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A Rare atypical Case of Low-grade Appendiceal Mucinous Neoplasm: A Case Report

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INTRODUCTION:

Appendiceal Mucinous Neoplasm is a very rare malignancy. In United States, only around 1000-2000 cases are diagnosed annually [1]. Both benign and malignant appendicular mucinous lesions have a slight female predominance and are usually diagnosed in the 5th and 6th decades. According to the Peritoneal Surface Oncology Group International (PSOGI) Neoplastic Appendiceal Mucinous Lesions are broadly classified into three main types [1],

1. Serrated Polyps of the Appendix

2. Mucinous Appendiceal Neoplasm- which is further classified into Low-grade appendiceal mucinous neoplasm (LAMN) and High-grade appendiceal mucinous neoplasm (HAMN).

3. Mucinous Adenocarcinoma of the Appendix.

There are no specific clinical features of LAMN, most of these patients present with features of acute appendicitis and histopathology later reveals this pathology. Among the appendiceal mucinous neoplasms, the serrated polyps or simple mucocele are not typically known to recess once they rupture, but on the other hand. LAMN or HAMN, or Mucinous Adenocarcinoma can lead to disastrous consequences such as progressive intraperitoneal spread and accumulation of both mucinous ascites and neoplastic cells, often referred to as a clinical syndrome known as Pseudomyxoma Peritonei.

CASE PRESENTATION:

A 65 year old female, known hypertensive for more than a decade, non-diabetic presented to the OPD on February 10th, 2018 with complaint of lethargy, loss of appetite, generalized body weakness, nausea and intermittent low grade fever for 10 days. On examination abdomen was soft and non-tender. Bloods were performed showing high CRP (146.09 mg/L). Urine showed few RBCs, bilirubin + trace protein, blood +. Rest of blood tests were within normal limits. Blood culture showed no growth. Ultrasound KUB was normal .She was advised oral antibiotics for a week. She improved a little but low grade fever and generalized weakness continued. Bloods were repeated and CAT scan with IV contrast was done.

The lab findings included a raised TLC of 16,100/ ul , raised CRP 140.04 mg/dl with unremarkable Urine R/E, Urine and Blood C/S. CT with contrast showed an approximately 4.9×7.5×6.7cm, irregular complex fluid collection in the Right Iliac Fossa extending up to the midline with excessive surrounding stranding changes and reaction.(Fig:1 & Fig :2) The stranding changes were reaching up to the right hemi pelvis merging with adnexa. The collection was closely abutting the Cecum and ileocecal junction. The Cecum and Ileoceal junction were however thick walled and the Appendix was not separately visualized. Distal ileal loops were inseparable and adherent to the collection. Few surrounding enhancing nodes were visualized, the largest one being 7mm. The radiologist commented that because the Appendix was not separately visualized and keeping in view the lab findings, the above described finding represented ruptured appendix with abscess formation, mucocele/pyocele of the appendix remained in the differentials. However, keeping in view patient's age possibility of neoplastic disease process could not be ruled out. There was also an incidental finding of Cholelithiasis.

CAT Scan Images:

