

Intestinal Obstruction Secondary to Appendiceal Mucoceles

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Rezumat

Ocluzie intestinală prin mucocel apendicular invadant în două anse enterale

Puține cazuri de ocluzie intestinală secundară unui mucocel apendicular sunt citate în literatura de specialitate. Prezentăm cazul unei paciente care se prezintă în urgență pentru dureri abdominale difuze, grețuri, vărsături și întreruperea tranzitului intestinal. Examenul obiectiv: abdomen destins, sensibil difuz la palparea profundă, cu timpanism la percuție. Radiografia abdominală pe gol evidențiază anse enterale dilatate cu nivele hidro-aerice, iar probele de laborator prezența leucocitozei. Se intervine chirurgical de urgență cu diagnosticul de ocluzie intestinală. Intraoperator se decelează o formațiune tumorală apendiculară invadantă în două anse enterale. Se practică apendicectomie în bloc cu două rezecții segmentare enterale și restabilirea continuității digestive prin două anastomoze entero-enterale T-T. Examenul anatomopatologic pune diagnosticul de chistadenom mucinos apendicular. Evoluția postoperatorie este favorabilă, pacienta fiind externată în ziua 7 postoperator. Controalele periodice efectuate la 3, 6, 12 și 24 de luni (inclusiv colonoscopia) relevă relații normale, fără semne de recidivă.

Cuvinte cheie: apendice, mucocel, ocluzie intestinală, chistadenom mucinos

Abstract

Few cases of intestinal obstruction complicating an appendiceal mucocele have been recorded. We report the case of a young woman who presented to the emergency room with diffusely abdominal pain, nausea, vomiting and disruption of bowel movements. Her abdomen was mildly distended and tympanic. A flat film of the abdomen revealed dilated small bowel loops with air-fluid levels suggestive of small bowel obstruction. She also had leukocytosis. An emergency operation was performed under the diagnosis of intestinal obstruction. The intraoperative findings showed a tumoral appendiceal mass permeated into the ileum in two distinct points, causing an enteral stenosis. We performed an appendectomy "en bloc" with two enteral loop resections of the permeated ileum followed by two T-T enteral anastomoses. The pathologic examination revealed appendiceal mucinous cystadenoma. Postoperative course was favorable, the patient being discharged on the seventh postoperative day. Postoperative checks performed at 3, 6, 12 and 24 months (including colonoscopy) have not showed pathological changes.

Key words: appendix, mucoceles, intestinal obstruction, mucinous cystadenoma

Introduction

Mucocele is an uncommon pathology of the appendix characterized by a cystic dilation of the lumen as a result of abnormal accumulation of mucus. The incidence ranges between 0.2% and 0.3% of all appendectomies. It can be asymptomatic, but acute or chronic pain in the right iliac fossa is the most frequent symptom (1). Intestinal obstruction complicating an appendiceal mucocele has rarely been reported (2,3). We

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describe the case of a young woman who came into the emergency room with signs and symptoms of intestinal obstruction due to appendiceal mucocoele.

Case presentation

We report the case of a 39 years old, previously healthy woman, who presented to our emergency room with a 16-hour history of acute and diffusely abdominal pain, nausea, vomiting and disruption of bowel movements. Her abdomen was mildly distended and tympanic with diffuse tenderness, especially in the periumbilical areas. She had no bowel sounds. A flat film of the abdomen revealed dilated small bowel loops in the mid-abdomen and left upper quadrant with air fluid levels suggestive of small bowel obstruction. The leukocyte count was 13 300 with 85.3% neutrophils. Under these conditions, we did not carry out any other paraclinical examinations and an emergency operation was performed under the diagnosis of acute intestinal obstruction.

After a quick preoperative treatment, the patient underwent a laparotomy that revealed dilated enteral loops with air-fluid content and a whitish mass with a glistening outer surface extending from the distal half of the appendix to the terminal ileum (Fig. 1). This tumoral mass permeated the ileum in two distinct points at a 50 cm distance between them, causing an enteral stenosis (Fig. 2).

We performed an appendectomy “en bloc” with two enteral loop resections of the permeated terminal ileum (Fig. 3) followed by two T-T entero-enteral anastomoses (in both areas of enteral resection).

The pathological examination revealed that the intestinal obstruction was secondary to an appendiceal mucinous cystadenoma permeated into two enteral loops, causing an extrinsic stenosis of the terminal ileum.

Postoperative course was favorable, the patient being discharged on the seventh postoperative day. Postoperative checks performed at 3, 6, 12 and 24 months (including colonoscopy) have not showed pathological changes.

Discussion

The term “appendiceal mucocoele” belongs to a heterogeneous group that includes 4 histopathological findings: simple mucocoele or retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma (1,4).

The first three types are the most common, comprising over 90% of all cases, while mucinous cystadenocarcinoma is less frequent. Mucinous cystadenomas and cystadenocarcinomas of the appendix are often referred to as neoplastic mucocoeles of the appendix. (5,6)

Appendiceal mucocoele is more frequent in individuals over 50 years of age (4,5) although our patient's age was well below this limit.

