

Case Report

Mucocele of appendix unmasking appendiceal neoplasm

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ABSTRACT

An appendiceal neoplasm is rare among all the gastrointestinal tumors. An appendiceal mucocele is a dilatation of the appendix due to a benign or malignant disease, which causes the obstruction of the appendix and consequent accumulation of mucus secretion. Mucinous malignancy of the appendix represents a low occurrence among appendiceal neoplasms. The preoperative diagnosis is difficult due to non-specific clinical manifestations of the disease. Preoperative investigations may be inconclusive for diagnosis at times. Management is based on the size and grade of the tumor. We had two female patients who presented with poorly defined symptoms in whom, radiological evidences showed appendiceal dilatation. Both the patients when proceeded with laparotomy, had no peritoneal spillage, and were subjected to right hemicolectomy. Based on the grade of the tumor, adjuvant management was planned. Both the patients had favorable outcomes following treatment.

Keywords: Appendix, Mucocele, Mucinous appendiceal neoplasm

INTRODUCTION

Appendicular neoplasm is a rare occurrence among gastrointestinal tumors which comprises approximately <1.4 percentages of appendectomies.^{1,2} Histologically, 65 percentages of appendiceal tumors are neuroendocrine in origin while 20 percentages of tumors are adenocarcinoma.^{1,4} Among these, mucinous cell tumors of appendix are rare (<0.3%). Primary mucinous neoplasms of the appendix vary from simple mucocele to invasive adenocarcinoma.⁴ A mucocele of the appendix is a rare entity contributing to 0.2 percentages to 0.7 percentages of appendiceal pathologies as well as is more frequent among individuals aged 50 years or more.³ Occurs more common in women.^{1,4,6} Patients are often asymptomatic or present with the right lower quadrant abdominal pain mimicking appendicitis, intestinal obstruction, perforation, bleeding. Despite extensive preoperative investigations, the diagnosis may remain elusive as well as may only be made at the time of the surgery.⁵

CASE REPORT

Case 1

A 47 years old female, presented with right iliac fossa pain for past 1 month aggravated for past 1 week associated with vomiting. On examination, tachycardia was seen and per abdomen palpation presented a vague tender mass of size 4×4 cm with irregular surface and firm consistency in the right iliac fossa. Ultrasonogram showed 5×4 cm cystic mass with internal debris with uncertain origin. CT showed well-defined 6×4.5 cm hypodense cystic lesion with peripheral rim of calcification arising from base of caecum with a possibility of appendiceal mucocele / mesenteric cyst. As the patient showed picture of acute appendicitis/appendicular abscess, patient was taken up for emergency laparotomy where appendix was distended with mucin to a size of 7×5×5 cm and there was no peritoneal deposits and spillage. Hence, proceeded with right hemicolectomy and ileo-transverse anastomosis.



Figure 1: USG showing cystic mass in the right ileac fossa.

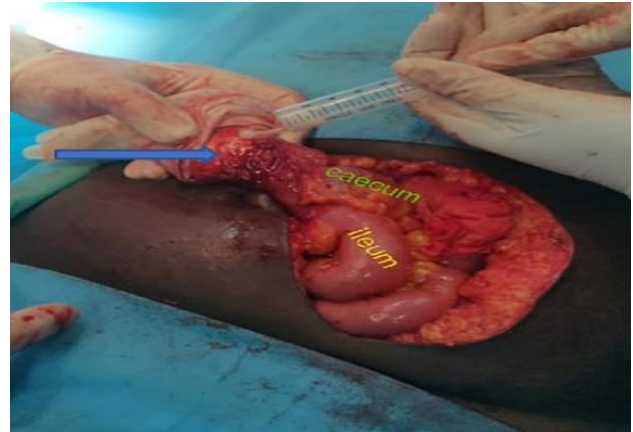


Figure 4: Aspiration of content from the distended appendix-mucin.

