Ileocecal Intussusception due to Appendiceal Mucocele in a Young Male Patient

PO-CHANG WANG¹, YI-FANG WU², I-CHUAN CHEN², CHENG-TING HSIAO²

Intussusception caused by appendiceal mucocele is extremely rare. Only few case reports were documented and most of those cases were female. We presented a 28 year-old male patient presented to our emergency department with intermittent epigastric and right lower abdominal pain for one week. Vomiting and bloody stool occurred one day prior to his arrival at our ED. Abdominal CT showed one appendiceal mass with intussusception, which was confirmed as an appendiceal mucocele with ileocecal intussusception in a laparotomy. The patient is free of illness for eleven months after operation.

Key words: appendiceal mucocele, intussusception, right lower abdominal pain

Introduction

Intussusception in adults is usually associated with pathological origins. Intussusception caused by appendiceal mucocele is extremely rare, accounting for only 0.25% of patients undergoing an appendectomy (1) and 0.4% of Taiwanese population in a 10-year study included 4038 patients (2). About 20% to 50% of appendiceal mucocele patients may present features indicative of acute appendicitis (3). Here, we report a young male patient presenting abdominal pain that migrated from the epigastric to the right lower abdomen. Abdominal CT showed one appendiceal mass with intussusception, which was confirmed as an appendiceal mucocele with ileocecal intussusception in a laparotomy. The patient is free of illness for eleven months after operation.

Case Report

A 28 year-old male patient presented to our emergency department (ED) with intermittent epigastric and right lower abdominal pain for one week. Symptoms of vomiting and bloody stool occurred one day prior to his arrival at our ED. He did not have any previous surgeries or systemic diseases, and vital signs on presentation were stable. Physical examination revealed epigastric abdomen and McBurney’s point tenderness, while laboratory studies showed leukocytosis. Plain X-rays of abdomen showed no mass and normal gas patterns. Abdominal CT revealed swelling of the appendix (14 mm) with intussusception (Fig. 1) and one appendiceal mass (Fig. 2). A laparotomy revealed appendicitis and ileocecal intussusception caused by an appendiceal mucocele. Appendix swelling of 1.5 cm and a tumor of 5 cm at the appendiceal opening were noted during operation (Fig. 3). The pathological examination of the specimen showed ruptured mucinous cystadenoma with mucus seeding on the adjacent mesentery. Right hemicolecotomy with side to side ileocolic anastomosis was performed.
Fig. 1, 2 Abdominal CT revealed swelling of the appendix with intussusception (Fig. 1) which was probably due to appendix mucocele (Fig. 2)

Fig. 3 Appendix swelling of 1.5 cm and a tumor of 5 cm at the appendiceal opening were noted during operation
anastomosis was performed. The patient is free of illness for eleven months after operation.

**Discussion**

Abdominal pain is a main presenting symptom in ED patients. An accurate diagnosis of appendicitis in patients with acute abdominal pain that migrate from the umbilicus to the right lower quadrant has been estimated to be nearly 95%. Patients with typical symptoms may be referred for an emergency appendectomy. Appendiceal mucocele accounts for 0.25% of patients undergoing an appendectomy and 0.4% in Taiwanese population in a 10-year study included 4038 patients.

Appendiceal mucocele (AM) is a rare disease which causes a distension of the appendix via an abnormal mucus accumulation. It has been histologically divided to four subtypes: retention cysts, mucosal hyperplasia, cystadenoma, and malignant tumors as cystadenocarcinoma with relative frequencies of 18, 20, 32, and 10%, respectively. Patients with AM are commonly diagnosed using a sonogram by accident, since one quarter of patients have no symptoms prior to diagnosis. Clinical evidences from other studies have demonstrated that about 20% to 50% of AM patients presented with features indicative of acute appendicitis. However, an accurate pre-operative diagnosis is essential for resection to prevent rupture during surgery. A rupture of AM may result in peritoneal contamination with development of pseudomyxoma peritonei, which has a 5-year survival rate of only 53% to 75%.

