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Mucocele of the Appendix Causing Acute Appendicitis with a Rare Phenomenon- A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Acute appendicitis is a very common emergency encountered in surgical practice. The common cause of acute appendicitis is obstruction to the lumen of the appendix due to fecolith, lymphoid hyperplasia or worms. Very rarely an uncommon cause of appendicitis is encountered during surgery. One such rare cause of appendicitis due to a mucocele of the appendix was encountered in our surgical practice. The mucocele also unexpectedly presented with multiple spherical balls of mucin appearing like Sago Pearls, which by itself is a very rare phenomenon. It is also termed as myxoglobulosis a morphological variant of mucocele.

Case Report: A 62 years old male patient presented with clinical features of acute appendicitis. An ultrasound abdomen was done, which showed a dilated and elongated appendix with probe tenderness. Due to the enlarged size of the appendix, surgery was planned by open method. The appendix was removed intact and bench dissection was done. Mucin was present inside the lumen of the appendix along with multiple spherical balls of mucin similar in appearance to "Sago Pearls", usually used in cooking. The histopathological type was simple mucocele or retention cyst, which is not a common presentation.

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Conclusion: Mucocele of appendix presenting as a cause of acute appendicitis is very rare. Moreover, mucocele presenting like spherical mucin balls similar to "Sago Pearls" is by itself, a very uncommon phenomenon. We present this manuscript for the rarity of the presentation of the mucocele causing acute appendicitis with an unusual Sago Pearls like appearance, which is an unusual phenomenon.

Keywords: Mucocele; sago pearls; acute appendicitis; spherical balls.

1. INTRODUCTION

Acute appendicitis is a common emergency in surgical practice. The causes are obstruction to the lumen of the appendix due to fecolith, lymphoid hyperplasia or worms. It is unusual to encounter a rare cause causing acute appendicitis. One such rare cause of appendicitis was encountered in our surgical practice. It was a mucocele of the appendix presenting as acute appendicitis [1]. Appendicectomy was done in the open surgical method. Inside the lumen of the appendix, along with mucin, there was an unusual presence of multiple spherical balls of mucin similar in appearance to "Sago Pearls", usually used in cooking. The histological type was simple mucocele or retention cyst, which is also not a common presentation. We present this manuscript for the rarity of the presentation of acute appendicitis due to an unusual cause of mucocele, manifesting with a rare appearance similar to "Sago Pearls".

2. CASE REPORT

A 62 years old man presented with acute abdominal pain of 2 days duration to the emergency room in our hospital. He had pain in the right iliac fossa for the past 2 days. The pain was intense, continuous and not radiating. He also had nausea and vomiting. He had a painkiller injection once in a nearby clinic with minimal relief. The patient is not a known diabetic or hypertensive. He has not undergone any surgeries in the past. On examination of the abdomen he had tenderness in the right iliac fossa and Mcburney's point. He did not have the classical guarding, rigidity and rebound tenderness. The clinical suspicion was acute appendicitis.

The blood investigations were normal. An abdominal ultrasound scan was done subsequently. The ultrasound scan of the abdomen revealed a dilated fluid filled structure of size 5.6 x 2.3cms in the right iliac fossa suggestive of the appendix (Fig. 1). Probe tenderness was present in the Mcburney's point suggesting acute appendicitis. In view of the clinical picture and with sonological evidence, surgery was decided as the ideal treatment. Laparoscopic appendicectomy was the usual mode of surgical treatment. But the patient was planned for open surgery, since the suspected appendix was very much dilated and enlarged.

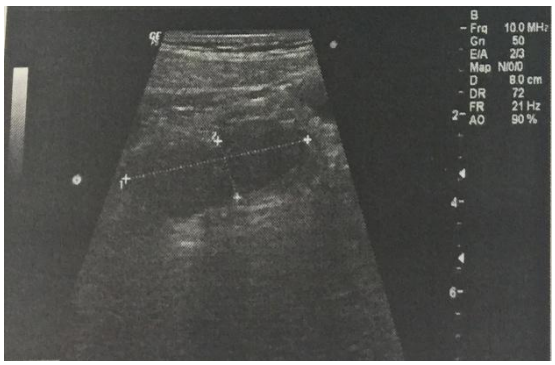

Fig. 1. Ultrasound scan image of the mucocele of the appendix

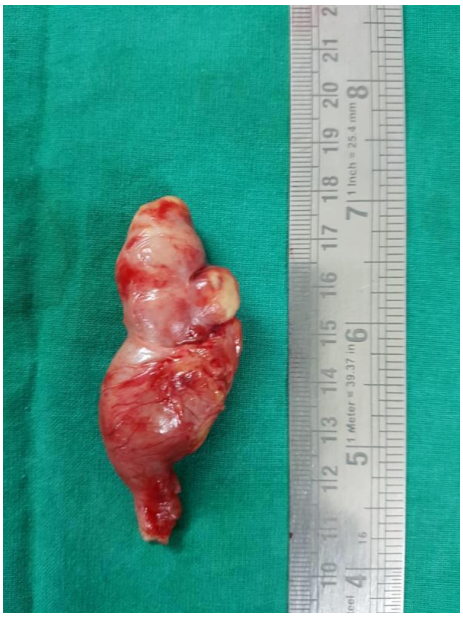

Fig. 2. Dilated and fluid filled appendix specimen after appendicectomy

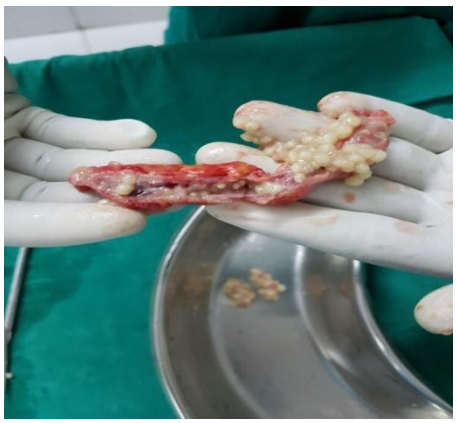

Fig. 3. Mucin laden "Sago Pearl" like spherical balls (red arrow) in the lumen of the appendix

