinical manifestations are variable and nonspecific [2,5]. Historically, 50% of patients are asymptomatic, while symptomatic cases present with right iliac fossa pain, palpable abdominal mass, nausea, vomiting, altered bowel habits, gastrointestinal bleeding, or, occasionally, intestinal obstruction [4].

Radiologic findings vary. Abdominal radiography may show a right lower quadrant mass with curvilinear wall calcification in up to 50% of cases [6]. Ultrasound typically demonstrates a cystic mass with posterior enhancement, an ill-defined wall, and variable echogenicity depending on mucin content; in giant mucoceles, an "onion-skin" appearance may be observed [6]. CT remains the most specific imaging modality, revealing a well-encapsulated, low-attenuation cystic mass adjacent to the cecum, often with wall calcifications [6].

Definitive diagnosis relies on histopathologic examination [6,9]. Preoperative diagnosis is challenging due to the lesion's rarity, nonspecific clinical presentation, and the overlap of imaging findings with other conditions [5]. Treatment is always surgical [3]. Appendectomy is indicated for non-malignant lesions, while right hemicolectomy is recommended in the presence of features suspicious for malignancy, such as positive lymph nodes, perforation, enlarged mesenteric nodes, positive cytology, or involvement of the ileum or cecum [8,9]. Thorough abdominal exploration is essential to exclude other mucin-secreting malignancies, including colon and ovarian cancer, as the risk of colon adenocarcinoma is six times higher in patients with appendiceal mucocele [4,8].

If rupture occurs intraoperatively, minimal handling is advised, with complete removal of the appendix including the mesoappendix, clear surgical margins, and histologic sampling of mucin [7,8]. Referral to a specialized peritoneal carcinomatosis center is warranted for cytoreductive surgery combined with heated intraperitoneal chemotherapy (HIPEC) and, in some cases, early postoperative intraperitoneal chemotherapy (EPIC) [7].

Conclusion

Appendiceal mucinous neoplasms are an uncommon condition with often nonspecific presentation and frequent incidental detection. Prompt diagnosis through appropriate imaging and surgical intervention is crucial to prevent complications such as pseudomyxoma peritonei. Appendectomy remains the treatment of choice in most cases, provided there is no

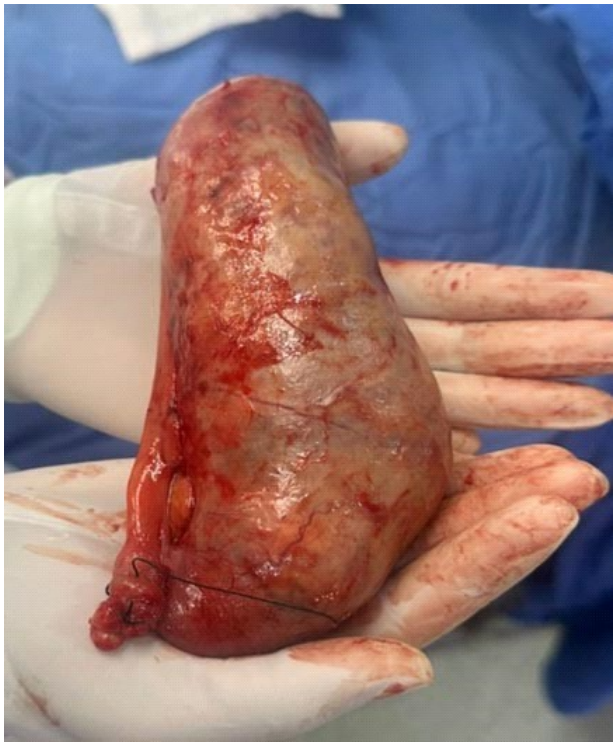

Figure 3. Surgical specimen was obtained, consisting of an appendiceal mucinous neoplasm measuring approximately 15 x 8 cm.

suspicion of malignancy. In complex cases involving perforation or neoplastic suspicion, right hemicolectomy and oncologic follow-up are recommended. Early recognition and intervention improve prognosis and reduce the risk of recurrence or progression.

Conflicts of interests

The authors declare that there are no financial, personal, or institutional conflicts of interest that could have influenced the work reported in this manuscript.

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