

ORIGINAL RESEARCH ARTICLE

Appendiceal Mucinous Neoplasm- An Institutional Study**¹Dr. Gitimadhuri Dutta, ²Dr.Sunanda Nayak, ³Dr. Radhamohan Gharei, ⁴Dr. Anupa Toppo, ⁵Dr. Binod Kumar Sahu, ⁶Dr. Sna Khatoon**

¹Associate Professor, Department of Pathology, Veer Surendra Sai institute of Medical Sciences and Research (VIMSAR), Burla, Odisha, India.

²Associate Professor, Department of Pathology, Veer Surendra Sai institute of Medical Sciences and Research (VIMSAR), Burla, Odisha, India.

³Assistant Professor, Department of Pathology, Veer Surendra Sai institute of Medical Sciences and Research (VIMSAR), Burla, Odisha, India.

⁴Associate Professor, Department of Pathology, Veer Surendra Sai institute of Medical Sciences and Research (VIMSAR), Burla, Odisha, India.

⁵Assistant Professor, Department of Pathology, Veer Surendra Sai institute of Medical Sciences and Research (VIMSAR), Burla, Odisha, India.

⁶Junior Resident, Department of Pathology, Veer Surendra Sai institute of Medical Sciences and Research (VIMSAR), Burla, Odisha, India.

Corresponding Author:Dr. Anupa Toppo

Article History: Received: 10.06.2023 Revised:07.07.2023 Accepted: 20.07.2023

ABSTRACT

Background: Appendiceal mucinous neoplasm (AMN) are rare neoplasm of appendix, accounting for less than 1% of neoplasm of appendix. These are group of epithelial neoplasm of appendix characterised by proliferation of mucinous epithelial cells with extracellular mucin and pushing invasion pattern or pushing tumour margin. It may cause Pseudomyxoma peritonei(PMP). Mucocele of appendix is the clinical term used to describe dilated appendix due to accumulation of mucin in the lumen of appendix. Recently peritoneal surface oncology group International (PSOGI) given a modified Delphi consensus on classification and diagnostic terminology used for AMN. Most of the time mucocele of appendix is accidental finding, diagnose most commonly in women in their 5th and 6th decade of life.

Methods: The present simple observational study was conducted in the Department of Pathology VIMSAR Burla, from January 2016 to May 2023 we found 8 cases of appendiceal mucinous neoplasm. It is an observational study. Data were collected from histopathology records and clinical records. We had done this case series because it is a rare neoplasm.

Result: Total 8 cases were included in the study. 7 cases were low grade mucinous neoplasm of appendix and one case was high grade mucinous neoplasm of appendix.

Conclusion: Mucinous neoplasm of appendix is usually confined to appendix and need surgical resection for better prognosis. when patient develop PMP treatment modalities are change. Mucinous neoplasm of appendix needs to follow up to prevent serious complication, like PMP. Early diagnosis can prevent fearful complication and provide good outcome.

Key words: Appendix, mucocele, low grade mucinous neoplasms[LAMNs].

INTRODUCTION

Low grade mucinous neoplasm of the appendix is a rare presentation. accounting for less than 1% of neoplasm of appendix[1,2]. These are group of epithelial neoplasm characterised by proliferation of mucinous epithelial cells with extracellular mucin and pushing invasion or pushing tumor margin.It's a heterogenous group of disease with varying malignant potential[3]. Its timely management is very important to prevent serious complications. Low grade mucinous neoplasm (LAMN) of the appendix are rare, low grade, non invasive epithelial tumor of the appendix that produces mucin. Mucocele refers to cystic dilation of appendix because of accumulation of mucin. Most of the time mucocele of appendix is incidental finding. Patient usually presents with pain in right lower quadrant, mimicking acute appendicitis[3]. Most severe complication seen in LAMN and high grade appendiceal mucinous neoplasm (HAMN) is due to rupture of appendix leading to seeding of mucin and neoplastic cells into the peritoneum result in pseudomyxoma peritonei [PMP][4]. (approximately 20% patients develop PMP) and intraperitoneal dissemination, making poor outcome and low survival rate. Recently Peritoneal Surface

Oncology Group International (PSOGI) reached a modified Delphi consensus on classification and diagnostic terminology used for appendiceal mucinous neoplasm.[5].

