

# Giant Appendiceal mucocele: A case report

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## Abstract

Appendiceal mucocele is a rare disease. It is resulted from accumulation of mucoid material due to obstruction of the lumen. It has multiple pathologic forms and classification is controversial. Recently a consensus reached on the pathologic reporting checklist and classification which is helpful for standardization of diagnosis, management plan and comparison of different centers outcome.

**Case:-**We presented a 41 year Ethiopian male patient, who has presented with vague abdominal pain, after imaging study he was diagnosed with appendiceal mucocele. Patient was admitted and counseled about the possibility of Right hemi-colectomy.

Laparotomy performed through a vertical mid line incision. Intraoperative finding was tense pear-shaped 10x6cm appendicular mass which contain fluid with 3cm fibrous base with no luminal communication with the cecum and no surrounding tissue infiltration or dissemination of the content to the surrounding (figure2), appendectomy was performed and specimen sent for histopathologic examination. His post-operative stay was smooth and patient was discharged on the 2<sup>nd</sup> postoperative day. Histologic examination revealed low grade appendiceal mucinous neoplasm. The presentation, imaging findings, management and histological appearance of the case discussed with review of literature

**Conclusion:-** Appendiceal mucocele is a rare disease .A correct diagnosis before surgery is very important for the selection of surgical technique to avoid severe intraoperative and postoperative complications. Imaging studies should be used extensively for this purpose.

## **Introduction**

Appendiceal mucocele is a rare case accounting for about 0.2 to 0.7% of appendectomies. It is an obstructive dilatation of the appendix caused by the intraluminal accumulation of mucoid material. The disease was described by Rokitsansky in 1842, but the classification mucinous tumor of the appendix is controversial to date. (1). Mucocele of the appendix is dangerous because if ruptured it causes Pseudomyxoma peritonei (PMP) which is associated with significant mortality & morbidity(1). In this paper we present a case of huge appendicular mucocele. Clinical presentation, imaging investigation and pathologic features are discussed with literature review. We used the current classification of Pseudomyxoma peritonei (PMP) to describe the pathologic diagnosis. (2)

Pathologic result will govern further treatment plan and follow up of the patient.

Key words: - Appendix, appendiceal mucocele, Mucinous neoplasm, Mucinous adenoma

## **Case report**

A 41 year old male patient referred from a private clinic to our hospital with complaining of abdominal pain of 10 month duration .the pain was vague and non-localized. At presentation his vital sign was within normal range & he had mild right lower quadrant abdominal tenderness. His white blood cell count, organ function test, & Urine analysis was all within the normal range. He came with an Abdominal ultrasound which showed a 5X6cm well defined, blind ended echo lucent lesion in RLQ with the index of appendicular mass to rule out mucocele and recommended to have CT scan. Contrast-enhanced computed tomography (CT) was ordered and the result showed ~46X83mm retrocecal tubular homogenous, hypo dense & non enhancing mass with close proximity with the caecum. Conclusion was appendiceal mucocele (Figure 1).

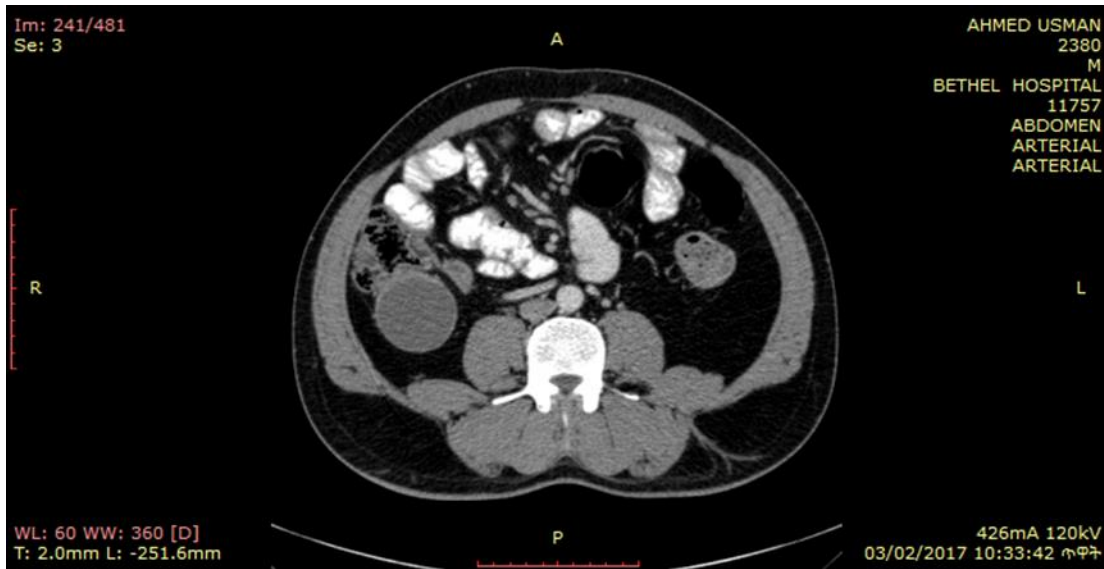


Figure 1 CT scan showing 4.6x8.3cm homogenous, hypo-dense and non-enhancing mass with close proximity to the cecum (white arrow head)

