Mucocele of the appendix, therapeutic attitude

A case report

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Abstract

The appendicular mucocele is a rarely encountered pathology in clinical practice, with an occurrence rate of 0.2% of all appendectomies, it usually affects people older than 50 years. We present the case of a patient, 40 years old, with no significant pathological and heredocolaterale history, which was hospitalized in our clinic for abdominal pain in the right iliac fossa, and finally we remember the principles of the terapeutical attitude for the appendicular mucocele.

Keywords: *appendicular mucocele, peritoneal pseudomixoma, chemotherapy*

Introduction

The present case try to analyze the medical literature regarding the appendix mucocele linked to diagnosis and treatment. First we present the case of a patient diagnosed with simple appendicular mucocele.

The appendicular mucocele is a rarely encountered pathology in clinical practice, with an occurrence rate of 0.2% of all appendectomies, affecting usually people older than 50 years[(1)]. There are controversies regarding the gender it most often affects, most likely caused by geographical areas[(2)].

There are regions where the incidence of disease is higher in women and others where the ratio is changing and men are more commonly affected[(3)]. The appendicular mucocele is defined as a slow distension of the appendix filled with sterile mucoid collection[(4)]. The pathophysiologic mechanism includes the obstruction of the appendix lumen towards the base by a foreign body, a tumor or stenosis. If in the lumen the content is sterile, the mucous cells continue to secrete until, due to the overgrowth of pressure, the wall is thinning and affects cell nutrition[(5)].

The appendicular mucocele can be divided into three categories: (a) mucocele retention with normal mucosa found in 5-25% of cases; (b) mucinous cystadenoma with modified surface of the epithelium, similar to that of the adenomatous polyps and villous adenomas in 63-84% of cases; (c) mucinous cystadenocarcinoma with epithelium, similar to that observed in the colon adenocarcinoma, in 11 to 20% of cases[(6)]. The tumor formation, in terms of which it is presented, can grow until it is transabdominal palpable[(7)].

The rupture of the mucocele can cause gelatin pseudomixom (i.e. gelatin peritonitis), which is characterized by the implantation of mucinous epithelial cells in the peritoneum and mucus accumulation in the peritoneal cavity, which often leads to the development of adherence clamps and intestinal obstruction. In a study by Ronnet on a number of 109 cases, peritoneal pseudomixoma had a survival rate of 84% for histopathological diagnosis of mucinous adenoma and of 6.7% for mucinous adenocarcinoma[(6)].

A guiding criterion regarding the etiology of the mucocele is the size of the base of the appendix[(8)]. Benign forms have a smaller diameter, generally less than 2 cm, while the base of the appendix that is greater than 2 cm presents an increased risk of malignancy[(9)]. As useful laboratory investigations we mention abdominal ultrasound, computed tomography (CT), and colonoscopy[(10,11)]. Imaging investigations reveal the presence of a well-defined tumor formation, with fluid. Colonoscopy reveals an erythematous area, protrusion, with a crater in the center, a sign that is known in the literature as volcano sign[(11)].

Treatment of mucocele appendix is surgery. Surgical attitude is dictated by several factors: integrity of the appendicular wall, the base diameter of the appendix, and extemporaneous examination result.

Case Report

We present the case of a patient, 40 years old, with no significant pathological and heredocolaterale history, which was hospitalized in the Department of Surgery from Central Military University Emergency Hospital "Dr. Carol Davila" from Romania for abdominal pain in the right iliac fossa.

Clinical examination of the abdomen reveals a palpable tumor formation in the right iliac fossa, approximately 7/5 cm in size, well defined, mobile in regards to the superficial and deep plans, painful.

Paraclinic exams included routine laboratory tests, with exception of a leukocytosis (12.000/mm^3), which do not showed other pathological deviations, tumor markers (carcinoembryonic antigen, CEA 125) - negative.

