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Review

Mucocoele and mucinous tumours of the appendix: A review of the literature

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HIGHLIGHTS

- Mucocoele of the appendix contributes 0.2–0.7% of all appendiceal pathologies.
- Benign causes are epithelial hyperplasia or obstruction by stricture or faecolith.
- Neoplastic lesions include cystadenomas and malignant mucinous cystadenocarcinomas.
- The most common symptoms are abdominal pain and a right iliac fossa mass.
- Malignant mucocoele can progress to pseudomyxoma peritonei if incorrectly managed.

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ABSTRACT

Introduction: Mucocoele of the appendix is rarely encountered but consultant general surgical and trainees must be aware of it as a differential diagnosis, due to the nuances associated with its management. We aimed to provide a comprehensive review of the current literature concerning this rare surgical pathology.
Methods: Search terms "appendi*", "tumour", "malignancy", "mucino*" and "cystadenoma" were used in combination to identify papers from PubMed. Abstracts and full text were manually reviewed to identify suitable papers.
Results: Full search results included 311 articles. Review of titles and abstracts led to further full text review of 46 articles. Of these 30 were selected for inclusion based on relevance, adequate sample size and recent publication date.
Discussion: Mucocoele of the appendix describes dilatation with associated luminal mucin and can result from benign and malignant processes. It contributes 0.2–0.7% of all appendiceal pathologies. The most common presenting symptoms are abdominal pain and a palpable mass in the right iliac fossa. Computed tomography of the abdomen and pelvis is key in facilitating diagnosis, although CEA and CA19-9 also have a role. The major complication of malignant causes of mucocoele is progression to pseudomyxoma peritonei. Treatment is surgical with or without chemotherapy depending on the underlying cause. Prognosis depends on aetiology.
Conclusion: Mucocoele of the appendix is a rare diagnosis. However, given the possibility of neoplastic peritoneal dissemination, it should be considered as a diagnosis, especially in older females with non-specific symptoms similar to appendicitis.

1. Introduction

Mucocoele of the appendix is a rare diagnosis but given the high volume of acute surgical patients seen by on call teams, it is likely to be encountered by consultant and trainee general surgeons. Malignant causes of appendiceal mucocoele must be managed differently from acute appendicitis, which symptomatically mimics this condition. Here, we aim to provide a comprehensive overview of this rare pathology to ensure good background knowledge of this heterogeneous entity, which is vital for trainees so that it can be

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included as a differential diagnosis where appropriate and accurate management plans can be formulated. The malignant sequela, pseudomyxoma peritonei, is also discussed.

2. Methods

PubMed was searched using the terms "appendi*", "tumour", "malignancy", "mucino*" and "cystadenoma". Papers not in English were excluded. This yielded 311 results initially. One reviewer manually screened appropriate articles by review of titles and abstracts. Forty-six full-text articles were subsequently downloaded. Of these 30 were selected for inclusion based on their relevance, adequate sample size and recent publication date.

3. Findings and discussion

3.1. Mucocoele

A mucocoele of the appendix describes dilatation of the appendix with associated filling of the lumen with mucin [1]. The term mucocoele encompasses several causative pathologies. Benign causes are epithelial hyperplasia or simple obstruction by stricture or faecolith. In more sinister cases, neoplastic processes can result in luminal obstruction and unregulated mucin production leading to mucocoele with potential for progression. Benign lesions are referred to as mucinous cystadenomas and malignant as mucinous cystadenocarcinomas [1–3]. Other malignancies of the appendix include carcinoid tumours, non-mucinous adenocarcinoma, goblet cell carcinoma and signet ring cell carcinomas, although these rarely lead to mucocoele [4].

3.2. Epidemiology

Mucocoele of the appendix is a rare entity, contributing to only 0.2–0.7% of appendiceal pathologies [2,5,6]. Simple mucocoele is seen in approximately 29% [6]. Epithelial hyperplasia and mucinous cystadenoma contribute to 31–34% of appendiceal mucocoeles; cystadenoma is often cited as the more common of these two [5,6]. Mucinous cystadenocarcinoma is rarer, causing 5% of mucocoeles and is the more common malignant tumour of the appendix, contributing 23–55% of all appendiceal malignancies [7–9].

