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Case Report

Peritoneal Pseudomyxoma Complicating an Appendiceal Mucocele

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Abstract

Appendiceal mucocele (AM) is a rare condition characterized by mucinous distension of the appendix, which can remain asymptomatic for a long time and only be incidentally discovered during surgery or when a complication occurs. Intra-peritoneal rupture and the spread of mucinous fluid into the peritoneal cavity constitute a rare and serious complication. Here, we report a case of a 52-year-old patient diagnosed with a complicated appendiceal mucocele with pseudomyxoma peritonei and we emphasize the importance of considering this condition in the presence of an appendicular syndrome.

Keywords: *Mucocele; Appendix; Pseudomyxoma Peritonei; Imaging*

Introduction

Appendiceal mucocele (AM) is a rare entity of the appendix characterized by obstructive distension of the appendix due to the intraluminal accumulation of mucinous fluid, with an incidence ranging from 0.2% to 0.7% in all appendectomies, and a higher frequency in women and patients around 40 years of age [1]. The clinical presentation of appendiceal mucocele is nonspecific and it is often discovered incidentally during surgery for acute appendicitis. Rupture of the appendiceal mucocele and the spread of mucinous material into the peritoneal cavity leads to pseudomyxoma peritonei, which is the most feared complication and is associated with high mortality. CT scan is the intraoperative diagnostic method. Through this case, we report the imaging aspects of this pathology.

Case Report

A 52-year-old patient with no medical history was admitted to the emergency department for progressive abdominal distension developing over the past 8 months, associated with low-intensity abdominal pain localized in the right iliac fossa, intermittent postprandial vomiting, and a deterioration in general condition in an afebrile context. Laboratory tests show leukocytosis with a white blood cell count of 14,000 per mm^3 and an erythrocyte sedimentation rate of 36 mm in the first hour.

An abdominal ultrasound was performed, revealing a cystic distension of the appendix measuring 22 mm of diameter associated with significant ascites.

An abdominal-pelvic CT scan with contrast injection showed extensive peritoneal effusion causing scalloping on the liver and central displacement of digestive structures (Figure 1), along with a well-defined cystic mass in the right iliac fossa containing curvilinear mural calcifications and measuring 23 mm of transversal diameter (Figure 2). Omental infiltration in the left hypochondrium was also observed (Figure 3).

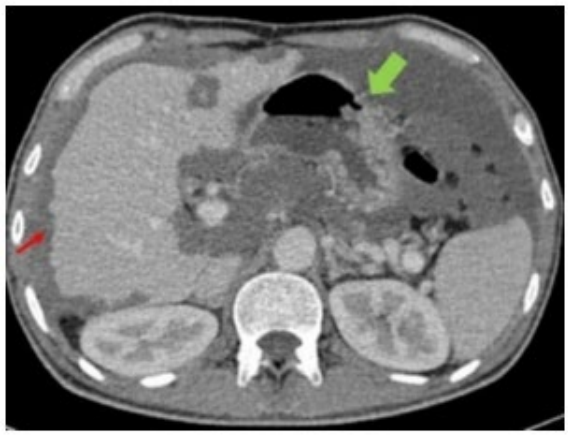

Figure 1: *Axial CT scan with contrast injection showing significant ascites causing hepatic scalloping (Red arrow) and central displacement of digestive structures (Green arrow).*

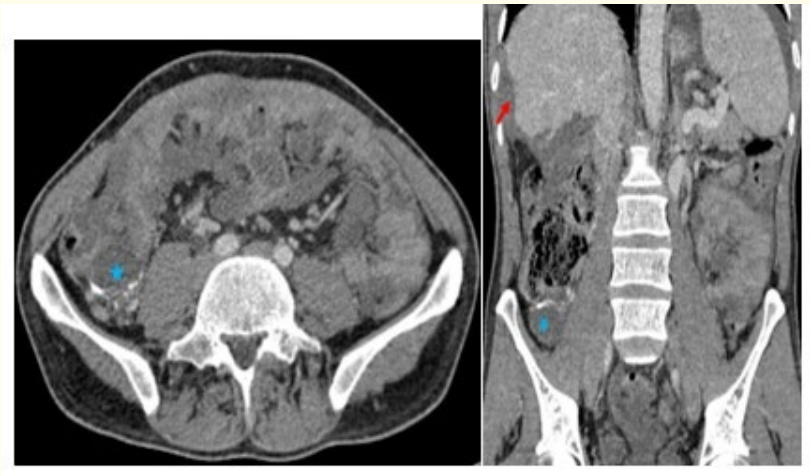

Figure 2: *Axial (A) and coronal (B) CT scan with contrast injection showing a Well-defined cystic mass in the right iliac fossa (Blue star) containing parietal calcifications, along with ascites causing hepatic scalloping (Red arrow).*