Ultrasonography showes a heterogeneous cystic structure, hipoecogenic, transonic. CT described a rectrocecal mass, well defined, encapsulated, with re-

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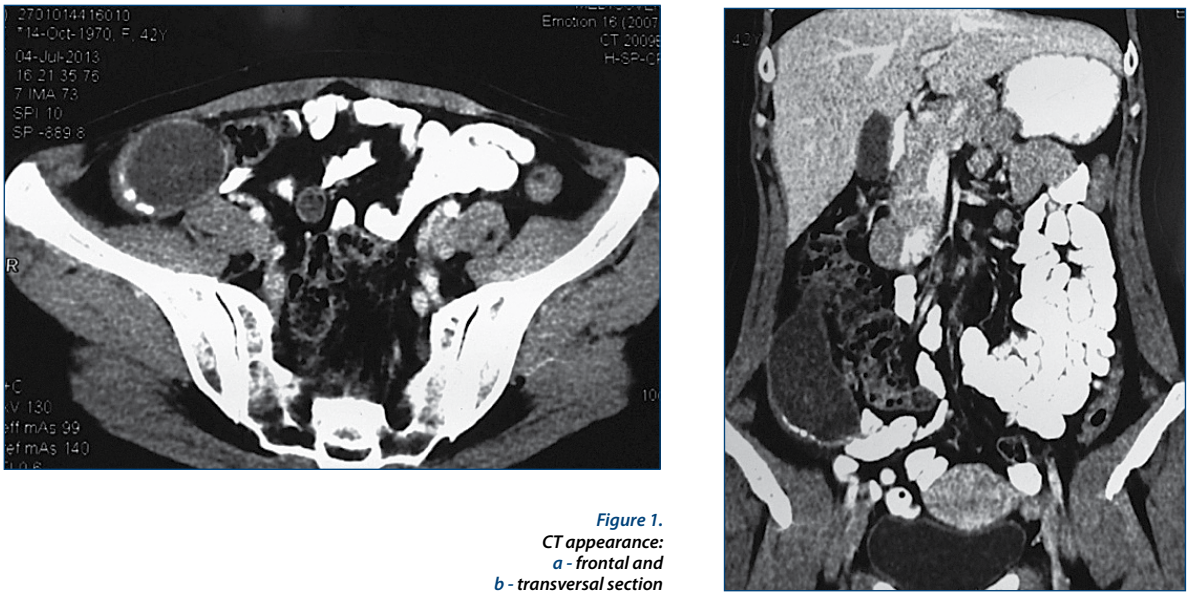

