Case Report

Appendiceal Mucocele: Report of a Case and Brief Review

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Abstract

Appendiceal mucocele (AM) is a lesion of the appendix, characterized by a gross enlargement of the appendix from accumulation of mucoid substance within the lumen. It may be caused by benign or malignant diseases. AM is a rare condition, encountered in only 0.1-0.4% of all appendectomies with a female predominance. Clinically it can remain either asymptomatic for a long time, or it can manifest with abdominal pain that can be associated with the presence of a palpable mass. The most common clinical manifestation is pain in the right iliac fossa. Preoperative diagnosis is rare, while it is more frequently intraoperative. Therapy is fundamentally surgical. We report the case of a 10-year male with right lower abdominal pain, anorexia and low-grade fever. A complete blood count, urine routine examination and ultrasonogram of the whole abdomen was advised to the patient.

Keywords: Right lower abdominal pain, Appendiceal mucocele, Surgical management.
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Introduction

Mucocele of the appendix is a descriptive term that refers to dilation of the appendiceal lumen because of mucin accumulation and is based on the gross or macroscopic appearance of the appendix[1,2]. It is an uncommon condition, accounting for 0.2% to 0.7% of all appendectomy specimens[3-5]. Mucocele formation is primarily due to epithelial proliferation, which can be either benign or malignant. Much less frequently, inflammatory or obstructive causes, including appendicitis and obstruction by a fecalith or appendicolith, are the causes of mucocele formation[1]. Appendiceal mucocele can be classified into four histological types: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma[6,7].

The mucocele of the appendix was first identified by Rokitansky. The mucosa of appendix is lined by columnar epithelium containing goblet cells more than that of the colon. As a result of this, most appendiceal epithelial tumours are mucinous and start as mucocele[8]. The clinical progression of the disease does not present a distinct pattern and often remains asymptomatic. In approximately 50% of cases, it is incidentally found during radiological or endoscopic evaluations, or during surgery. It often progresses without symptoms. In approximately 50% of cases, it is found accidentally during radiologic or endoscopic examinations, or during surgery. A patient may present with clinical symptoms such as pain in the right lower quadrant of the abdomen, a palpable abdominal mass, nausea, vomiting, weight loss, gastrointestinal bleeding, and signs of intestinal intussusception[9-11].

Case Presentation

A 10-year-old male child hailing from Chandina, Cumilla came to paediatrics surgery department with the complaints of sever right lower abdominal pain and anorexia for 1 day. He also gave the history of fever for 2 days. His bowel and bladder habits were normal. On abdominal examination there was severe tenderness in McBurney's point. There was no palpable mass in the abdomen. His temperature was 101° F. A complete blood count (CBC), routine examination of urine and ultrasonogram (USG) of whole abdomen was advised to the patient.

***Laboratory investigations finding**:* In CBC report there was leukocytosis with increased neutrophil count. Other parameters were within normal range. Routine examination of urine was also normal. The USG of whole abdomen showed a tubular inflamed lesion in the right lower quadrant.

Diagnosis and management: Clinically and from investigation reports, he was diagnosed with a case of acute appendicitis. After clinical diagnosis we decided to do appendicectomy under general anesthesia. A large incision was made in the right iliac fossa through McBurney's point. During the exploration of the right lower abdomen, we found huge intra-abdominal clear mucinous fluid and the lesion was similar to inflamed small intestine. After the appendicectomy it looked like appendiceal mucocele. The length of the organ was 10 cm and diameter was 2.4 cm. Surgery was completed, and the specimen was sent for histopathology.

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On the first post-operative day, the patient became asymptomatic. His CBC report was within normal range. On the third post-operative day, the patient was discharged with advice.

Follow-up and histopathology report: On the tenth post-operative day patient came to follow-up. His stitch was good without any sign of infection. His histopathology report showed acute gangrenous inflammation of appendix. Some segments of specimen were hyperplastic with mucous retention which closely resembled the characteristics of appendiceal mucocele.

