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Low-Grade Appendiceal Mucinous Neoplasm Presenting as Acute Abdomen: A Case Report

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Abstract: *Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy with symptoms varying depending on the clinical manifestations. The most important complication of this particular neoplasm is seeding of mucin into the adjacent peritoneum leading to pseudomyxoma peritonei (PMP). We present the case of a female patient came to our emergency department with acute abdominal pain mimicking an acute appendicitis. CT and ultrasound scan showed a heterogenous mass located in the right iliac fossa. The patient was submitted to laparotomy. The operative findings were suggestive of an appendiceal mucocele. The histology report revealed a low-grade appendiceal mucinous neoplasm.*
Keywords: *Low-grade appendiceal tumor, Acute abdomen.*

I. Introduction

Mucocele of the appendix is a dilation of the appendix, secondary to obstruction and mucous accumulation intraluminally. They are quite rare and only account for 0.2-0.3% of all appendectomy specimens. (1). A mucocele of the appendix can be identified on a variety of imaging modalities. The test of choice is a CT scan as it has the advantage of better delineating the anatomic relationship to the right ovary than ultrasound and is superior than magnetic resonance imaging at finding mural calcifications. (2).The most common presentation of an appendiceal mucocele is an asymptomatic mass found incidentally on imaging, although less commonly the symptoms can includes weight loss, nausea, vomiting, acute appendicitis, change in bowel habits or anemia.(3) In this report, we present a rare caseof a low-grade appendiceal mucinous neoplasm referred to our department with the clinical picture of an acute abdomen and a palpable mass which was managed as a surgical emergency.

II. Case report

A 60-year-old saudi female patient was transferred to our hospital with acute severe right iliac fossa pain of 48 h duration and a fever of 37.6°C. The pain was continuous and confined to the lower abdomen. The patient had vague lower abdominal pain for seven days. On examination the general appearance of the patient was good. There was rebound tenderness and a tender mass was felt in the right iliac fossa on palpation. After admission, the patient had routine blood tests and plain X-rays of the abdomen (erect and supine) as well as a chest X-ray. At that time, she was also submitted to ultrasound and a CT scan of the abdomen. Ultrasound examination of the mass showed dilated appendix with multi-septated cystic lesion at iliac fossa (Fig 1, Fig 2).

Computed tomography (CT) showed awell-circumscribed, low-attenuation, spherical mass contiguous with the base of the cecum. Few calcifications are seen as well as mild wall thickening. No low attenuation deposits in the peri -appendiceal space, peritoneal cavity or at the surface of abdominal viscera, including ovaries and bowel. (Fig. 3, Fig 4).

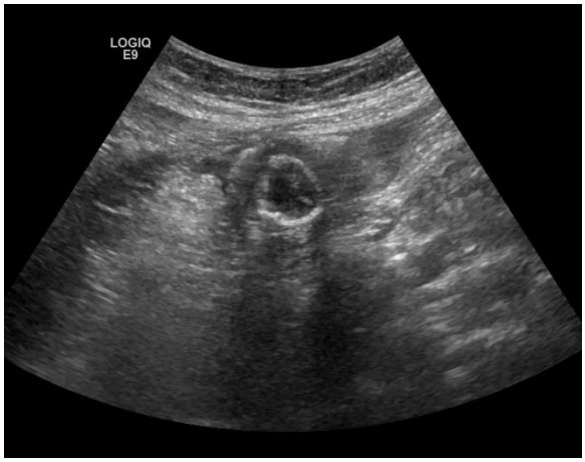

Figure 1: Ultrasound study showed right iliac fossa dilated appendix reach up to 1.6 cm with mild vascularity .

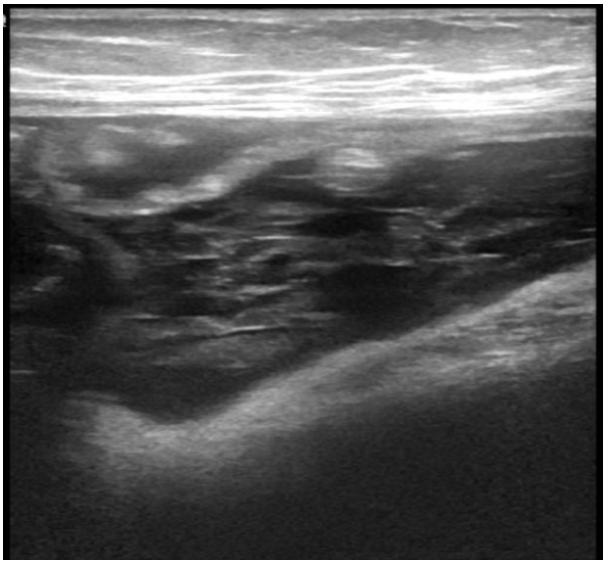

Figure 2: Ultrasound study showed right iliac fossa muti-sepated collection which close to the dilated appendix. No significant vascularity.

