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Low Grade Appendiceal Mucinous Neoplasm Presenting as a Mucocoele of the Appendix

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Abstract

Low grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy with incidence of 0.7% to 1.7% of all appendicectomies [1]. LAMN has the worst complication of mucin seeding into adjacent peritoneum leading to pseudomyxoma peritonei. LAMN lacks standardised treatment approach where only appendicectomy is preferred for resection of non metastatic disease. Case Presentation A 38 yr old female presented with right lower quadrant where CT Abdomen revealed Mucocele of Appendix. Patient underwent Laparoscopic Appendicectomy and Histopathological examination revealed Low grade appendiceal mucinous neoplasm. Patient is on regular follow up and is disease free. Conclusion low grade appendiceal mucinous neoplasm is usually an incidental finding where treatment depends on staging and histology of the tumour.

Keywords

Low Grade Appendiceal Mucinous Neoplasm, Pseudomyxoma Peritonei, Appendicectomy

1. Introduction

Primary mucinous neoplasms of the appendix vary from simple mucocele to invasive adenocarcinoma. Histologically, 65% of appendiceal tumours are neuroendocrine in origin while 20% of tumours are adenocarcinoma [2]. Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy surgical treatment usually includes appendicectomy with the removal of mesoappendix or right hemicolectomy. About 20% of the appendicular mucinous mass presents with the most dreaded complication called pseudomyxomaperitonei (PMP), which is diffuse collection of gelatinous material occurs due to mucin seeding into the peritoneal cavity [3]. LAMN is associated with herniation, diverticula, dissection and

rupture [4]. LAMN is usually an intraoperative incidental finding or usually present as Right lower quadrant symptoms. Diagnosis is difficult more commonly in female patients where ovarian mucinous neoplasm and Appendiceal neoplasm share common presentation. Here we describe 38 yr old female a case of low grade appendiceal mucinous neoplasm presenting as mucocele of appendix.

2. Case Report

A 38 year old male presented with complaints of lower right sided abdominal pain for 2 weeks which was gradual in onset, progressive in nature, not associated with nausea, vomiting and fever. Per Abdominal examination showed mild tenderness present over right iliac fossa, with no guarding or rigidity. CT abdomen showed a blind ending fluid filled bowel loop seen arising from appendix 2.7 cm from base of appendix measuring 26 mm in diameter and 10 cm in length with evidence of peripheral calcification suggested as appendiceal mucocele (**Figure 1**). CEA and CA125 were normal.

There were no lymph nodes, no perforation and the base of the appendix (as per the algorithm of Dhage-Ivatury and Sugarbaker) and there was no intra peritoneal clump of mucin. Laparoscopic appendicectomy was performed (**Figure 2**) and specimen (extracted with an endo bag) sent for histopathology. During laparoscopic dissection, pneumoperitoneum, grasping of the appendix specimen, or retrieval of the specimen through the abdominal wall might contribute to peritoneal dissemination of a tumour. This setback is usually avoided by taking precautions like usage of non traumatic bowel holding graspers to handle the mucocele, and using a non-permeable bag to deliver the specimen out of the port. Section from appendix showed walls lined by columnar cells with apical mucin and oval to elongated mild pleomorphic vesicular nuclei with focal areas of stratification forming billions structure. Lumen was filled with abundant mucinous material (**Figure 3**). With clinical, radiological and histopathological

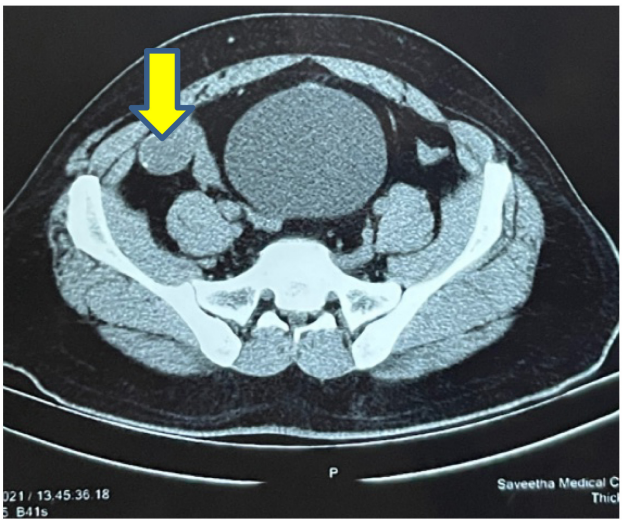

Figure 1. Showing CECT abdomen picture of appendiceal mucocele.

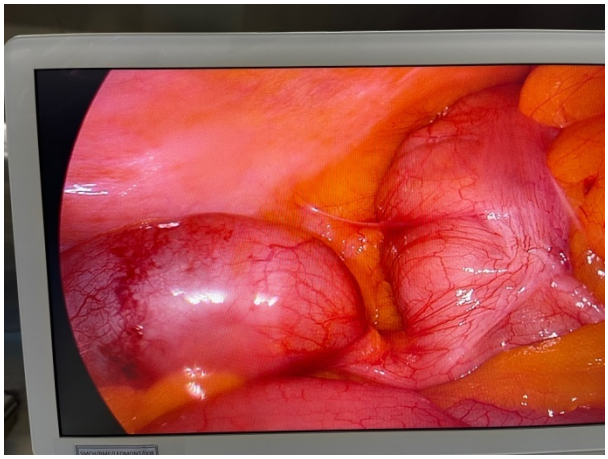

Figure 2. Showing the laparoscopic view of the mucocoele.