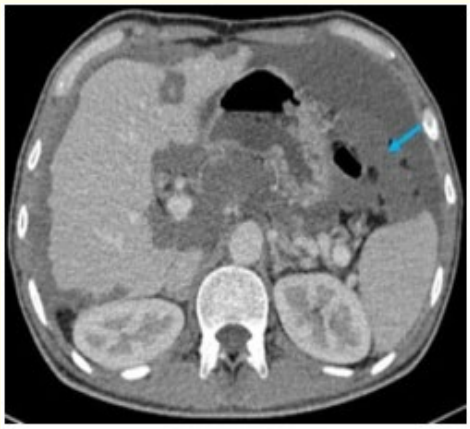

Figure 3: *Axial CT scan with contrast injection showing omental Infiltration in the left hypochondrium, with the "omental cake" appearance (Blue arrow).*

Based on this semiological analysis, the proposed diagnosis was a ruptured appendiceal mucocele with pseudomyxoma peritoneal. A laparoscopic biopsy of the peritoneal nodules confirmed the diagnosis of appendiceal mucinous cystadenoma complicated by pseudomyxoma peritonei. An appendectomy, omentectomy, and localized peritoneal resection were performed.

Discussion

Appendiceal mucocele is a rare condition found in 0.2 to 0.7% of appendectomies [1], with the most feared complication being pseudomyxoma peritonei. Described for the first time by Rokitansky in 1842 [2], it corresponds to mucinous dilatation of the appendix, which can result from obstruction by a fecalith, inflammatory stenosis, or extrinsic compression. Etiologies also include villous epithelial hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. The cystadenoma and cystadenocarcinoma are neoplastic appendiceal mucoceles, comprising approximately 35% of all primary tumors of the appendix [3,4]. These conditions can develop de novo or from preexisting simple mucoceles.

It mainly affects adults, with an average age between 50 and 60 years [5], as observed in our case. However, appendiceal mucocele can also affect children, as reported by Duquenoy [6], who described this condition in 5 children aged 4 to 13 years with cystic fibrosis. Regarding gender, the male-to-female ratio varies across studies, with a predominance of females in recent research [7,8]. However, a male predominance was noted by Souei-Mhiri [5], as observed in our case.

The clinical presentation is nonspecific; appendiceal mucocele may present with acute or chronic abdominal pain localized to the right iliac fossa or sometimes with a palpable mass. It is most often discovered incidentally during imaging or upon the occurrence of a complication [9]. In cases of a tumor-related cause, the presentation is more pronounced.

On ultrasound, an appendiceal mucocele presents as a well-defined, oblong cystic mass with a partially calcified wall and echogenic, layered content, resembling an “onion-skin” appearance. A transverse diameter greater than 15 mm has a sensitivity of 83% and a specificity of 92% for diagnosing an appendiceal mucocele [10]. On computed tomography (CT), an uncomplicated appendiceal mucocele appears as a well-defined, cystic mass in the right iliac fossa with liquid density. Arcuate mural calcifications are not always present, but their presence supports the diagnosis of an appendiceal mucocele [11]. The histopathological examination confirms whether the Mais benign or malignant.

In MRI, the contents of the MA appear as hypointense on T1-weighted images and hyperintense on T2-weighted images. Calcifications are visualized less distinctly than in CT scans.

Intraperitoneal rupture is the most feared complication responsible for pseudomyxoma peritonei (PMP). It is defined by the dissemination of the mucous content of the appendiceal mucocele into the abdominal cavity. Abdominal CT scan is the preferred examination for diagnosing PMP and specifying its characteristics: the typical appearance of PMP is "scalloping" on the surface of solid organs, notably the liver and spleen [12]. It may contain fine septa and curvilinear calcifications. Peritoneal implants are less typical, often located in areas of dependent stasis, particularly the Douglas pouch and paracolic gutters. These nodules may coalesce to form the classic "omental cake" at the greater omentum. The definitive positive diagnosis is obviously anatomopathological, relying on a biopsy or transcutaneous puncture yielding gelatinous fluid, or through biopsies during laparoscopy.

In the case of an unruptured mucocele, surgical treatment is recommended with a favorable prognosis, whether it is benign or malignant [13]. It is recommended to perform an excision of an appendicular mucocele during an appendectomy, absolutely without breaking its wall [14] in order to avoid its rupture which is a serious complication.

The treatment of pseudomyxoma peritonei is similar for all types: radical tumor reduction surgery including omentectomy, localized peritoneal resection, and right hemicolectomy, with splenectomy, cholecystectomy, and bilateral ovariectomy in women [15]. Occasionally, it is combined with intraperitoneal chemotherapy-hyperthermia in cases of malignancy. The treatment of malignant PMP is managed by specialized centers.

Conclusion

In the presence of a discovered or persistent mass in the right iliac fossa or an appendicular syndrome, the diagnosis of appendiceal mucocele, although rare, deserves consideration. However, an early preoperative diagnosis of unruptured MA by the radiologist through the performance of an ultrasound or a CT scan alerts the surgeon to the risk of rupture during surgery and helps avoid pseudomyxoma peritonei. The management typically involves a simple appendectomy in the majority of cases, but a right hemicolectomy should be systematically performed when clear signs of local malignancy are present or confirmed by the histopathological analysis of the surgical specimen. The occurrence of pseudomyxoma peritonei renders the prognosis much graver and encourages an expanded therapeutic approach.

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