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Incidentally discovered ruptured mucinous appendiceal neoplasms. What about the surgical approach?

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Abstract

Mucocele of the appendix is an uncommon pathological condition often discovered incidentally during surgery. It may arise from non-neoplastic or neoplastic lesions, necessitating histopathological examination for definitive diagnosis. Failure to identify and manage this condition preoperatively can result in rupture, spilling mucin into the peritoneal cavity, and causing Pseudomyxoma Peritonei (PMP)—a severe complication with poor prognosis. This literature review aims to consolidate information regarding ruptured mucinous appendiceal neoplasms to minimize the risk of PMP and provide guidance on evidence-based surgical management. The review evaluates epidemiology, clinical manifestations, complications, pathology, diagnostic approaches, and treatment strategies. A meticulous surgical excision remains the cornerstone of treatment, with open surgery preferred over laparoscopy to reduce the risk of spillage. Preoperative diagnosis, using imaging modalities such as Computed Tomography (CT) and ultrasonography, is critical to detect potential malignancies. Primary care physicians play a pivotal role in early recognition and timely referral to prevent complications and optimize outcomes. In cases with advanced patients with diffuse mucinous dissemination after mucocele rupture, Cytoreductive Surgery (CRS) and Hyperthermic Intraperitoneal Chemotherapy (HIPEC) are recommended, however the benefit of prophylactic CRS + HIPEC in high-risk patients without peritoneal spread is controversial.

Keywords: Appendix; Neoplasms; Mucinous; Pseudomyxoma peritonei; CRS; HIPEC.

Abbreviations: PMP: Pseudomyxoma peritonei; CRS: Cytoreductive surgery; HIPEC: Hyperthermic intra-peritoneal chemotherapy.

Introduction

Ruptured acute appendicitis is a common surgical emergency often accompanied by findings such as fluid collections and a dilated appendix in the right pelvis on imaging [1,2]. These findings typically necessitate urgent surgical intervention to prevent complications such as abscess formation or peritonitis

[3]. However, one of the most challenging intraoperative scenarios is the identification of a ruptured appendiceal mucocele, a rare condition arising from mucinous neoplasms of the appendix [4]. This situation demands careful surgical decision-making, as it has distinct treatment approaches and prognostic implications. Surgeons must adopt a comprehensive and specialized approach, especially given the risks associated with a ruptured mucocele, such as pseudomyxoma peritonei, to ensure optimal outcomes [5]. In these cases, cytoreductive surgery (CRS) and Hyperthermic Intraoperative Chemotherapy (HIPEC) are recommended.

Discussion

Appendiceal mucoceles, while often incidental findings, pose a significant concern due to their potential to rupture. If rupture occurs, mucinous material can disseminate into the peritoneal cavity, leading to Pseudomyxoma Peritonei (PMP). This condition can result in extensive abdominal involvement and severe morbidity if untreated.

When an appendiceal mucocele is identified intraoperatively, the following considerations apply:

Intact mucocele: If the mucocele appears intact, a simple appendectomy is generally sufficient for treatment [6]. Careful handling is crucial to prevent rupture.

Suspected or confirmed rupture: If there is evidence of rupture or mucinous material within the peritoneal cavity, the surgical approach must change to prevent further spread of mucinous material and reduce the risk of PMP.

- Conversion to an open procedure is recommended to allow thorough exploration.
- Complete resection of the appendix and meticulous removal of all mucinous material is essential [7,8].

Prognosis and postoperative considerations

- **Localized mucocele without perforation**: Patients with mucinous neoplasms confined to the appendix and without perforation have a better prognosis. A simple appendectomy is often curative in these cases.
- **Mucinous neoplasm with perforation**: Perforation may lead to aggressive mucinous dissemination, increasing the risk of PMP and requiring more extensive intervention.
- **Postoperative discovery**: If a mucinous neoplasm is identified postoperatively in the histopathological specimen, the treatment strategy can be controversial.
  - Some surgeons argue against preventive extended resection if the appendectomy margins are negative, as it may not improve survival [9].
  - In cases with advanced disease or diffuse mucinous dissemination, additional treatments, such as Cytoreductive Surgery (CRS) combined with Hyperthermic Intraperitoneal Chemotherapy (HIPEC), may be necessary.

Timely identification and appropriate surgical management are critical in preventing complications associated with PMP, especially in cases involving perforated mucinous neoplasms.

When an appendiceal mucocele ruptures, the risk of peritoneal dissemination and subsequent Pseudomyxoma Peritonei (PMP) increases significantly. CRS and HIPEC are often recommended in cases where there are confirmed peritoneal dissemination or a high likelihood of progression to PMP. The goal is to improve long-term survival by achieving complete cytoreduction and addressing residual mucinous disease. The benefit of prophylactic CRS and HIPEC in high-risk patients without evidence of peritoneal spread remains controversial. CRS combined with HIPEC is typically not performed during the initial surgery unless the procedure occurs at a specialized center equipped with the resources and expertise necessary to manage peritoneal surface malignancies [10]. Given the complexity of appendiceal neoplasms, particularly those with malignant transformation, outcomes depend on several factors such as extent of peritoneal spread, aggressiveness of the neoplasm and initial treatment strategy.

Conclusion

It is generally recommended that patients with appendiceal neoplasms, especially those with malignant potential or peritoneal involvement, be referred to specialized centers. These centers can evaluate the need for CRS and HIPEC and optimize outcomes through expert care. Also, timely and comprehensive management is key to reducing recurrence and improving long-term survival in patients with appendiceal neoplasms [11].

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