

Case Report of Mucocele of Appendix

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Abstract: Appendiceal Mucinous neoplasms are a heterogeneous group of neoplasms ranging from simple mucoceles to complex pseudomyxoma peritonei. Low - grade appendiceal mucinous neoplasms (LAMN) are detected in 0.7 to 1.7% of all appendicectomies. LAMN can present with abdominal pain, vomiting, distention, palpable mass, intestinal obstruction, and rarely with urological symptoms. **Case Report:** A 65 - year - old patient presented with mass in right iliac fossa since 6month. His USG Abdomen showed hypoechoic cystic lesion with internal echoes in right iliac fossa. His CECT abdomen showed - Mucocele of Appendix. Right hemicolectomy with ileo - transverse anastomosis was done. Histopathology of Appendix showed Low grade appendiceal mucinous neoplasm. Patient advised to remain in follow - up.

Keywords: LAMN, Appendiceal mucocele; Pseudomyxoma peritonei; Appendiceal Mucinous neoplasm; Case report; Hemicolectomy

1. Introduction

A mucocele of appendix is the result of obstruction of appendiceal orifice with distension of appendix caused by accumulation of mucoid material [1]. incidence is 0.2 - 0.3% in appendicectomies reviewed [2]. Histologically mucoceles can be divided into mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma [3]. Surgery is the treatment of choice

2. Case Presentation:

65 year old male patient admitted with chief complains of right iliac fossa mass since 6 month. No history of abdominal pain, nausea, vomiting, anorexia, fever, weight loss. He had no other symptoms or comorbidities. Clinical examination was suggestive of palpable lump in right iliac fossa. (Figure 1) Ultrasonological evaluation showed a tubular non peristaltic, non compressible structure is noted in right iliac fossa measuring approx 50* 104* 58 mm size. It appears content filled with thick echogenic wall (measuring 5 - 6mm) and without internal vascularity with presence of onion skin appearance with posterior acoustic enhancement is noted in above mention tubular structure. Wall of terminal ileum and ileocecal junction appears normal. possibility of mucocele of appendix more likely / mesenteric cyst. CT scan revealed a tortuous tubular fluid filled distended structure with internal calcification noted in subcecal and right iliac fossa region. Maximum diameter of lesion measures 38 mm. Appendix was not separately made out. Rest of the abdomen was normal. Mucocele or benign cystic lesion was suspected. . On exploration, a cystic lesion of appendix size 8cm*5 cm with broad base and inflamed walls communicating with caecum but no perforation was discovered in right iliac fossa. (figure 2) Right hemicolectomy with ileo - transverse anastomosis was done. Histopathological diagnosis of Low grade Appendiceal Mucinous neoplasm is made. After 6 month of

surgery patient is doing well with no postoperative complications.



Figure 1: Pre op picture showing mass in RIF



Figure 2: Intra op picture showing mucocele of appendix



Figure 3: Right hemicolectomy specimen

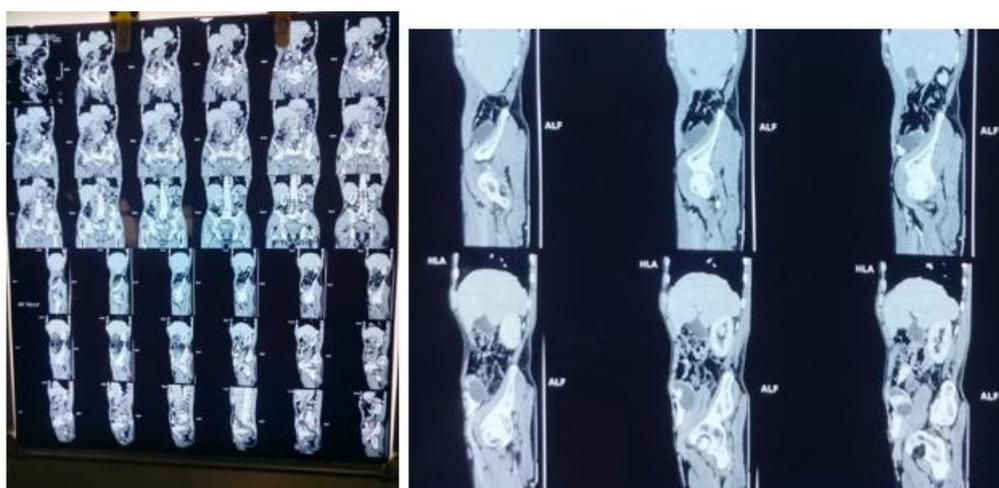


Figure 4: CECT (A+P) s/o mucocele of appendix

3. Discussion

Appendicular mucocele is a rare condition where there is a cystic tubular dilatation of the appendiceal lumen. It occurs in patients in their 5th or 6th decades and there is a slight female preponderance. They are often asymptomatic, however, they can present as acute appendicitis. Rarely they can present with bowel obstruction or gastrointestinal bleeding due to intussusception, genitourinary symptoms due to obstruction of the right ureter or bladder or generalized peritonitis from a rupture [1, 4]. The relevance of appendiceal mucocele in the spectrum of appendiceal tumours was not clearly defined until recently when the Peritoneal Surface Oncology Group International (PSOGI) developed a consensus classification that has helped to resolve much of the confusion surrounding the diagnostic terminology. Accordingly, mucinous lesions of the appendix are divided into 1) Non - neoplastic appendiceal mucinous lesions (also referred to as inflammatory or obstructive mucoceles) like simple mucoceles or retention cysts characterized by degenerative epithelial changes secondary to obstruction due to faecolith and 2) Neoplastic appendiceal mucinous lesions which include a) Serrated polyp with or without dysplasia, b) Mucinous neoplasms – either a low - grade appendiceal mucinous neoplasm (LAMN) or as high - grade appendiceal mucinous neoplasm (HAMN)

and c) Mucinous adenocarcinomas which can be well, moderately or poorly differentiated mucinous adenocarcinomas [1].