Figure 1.
CT appearance:
***a** - frontal and*
***b** - transversal section*

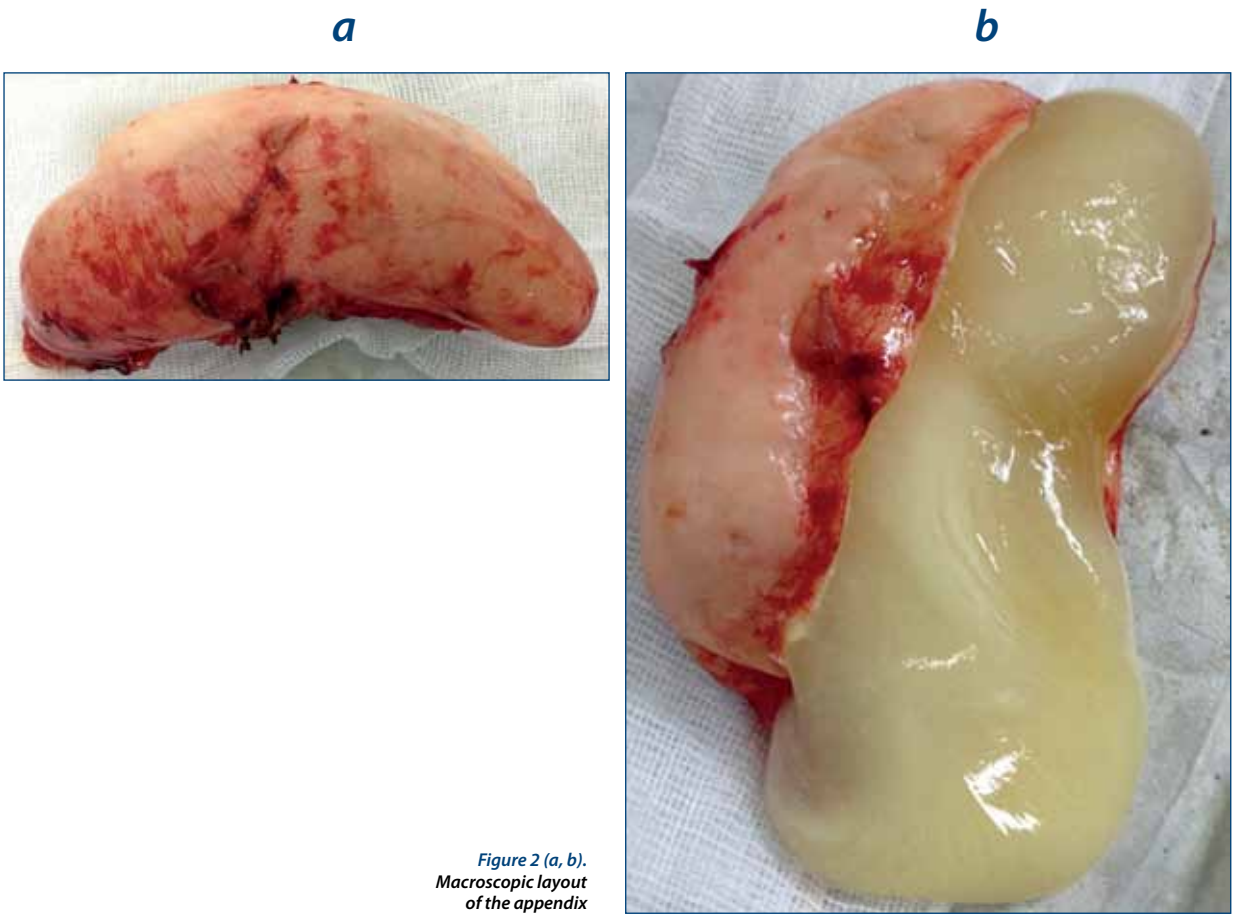

Figure 2 (a, b).
Macroscopic layout of the appendix

gular margins, but without a clear origin, and it was contained fluid, the approximate dimensions were of 13.5/8 cm, without signs of local invasion (Figure 1).

Exploratory laparoscopy was executed. Intraoperatory, the appendix was observed and it was increased in volume, approximately 14/8 inches, with a small base, of approximately 1.8 cm in diameter, without invasion of other organs (Figure 2). Laparoscopic appendicetomy was executed and the extracted part was sent to histopathological examination, confirming thus the diagnosis of appendicular mucocele with normal mucosa. Postoperative evolution was favorable with the resuming of gas transit after 48 hours.

The patient was discharged on the third postoperative day in good general condition. The patient was further clinically reevaluated 7 days postoperatively when she returns for removal of suture threads.

Discussion

Appendectomy with perpendicular lymphadenectomy and mezoappendix excision is recommended in the case of simple mucocele[12].

If the appendix base is more than 2 inches in diameter, or when mucocele appendix is accompanied by a cecal tumor, segmental colectomy and orientation of the surgical attitude is recommended, depending on the result of the extemporaneously histopathological examination. If invasion of the resection margins is observed, then right hemicolectomy with regional lymphadenectomy is executed[13,14].

If the integrity of the appendix walls is compromised and the content of the appendix lumen reached the abdominal cavity, the disease called pseudomixoma peritoneal is installed. This disease is characterized by the seeding of the peritoneal with mucinous tissue, and development of the mucinous ascites mainly in the lateral areas in peritoneal sacs.

Treatment of this condition is also oriented according to histopathological diagnosis[15]. The maximum size of the secondary determinations in order for chemotherapy to be effective should not exceed 2.5 mm in diameter[16]. Chemotherapy with mitomycin or oxaliplatin is performed at 420C for 60 or 120 minutes. If the examined fluid in the peritoneal cavity does not contain mucinous cells, along with the treatment of mucocele, intraperitoneal hyperthermia chemotherapy is also executed[17].

If there is a positive result, cytoreduction is added to the therapy mentioned above and this may include: omentectomy, splenectomy, cholecystectomy, partial or total colectomy, subtotal or total gastrectomy, hysterectomy, and ovariectomy[18,19]. Postoperatively systemically chemotherapy treatment is administered with 5-fluorouracil, in the first 5 days after surgery[20].

Conclusions

Currently there is controversy regarding the indication or contraindication to laparoscopy. Regardless of the surgical approach, dissemination of the appendix contents should be prevented. *In situ*ations in which the surgeon skills on laparoscopic technique does not allow the development of surgery in conditions of maximum safety, conversia of the surgery is recommend. Monitoring the postoperative evolution involves dosing of the CEA 125 every 3 months and tomographic surveillance at 6 and 12 months. ■

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