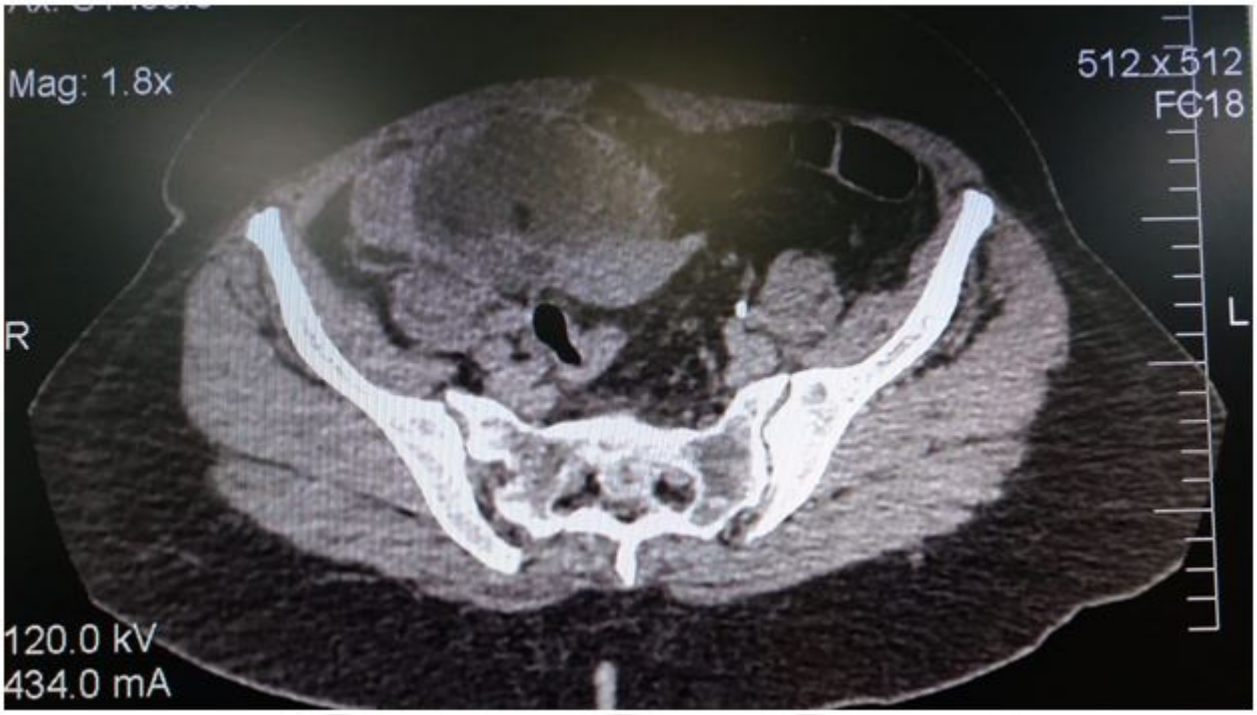

Figure 1: Irregular complex fluid collection in the right iliac fossa extending up to the midline with extensive surrounding strandy changes and reaction.

