Case report

Appendiceal mucocele presenting as a leading point in ileocolic intussusceptions: “Case report”

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ABSTRACT

Introduction and importance: Appendiceal mucocele is a rare clinical scenario, which is found in 0.2–0.7 % of appendectomies. Ileocecal/ileocolic intusception caused by appendiceal mucocele is an extremely rare condition with few case reports in literatures. The treatment is surgery with the extent determined by intra-operative findings. Ultrasound and CECT scan can suggest the diagnosis, but definitive diagnosis is by histopathology. The aim of this presentation is to discuss appendiceal mucocele in terms of clinical features, diagnostic imaging and treatment. This case report can create awareness to primary care physicians, radiologists, surgeons and pathologists aiding in accurate diagnosis and early surgical intervention to prevent rupture.

Presentation of the case: A fifty years old woman presented with intermittent colicky peri umbilical abdominal pain of one-week duration. She had nausea, vomiting, mild abdominal distension, and failure to pass feces and flatus. Physical examination was normal. Imaging suggested ileocolic intussusceptions with cystic leading point on ultrasound, but on CECT scan, no leading point reported. Appendiceal mucocele diagnosed intra operatively and confirmed by pathology.

Conclusion: Appendiceal mucocele is rare and can be benign or malignant. Preoperative diagnosis is often difficult. Definitive diagnosis is by histopathology. Appendiceal mucocele can rarely present with ileocolic intussusceptions. Radiologists, pathologists, primary care physicians and surgeons must be aware of this condition. Accurate preoperative diagnosis and early surgical treatment of appendiceal mucocele is important to prevent complications like pseudomyxoma peritonei(PMP), which has poor prognosis.

1. Introduction

Appendiceal mucocele is a descriptive term, given to cystic dilatation of the appendix as a result of obstruction by benign or malignant pathology and seen in 0.2–0.7 % of appendectomies [1]. It is classified into retention cysts, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma [2]. The term “mucocele” is recently discouraged by some authors for more specific pathological terms, though it is still in use by radiologists and surgeons [3]. Appendiceal mucocele is more common in women and age of occurrence ranges from 50 to 60 years [4].

Clinical presentation is often delayed, as symptoms may be absent or mild. Lower abdominal pain or mass can be confused for appendicitis or tubo ovarian mass in female [5]. Diagnosis of appendiceal mucocele is often difficult preoperatively, even with the use of imaging. Definitive diagnosis is by histopathology. Treatment is surgical resection with care not to spill the contents, to prevent pseudomyxoma peritonei which has poor prognosis [6]. Here, a rare case of ileocolic intussusceptions caused by appendiceal mucocele is presented. This work is reported in line with the SCARE 2020 criteria [7].

2. Presentation of the case

A 50 years old homemaker, married woman from Adama town, East Shoa, Ethiopia presented with complaint of intermittent colicky periumbilical abdominal pain of one-week duration. In association with the pain, she had nausea, three episode of vomiting of ingested matter, mild abdominal distension, and failure to pass feces and flatus of one-day duration. She had history of mild abdominal discomfort that was not worrisome for the last 4 months. She has no history of smoking, alcohol ingestion, diabetes or hypertension. Physical examination showed normal vital...
signs, soft abdomen with no mass or tenderness, and digital rectal examination was not revealing.

Complete blood count (CBC), serum electrolytes, liver function tests and renal function tests were normal. Abdominal ultrasound showed right side concentric ring of bowel with diameter of 5.6 cm and length of 10 cm and 4.4 cm cystic mass as a leading point. Ultrasound diagnosis was ileocolic intussusceptions with cystic mass as a leading point. Differential diagnosis suggested by US was Meckel’s diverticulum or Duplication cyst. Abdominal CECT scan showed telescoping of terminal ileum in to the cecum and ascending colon 15 cm long and 7 cm wide, no leading point seen concluding long segment ileocolic intussusceptions. CT scan reevaluated post operatively and showed a teardrop shaped cystic mass as a leading point (see Fig. 1 below). Chest X-ray and ECG are normal.

Exploratory laparotomy showed minimal reactive fluid; partly reduced intussusceptions, edematous cecum, and whitish distended tense appendix with its base and edematous cecum in the ascending colon (see Fig. 2 below). Right colectomy with ileotransverse end-to-end anastomosis was done, as malignancy could not be ruled out (see Figs. 3 and 4 below). Histopathology revealed appendiceal mucocele as a lead point and no malignant cells seen. Postoperative recovery was uneventful. Patient discharged on the sixth postoperative day.

3. Discussion

Appendiceal mucoceles are rare tumors commonly seen in women of 50 to 60 years of age [4]. They are encountered in 0.2–0.7 % of appendectomy specimens [1]. Most of appendiceal mucoceles are an incidental encounter at the time of imaging, colonoscopy or surgery for unrelated conditions or during pathological assessment of appendectomy specimens [6].

