

# Low-Grade Appendiceal Mucinous Neoplasm; A lesser known Entity with Significant Morbidity: Case Report

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## Abstract

Mucinous dilatation of the appendix is known as a mucocele. This could be due to mucinous neoplasms of the appendix. They involve a wide variety of tumors ranging from benign to malignant tumours. Whether to consider them benign or malignant has been the subject of debate, But WHO (World Health Organisation) gave them the official name of low-grade appendiceal mucinous neoplasms (LAMN). They are difficult to diagnose pre-operatively although radiology, pelvic examination, colonoscopy and serum tumor markers may help in the diagnosis. Pseudomyxoma Peritonei is the most dreaded complication. Most of the times a simple appendectomy is sufficient but be careful to prevent the spillage of mucin out of the appendix.

**Keywords:** LAMN; Mucocele; Mucinous neoplasm; Pseudomyxoma Peritonei

## Introduction

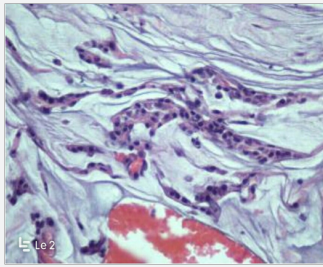
Mucocele of the appendix is a rare condition which is caused by the either mucus cysts or mucinous neoplasms of the appendix. The incidence of mucoceles is less than 2%. About 50% of the times mucoceles are diagnosed incidentally at the time of surgery or confirmed at histopathology. They may present with features of acute appendicitis like right lower quadrant abdominal pain, a tender lump, intestinal obstruction, intussusceptions, or gastrointestinal bleeding. Radiological investigations can help in diagnosis [1]. There has been a controversy about the nomenclature of mucinous neoplasms of the appendix. The 4th edition of world health organisation (WHO) has accepted the low-grade appendiceal mucinous neoplasms (LAMN) as the official nomenclature. The mucinous tumors of appendix include a wide variety of tumors amongst which the LAMN are most controversial. They could be localised to the appendix only or they can invade and perforate the appendix disseminating in the peritoneal cavity producing pseudomyxoma peritonei thus acting

like a low grade malignancy. About 15-20% of LAMN are detected incidentally in patients undergoing surgery for other diseases. The treatment is surgical removal of the appendix with care to prevent the spillage of mucin which can lead to Pseudomyxoma peritonei [2].

## Case Report

We present here a case of 39 year female who presented to us with a history of pain right iliac fossa for two months. There was no history of weight loss, loss of appetite. There was no history of discharge per vaginum or any menstrual abnormalities. Urinary and bowel habits were normal. Patient was admitted to another hospital about two months back for similar complaints where she had undergone abdominal sonography and was diagnosed with apendicular lump and was managed conservatively. Patient had no pallor, lymphadenopathy. On examination there was a tender lump of about 3x2 centimeter (cm) size in right iliac fossa. A contrast enhanced computerised tomogram (CECT)

of the abdomen and pelvis was done which revealed acute appendicitis with appendicular mass. Colonoscopy was done which revealed no abnormality. The patient was taken up for surgery. The appendix was inflamed, about 4cm in length, pelvic in position with the omentum adherent to the tip of appendix. Base of appendix was healthy. Appendectomy was done and specimen was sent for histopathological examination. No other abnormality was noted intraoperatively. Histopathological report was suggestive of low- grade appendiceal mucinous neoplasm with extravasation of mucin. Resection margins were free. Subsequently carcinoembryonic antigen levels of the patient were found within normal limit. Patient has been under follow up for last one year with no signs of recurrence (Figure 1).



**Figure 1:** Showing features of low grade appendiceal mucinous neoplasm along with extravasation of mucin.

## Discussion

Primary malignancies of the appendix are a rare entity noted only in almost 0.9-1.4% of the appendectomy specimens. They are hard to diagnose before surgery and only 50% are diagnosed at surgery. Data from a review by the national cancer institute's surveillance, epidemiology and end results (SEER) program found 0.12 cases of appendiceal malignancies per 1000000 people per year. The same data also suggested that mucinous adenocarcinoma was the most common histological type followed by adenocarcinoma, carcinoid, goblet cell carcinoma and signet - ring cell carcinoma. Incidence of mucocele of appendix is very low [3]. A cystic dilatation of the appendix due to intraluminal accumulation of the mucoid material has been known as the mucocele. Mucocele of the appendix was first described by Rokitsky in 1842 but named by Feren in 1876 [4]. They are more common in elderly females usually above 50 years of age. Usually the presentation is nonspecific and diagnosis is made at the time of surgery. A mucocele of the appendix could be formed because of retention cyst, mucosal hyperplasia, cystadenoma or cystadenocarcinoma [3]. These tumors may present as intestinal obstruction, gastrointestinal bleeding or intussusception very rarely. These tumors possess the potential of malignant spread so it would be unfair to call them benign mucinous adenomas.