MATERIALS AND METHODS

The present simple observational study was conducted in the Department of Pathology VIMSAR Burla. It is an observational study. Data were collected from histopathology records and from clinical records . Archived slides of all cases of appendiceal mucinous neoplasm were studied again. Histopathological blocks were recut stained and studied. All the data of appendiceal mucinous neoplasm were collected from period between January 2016 to May 2023. We found total 8 cases of appendiceal mucinous neoplasm. This case series study was done because of its rarity.

RESULT

CASE-1

History:-A 80 years old male came to OPD, complaining of pain in right lower abdomen since last 1 month, which was not associated with vomiting. There was no history of anorexia and fever.

On Examination: Vitally patient was stable. On per abdomen examination, abdomen was soft ,a palpable mass was palpable in right iliac fossa, mild tenderness and rebound tenderness present. Guarding and rigidity was absent. On USG -Tubular, non peristaltic, non compressible structure of approximate size measuring was [10x6x5]cm noted in right iliac fossa with internal ring like echoes in the lumen. most likely suggestive of mucocele of appendix.

Intra operative findings: Omentum was found to be attached to the wall of mucocele of appendix. Adhesiolysis of attached omentum was done. Marsupialisation of cyst was done. Content of mucocele and wall of the cyst sent for Histopathological examination.

Pathology

On GROSS examination:- 2 containers received container-1 labelled as "APPENDIX"-contain greyish white, dilated, elongated cyst like structure measuring [10x6x5]cm. On cut sections -cyst contain mucinous material. cyst wall thickness was 0.5cm. focal area showing papillary like projection in lumen (3 sections are given from different areas and 1section from cut margin) .container-2 labelled as-"OMENTUM": contains fibrofatty grey white tissue measuring [5x3x1]cm (2 sections from thickened area).

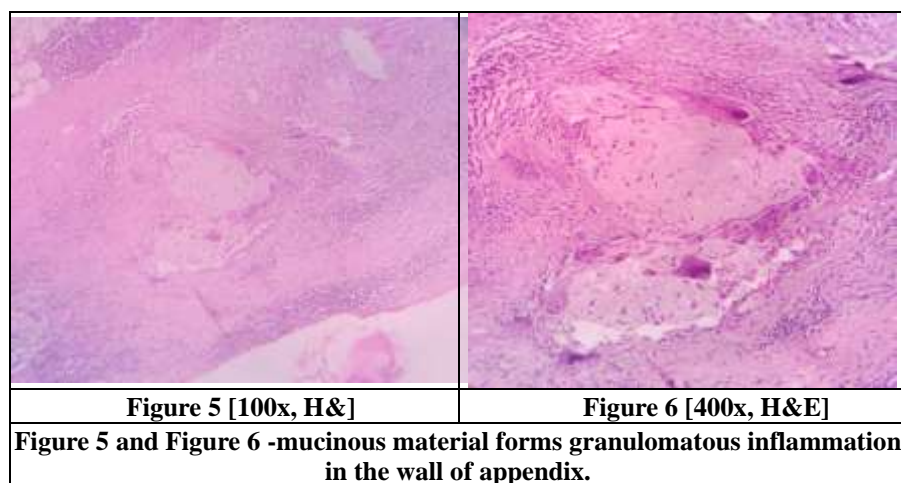
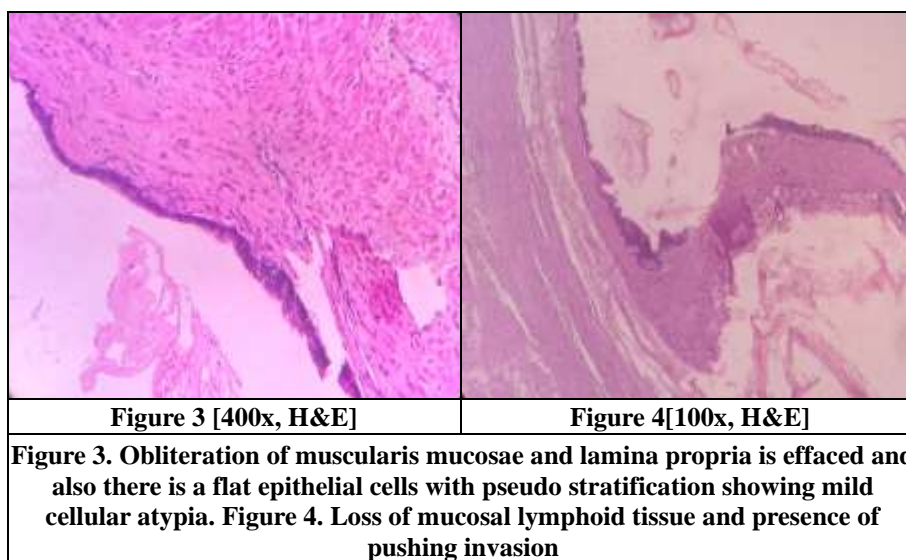


Figure 1: (case1) Gross specimen of mucocele of appendix. grey white cystically dilated tissue measuring(10x6x5)cm.



