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Appendicular mucocele - the vaginal way

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Abstract

Appendiceal mucocele (AM) is a rare pathology and is characterized by accumulation of mucin in the appendiceal lumen. Clinically, it can manifest with abdominal pain in the presence of a lower abdominal palpable mass. It is usually discovered intraoperatively. Proper preoperative diagnosis is mandatory since it requires surgical treatment and it must always be extracted intact. Effraction of AM could lead to development of pseudomyxoma peritonei, a pathology with a severe prognosis. Differential diagnosis with an adnexal mass is essential since an ovarian benign pathology could be extracted using the vaginal approach in selected cases. If the abdominal mass is an AM, then laparotomy or laparoscopy is elective. We report the case of a 43-year-old woman with a preoperative diagnosis of an ovarian mass that during surgery turned out to be an AM.

Keywords: *appendicular mucocele, ovarian mass, vaginal, hysterectomy, pseudomixoma peritonei*

Introduction

The term mucocele is used to describe a luminal dilation of a cavitary organ (appendix, gallbladder, paranasal sinuses or salivary glands) secondary to mucus accumulation[(1)]. Appendiceal mucocele (AM) is an extremely rare pathology and it is most frequently discovered during surgery.

Case report

Clinical data

A 43-year-old gravida 3 para 1 referred with the complaint of menorrhagia and persistent right lower quadrant pain for the preceeding three months, symptoms that intensified during the last month.

Pelvic examination revealed an enlarged uterus corresponding to a 6-week pregnancy size and a palpable 7/5 cm large, renitent, mobile tumoral mass located in the right iliac fossa.

The transvaginal sonographic examination showed an anterior-flexed-uterus 79 x 27 x 45 mm in size without parietal asymmetry or any present myomas and a round oval hypoecogenic cystic mass (74x50x52 mm) (Figure 1), with regulated thin walls and posterior enhancement in the right iliac fossa. Near its inferior pole ovarian tissue was observed. The left ovary was normal and no other pelvic masses were seen and no fluid was found in the cul de sac (literally 'back of the bag'). Computed tomography scan was not available at the time of diagnosis.

The laboratory data, including tumor markers, and both endometrial and cervical cytology were within normal limits. Immunologic pregnancy test was negative and beta-human chorionic gonadotropin was undetectable.

A transvaginal hysterectomy with salpingo-oophorectomy was decided according to preoperative evaluation.

Surgery

The operation was performed under spinal anesthesia using the vaginal route. Uterine enlargement was confirmed, but the right anexial mass turned out to be an appendicular ovalar mass with tensed thin walls 50x48x24 mm, adherent to a normal ovary, suggestive for appendicular mucocele (Figures 2 and 3). We performed a transvaginal hysterectomy with bilateral salpingo-oophorectomy and a vaginal appendectomy with double ligation of the appendiceal stump using a slowly 3-0 absorbable suture. We mention that the appendicular mass was extracted intact, without effraction of the capsule. Pathology confirmed intraoperative diagnosis, establishing the simple mucocele form.

Postoperative evolution

Postoperative evolution was favorable with passage for stool present at 72 hours postoperatively and the patient was discharged well in day 7 postoperative.

Discussion

We present the case of a patient with persistent pelvic pain, most likely due to an ovarian cystic mass associated with uterine myoma.

Taking into consideration ultrasound, uterine mobility, wide vaginal access, benign cervical and endometrial cytology, we offered the patient a vaginal hysterectomy since AM was not a diagnosis to consider at the time.

Preoperative diagnosis of AM was impossible due to non-specific ultrasound in the presence of gynecologic symptomatology and lack of more specific preoperative imaging. The major risk in the case of vaginal approach of such large probable ovarian mass, in reality an appendicular one, was mucocele effraction with subsequent development of pseudomyxoma peritonei (a pathology with a severe prognosis).

Appendicular mucocele, a rare pathology, was first described in 1842 by Rokitansky and formally defined by Feren in 1876. In 1915 Castle reports a 0.2% incidence in a series of 13158 autopsies[(2)]. In 1973 Higa and contributors established three different entities of appendicular mucocele[(3)]. The gross appearance of AM is the distension of the appendix by mucin.