There has been a controversy regarding their classification and their concomitant association with the ovarian tumors [2]. Woodruff & McDonald [5] classified the cystic mucinous neoplasms of appendix into benign mucoceles and cytadenocarcinomas grade 1 in 1940 [5]. Higa et al. [6]

classified these tumors as cystadenocarcinomas when they were associated with pseudomyxoma peritonei, and cystadenomas otherwise [6]. Carr et al. [7] classified them as adenomas, mucinous neoplasms of uncertain malignant potential and adenocarcinomas in 1995. Pai & Longacre [8] divided the mucinous neoplasms of the appendix into four groups. Group 1 were known as adenomas where the disease was confined to the appendix. These tumors never occurred after appendectomy. Group 2 tumors were associated with low grade dysplasia along with extra- appendiceal acellular mucin. These tumors rarely recurred. Group 3 tumors contained extra appendiceal neoplastic epithelium along with low grade dysplasia.

The 5 year disease free survival rate was 25%. Group 4 included mucinous cystadenocarcinomas with high grade cytology, complex architecture or invasion [8]. Misraji et al. [2] classified them as low-grade appendiceal mucinous neoplasms (LAMN), LAMN with peritoneal spread and invasive adenocarcinoma. The typical feature of LAMN is the pattern of invasion of layers of the appendix which is known as 'pushing invasion'. It can have different patterns. There may be attenuated or absent muscularis propria, frequently it may be fibrotic or hyalinised. Neoplastic epithelium growing over hyalinised or fibrotic stroma rather than lamina propria or muscularis mucosae is a feature of pushing invasion. The 4th edition of world health organisation (WHO) classification has accepted low-grade appendiceal mucinous neoplasms (LAMN) as the official nomenclature [2]. These tumors if they rupture may lead to pseudomyxoma peritonei, a dreaded complication where the gelatinous like material can fill up the whole abdominal cavity and lead to repeated episodes of intestinal obstruction [3]. In 1901, Fraenkel was the first one to find the association of mucinous neoplasm of appendix and pseudomyxoma peritonei.

Pseudomyxoma is now thought to arise from appendiceal mucinous tumors and primary ovarian origin is found rarely [2]. So a careful handling of the appendix is recommended. If at laparoscopy a mucocele of the appendix is encountered conversion to open appendectomy is recommended [3]. These tumors have association with the ovarian and colon cancers so colonoscopy and a computerised tomogram (CT) scan should be done along with the serum tumor marker levels (CEA). Finding a mucocele at the time of surgery does not mandate the performance of radical surgery like right hemicolectomy. A simple appendectomy with careful removal of the mesoappendix will suffice until there is gross involvement of the mesentery, lymph nodes or the cecum. A limited resection is good enough sometimes [9]. A recurrent and aggressive disease warrants right hemicolectomy and hyperthermic intraperitoneal chemotherapy [2].

## Conclusion

Low grade appendiceal mucinous neoplasms may present as mucoceles at the time of surgery or they are diagnosed in

appendectomy specimens incidentally. They are difficult to diagnose preoperatively. One should be careful while performing appendectomy for a mucocele. Most of the time a simple appendectomy is good enough. But a spillage of mucin into the peritoneal cavity may cause pseudomyxoma peritonei, a dreaded complication which can make the life miserable for the patient and sometimes can cause death due to intestinal obstruction. Thus the patient should be carefully followed up with CT scan, pelvic examination, colonoscopy and serum tumor markers. Disease recurrences should be treated with aggressive intent.

### Conflict of Interest

Authors declare no conflict of interests.

### Author's Contributions

- A. Rajesh Chaudhary: Contributed substantially to the Conception, design, Acquisition of data, Analysis and interpretation of data, drafting the article, Critical revision of the article and final approval of the version to be published.
- B. Kulbhushan Sharma: Contributed substantially to the Conception and design, Acquisition of data, Critical revision of the article and final approval of the version to be published
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