Patient was admitted to surgical ward private wing and counseled about the possibility of Right hemi-colectomy. On the next day of the admission laparotomy performed through a vertical mid line incision. Intraoperative finding was tense pear-shaped 10x6cm appendicular mass which contain fluid with 3cm fibrous base with no luminal

communication with the cecum and no surrounding tissue infiltration or dissemination of the content to the surrounding (Figure2), appendectomy was performed and specimen sent for histopathologic examination. His post operative stay was smooth and patient was discharged on the 2<sup>nd</sup> postoperative day.

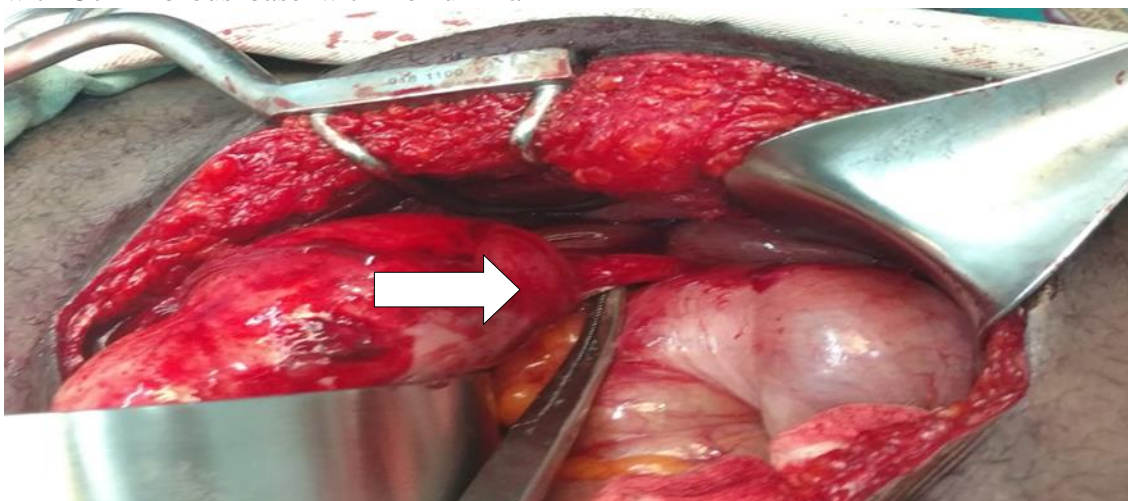


Figure 2 An intraoperative picture showing a distended appendix with long and fibrous base (black arrow head) and healthy looking cecum (white arrow head)

The histopathologic examination revealed cyst wall fragment lined by epithelial cells featuring low-grade nuclear atypia, acellular mucin dissecting through the wall, submucosal fibrosis, and loss of muscularis mucosa.

And the final diagnosis was in agreement with the radiologic impression of mucocele. (Figure 3a - 10x; 3b - 40x)

The patient was followed for 6months and he had uneventful course.

