Case Report

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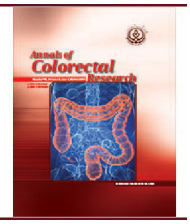

Pearls in the Appendix – Myxoglobulosis

***Vidhyachandra Gandhi*[1*], *Neeraj Rayate*[2], *Satish Pattanshetti*[2], *Pratik Gautam*[2]**

[1]*Department of Gastrointestinal & HPB Surgery, Ruby Hall Clinic, India*
[2]*Department of General Surgery, Ruby Hall Clinic, India*

Corresponding authors:
Vidhyachandra Gandhi,
Consultant GI & HPB Surgeon, Ruby Hall Clinic, Sasoon Road, Pune – 400001
India
Email: drgandhivv@gmail.com

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Abstract

We report a rare variant of mucocele of the appendix. An elderly gentleman was admitted with symptoms and signs of acute appendicitis. He underwent laparoscopic converted open appendicectomy. On exploration there were pearls or fish egg like globular material along with jelly in and around the appendix. On examination of the specimen a differential diagnosis of hydatid cyst or pseudomyxoma was considered. On microscopic examination, the globules consisted of eosinophilic laminations of mucin surrounding an amorphous granular core. After histopathological examination, our case was diagnosed as myxoglobulosis of the appendix.Acute appendicitis is a common clinical problem and appendisectomy a regular surgery. The clinician should be aware of this entity of myxoglobulosis, to avoid confusion with pseudomyxoma or hydatid cyst. Presence of typical fish eggs or pearls like globular structures in and around the appendix is suggestive of myxoglobulosis. Appendisectomy is curative treatment for patients with myxoglobulosis of appendix and there are no reports of its recurrence.

Keywords: Gastrointestinal surgery, General surgery , chronic abdominal pain, acute appendicitis

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Introduction

Appendiceal mucocele is not an uncommon finding during a routine elective appendectomy. It has an incidence of 0.2 - 0.7% amongst all appendectomy specimens (1-3), and occurs secondary to accumulation of mucus within the appendiceal lumen. Myxoglobulosis, a rare variant of appendiceal mucocele, may be asymptomatic or may present with non-specific chronic abdominal pain or symptoms simulating acute appendicitis (4, 5). We report an elderly gentleman who was diagnosed with myxoglobulosis of the appendix, otherwise known as caviar appendix, on histopathological examination after appendectomy.

Case Presentation

A sixty-two-year-old gentleman reported to the emergency medical services with abdominal pain, vomiting, and fever. The pain was in the form of a continuous dull ache in the lower abdomen,

predominantly in the hypogastrium and right iliac fossa. He had few episodes of non-bilious vomiting and intermittent fever spikes since two days beforehand. He denied a history of weight loss, loss of appetite, alteration of bowel habits, rectal bleeding, or urinary symptoms. He had similar symptoms six months ago, which were managed at a peripheral hospital with medications and without investigations. He was a diabetic on oral hypoglycemic medications.

Investigations

The ultrasound scan of the abdomen and pelvis revealed a dilated and thickened appendix with minimal free fluid, suggestive of acute appendicitis. His laboratory profile (hemogram, serum electrolytes, serum creatinine, blood sugar levels, and urine routine) was normal.

Treatment

He was posted for laparoscopic appendectomy under general anesthesia. On laparoscopy, the omentum and terminal ileum were adherent to the anterior abdominal wall; as we were unable to proceed, the decision was taken to convert to open surgery. The abdomen was opened with a McBurney's incision. On exploration and adhesiolysis, a short 6 x 3 cm turgid appendix was noted in the retrocecal position. Many globular egg-like structures were noted over the appendix (Figure 1a). The mesoappendix was transected via bipolar coagulation. The stump was suture ligated with 3/0 polygalactin, and the specimen was delivered. On opening the specimen, some jelly-like material along with egg-like globular structures were seen (Figure 1b). We were confused and thought that we were dealing with a hydatid cyst or pseudomyxoma. Peritoneal lavage was given and the incision was closed after placing a Jackson-Pratt drain. Microscopic sections from the appendix and periappendicular area showed mucosal ulcerations and rupture lined by granulation tissue, hemorrhage, mucinous material, and dense acute-on-chronic inflammatory infiltrate comprising of neutrophils, lymphocytes, eosinophils and histiocytes (Figure 2a). The surrounding mucosa showed hyperplastic, benign, columnar epithelium with infiltration of lamina propria by eosinophils and mild fibrosis. Sections from the mucinous globules showed an amorphous granular core surrounded by laminations of mucin (Figure 2b). There was no evidence of malignancy.

Outcome & Follow Up: The patient's recovery was uneventful and he was discharged on the third post-operative day. He remained asymptomatic at six-months follow-up.

