

MUCINOUS CYSTADENOMA OF THE APPENDIX - A RARE CASE REPORT

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ABSTRACT

Mucinous cystadenoma is rare but is the most common benign appendiceal tumor. Although a benign tumor, mucinous cystadenoma can be a devastating condition if not treated optimally, as it can lead to pseudomyxoma peritonei. However, with appropriate treatment this tumor has excellent prognosis. Thus accurate diagnosis and optimal treatment is very essential in case of this tumor. In this case report we describe the clinical and histopathological features of appendiceal mucinous cystadenoma.

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INTRODUCTION

Mucinous cystadenoma is rare but is the most common benign appendiceal tumor.¹ Although a benign tumor, mucinous cystadenoma can be a devastating condition if not treated optimally, as it can lead to pseudomyxoma peritonei.² Since this tumor has highly variable clinical presentation, it is important to consider the diagnosis of mucinous cystadenoma in cases of cystic lesions of right lower quadrant of the abdomen. Here, we describe a case of mucinous cystadenoma presenting as appendicitis and present a brief review of literature.

Case Report

A 55 year old lady came with a complaint of chronic lower abdominal pain which had aggravated suddenly. On examination there was right lower quadrant tenderness without peritoneal signs. Ultrasonography of the abdomen revealed a cystic mass. On exploratory laparotomy, the cystic mass turned out to be mucocele of the appendix. Since there was no caecal involvement only appendectomy was performed.

Surgical specimen consisted of appendix having a thinned out wall and a dilated lumen filled with mucoid material. On microscopy, the wall of the appendix was lined by crowded tall columnar adenomatous epithelium with basally located, hyperchromatic, pseudostratified nuclei and apical mucin. Epithelial cells exhibited mild degree of dysplasia. The lumen was filled with mucoid material. The case was signed out as mucinous cystadenoma of the appendix.

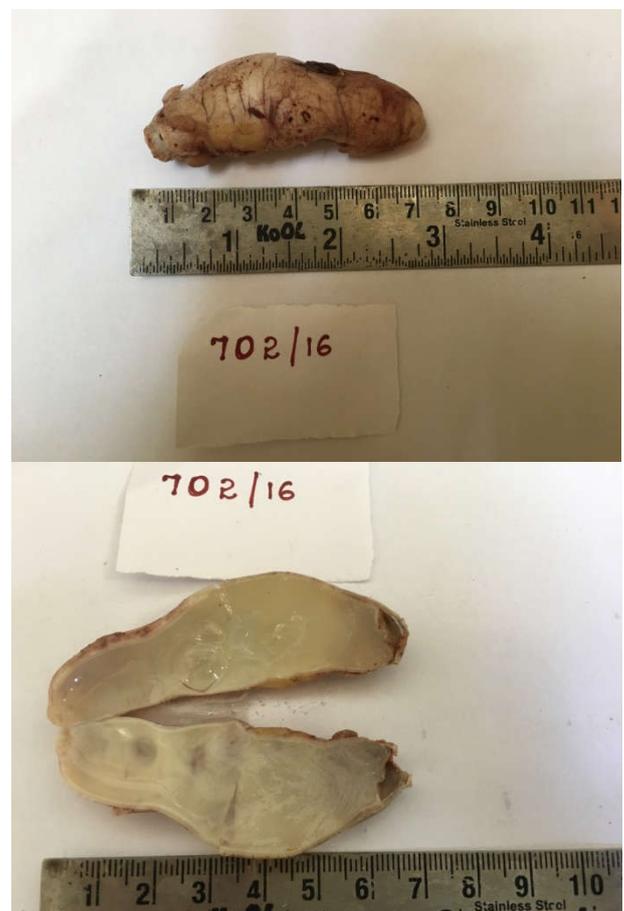


Figure 1 & 2 Surgical Specimen: Appendix With Thinned Out Wall And Dilated Lumen Filled With Mucoid Material.

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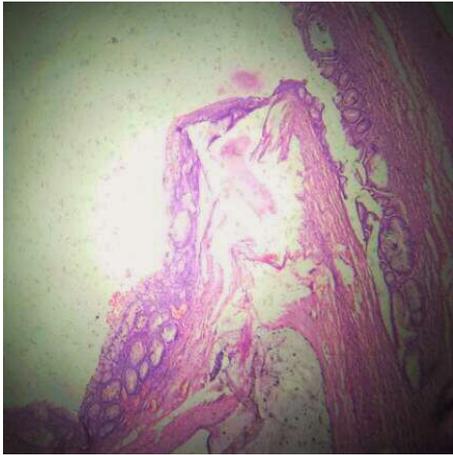


Figure 3 Microscopy: Thinned Out Wall of Appendix Lined By Mucous Glands And Tall Columnar Epithelium With Apical Mucin.