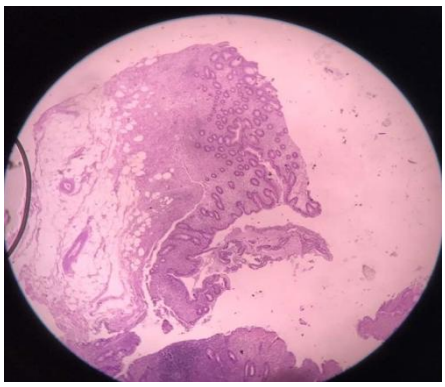

Fig. 4. Histopathology examination revealing the mucin pool in the submucosa and serosa

During surgery the appendix was found to be inflamed, grossly enlarged and oblong shaped with size 7 x 3cms confirming the findings of the ultrasound scan of the abdomen (Fig. 2). The base of the appendix and the caecum was normal. Mucocele of the appendix was suspected by the appearance. The distal ileum, ascending colon and the mesentery was normal. There were no pericolic or mesenteric nodes. Appendicectomy was done with utmost care to avoid spillage of the luminal contents. After surgery, bench dissection was done and the lumen of the appendix was opened. Multiple spherical mucin ball like structures, soaked in the mucus were seen (Fig. 3). They appeared like "Sago Pearls" which is usually used in cooking in the southern parts of India. This was a peculiar feature very rarely seen in mucocele. The specimen was sent for histopathological examination. The patient recovered well in the postoperative period and was discharged on the 3rd post-operative day. In the histopathological report, macroscopically there were no nodes and no evidence of perforation of the appendix. Microscopically, there was dense inflammatory cell infiltrates in the mucosa, submucosa and serosa. There were mucinous deposits in mucosa and submucosal layers (Fig. 4). There was no evidence of dysplasia or malignancy in the appendiceal tissues. The histopathological report confirmed it as a mucocele of the appendix, which was a simple mucocele or retention cyst type as per the classification.

3. DISCUSSION

Mucocele of the appendix is a rare condition. It was first described by Rokitansky [*2*]. It is characterized by dilatation of the lumen due to accumulation of mucus secreted by the goblet cells. The incidence is 0.2% - 0.7% of all appendicectomy specimens. The mucocele can be benign or malignant. The histological classification is of 4 types: retention cyst or simple mucocele, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma [3]. The mucinous cystadenoma is the commonest (52%), followed by mucinous hyperplasia (22%), retention cyst (18%) and mucinous cystadenocarcinoma (10%) in order [4]. Mucocele are also differentiated by size. Those that are less than 3 cm are rarely malignant and if more than 6cm has higher incidence of malignancy and tends to rupture [5]. Mucocele commonly presents asymptomatically and identified either during radiological investigations or incidentally during surgery. Clinical symptoms are present rarely and it may be pain in right iliac fossa, gastrointestinal bleeding, vomiting, intussusception of intestines and weight loss [6]. Rarely the mucocele can rupture and produce pseudomyxoma peritonei for which treatment is very difficult [7]. Myxoglobulosis is a rare morphologic variant of appendiceal mucocele characterized by intraluminal mucinous globules of the appendix [8]. The incidence of myxoglobulosis constituted 0.35% to 0.8% of appendiceal mucocele [9]. It develops due to obstruction to the proximal appendiceal lumen with continued production of mucin distally [10]. The most frequent complication of myxoglobulosis is pseudomyxoma peritonei [11].

In our patient, the presentation was with acute abdominal pain without classical signs of acute appendicitis. Ultrasonogram scan of the abdomen revealed the dilated, fluid filled appendix. Hence the decision to go for open

surgery instead of laparoscopy surgery was made. During the surgery, the appendix was found to be grossly dilated and so mucocele was also suspected due to the presence of fluid in the ultrasonogram scan. As a result, appendicectomy was done with utmost care, without spillage of the contents. The unusual feature noted in our patient was the presence of multiple mucin laden spherical balls like structures similar to “Sago Pearls”. These spherical structures are not mentioned as a type in the classification of mucocele. But, it is termed as myxoglobulosis by von Hansemann in 1914, which is a rare morphologic variant of mucocele [12]. This “Sago Pearls” like appearance of the mucin is not seen in most of the literature reviews and is a very unique phenomenon noted in our patient. The histopathological presentation in our patient is of simple mucocele or retention cyst type, which has occurred due to the obstruction of the lumen of the appendix by the thick mucin. There is no evidence of dysplasia or malignant cells in the histopathology report. . The patient was followed up for two years with ultrasound abdomen once a year. There was no evidence of ascites or mesenteric adenopathy in the ultrasound abdomen scan.

4. CONCLUSION

Mucocele of the appendix is a rare cause of appendicitis. Ultrasonogram scan of the abdomen gives a clue to the diagnosis, but if in doubt a CT scan abdomen should also be considered to confirm the diagnosis. Laparoscopic surgery can be started initially but conversion into an open procedure should be done without hesitation to retrieve the specimen intact without spillage of mucin. Mucocele with mucin appearing as “Sago Pearls” which is also termed as myxoglobulosis, is a very unique phenomenon. We present this patient, due to the rarity of the mucocele of the appendix, presenting as acute appendicitis and also due to the presence of the “Sago Pearls” like mucinous balls, which was a rare feature encountered.

CONSENT

As per international standard or university standard, patients’ written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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