In our case, the patient is a 28 year-old young male, which is unusual. Based on previous studies, AM was predominantly diagnosed in female patients with a female to male ratio of 3:1. Also, the average age at the time of diagnosis is 54 years old for benign mucoceles and 64 years old for malignant ones. To our knowledge, there are no case reports of AM with intussusception in a young male. He had epigastric and right lower abdominal pain for a week, which could be mistaken as appendicitis. However, upon reviewing his medical history and noting the presence of bloody stool, which is an uncommon symptom of appendicitis, we suspected a different diagnosis. The most common symptom of AM is a right lower quadrant abdominal pain (64%) and about 25% of patients are asymptomatic at the time of diagnosis. Other symptoms include an intestinal intussusception, torsion, urethral obstruction, and hematuria. Because AM may present symptoms of appendicitis, more attention should be paid to unusual symptoms when managing patients with RLQ pain, including bloody stool, which could be indicative of intussusception.

Modalities in helping diagnosis of AM include abdominal sonogram, barium enema, colonoscopy and computed tomography. An appendiceal threshold diameter of 15 mm in a sonogram is the optimal threshold for AM diagnosis, with a sensitivity of 83% and a specificity of 92%. Compared with AM, an outer diameter threshold of 6 mm has been established for an acute appendicitis diagnosis. On a CT, the presence of curvilinear or punctate wall calcifications in a right lower quadrant cystic lesion strongly suggests a diagnosis of the mucocele of the appendix. Displacement of the adjacent bowel without periappendiceal inflammation or abscess is the key differential point in discriminating AM from acute appendicitis. There are only limited reports comparing the diagnosis of AM by sonography, barium enema, colonoscopy and abdominal CT.

Appendectomy is the treatment of choice. Non-surgical management cannot be accepted, because apparently benign lesions can progress to mucinous cystadenocarcinoma, and the rupture of mucocele may determine the development of pseudomyxoma peritonei. Patients with simple
mucocele, mucosal hyperplasia and mucinous cystadenoma have shown an excellent prognosis with 5-year survival rates of 91-100%. However, the 5-year survival rate is markedly decreased to 25% in malignant mucoceles due to complications of pseudomyxoma peritonei (10). It has been reported in some cases that pseudomyxoma peritonei originated from mucoceles other than cystoadenocarcinoma. To prevent the rupture of the mucocele and to evaluate the presence of mucoid fluid accumulations, an open laparotomy is preferred to a laparoscopy when AM is suspected (11). A simple appendectomy is reliable with uncomplicated and unruptured mucoceles (12). In mucinous cystoadenocarcinoma and mucocele with invasion to the cecum or ileum, a right hemicolectomy may be needed. If a ruptured appendiceal mucocele is suspected, the primary resection should be accompanied by the removal of all gross implants (13). Follow-up is recommended, because there are cases of recurrences as pseudomyxoma peritonei and instances of metachronic colonic neoplasms.

Appendiceal mucocele is related to tumors and about 11-20% of patients have adenocarcinoma of the colon (14). A correlation with tumors and the possibility of a major complication during surgery have put emphasis on the importance of a correct diagnosis prior to operation. We should pay more attention to unusual presentations and keep appendiceal mucocele in mind when approach patients with right lower abdominal pain.

References
年輕男性的闌尾黏液囊腫
引起迴腸盲腸套疊病例報告

王博章1  吳宜芳2  陳怡娟2  蕭政廷2

因闌尾黏液囊腫而引起的腸套疊非常稀少，文獻上只有幾個病例報告且大部分是女性病患。我們報告一個二十八歲男性病患，因為上腹部與右下腹痛而至急診就診。病患於就診前一天開始出現嘔吐與腹瀉，且伴隨有血便。腹部電腦斷層發現闌尾腫瘤伴隨腸套疊，手術中證實為闌尾黏液囊腫。

關鍵詞：闌尾黏液囊腫，腸套疊，右下腹痛

---

收到：98年5月20日   接受刊載：98年12月30日
長庚醫療財團法人嘉義長庚紀念醫院1心臟內科   2急診醫學科
通訊及摘要索取：吳宜芳醫師
嘉義縣朴子市嘉朴西路6號   長庚醫療財團法人嘉義長庚紀念醫院急診醫學科
電話：0937-790909, 0982-630274
E-mail: yvonnearea@yahoo.com.tw