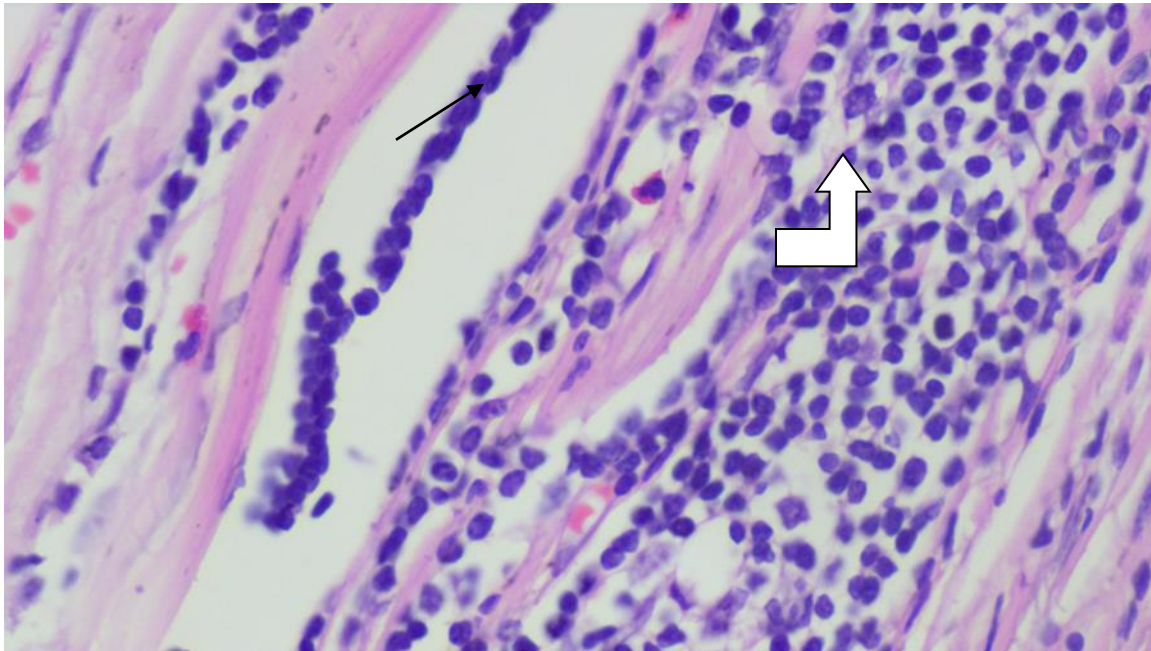


Figure 3:-Cyst wall fragment lined by epithelial cells featuring low-grade nuclear atypia, acellular mucin dissecting through the wall, submucosal fibrosis, and loss of muscularis mucosa (Figure 3- 40x)

## Discussion

Appendiceal mucocele refers a group of lesions characterized by a distended, mucous filled appendix. It affects both sexes between 5<sup>th</sup> and 7<sup>th</sup> decades of life. (1) Appendiceal mucocele accounts for only 8% of primary Appendiceal tumors. Most tumor types seen in the appendix are mucous producing; but the excessive production of mucus by adenomatous

tumors leads to the formation of mucocele. (1)

It is one of the disease entities where preoperative diagnosis is difficult, as most cases are asymptomatic or lack specific clinical symptoms. It is an incidental finding during laparotomy or radiological evaluation or endoscopic procedures (3). Clinical presentation includes right lower quadrant abdominal

pain, intermittent colicky pain, lower abdominal mass, bowel obstruction, anemia, weight loss and chronic abdominal pain (3)

The threshold for performing imaging studies in patients with suspected appendicitis and atypical disease presentation should be low, which will help for planned intervention and avoiding complications and the need for relaparotomy (4).

Ultrasound(US) and Computed tomographic Scan (CT) findings suggestive of mucocele of the appendix is described well in the literatures. The US findings may be variable depending on the content but the presence of onion skin sign is considered specific (5). Features suggestive of Appendiceal mucocele computed tomography include cystic dilatation of the appendix, luminal diameter of greater than 1.3cm and mural calcification; The presence of calcification differentiates it from appendicular abscess (5).

There is no consensus regarding the optimal management, while surgery remains the only known potential curative treatment. But still laparoscopic approach is not advised due to risk of manipulation which leads to rupture and PMP. (3)The extent of surgery ranges from appendectomy, typhlectomy, and right hemicolectomy to cytoreductive surgery; and it depends on the following findings:-Involvement of the base, integrity of the wall, and histopathologic examination of the cause of the mucocele (5)

We performed appendectomy with removal of the mesoappendix as the base was long (>2cm) and not involved in the dilatation.

Although widely used the term mucocele is inherently imprecise and inconclusive

of both benign and malignant lesions. Histopathologic level the diagnosis is not usually straight forward and the terminology has not been completely settled. There are authors who are suggesting to continue using the term mucocele for reasons that being deeply rooted in the literature, its meaning is understandable for all and it is a simple descriptive term, which enables the surgeon or radiologist to report on the lesions before pathologic examinations(4) .

The current consensus for classification and pathologic reporting of Pseudomyxoma Peritonei and associated appendiceal neoplasia has developed a checklist for the pathologic reporting of PMP and Appendiceal mucinous neoplasm. The agreed classification will be able to bring better diagnosis and management plan for individual patients and allow meaningful comparison among different centers. (2) We used both terms to address common understanding while maintaining the exact pathologic description for future pathologic review of similar cases.

In conclusion, appendiceal mucocele is a rare disease .A correct diagnosis before surgery is very important for the selection of surgical technique to avoid severe intraoperative and postoperative complications. Imaging studies should be used extensively for this purpose. Pathologic result will govern further treatment plan and follow up of the patient.

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