



## MUCOCELE OF APPENDIX: AN ELUSIVE HISTOPATHOLOGIC DIAGNOSIS

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**ABSTRACT** Appendiceal mucocele is an incidental finding with an incidence rate of 0.2-0.4% of all appendectomy specimens. It can be asymptomatic or clinically present with appendicitis-like symptoms. It is defined as dilatation of the appendix which can be either benign or malignant in nature and spontaneous or iatrogenic rupture of mucinous appendiceal tumors may lead to spread to the peritoneum resulting in a condition known as pseudomyxoma peritonei. Here we present a case of 32 year old male with complaints of pain in the right iliac fossa since one day. Clinical suspicion of acute appendicitis was established and patient was operated laproscopically for the same. On histopathological examination, the diagnosis of appendiceal mucocele was established based on gross and microscopic findings.

**KEYWORDS** : Appendiceal Mucocele, Pseudomyxoma peritonei, Mucinous neoplasms

## INTRODUCTION

Appendiceal mucocele is a progressive dilatation of the appendix from intraluminal accumulation of mucoid substance usually resulting from either benign (hyperplasia or mucinous cystadenoma) or malignant (mucinous cystadenocarcinoma) epithelial proliferation<sup>1</sup>. The appendiceal mucocele first described by Rokitsky in 1842<sup>2</sup>, is an infrequent disease entity having a clinical picture that resembles acute appendicitis or is discovered as an incidental surgical finding. The incidence is 0.2 –0.4 % of all appendectomy specimens and is most likely seen after the age of 50 years, with female preponderance<sup>3,5</sup>.

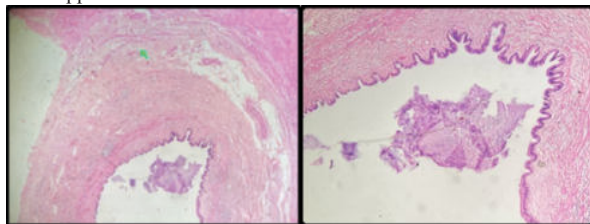
## CASE REPORT

We present a case of 32-year-old male referred by Department of Surgery, admitted with chief complaints of pain in abdomen radiating towards the umbilical region since one day. The appendix was removed laproscopically and submitted for histopathological examination.

Gross examination: The appendix was tubular, soft to firm, measuring 5 x 2 x 2 cm. On cut opening, a single cystic structure measuring 3 x 1 cm was noted which was filled with mucinous material (Fig 1).

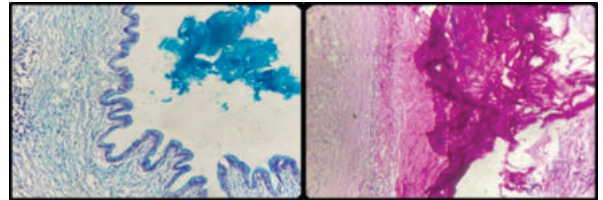


**Fig 1:** A single cystic structure measuring 3 x 1 cm noted in the lumen of the appendix.



**Fig 2:** Lumen of the appendix is cystically dilated and contains abundant mucin (H&E stain 10X)  
**Fig 3:** Appendiceal wall lined with monolayered mucin secreting columnar epithelium with apical mucin (H&E stain 40X)

Microscopic examination: H&E stained sections studied from the appendix showed mucosa, submucosa, muscularis and serosa. The mucosa showed crypt loss and blunting of lining epithelium at few places. Sections from the dilated cystic structure showed lining epithelium comprised of tall columnar cells with abundant intracellular mucin and apical snouts. Sections stained from Alcian Blue and Periodic Acid Schiff stain showed presence of abundant extracellular as well as intracellular mucin.



**Fig 4:** Alcian Blue stain (40X) **Fig 5:** PAS stain (40X) stained positive for both extracellular and intracellular mucin.

## DISCUSSION

Recent reports show a male predominance (4:1), suggesting that the sex distribution is changing<sup>6</sup>. The incidence of mucocele of the appendix in India is 0.15%, which is consistent with the worldwide incidence<sup>4</sup>.

Some regard this lesion as benign, a result of obstruction of the proximal lumen by mucin; others believe it to be a neoplasm of the appendix<sup>7</sup>. The stained sections show crypt loss and blunting of the lining epithelium at focal places in the mucosa<sup>8</sup>. The cystic structure shows tall columnar lining epithelium with intracellular and extracellular mucin. Sections stained with Alcian blue are positive for both intracellular and extracellular mucin.

Mucocele of the appendix is divided into four pathological subgroups based on the lining epithelium histopathological appearance<sup>9</sup>. The first group consists of a simple retention cyst secondary to occlusion of the appendix by faecolith, scar tissue from previous inflammation. It has a normal or flattened epithelium, moderate luminal dilatation up to 2 cm<sup>10</sup>. The case presented falls under this group.

The second group with hyperplastic epithelium and moderate luminal dilatation: This constitutes about 20% of all mucocele of the appendix<sup>11</sup>.

The third group is benign mucinous cystadenoma: This is characterized by tubular adenomatous epithelium with varying degree of epithelial atypia. The fourth group encompasses the malignant mucinous cystadenocarcinoma; characterized by glandular stromal invasion and/or tumour cells in peritoneal implants i.e. Pseudomyxoma peritonei which sometimes resembles mucinous

carcinoma of the colon<sup>12</sup>.

## CONCLUSION

Owing to its rarity, appendiceal mucocele continues to intrigue the pathologists and clinicians alike as the diagnosis being an incidental finding due to lack of clinical symptomatology and preoperative radiological confirmation, hence it is concluded that histopathological examination of all appendicectomy specimens should be performed and considered the gold standard to avoid missing its diagnosis and avoiding any clinical negligence.

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