Radiological studies, in particular, Contrast - enhanced computed tomography and ultrasound abdomen can diagnose appendiceal mucoceles, however, they cannot definitively distinguish between non - neoplastic and neoplastic lesions. On most occasions, neoplastic lesions are generally larger (measuring more than 2 cms) compared to non - neoplastic lesions. The presence of soft tissue thickening, wall calcifications and wall irregularity, but not an increase in wall thickness, are suggestive of malignancy. The presence of ascites with hypodense peritoneal lesions and scalloping of the liver surface suggests the intraperitoneal spread of neoplastic cells from a ruptured mucinous neoplasm [2].

Surgical resection is recommended for all appendiceal mucinous lesions. As there are no reliable criteria to exclude benign from malignant lesions, surgery should be pursued even for a benign - appearing appendiceal mucocele on imaging studies. An earlier laparoscopic approach was not advised because of the risk of rupture but now with increasing expertise, more surgeons are favouring the laparoscopic approach. The decision on whether to perform the surgery, laparoscopic or open is largely dependent on the surgeon's

expertise [3]. In either case, the principles followed have to be the same, which include resection of the appendix, wide excision of mesoappendix to facilitate retrieval of all periappendiceal lymph nodes and careful assessment of the base of the appendix to exclude any extension into the caecal wall. In patients with a positive margin at the appendix base or positive peri - appendiceal lymph nodes, a right hemicolectomy is warranted [3, 4]. A more aggressive approach such as a radical resection, removal of all gross implantations and hyperthermic intraperitoneal chemotherapy (HIPEC) is recommended in cases of ruptured appendiceal mucinous neoplasms that have to lead to Pseudomyxoma Peritonei (PMP) [1].

The prognosis of appendiceal mucinous lesions is closely associated with the histopathology, presence and extent of peritoneal spread. Prognosis in benign lesions is excellent with a 5 - year survival rate of 91% to 100% after standard appendicectomy. Neoplastic lesions, particularly high - grade appendiceal mucinous neoplasms (HAMNs) have a more guarded prognosis even after curative resection. Histopathologic features such as the presence of extraappendiceal neoplastic epithelium, high - grade cytology, architectural complexity and invasion are important predictors of recurrence. Prognosis is worse for mucinous adenocarcinomas, particularly if the appendiceal mucocele ruptures into the peritoneal cavity. Therefore all of these patients need to be under surveillance. Tumour markers such as Carcinoembryonic antigen (CEA), Ca 19 - 9 and Ca 125 can be elevated in patients with advanced appendiceal mucinous lesions and the levels correlate with treatment outcomes [5]. Currently, there are no guidelines for post - treatment surveillance but it is advised according to the histology, grade and completeness of surgery [1, 5].

4. Conclusion

Appendiceal mucoceles or appendiceal mucinous lesions can present as appendicitis. They are difficult to diagnose clinically. Computed Tomography of the abdomen is essential. Large sizes of mucoceles should raise suspicion of underlying malignancy. Appropriate and timely surgical resection with clear margins leads to excellent post - op recovery and oncological outcomes, however, these patients require close follow - up.

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References

- [1] Carr NJ, Bibeau F, Bradley RF, et al. The histopathological classification, diagnosis and differential diagnosis of mucinous appendiceal neoplasms, appendiceal adenocarcinomas and pseudomyxoma peritonei. *Histopathology* 2017; 71: 847 – 858. doi: 10.1111/his.13324. Epub 2017 Sep 19.
- [2] Wang H, Chen YQ, Wei R, et al. Appendiceal mucocele: A diagnostic dilemma in differentiating malignant from benign lesions with CT. *AJR Am J Roentgenol* 2013; 201: W590 – W595. doi: 10.2214/AJR.12.9260.

- [3] Palanivelu C, Rangarajan M, John SJ, Senthilkumar K, Annapoorni S. Laparoscopic right hemicolectomy for mucocele due to a low - grade appendiceal mucinous neoplasm. *JLS*.2008; 12: 194–7. PMID: 18435897; PMCID: PMC3016193.
- [4] Stocchi L, Wolff BG, Larson DR, Harrington JR. Surgical treatment of appendiceal mucocele. *Arch Surg* 2003; 138: 585 - 9. doi: 10.1001/archsurg.138.6.585
- [5] Asare EA, Compton CC, Hanna NN, et al. The impact of stage, grade, and mucinous histology on the efficacy of systemic chemotherapy in adenocarcinomas of the appendix: Analysis of the National Cancer Data Base. *Cancer* 2016; 122: 213 - 21. doi: 10.1002/cncr.29744. Epub 2015 Oct 27