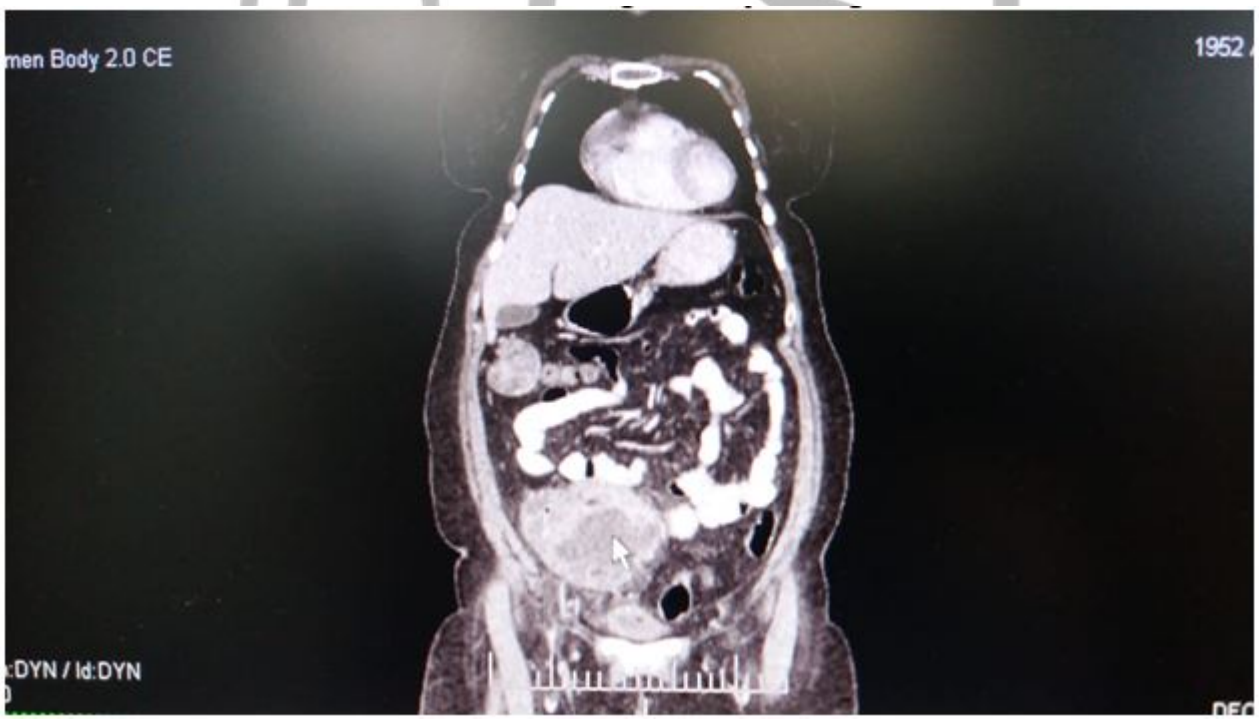

Figure 2: Sagittal view showing right iliac fossa collection

Keeping in view patient's clinical picture, lab reports, and imaging, the patient was treated conservatively as a case of Appendicular Abcess with I/V Antibiotics for 10 days. Afterwards one week course of oral antibiotic was given. Interval CT scan and Ultrasound RIF were performed to follow disease progression. Follow up CT scan showed marked interval resolution of previously described irregular complex fluid collection in Right Iliac Fossa and thickening of the Cecum and Ileocecal junction with mild residual nodularity and stranding changes. Collection size was reduced down to a 2.1×1.1 cm. Appendix was not separately visualized and overall findings were likely suggestive of Infective/Inflammatory etiology since the disease responded to antibiotic. No image-guided aspiration/ drainage was considered as she was showing marked improvement with conservative treatment. The same management was continued with 2 weekly ultrasound RIF which showed consistent reduction in the collection and strand changes. Four months after initially observed RIF collection CT scan was done which showed almost a complete resolution of the RIF's collection, stranding and congestion. The patient was advised an elective appendectomy to prevent any future episodes. On laparoscopy, she had a mass of omentum and ileum containing thick-walled, inflamed appendix. With careful dissection the appendix was removed laparoscopically along with cholecystectomy (for silent cholelithiasis). Both the organs were sent for histopathology. The salient histological features were described as follows, low grade Appendicular Mucinous Neoplasm, Histological type: G1 undifferentiated with a tumor size of 1.9×0.7cm. The tumor was seen invading through the muscularis propria into the mesoappendix but did not extend to the serosal surface, no lymphovascular or perineal invasion was identified. The proximal margins of the specimen were not involved. A cystic lesion containing extravasated mucin was seen lined focally by columnar cells within and adjacent to the vessel. The gallbladder specimen showed typical features of chronic cholecystitis.