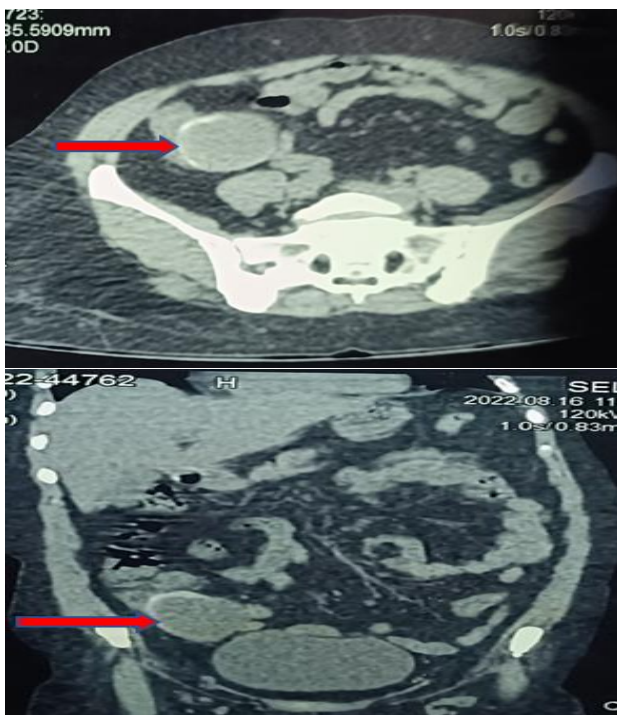


Figure 2: Arrowhead showing cystic lesion with rim of calcification arising from base of caecum.



Figure 3: Distended appendix.



Figure 5: White jelly material from the specimen.

Histopathological examination revealed diffusely involving low grade appendiceal mucinous neoplasm confined by muscularis propria and 17 nodes submitted showed reactive hyperplasia.

As it was a low-grade tumor without spread, no adjuvant therapy was contemplated. Post op follow up for 18 months showed no evidence of recurrence or any other metachronous lesion.

Case 2

A 69 years old female, hypertensive and post total thyroidectomy status presented with a past history of right iliac fossa pain for 2 days and got evaluated with CECT abdomen outside at a different hospital showing appendix distended to 2.3 cm with non-enhancing hypodense content. On admission, patient had no complaints. On examination, vitals were stable and a vague non tender, non-mobile mass of size 3×3 cm with irregular surface and firm in consistency was palpable in the right iliac fossa, with a palpable lower border. As there were no signs of requirement of emergency intervention, patient was evaluated further with colonoscopy which showed mucin protruding from appendicular opening with mucosal bulge in caecum. A

diagnosis of mucocoele of appendix was made and was planned for elective laparotomy. Intraoperatively, dilated appendix till the base with mucin as content was seen. There was no peritoneal deposits and spillage. Hence, proceeded with right hemicolectomy and ileo-transverse anastomosis.



Figure 6: CECT showing distended appendix with non-enhancing hypodense content.

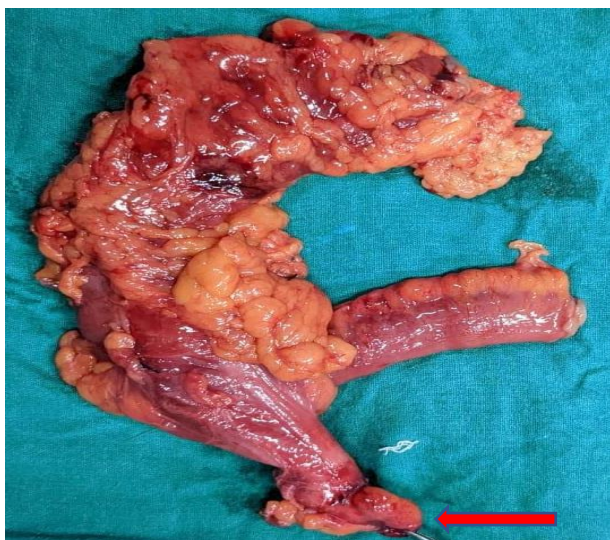


Figure 7: Arrowhead showing distended appendix in the resected right hemicolectomy.