In cystadenocarcinoma, females are more commonly affected than males, which is the only appendiceal tumour to show this preponderance [7]. The mean age of onset ranges from 62 to 66 years [7,8].

A retrospective analysis of 1621 appendicectomy specimens removed for acute appendicitis, without the suspicion of neoplasm, identified 134 that had pathologies other than appendicitis [5]. Of these 8 were cystadenomas and 1 was a cystadenocarcinoma [5]. These patients had a mean age of 49.1 years and showed a female preponderance, whereas those with confirmed appendicitis had a lower mean age of 36.7 years and showed a slight male preponderance [5]. These basic epidemiological findings indicate we should heighten our awareness of this differential diagnosis in older females presenting with symptoms of acute appendicitis.

3.3. Pathology

The pathology of simple mucocoele and epithelial hyperplasia is self-limiting and luminal dilatation is usually mild. As pressure rises within the appendicular lumen, atrophy of the epithelial cells occurs, resulting in diminished mucin production [1,9].

Cystadenoma often exhibits more extensive dilatation with epithelial atypia and villous changes replacing normal appendiceal epithelium. Perforation occurs in approximately 20% leading to mucin distribution either locally or throughout the peritoneal cavity. There is however no malignant spread associated with perforation of cystadenoma [1,9].

Some low-grade appendiceal mucinous neoplasms (LAMNs) do have the potential to progress to pseudomyxoma peritonei (PMP) via peritoneal spread. The presence of mucin or neoplastic epithelium outside the lumen, in locations such as the appendiceal submucosa, wall or periappendiceal tissue, is associated with PMP [10,11]. Lesions of this type can be labelled as LAMN Type 2, whereas lesions with disease confined to the lumen are defined as LAMN Type 1.

The characteristics of mucinous adenocarcinoma have been summarised by a large American analysis, which included 2101 tumours. It showed that these neoplasms are usually moderately or well differentiated and are likely to be greater than 4 cm by the time of operation. These are often found late and therefore often present as stage T3/4. Nodal spread is seen in only approximately 20% but metastasis is found in almost half [12]. This high proportion of metastatic disease is predominantly due to appendix rupture and peritoneal dissemination but may rarely be caused by haematological dissemination [9].

3.4. Signs and symptoms

In 50% of cases, mucocoele of the appendix is asymptomatic and picked up as an incidental diagnosis [9,13,14]. It is often referred to as a "diagnostic dilemma" due to crossover of symptoms between this pathology and those of the much commoner acute appendicitis. The most common complaint of symptomatic patients, present in over 80%, is abdominal pain [9,15]. Other common symptoms include a palpable mass in the right iliac fossa, nausea, vomiting and weight loss [9,13]. Symptomatology is unable to differentiate between benign and neoplastic causes of mucocoele.

Other presentations include generalised peritonism and intestinal obstruction [15,16].

3.5. Investigations

Raised serum carcinoembryonic antigen (CEA) and CA19-9 are known to be associated with both mucinous cystadenoma and cystadenocarcinoma [17,18]. An analysis of 532 patients with mucinous adenocarcinoma found 56.1% to have raised CEA and 67.1% exhibited raised CA19-9 [18]. Inclusion of these markers in diagnostic investigations is recommended. However, approximately 40% of those with mucinous adenocarcinoma will have a normal CEA, and 26% a normal CA19-9 level [18]. It is therefore important to progress to computed tomography (CT) of the abdomen and pelvis if clinical suspicion is high.

CT is the radiological investigation of choice as dilatation of the appendix is easily evidenced. Mucinous tumours of the appendix appear as well-encapsulated, cystic masses filled with low-attenuation material. Calcification of the wall of the appendix is seen in approximately 50% [19]. Although CT is unable to absolutely differentiate benign tumours from malignant; features of malignancy include irregularity in the appendiceal wall and soft-tissue thickening. The presence of calcification and peri-appendiceal fat stranding cannot predict malignancy [20].

Although diagnosis of appendiceal tumours by colonoscopy is reported in the literature, it is often incidental and only sensitive for large tumours extending into the caecum [21]. It is not recommended as a diagnostic investigation should mucocoele be suspected.

3.6. Pseudomyxoma peritonei

Neoplastic causes of mucocoele have the ability to progress and,