Patients with AM, are mostly asymptomatic or present with acute or chronic lower abdominal pain and palpable right lower quadrant mass creating diagnostic confusion with appendicitis [8]. In women, it may be difficult to distinguish AM from adnexal pathology with palpable pelvic mass, on imaging or even intraoperatively [9–11]. Patients with AM can also present with change in bowel habits, nausea, vomiting, genitourinary symptoms from local compression [12]. AM can also present with intussusceptions, bleeding per rectum and bowel obstruction [5,13]. Ileocecal/ileocele intussusception secondary to AM is a rare clinical scenario with limited case reports [13–16]. The case presented in here also came with abdominal pain, nausea, vomiting and ileocolic intussusceptions on imaging.

AM is a diagnostic challenge as a result of atypical clinical presentation and its rare occurrence, leading to misdiagnosis or delayed diagnosis [5]. This in turn can cause inappropriate treatment or delayed treatment with the chance of rupture and PMP with poor prognosis [3,6]. Hence, in-depth knowledge and awareness of this condition is important to make early diagnosis and suggest appropriate management.

Differential diagnosis of AM can include mesenteric cyst, ovarian tumors, tubo-ovarian abscess, peri appendiceal abscess, intussusceptions due to colonic mass and enteric duplication cyst [14].

CT scan and ultrasound can propose AM with acceptable level of
confidence of neoplasm [5]. However, definitive diagnosis is by histopathological examination. Ultrasound characteristic of AM includes “onion skin” appearance, nodular enhancement of the wall and diameter greater than 1.5 cm. Appearance of AM on CT scan is commonly a low attenuating lesion often with curvilinear calcification is considered the investigation of choice. Once mucocele is seen on imaging, colonoscopy should be done to rule out synchronous colonic lesions which can be seen in 13–42 % of appendiceal neoplasms and cecal invasion from appendiceal tumors [5]. Both ultrasound and CT scan failed to suggest AM in the case presented here.

CT scan appearance of intussuscepted AM is rarely reported with few reports in the literature [17]. The finding consistent with ileocolic intussusceptions with AM acting as a lead point is tubular low density mass in the colon with peripheral calcification and a portion of small bowel mesentery pulled into the colon. Coronal reformations can improve visualizations of the AM lead point, and increase diagnostic accuracy of intussusceptions.

Even though, not specific, tumor markers like CEA, CA 19-9, and CA-125 needs to be determined after diagnosis of AM and repeated to monitor disease progression [5].

Once the diagnosis of AM is made, early surgical resection is recommended for all to exclude mucinous neoplasm, prevent spontaneous rupture and PMP which has poor prognosis [18]. There is no consensus regarding the optimum surgical procedure for AM [19]. Currently the standard surgical treatment for AM is open surgery despite the growing evidence is in favor of laparoscopic approach. The extent of surgery should be guided by pathologic diagnosis and depends on several factors like tumor size, location, mucin content, ileal and cecal involvement, lymph node involvement, and margin status [6]. Abdominal exploration need to exclude co-existing ovarian and colonic tumors additionally [5].

Treatment of AM based on intra operative finding can be simple appendectomy if the base is free, partial cecectomy, ileocecectomy or right colectomy can be performed if the base is involved following oncologic principles [5]. As reduction to allow limited resection is not recommended for intussusceptions caused by AM to prevent rupture, right colectomy is the treatment [13]. This is also true for the case presented here.

For ruptured and walled off lesion right colectomy, thorough peritoneal wash and referral to specialized centers if positive pathologic findings of the spillage for subsequent management. If PMP is diagnosed, the treatment is cytoreductive surgery and heated intra peritoneal chemotherapy (HIPEC) [3].

For non-neoplastic mucinous lesions appendectomy is enough even if it get ruptured [5]. Prognosis is dependent on the presence and extent of peritoneal invasion and spread that can determine the recurrence. Five years survival rate for the simple AM is 91–100 % that drops to 25 % for the malignant AM after appendectomy [5].

4. Conclusion

AM is a rare clinical scenario that can be benign or malignant. Even though, extremely rare AM can present as a lead point in intussusceptions. Radiologists, pathologists, primary care physicians and surgeons must be aware of AM, as accurate preoperative diagnosis is essential for early appropriate surgical intervention and postoperative care. Though laparoscopy is increasingly in use, open surgery is the standard of care for AM. Right colectomy is needed for ileocolic intussusceptions caused by AM. Delay in diagnosis and treatment of AM can lead to complications like PMP, which has poor prognosis.

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Ethical approval

The study is exempt from ethical approval in our setup.

Author contribution

Single author did all the work.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

Not applicable.

Guarantor

Gosa Bejiga.

Declaration of competing interest

There is no conflict of interest.
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