Histopathological examination showed well differentiated infiltrating mucinous adenocarcinoma involving distal half of appendix invading visceral peritoneum and 13 nodes showing follicular hyperplasia (pT4a N0 Mx).

Patient was put on adjuvant chemotherapy-T. capecitabin. Follow up of the patient for 6 months showed no evidence of recurrence.

DISCUSSION

Appendiceal mucocoeles are rare and can be due to neoplastic and non-neoplastic causes. In 2012, the peritoneal surface oncology group international classified appendiceal mucinous lesions into two categories: non-neoplastic lesions- simple mucocoeles or retention cysts and neoplastic lesions-(a) serrated lesions with or without dysplasia, (b) mucinous appendiceal neoplasms, (c) mucinous adenocarcinomas of the appendix.⁹ Mucinous cystadenomas are noninvasive epithelial proliferation with cellular invasion confined to the mucosa, without mucin or cells penetrating the muscularis mucosa which may be high grade (HAMN) or low grade (LAMN).^{6,7} Mucinous cystadenocarcinomas are invasive tumors with cellular invasion beyond the muscularis mucosa.^{6,7} Low-grade appendiceal mucinous neoplasms (LAMN) which are rare mucinous tumors of the appendix, are relatively a common cause for mucocoele.

In our study, first case was LAMN, where tumor was confined to muscularis propria and the next case was an infiltrating adenocarcinoma of mucinous type with no extra serosal involvement. The gold standard treatment for appendicular neoplasm is surgical resection. All the mucinous tumors are at risk of rupture and developing pseudomyxoma peritonei (PMP).¹⁰ Hence, early surgical resection is recommended for all AM to exclude mucinous neoplasm and to prevent spontaneous rupture in the future. The depth and extent of surgical resection relies on the grade of tumor and the size of mucocoele. Most commonly the tumor is detected intraoperatively. If the tumor is confined to the appendix without involving the base of the appendix, <2 cm, appendectomy along with removal of mesoappendix is sufficient, provided the margin is negative. If the tumor is extending beyond the appendix, >2 cm or involving the base of the appendix, or the margin is positive, right hemicolectomy is done because of the potential for lymph node metastases. And if there is rupture or spillage of mucin deposit resulting in pseudomyxoma peritonei (PMP), cytoreductive surgery is planned along with HIPEC treatment.⁹ Previously, it was said that open surgical approach is better when compared to laparoscopic approach as it allows a better damage control of the potential rupture and iatrogenic lesions. But with recent advances in instrumentation and better surgical hands, laparoscopic management is preferred. Adjuvant chemotherapy is provided for adenocarcinoma, pseudomyxoma peritonei (PMP).

Preoperative diagnosis is essential for planning operative procedure. The most widely used imaging study is CT. It is typically seen as a well-circumscribed, low-attenuation, spherical, or tubular mass contiguous with the base of the cecum. Curvilinear mural calcification if present suggests the diagnosis but is seen in less than 50% of cases.⁸ In CT, an appendix with a diameter of more than 15 mm, soft tissue mass, wall thickening or irregularity should raise the suspicion of mucinous neoplasm.⁷ Biopsy is not

indicated in mucinous tumors in the fear of seeding of mucin into peritoneal cavity.

CONCLUSION

Mucinous appendiceal neoplasms are rare pathology that present with unspecific clinical signs and, in many cases, are asymptomatic. Preoperative diagnosis or suspicion is important for the operative strategy and choice of operating technique for a R0 resection and proper management. The mainstay management for mucinous appendix is surgical resection. Early surgical resection is recommended for all AM to exclude mucinous neoplasm and to prevent spontaneous rupture in the future.

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