Histopathological features

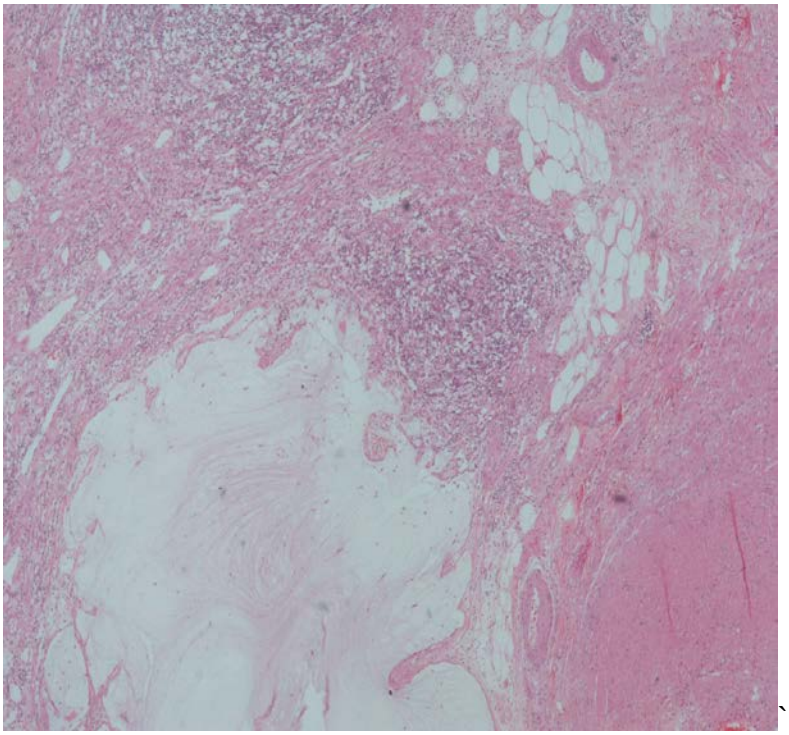

Figure 3: Mucin filled appendix

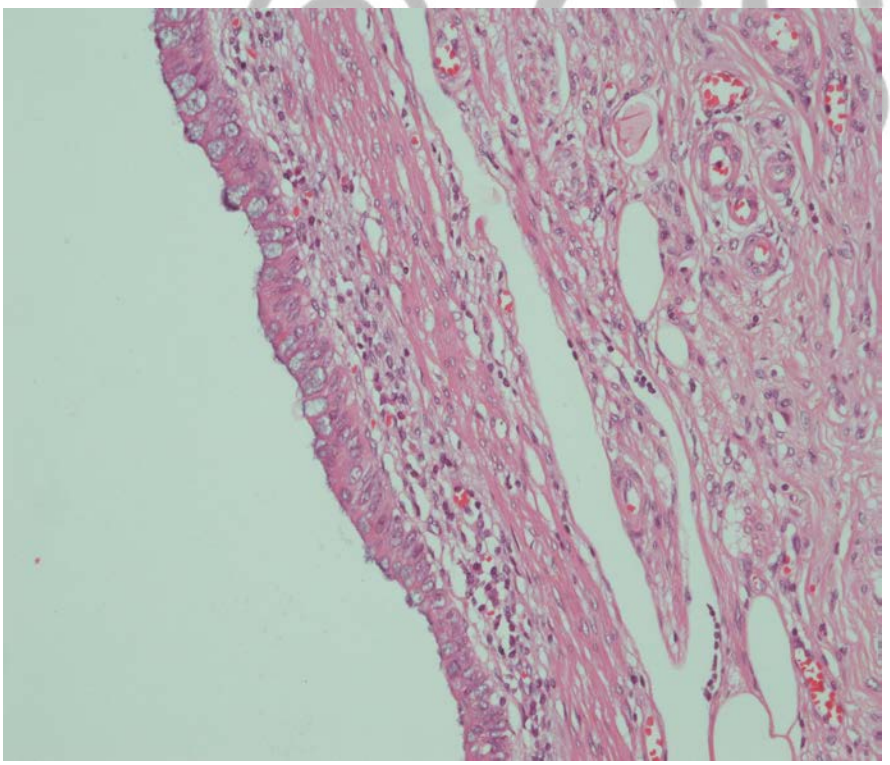

Figure 4: Appendiceal epithelium showing elongated nuclei and low grade nuclear atypia

The patient had an uneventful recovery after surgery. After one month she had a colonoscopy in which biopsies were taken from the appendiceal orifice. The biopsy revealed large bowel mucosa with mild inflammation, negative for any dysplasia or malignancy. A repeat Ultrasound five month after the surgery did not indicate any previous or new collection. She was advised regular CT and colonoscopy after six months but she did not follow. Her last contact after two years of surgery, revealed no clinical abnormality. She was again advised to have CT scan and colonoscopy that she promised to get done after situation of COVID-19 pandemic is over.

DISCUSSION:

LAMNs are rare adenomas localized in the appendix or the surrounding appendiceal mucosa wall. Patients with LAMN can present with abdominal pain, intussusception, and obstruction. However, LAMNs are often incidentally found in asymptomatic patients. Complications of LAMN include intussusception, ureteral obstruction, volvulus, small bowel obstruction (SBO), rupture, and PMP [2-3].

Low-grade and high-grade tumors differ from management perspective too. The treatment of low-
grade tumors includes surgical resection in early stage and peritoneal debulking and hy perthermic intraperitoneal chemotherapy (HIPEC) in advanced stage disease. Likewise, high-grade tumors benefit from debulking surgery and HIPEC, with or without preoperative chemotherapy [4]. Imaging modalities for diagnosis include ultrasound(US) and CT, with CT as the most commonly used radiographic interpretation for preoperative diagnosis. The common abdominal CT findings include cystic dilation within the appendiceal lumen with wall calcifications and irregular appendiceal wall thickening as demonstrated in our case. Grossly, specimens of LAMN include hyalinization and fibrosis of the appendiceal wall with a grossly swollen appendix secondary to mucinous accumulation. LAMNs less than two centimeters (cm) are rarely malignant and are classified as benign simple or retention Mucoceles. Masses larger than 6 cm present with a higher risk of malignant cells, a higher risk of appendiceal perforation, and development of PMP. Histological evidence of LAMN includes atypical glandular cells and epithelial cells with “pushing invasion” of malignant cells creeping into the appendiceal wall with possible diverticular formation. Mucinous, colonic, and goblet cells are also often identified within LAMN. Elevated CEA, Ca -19-9, and Ca-125 may be detected in 56.1-67.1% of patients with LAMN. These tumor markers can also be used for the surveillance of peritoneal malignancy following surgical or medical intervention. There is also a 35% risk of a concurrent GI malignancy in patients with LAMN. Surveillance of patients with LAMN incorporates radiographic imaging every six months post appendectomy for two years for adequate monitoring of tumor recurrence and complications associated with PMP. Accurate pathological assessment and classification of LAMN are important to assess for malignancy risk, seeding, recurrence, and patient prognosis. For patients with a high risk of disease progression, follow-up

should continue for the first five years after diagnosis of LAMN [5-6]. High-risk patients include those with evidence of infiltration of malignancy into submucosa or with the presence of lymph node metastasis. Additional surveillance and treatment studies are needed, but until then, the treatment for LAMNs will remain inconsistent due to a lack of standardized interventions based on diagnostic criteria. The five year survival rate for localized LAMN is 95%.

CONCLUSION:

A high index of suspicion is needed to diagnose rare cases of malignancies of appendix especially if the symptoms are not typical of acute inflammation.

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