Appendicular mucocele - the vaginal way

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 preceding 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 hypoechoic 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 hysterecctomy 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 pseudomixoma 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...
A differential diagnosis of a right lower quadrant cystic mass is complex and should be established with adenocarcinoma of the appendix, carcinoid tumor, mucinous cystadenocarcinoma, lymphoid hyperplasia, lymphoma, periaortic abscess and ovarian tumors in women.

The most serious complication of AM is pseudomyxoma 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 secondary to intraperitoneal effraction of mucin, or, more commonly, by diffuse proliferation of neoplastic cells along the peritoneum. Pseudomyxoma peritonei is often associated with ovarian tumors. Intussusception is rare, but often described pseudomyxomuloi 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 cystadenocarcinoma.

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.

References