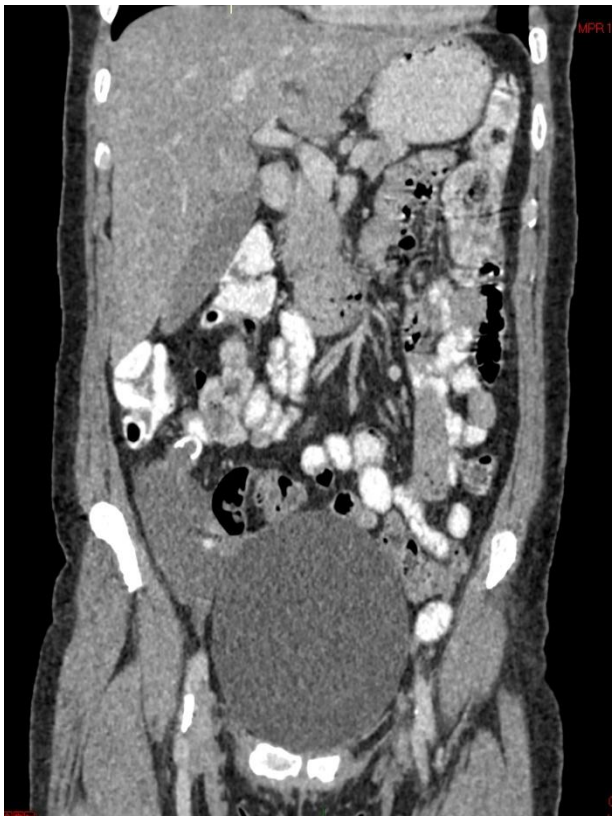

Figure 3: Coronal CT scan with intravenous and oral contrast elicit right iliac fossa low attenuation mass with calcification at its upper portion as well as wild wall thickening. No signs of peritoneal deposits.

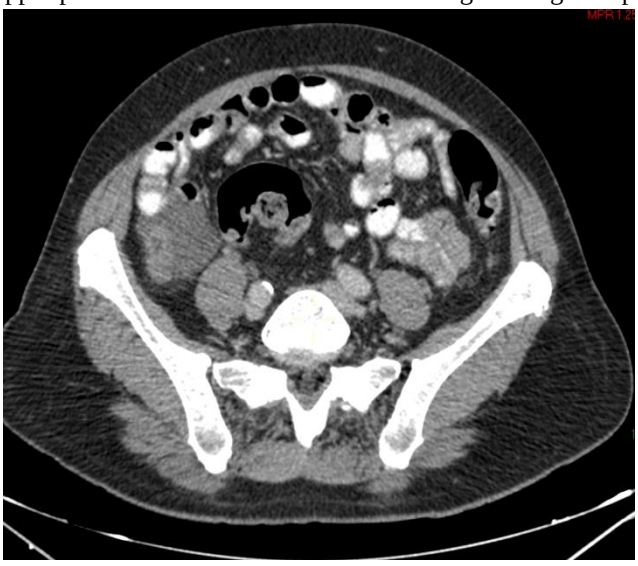

Figure 4: Axial CT scan images with intravenous and oral contrast: showedwell-circumscribed, low-attenuation, spherical mass contiguous with the base of the cecum.

The patient underwent a laparotomy through a midline incision. Based on CT finding and the experience of the surgeon, a mucocele of the appendix was suspected. Stomach, caecum and large bowel were found normal. The entire small bowel was also normal in appearance with no signs of deposits.

Pathologic analysis revealed a low-grade mucinous neoplasm. There was no perforation of the appendix, but there was extensive subserosal mucin accumulation, with mucin focally present at the inked serosal surface. The

surgical margin was negative for tumor.
Patient has been followed clinically as well as with serial imaging via CT and ultrasound for 2 years, with no evidence of any recurrence.

III. DISCUSSION

Mucinous neoplasms of the appendix are a heterogeneous collection of appendiceal epithelial neoplasms that can obstruct the lumen of the appendix while secreting excess mucin, and are thought to cause appendiceal mucocele. They are rare lesions, found in approximately 0.2-0.7% of pathology specimens following appendectomy (4).

A mucocele of the appendix typically presents as a pelvic mass. The mean age of diagnosis is 55 years, with a male to female preponderance of 4:1(5). Often, this malignancy is misdiagnosed as acute appendicitis, retroperitoneal tumors in the right iliac fossa, or an adnexal mass (6). our case was 60 years old femaleand was admitted as a case of cute appendicitis s after clinical presentation but the CT imaging support the diagnosis of appendicealmucocele.

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. 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 (6).

Most authors now agree that rupture of a mucinous neoplasm (mucocele), mainly of the appendix, followed by the ovaries, would result in the development of pseudomyxoma peritonei. Preoperative recognition of a possible mucocele based on CT scan findings can be done as long as a well-encapsulated mass with smooth regular wall is seen in the right lower quadrant with no periappendiceal inflammation or abscess (7)

In our case, there was no pathological evidence of malignancy infiltration into the bowel submucosa or lymph node metastasis and no evidence of malignant cells in the mucin pools in the peri-appendiceal tissue. Thus, further surgical and adjuvant therapies were not required in our patient.

In the setting of suspected appendicitis, when an appendiceal mucocele is encountered, the surgeon assumes extreme caution while handling the mucocele to avoid rupture and dispersion of mucus or epithelial cells into the peritoneal cavity as this is associated with a poorer prognosis (8).

IV. CONCLUSION

The mucinous neoplasm of the appendix is very rare condition, so the preoperative diagnosis by imaging modalities like CT in very important toavoid rupture .
Clear communication between the radiologist, pathologist and surgeon is important for optimal patient management.

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