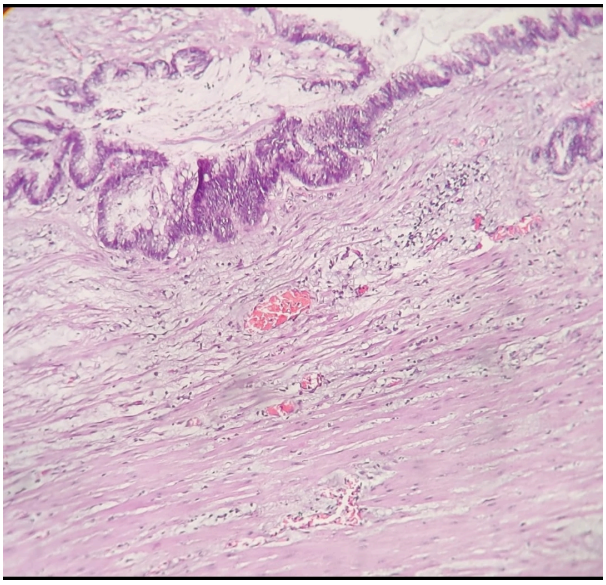

Figure 3. Histopathological report showing walls lined by columnar cells with apical mucin and oval to elongated mild pleomorphic vehicular nuclei with focal areas of stratification.

evidence diagnosis of low grade appendiceal mucinous neoplasm was made and the base was found to be free of carcinoma. Postoperative period was uneventful. Patient is on regular follow up and showed no evidence of disease progression.

3. Discussion

Mucocele of the appendix was described by Rokitansky. This disease is characterised by dilatation of a lumen as a result of excessive mucus accumulation. The mucosa of appendix is lined by columnar epithelium containing goblet cells more than that of the colon. As a result of this, most appendiceal epithelial tumours are mucinous and start as mucocele. With an incidence between 0.2% and 0.7% of all appendicectomies, it is categorised as a rare disease. Female preponderance of about 77% in Low-grade appendiceal mucinous neoplasm (LAMN) and is seen

in patients older than 50 years of age. Common clinical diagnosis includes lower right quadrant pain, nausea, vomiting, change in bowel habits, weight loss, and intestinal obstruction.

Diagnosis of acute appendicitis with ultrasound abdomen shows the outer diameter of the appendix is 6 mm - 15 mm which indicates the presence of a mucocele (with 83% sensitivity and 92% specificity). On CT Abdomen, appendix lumen more than 1.3 cm, cystic dilatation, and wall calcification confirm the presence of a mucocoele. Colonoscopic examination revealed an elevation of the appendiceal orifice and a yellowish mucous discharge would be visible from this orifice.

Mucocele of appendix represents an obstructive dilatation of the lumen due to excessive abnormal accumulation of mucus either due to a retention cyst, mucosal hyperplasia, adenoma, and adenocarcinoma. Simple mucocele (inflammatory, obstructive or retention cyst) is characterised by degenerative epithelial changes and results in the obstruction and the distension of the appendix. In hyperplastic mucocele, the appendix dilatation occurs due to the hyperplastic growth of the appendix or ceacal mucosa. Simple and hyperplastic mucoceles correspond to 5% to 25% of the cases, and mucus is usually acellular. The mucinous cystadenoma is an appendix neoplasm with dysplastic epithelium and corresponds to about 63% to 84% of the cases. The mucinous cystadenocarcinoma presents high grade cellular dysplasia and stromal invasion, besides muscularis mucosa, and represents 11% to 20% of the cases.

LAMN can also present as an incidental finding during surgery. Complications of LAMN include intussusception, ureteral obstruction, volvulus, small bowel obstruction, fistula and pseudomyxoma peritonei. Histologically mucinous, colonic and goblet cells associated with LAMN [5]. Elevated levels of carcinoembryonic antigen and carbohydrate antigen 19-9 act as useful markers for the diagnosis of LAMN. Elevated CEA, Ca 19-9, and Ca-125 may be detected in 56% - 67% of patients with LAMN [6]. These tumour markers can also be used for the surveillance of peritoneal malignancy following surgical or medical intervention. There is a risk of about 35% for concurrent GI malignancy in patients with LAMN.

The goal of management of LAMN includes the prevention of rupture, seeding, and development of pseudomyxoma peritonei. In the absence of invasion or lymph node metastasis, appendicectomy with the removal of mesoappendix should be performed as per the algorithm of Dhage-Ivatury and Sugarbaker [7]. Lymph node metastasis is about 4.2% of patients which is rare but would require an aggressive treatment [8]. Right hemicolectomy is the treatment of choice in patients with infiltration of submucosa or lymph node metastasis. LAMN < 2 cm is rarely malignant as compared to masses larger than 6 cm, which can show malignant cells and an association with pseudomyxoma peritonei. CK 20, MUC-2, and β-catenin are useful for the confirmation of the diagnosis. Earlier there were concerns about the role of laparoscopy [9] in LAMN, however safe laparoscopy can be offered today in select cases [10].

LAMN associated with pseudomyxoma peritonei can be diagnosed with the help of imaging modalities such as ultrasonography, CT scan, and magnetic resonance imaging and can be confirmed on the histopathological examination which will demonstrate epithelial cells and mucin in the peritoneum. The 5-year survival rate of pseudomyxoma peritonei is 25%. Close follow-up is recommended for all LAMN patients for 5 - 10 years. The 5-year survival rate for localized LAMN is 95%.

4. Conclusion

Low grade appendiceal mucinous neoplasm are heterogeneous group of tumors where management is based on staging of tumour and histological grading. In early stage low grade tumours surgical resection of primary site is preferred. In Advanced disease debulking surgery, Hyperthermic intraperitoneal chemotherapy (HIPEC) with or without chemotherapy is usually preferred.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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