Discussion

Appendiceal mucocele is commonly encountered during appendectomy, whereas myxoglobulosis is a rare variant of appendiceal mucocele accounting for 0.35% to 0.8% of all mucoceles (4, 5). Collins reported a single case of myxoglobulosis amongst 50,000 appendectomy specimen (6). Myxoglobulosis of the appendix occurs mostly in females who are in their 6th to 7th decades, but has also been reported in patients as young as 18 years of age (5, 7). The first report of myxoglobulosis was by Latham in 1897 (4, 8, 9). Since then, there have been a few reports of similar cases numbering less than 80 in 122 years. Myxoglobulosis due to mucocele is also seen in the oral cavity and has even been reported in the larynx (10, 11). It is also sometimes referred to as 'caviar appendix' or 'globoid body' due to the multiple globules that appear like fish eggs (9, 12). It is unclear how the transformation of mucin into globular bodies of myxoglobulosis occurs, although various authors have proposed various hypotheses: (a) there is formation of a core, acting as a nidus for concentric deposition of mucin, as the initiating event in the pathogenesis of the globules; (b) small mucinous masses are putatively formed in dilated glandular crypts after origination of bacterial and necrotic epithelial debris; (c) globules are organizing masses of mucin and granulation tissue

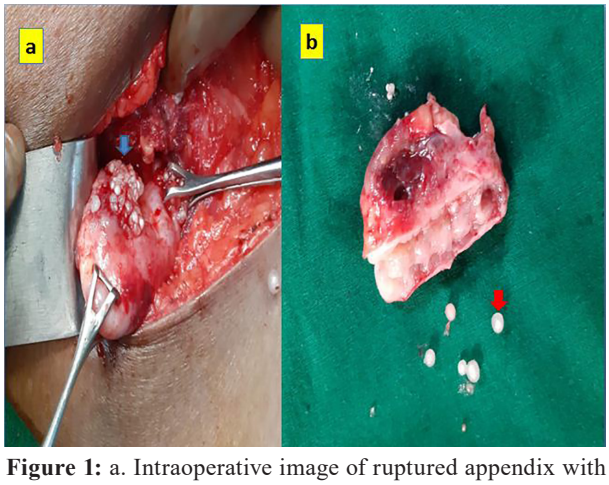

Figure 1: a. Intraoperative image of ruptured appendix with pearly globules marked with a blue arrow; b. Cut specimen of appendix showing jelly-like material inside with multiple globules marked with red arrows.

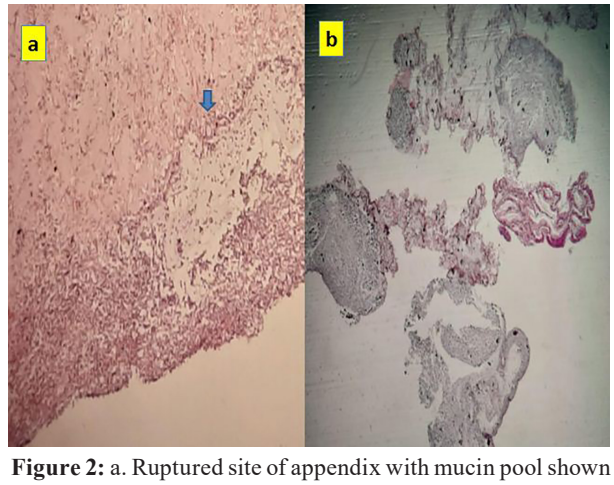

Figure 2: a. Ruptured site of appendix with mucin pool shown by the blue arrow; b. Mucinous globular structures consisting of eosinophilic laminated material with granular cores.

that originate in and break off from the appendiceal wall, subsequently undergo necrosis (4, 13, 14). Microscopic examination of the globules in our case revealed amorphous granular cores surrounded by faintly eosinophilic laminations of mucin (Figure 2b). The pathogenic mechanisms of myxoglobulosis are similar to that of an appendiceal mucocele, which include (a) partial or complete obstruction of the lumen of the appendix and (b) continued mucin production by a normal or altered epithelium (5).

Complications of myxoglobulosis can be intussusception, bleeding, perforation, peritonitis and pseudomyxoma peritonei, which are similar to that of mucocele (4, 7, 15, 16). Even a case of Amyand's hernia with myxoglobulosis has been reported (17). Association with a ruptured peptic ulcer, ruptured diverticulum, inguinal hernia, and imperforate occlusive membrane have also been reported (5, 16, 18, 19).

Diagnosing a case of myxoglobulosis preoperatively is extremely difficult. Almost all reported cases have been diagnosed incidentally intraoperatively or on autopsy (4, 8, 12, 15). Most of them did not give rise to symptoms or else mimicked acute appendicitis (16, 20, 21). Alcalay et al in 1985 published barium enema and lumbar spine radiographs of myxoglobulosis (12). Computed tomography and ultrasonography can be used for diagnosing myxoglobulosis preoperatively but are rarely picked up (22). The treatment for myxoglobulosis is appendectomy. However, a right hemicolectomy may be required in the case of rupture or any suspicion of malignancy (7-9, 21).

Acute appendicitis is a common clinical problem and appendectomy is a regular surgery. The clinician should be aware of this entity of myxoglobulosis to avoid confusion with pseudomyxoma or hydatid cyst. The presence of typical fish eggs or pearl-like globular structures in and around the appendix is suggestive of myxoglobulosis. Appendectomy is curative.

Conflict of Interests: None declared.

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