Clinical presentation is quite variable. A quarter of patients are asymptomatic and the tumor is discovered incidentally, although the most common presentation is acute or chronic right lower quadrant pain (64%). An abdominal palpable mass



Figure 1. Cystic dilatation at the top of the appendix (Appendiceal mucocoele)

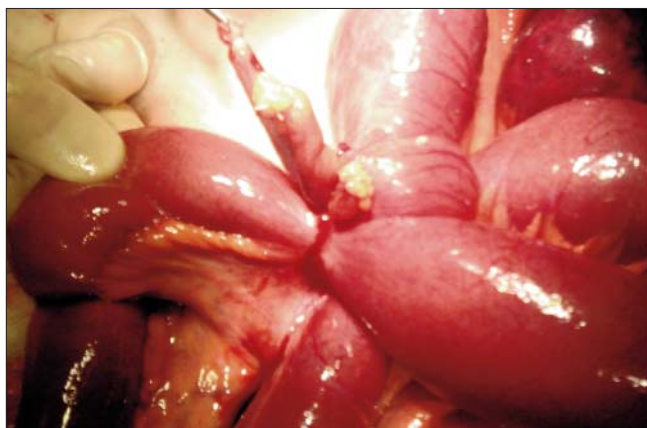


Figure 2. Appendiceal mucocoele permeated into the ileum in two distinct points

can be found in 50% of cases, whereas low gastrointestinal bleeding or genitourinary symptoms may be present (1,6,7).

In our case, the patient came into the emergency room with signs and symptoms of intestinal obstruction, and the differential diagnosis at presentation included colonic tumor, diverticular disease, benign small intestinal stricture secondary to Crohn's disease or tuberculosis. This is a particular aspect of this case, because intestinal obstruction complicating an appendiceal mucocoele has rarely been reported (2,3). Moreover, the mechanism of obstruction is different from those described in literature (compression of ileocecal junction by mucocoele or intussusceptions of the tumor). In our case, the tumoral mass was permeated into two distinct enteral loops, causing an extrinsic stenosis of the terminal ileum that required an enteral resection “en bloc” with an appendectomy.



Figure 3. Resected specimen

The worst complication of appendiceal mucocoele is pseudomyxoma peritonei ("the jelly belly"). It is characterized by peritoneal dissemination of mucoid material caused by spontaneous or iatrogenic perforation of the appendix. This mucoid material may be acellular or can contain cells with different grades of dysplasia (8). One has to keep in mind that as long as the appendix is intact, the outcome of the patient is secure. That's why we prefer to perform an "en bloc" resection of two enteric loop and appendix, in order to prevent a rupture of the appendix with leakage of the mucoid material into abdominal cavity.

Preoperative diagnosis of mucocoele has been considered exceptional, in the past. Today the number of cases diagnosed preoperatively has increased due to modern imaging techniques. On barium enema, there is usually no filling or partial filling of the appendix with contrast. Ultrasound reveals a cystic mass in the appendix area. Multiple echogenic layers along with a dilated appendix produce the appearance of "onion-skin" circles on US. CT of the abdomen usually shows a cystic well-encapsulated mass. Colonoscopy findings include the 'volcano sign', moving in and out with respiration. Increased levels of tumor markers (CEA, CA 19.9) have been reported in primary neoplastic mucocoeles and disease recurrence (1,3,4,6,7).

In our case, the US had no clinical value due to the large amount of gas in the gut; a plain radiography of the abdomen showed multiple gas-fluid levels with distended bowel. Under these conditions, we did not carry out any other paraclinical examinations and an emergency operation was performed under the diagnosis of acute occlusive abdomen.

Although it was not our case, it is important to keep in mind that other tumors are associated with appendiceal mucocoele. In the Mayo Clinic series, the rate of synchronous tumors was as high as 29% (8). The most common synchronous neoplasms occur in the large bowel, although they can also be found in other locations, such as the gallbladder, breast, kidney, ovary and thyroid (5,7,9).

The only accepted therapy is surgical. The main concern of surgical treatment is to keep the appendiceal mucocoele intact. Laparoscopic approach is possible, as long as the integrity of the appendiceal lumen is not jeopardized, although the laparoscopic dissection and grasping of the appendix specimen increase the risk of peritoneal dissemination (11).

The type of surgical treatment is related to the dimensions of the mucocoele, the extent of the disease, and the pathology findings (1,5,6). Appendectomy is performed for simple mucocoele or for cystadenoma, when the appendiceal base is intact. Right hemicolectomy is recommended if the appendix shows histology of mucinous cystadenocarcinoma and if lymph nodes metastases are present. Aggressive debulking, adjuvant radiotherapy and chemotherapy are recommended mostly after rupture of the malignant mucocoele and pseudomyxoma peritonei (1,5,6,11,12).

Mucocoele of the appendix is a rare clinical finding, especially in a young patient. The particularity of the presented case derives from the clinical presentation (acute intestinal obstruction) and the surgical management of this lesion (an appendectomy "en bloc" with two enteric loop resections, in order to avoid breaking the appendix and the intraperitoneal spillage of mucoid content).

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