Figure 2: Cut open appendix showing mucinous material and papillae like projection in the lumen[blue arrow].

MICROSECTION reveals proliferation of mucinous epithelial cells originated from appendiceal lumen. Abundant apical mucin, elongated nuclei [columnar epithelial cells] along with mild cellular atypia. There is a flat as well as pseudo stratification of epithelial cells showing low grade cellular atypia. Mucinous material forms granulomatous inflammation in the wall of appendix. Omentum showed fibrofatty tissue only, free from metastatic deposits.

**CASE-2**

46 years male presented with abdominal pain and vomiting. On physical examination tenderness was present over right iliac fossa. Patient was complaining of nausea, vomiting and low grade fever. Ultrasonography reports revealed, cystic tubular structure measuring (7.5x2x2)cm with ring like echogenicity. Gross specimen received was appendix with mesoappendix measuring (7.5x2x2)cm, cut section showed mucinous materials within the lumen along the entire length. In the small papillae like growth seen. Multiple sections given from tip of the appendix, body and from cut margins. Microscopic examinations was done and diagnosis of Low-grade mucinous neoplasm was given.

CASE-3

50 years male presented with right sided lower abdominal pain. On physical examination tenderness over right iliac fossa was present. Ultrasonography was done with a radiological diagnosis of mucocele was given. Appendicectomy was done and the specimen was sent to Department of pathology of histopathological examination.

Gross pathology: Appendicectomy specimen with attached mesoappendix measuring(5x2.5x1.5)cm. Proximal half the appendix was swollen measuring (2.5x2.5x1.5)cm. Outer surface was greyish white and smooth . Cut section showed mucinous material. Histopathological diagnosis of Low-grade mucinous appendiceal neoplasm was given.

CASE -4

55 years female presented with abdominal pain and vomiting admitted in the surgery ward , on physical examination there was tenderness in the McBurney's point. Ultrasonography report was suggestive of mucocele of appendix. Specimen was sent for histopathological examination.

Gross examination: Appendix m (5X1.5X1) cm. Cut section showed mucinous materials. Multiple sections given from different areas along with tip and surgical margin. Microsection showed features of low grade appendiceal mucinous neoplasm.

CASE -5

80 years female complained of abdominal pain and vomiting since last one month with clinical diagnosis of acute appendicitis was advised to do ultrasonography (USG). USG diagnosis was suggestive of mucocele of appendix was given. Appendicectomy was done and the surgical specimen was sent to department of pathology.

Gross pathology- container -1 Appendix measuring (5x2x1)cm. cut section showed mucoid material in the lumen. Sections given from tip, mid portion and from cut margin. Histopathological examination was done and diagnosis of High grade appendiceal mucinous neoplasm was given. **Container-2** – labelled as omental fat- Fatty tissue measuring (1.5x1x1)cm. Microscopic examination revealed, omental fat was free from tumor infiltration.

CASE-6

35 years female presented with dull aching pain in the lower abdomen since last 2 months. Patient was admitted in the female surgery ward. On physical examination ,there was tenderness in the McBurney's point. USG was done. Diagnosis was suggestive of mucocele of appendix. Appendicectomy specimen was sent for histopathological examination. Pathological diagnosis of low grade appendiceal mucinous neoplasm was done.

CASE -7

58 years female was complaining of abdominal pain since last two weeks . Patient was admitted in the community health centre , treated there but as the pain was not subsiding ,she was referred to casualty of medical college. USG was done with a radiological diagnosis of mucocele of appendix. Appendicectomy specimen received in the Department of Pathology.

Gross examination: Appendix with attached mesoappendix measuring (7x3.3x3)cm. Cut section showed appendix was distended due to accumulation of mucin in the lumen. Histopathological diagnosis was low grade appendiceal mucinous neoplasm.

CASE-8

76 years male was complaining of abdominal pain and vomiting since last 10 days. He was admitted in the Department of Surgery for treatment. USG was done and diagnosis suggestive of mucocele was given. Surgical excision of appendix was done.

Gross examination: Appendix with mesoappendix measuring (6.5x2x2)cm. Cut section showed appendix was filled with mucoid material. Histopathological diagnosis was low grade appendiceal mucinous neoplasm.