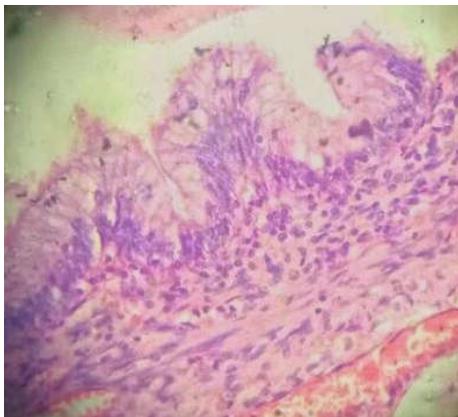
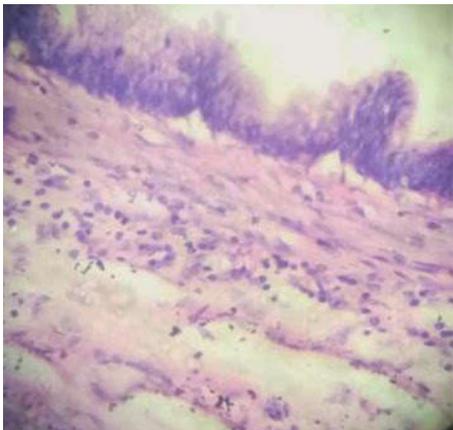


Figure 4 & 5 Microscopy: Wall Of The Appendix Lined By Pseudostratified Tall Columnar Epithelium With Basally Located, Elongated, Hyperchromatic Nuclei And Cytoplasmic Apical Mucin.

DISCUSSION

Appendiceal tumors are rare accounting for only 0.4% of gastrointestinal tumors.³ Appendiceal mucocele, which was first described by Rokitansky, represents a dilatation of appendix from the intraluminal accumulation of mucoid material. The incidence of mucocele varies between 0.2-0.7% among appendectomy specimens. Mucocele is the second most common type of appendiceal tumor after carcinoid. The causes of mucocele include retention cyst, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma. Mucinous cystadenoma is the most common cause of mucocele accounting for about 63-84%

cases.⁴ It develops as a result of proliferation of mucin secreting epithelium in an occluded appendix.⁵

Mucinous cystadenoma occurs more commonly in women and in patients aged more than 50 years.² The clinical presentation of appendiceal mucinous cystadenoma is highly variable.⁴ It may present as incidental finding, palpable abdominal mass or chronic right lower abdominal pain.^{1,4,6,7} Rare cases of mucinous cystadenoma presenting with bleeding, intussusceptions, umbilical hernia, volvulus, ureteral obstruction and hematuria have also been reported in literature.^{8,9,10,3,7} Synchronous cases of mucinous cystadenoma with colon cancer have been described with an incidence of upto 20%.⁹

Mucinous cystadenomas are associated with perforation in 20% of the cases.⁶ Pseudomyxoma peritonei is a potentially devastating complication of mucinous cystadenoma which occurs due to rupture of mucocele.⁴ It may either occur due to spontaneous rupture or spillage of contents into peritoneum during surgery.¹¹ Other complications include invasion into adjacent organs, recurrence and torsion.¹²

CT is the gold standard for radiological diagnosis. It is more informative than MRI and USG.⁴ However it is challenging to diagnose this condition pre-operatively as smaller lesions are often asymptomatic and larger lesions mimic normal or dilated small bowel on imaging studies.² Tumor marker studies like CEA levels can be useful in the diagnosis.¹³

Intra-operative frozen section is usually preferable in cases of appendiceal tumors, because it is extremely difficult to diagnose cystic appendiceal lesions pre-operatively and it is essential to distinguish between mucinous cystadenoma and cystadenocarcinoma as the latter needs an extensive procedure for optimum management.¹⁴

Mucinous cystadenomas show marked distention of lumen upto 6cm.⁴ On histopathology, they show epithelial villous adenomatous changes with some degree of epithelial atypia.¹⁵ It is now widely accepted that mucinous cystadenoma is the appendiceal counterpart of villous adenomas occurring elsewhere in the intestine.¹⁶ Mucinous cystadenomas generally tend to be low grade, with circumferential involvement. Progressive mucocele formation results in thinning of the wall due to pressure atrophy and progressive fibrosis.¹⁷

Surgical resection is the main modality of treatment of mucinous cystadenoma. The extent of surgery depends on thickness, dimension and involvement of caecal base by the tumor.⁴ In cases of mucinous cystadenomas without caecal involvement, resection of appendix is performed, followed by collection and cytological examination of peritoneal fluid and careful inspection of the base of the appendix.¹¹ Cecectomy is performed in cases of mucinous cystadenomas having thick base, involving caecum, patients with a positive margin at the base of the appendix or with positive lymph node.^{4,11} Laparoscopic approach has also been described for the management of appendiceal mucoceles in few recent studies.^{18,19,20,21}

The prognosis of mucinous cystadenoma is excellent. The five year survival rate is approximately 100%.²²

CONCLUSION

Mucinous cystadenoma remains a diagnosis that is rarely considered before elective surgery. However, accurate diagnosis is very essential to ensure optimal treatment and prevent complications like pseudomyxoma peritonei. Thus the diagnosis of appendiceal mucinous cystadenoma should always be kept in mind in cases of cystic lesions of right lower quadrant of the abdomen.

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