Current classification divides AM in four histological subtypes: (1) retention cysts or simple mucocele characterized by normal epithelium or epithelium with degenerative alterations due to simple obstruction; it often associates infection; luminal dilation is smaller than 2 cm; (2) mucous hyperplasia - focal or diffuse, with mild dilation of the appendicular lumen; it represents 5-25% of the cases[(3)] mucinous cystadenoma with

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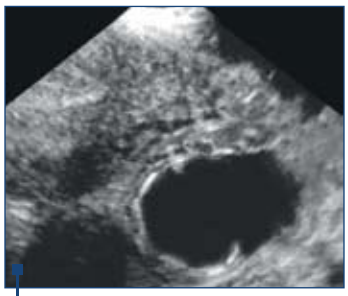

Figure 1. Ultrasound examination

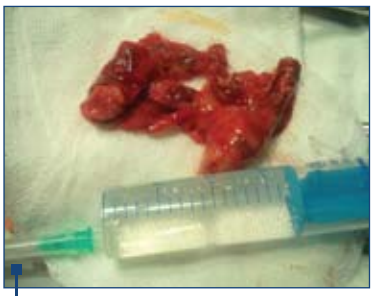

Figure 2. Gross aspect of the mucocele walls

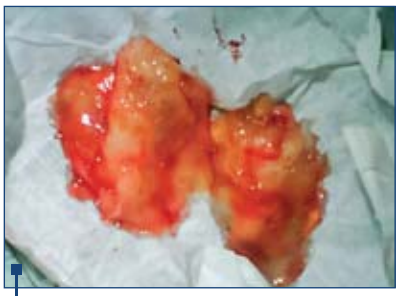

Figure 3. Gross aspect of the mucocele contents

dysplasic epithelium similar to adenomatous colon polyps or villous adenoma; neoplastic cells are absent and treatment consists of surgery; important dilation of the lumen; it includes 63-84% of the cases[4] mucinous cystadenocarcinoma characterized by neoplastic epithelium similar to colon adenocarcionma, with glandular stromal invasion; severe luminal dilation; is present in 11-20% of the cases[4].

Clinical manifestations include right lower abdominal pain, palpable abdominal mass. Preoperative diagnosis is mandatory in choosing the route for surgery, imaging being also important. In most of the cases, AM is an incidental finding during investigations or intraoperatively. It is unlikely for the patient to refer with acute appendicitis symptomatology, since it is based on a chronically obstruction. Other rare symptoms include those secondary to intussception or occlusion, gastrointestinal bleeding, passage of flatus, weight loss, vomiting or urological symptoms. Of all AM cases, 25% are asymptomatic[5].

When we suspect a AM, we could evaluate it by using barium enema, colonoscopy and angiography along with computed tomography examination and transabdominal/ transvaginal ultrasound.

In all histological types, ultrasound shows a visceral extrinsic mass located in the right iliac fossa, with variable size, hypoechgenic or inhomogeneous, sometimes with vegetations, with irregular shape, encapsulated if intact and with posterior enhancement. The typical feature that differentiates AM from appendicitis is the lack of appendiceal wall thickening of more than 6 mm. The suggestive feature for AM on ultrasound is the onion sign created by sonographic layering of the mucus within a cystic mass[6,7].

In contrast CT scan typical finding is a well-encapsulated mass with smooth regular walls localized in lower quadrant. AM density varies from that of water to that of soft tissues, according to mucin content. Sometimes it has a mass effect on enteric ansae, in the absence of periappendicular inflammatory process or abscess.

The differential diagnosis of a right lower quadrant cystic mass is complex and it should be established with adenocarcinoma of the appendix, carcinoid tumor, mucinous cystadenocarcinoma, limfoid hyperplasia, lymphoma, periapendicular abscess and ovarian tumors in women.

The most serious complication of AM is psudomyxoma peritonei, a rare pathology with an incidence of 2 cases per 10,000 laparotomies. Also called 'the gelatinous disease of the peritoneum', it consists of a spectrum of peritoneal lesions characterized by the accumulation of gelatinous material surface secondary to intraperitoneal effraction of mucin, or, more commonly, by diffuse proliferation of neoplastic cells along the peritoneum. Pseudomixoma peritonei is often associated with ovarian tumors. Intussesception is rare, is often described pseudomixomului association with ovarian tumors. Intussusception is rare and is found in less than 30 cases in the literature.

Surgical approach can be realized both laparoscopically or by laparotomy. Still, open surgery is recommended due to minor risk of effraction of the tumor with subsequent implants of mucinous epithelium on the peritoneal surfaces and mucus accumulation within the peritoneal cavity. Appendectomy is the definite treatment for simple mucocele, while as in the case of cystadenomas with wide appendicular base cecal resection is recommended. Right hemicolectomy is the election treatment for cystadenocarcinoma.

Conclusions

Despite perioperative evaluation it is still difficult to diagnose cystic lesions of the appendix and mucinos cystadencarcinoma.

Appendicular mucocele is a considerable differential diagnosis in cases of cystic ovarian mass. Proper preoperative diagnosis is important in establishing the surgical approach, in order to reduce the possibility of conversion to laparotomy, the risk of effraction of AM when using the vaginal route being considerable. ■

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