**Figure7****Figure 8**

Figure 7- (case-4) Gross specimen of mucocele of appendix. grey white cystically dilated, cut section-mucinous material. Figure 8. (case_5) Cut open gross specimen of appendix showing mucinous material

DISCUSSION

LAMNs are more commonly diagnosed in women in their sixth decade of life. patient can be asymptomatic or symptomatic with or without serious complications. These patients can be associated with urological symptoms like haematuria, Urinary tract infection, hydronephrosis, other like torsion, volvulus, small bowel obstruction, intussusception, PMP, and ovarian metastasis is also seen. LAMN and HAMN lack infiltrative invasion but later shows high grade cytological atypia. Mucinous appendix can be benign or malignant many etiological factors play a role like accumulation of mucus in appendix, inflammatory

stenosis, faecolith, stricture, mucosal hyperplasia, endometriosis, tumor, serrated polyp of appendix etc. [6]. Mucocele of appendix less than 2cm are rarely malignant it considered as benign, if it is more than 6cm there is high risk of malignancy and can be associated with serious complication. Gross examination cystically dilated appendix, enlarged, lumen filled with mucinous gelatinous material, wall may be hyalinized, thin, fibrotic or calcified [3]. Lining of appendix may be smooth, corrugated or granular lining. Appendiceal mucinous neoplasms are classified into LAMNs, HAMNs and mucinous adenocarcinoma [7]. The term, cystadenoma is outdated. LAMN and HAMN lesion lack infiltrative invasion with low and high grades cytological atypia respectively. LAMNs show mildly enlarged nuclei and few mitotic figures. HAMNs show pleomorphic cells with high nucleocytoplasmic ratio, pleomorphic nuclei, irregular nuclear border, hyperchromatic nuclei with atypical mitotic figure. HAMNS may show micropapillary or cribriform features. Mucinous adenocarcinoma is further classified into mucinous and non-mucinous. Furthermore it classified into well differentiated, moderately differentiated and poorly differentiated with features of infiltrative invasion along with desmoplastic stromal reaction. poorly differentiated type of mucinous adenocarcinoma is more likely to produce distant metastasis along with PMP. Luminal Epithelial lining of LAMN and HAMNS may become flat or villous[5]. Because of mucin accumulation in lumen Leads to flat epithelium, atrophy of crypts and lymphoid tissue and effacement of muscularis mucosae. CEA, CA19-9, CA125 tumor markers are most commonly used, latter two are used as recurrence predictive indicator, both have diagnostic and prognostic value[6,7]. Imaging plays a very important, valuable role to diagnose disease. Visualisations of appendix on Imaging more than 15mm in diameter around the caecum and most commonly behind the caecum is highly suggestive of appendiceal mucinous neoplasm. Ultrasound usually shows distended appendix and lamellated or concentric mucinous “onion skin” appearance in right iliac fossa which is specific for diagnosis of mucinous neoplasm[8,9]. Computed tomography is gold standard diagnostic tool, It can diagnose the disease and helps to prevent most dreadful complication means pseudomyxoma peritonei. it is also helpful tool in monitoring and surveillance of patients. Computed tomography imaging reveal enlarged appendix with wall calcification and thickening. MRI-hypointense with T1 weighted and hyperintense distended appendix with T2 weighted and intraluminal or peri appendiceal mucin appears bright on T2 weighted MRI[7]. Prognosis is poor, if tumor progress to pseudomyxoma peritonei. Approximatel 2% risk of PMP is seen in cystadenoma and 20 to 30% risk of PMP is seen in mucinous cystadenocarcinoma. Due to perforation risk, biopsy is not done. surgical removal of appendix is the main stay of treatment available. it also provide specimen for pathological examination for nature of disease to prevent perforation and progression to pseudomyxoma peritonei. If malignant, then examine for severity, grading, staging. Treatment depends on extent and severity of disease[8]. Biopsy and laproscopic approach is usually not recommended due to risk of perforation. Cytoreductive surgery (CRS) and Hyperthermic intraperitoneal chemotherapy (HIPEC) are done in ruptured appendix with PMP or disseminated peritoneal disease [10,11].

CONCLUSION

Mucinous neoplasm of appendix is usually confined to appendix and need surgical resection for good prognosis. when patient develop PMP treatment modalities are change. Mucinous neoplasm of appendix needs to follow up to prevent serious complication, like PMP. Early diagnosis can prevent fearful complication and provide good outcome.

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