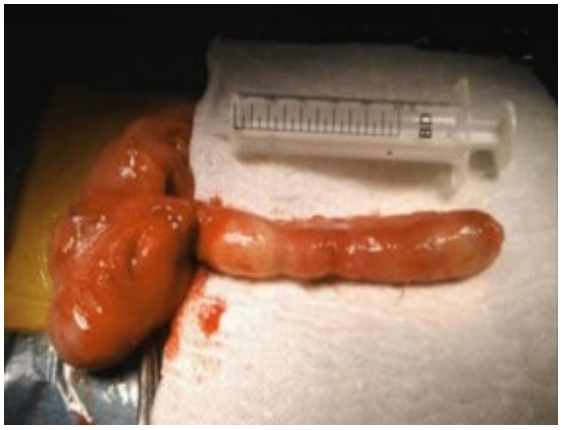

Figure-1: Appendiceal mucocele during the appendicectomy operation

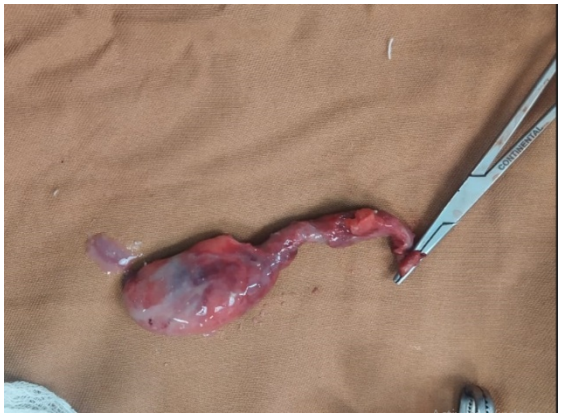

Figure-2: The Appendiceal mucocele after appendicectomy

Discussion

Appendiceal mucocele (AM) does not have a typical clinical picture. Sometimes the patient may experience pain in the lower right quadrant of the abdomen, which can lead a surgeon to misdiagnose it as acute appendicitis[7-9]. It is crucial to distinguish between these two conditions before surgery and choose the appropriate surgical approach. If not managed correctly, the mucocele can worsen, with epithelial cells potentially spreading into the peritoneal cavity, leading to pseudomyxoma peritonei, a condition associated with high mortality[12].

Due to the nonspecific nature of the disease, preoperative diagnosis is challenging. The lesion may be identified by abdominal CT scan or by ultrasound examination or colonoscopic means. CT scan findings of a mucocele typically show a distinct, well-defined, round, thin-walled cystic mass. Calcification is observed in 50% of cases[13]. Enhancing nodules in the mucocele wall are likely to suggest cystadenocarcinoma[14,15]. It is important to note that mucoceles less than 2 cm are rarely malignant. Larger mucoceles (6 cm or more) are linked to cystadenoma or cystadenocarcinoma and have a higher risk of perforation, with a rate of 20%[16]. Ultrasonography typically reveals a cystic, encapsulated lesion, tightly adhered to the cecum. The internal echogenicity varies, which is associated with the density of the mucus. In some patients, multiple echogenic layers along the dilated appendix produce the appearance of “onion skin-like” circles and are pathognomonic for mucocele[17]. Colonoscopy may show a pathognomonic image, the ‘sign of the volcano’, i.e., an erythematous, soft mass with a central crater, from which mucus is discharged[18].

Perforation leads to the spread of mucoid material into the peritoneal cavity. This material may be acellular or contain cells exhibiting varying degrees of dysplasia. Unlike colorectal cancer, appendiceal mucoceles are typically less aggressive and rarely metastasize to the lymph nodes or liver[13]. Surgery is recommended in all cases of mucocele. The type of surgical treatment is related to the dimensions and to histology of the mucocele[18]. Appendectomy is used for simple mucocele or for cystadenoma, when the appendiceal base is intact[19]. Cecal resection is indicated for cystadenoma with a large base and a right hemi-colectomy is recommended for cystadenocarcinoma[20].

Conclusion

Patients with mucocele of the appendix can be present with confusing symptoms and indeed may be asymptomatic. A preoperative diagnosis is crucial for guiding effective management and minimizing both intra-operative and post-operative complications. This case highlights the importance of considering appendiceal mucocele in differential diagnoses and the need for careful management to ensure favorable patient outcomes.

Conflict of Interest

The authors declared that they have no conflicts of interest.

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