



Appendix mucocèles and pseudomyxoma peritonei: Avoiding a sticky situation

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Summary

Appendix mucocèles and pseudomyxoma peritonei are commonly tested in surgical postgraduate examinations, but are rare to see in practice. Understanding their pathology, diagnosis, management, and prognosis will support decision making of surgeons in training via a colorectal multidisciplinary team. This article aims to serve as a short, accessible guide for medical students and surgical trainees preparing for examinations.

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Introduction

Appendix mucocèles and pseudomyxoma peritonei are complex pathological entities. A mucocèle is characterised by the accumulation of mucinous substance within the lumen of the appendix, which can lead to abdominal pain and appendicitis-like symptoms¹. This commonly occurs without corresponding rises in inflammatory markers. In modern practice, they are often seen on CT scans or but can be found incidentally during an appendicectomy for presumed simple appendicitis. Pseudomyxoma peritonei, on the other hand, is a rare form of cancer characterised by dissemination of mucinous material across the peritoneal cavity, often originating from a ruptured appendix mucocèle².

This article explores the nuances of these two conditions, with emphasis on their clinical significance, diagnostic approaches, and the evolving landscape of their management.

Pathophysiology and Etiology

Appendix mucocèles can be benign or malignant. They are typically classified based on their histopathological features, in accordance with a 2012 consensus

classification by the Peritoneal Surface Oncology Group International (PSOGI)²:

Non-neoplastic. This includes simple mucocèles, retention cysts, inflammatory and obstructive mucocèles. They characteristically have evidence epithelial degeneration without mucosal hyperplasia or neoplasia.

Neoplastic. This includes the following subclassifications:

Serrated polyps of the appendix, with or without dysplastic change

Mucinous appendiceal neoplasms, confined within the muscularis propria. They are not infiltrating and do not display desmoplastic reaction. They are cytologically subdivided into low- (LAMNs) and high- (HAMNs) grade appendiceal mucinous neoplasms.

Mucinous adenocarcinoma of the appendix, which demonstrate infiltration and desmoplastic stromal reactions, high-grade atypia and extracellular mucin in >50% of the lesion. The presence of signet ring cells is a feature of poor differentiation, with prognostic importance.

The progression from a mucocèle to pseudomyxoma peritonei involves the rupture of an appendix mucocèle, leading to the dissemination of the mucinous material. It results in a "jelly belly," where the peritoneal cavity fills with mucinous material, significantly impacting the patient's quality of life. Its prognosis is largely determined by the level of cellularity within the mucin⁴.

Clinical presentation and diagnostics

Patients with an appendix mucocèle may present asymptotically or with symptoms ranging from vague abdominal discomfort to those akin to acute appendicitis. Pseudomyxoma peritonei however often presents with increasing abdominal girth, pain, or changes in bowel habits, or even bowel obstruction.

Cross-sectional imaging using computed tomography scans are pivotal in differentiating and identifying mucocèles from more indolent pathologies. The typical features described include a large appendiceal cystic mass with diameter (i.e., >1.5cm) soft tissue thickening, with wall irregularity and eggshell rings of calcification. Pseudomyxoma peritonei is characterized by scalloping of the liver and of other coated visceral organs and omental caking due to mucinous deposits⁵.

The role of surgery

The surgical approach to appendix mucocèles depends on the type and extent of the lesion. Simple mucocèles may only require appendectomy with excision of the mesoappendix for staging, whereas cystadenomas and cystadenocarcinomas may require a right hemicolectomy to ensure clear margins and reduce the risk of pseudomyxoma peritonei⁶. The management of Pseudomyxoma peritonei involves cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC). This procedure aims to both remove all visible disease and treat microscopic deposits, respectively.

Prognosis

The prognosis for patients with appendix mucocèles is generally good if resected without perforation. The prognosis for pseudomyxoma peritonei varies widely based on the extent and grade of disease and completeness of cytoreduction achieved during surgery⁷. This highlights the importance of centralisation to high volume centres. The 5-year survival rate of Pseudomyxoma peritonei following right hemicolectomy, cytoreductive surgery and HIPEC has been estimated in the largest available series at around 60%, demonstrating the importance of early detection and aggressive treatment in specialised centres^{8,9}.

Conclusion

Appendix mucocèles and pseudomyxoma peritonei present challenges to surgeons and oncologists alike due to their pathological complexity and the need for judicious management. Advances in diagnostic imaging, surgical techniques, and adjuvant therapies have improved patient outcomes, but early detection remains crucial. As research in this area allows the field to evolve, it is important for trainees to stay up to date with latest developments to optimise patient outcomes.

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