Surgical approach to appendiceal mucocele mimicking an adnexal complex mass: case report

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Summary

Mucocele of the vermiform appendix is a rare disease of the appendix caused by mucoid substance retention in its lumen, due to obstruction or hyperproduction due to appendiceal retention cysts, mucosal hyperplasia, mucinous cystadenomas and cystadenocarcinomas. Therefore, also appendiceal malignancy can be the underlying cause, making accurate preoperative diagnosis imperative, even if this disease is often asymptomatic and an early diagnosis still remains very difficult on imaging studies. In women, appendiceal mucoceles can sometimes present on clinical and instrumental findings as a right adnexal mass mimicking an ovarian neoplasm. A rare case of appendiceal mucocele in a 36-year-old women with a right-sided painful pelvic mass is presented. The mucocele was misdiagnosed as a cystic complex mass of the right adnexa both clinically and ultrasonographically. Serum levels of CEA and CA19-9 were increased. Explorative laparoscopy was performed revealing an enlarged vermiform appendix with the uterus and adnexa macroscopically normal, and no signs of intraperitoneal metastasis or adnexal torsion. Laparotomic appendectomy followed. Histological examination revealed a mucinous cystadenoma of the vermiform appendix. This clinical entity of appendiceal mucocele should always be considered by gynaecologists as well as gastroenterologists in the differential diagnosis of patients presenting a right-sided adnexal mass on ultrasound in order to choose the best surgical approach.

Key words: Mucocele; Vermiform appendix; Adnexal mass.

Case report

A 36-year-old Italian woman was admitted to our department in August 2005 because of lower right abdominal pain. On pathological anamnesis no systemic disease was found and no history of abdominal or pelvic surgery was noted. The gynaecological anamnesis showed menarche had occurred at age 16, with oligomenhorrea; she was gravida 2, para 0, and had had a first trimester spontaneous abortion.

Her blood pressure, cardiac pulse and body temperature were regular; her body mass index (BMI) was 19.8. All routine laboratory findings were normal but the serum levels of tumoural markers CEA and CA19-9 were increased, measuring respectively 11.8 ng/ml (normal range 0-5) and 38 U/ml (normal range 0-37) while β-hCG, CA125 and α-FP serum levels were within normal range.

On abdominal examination a painful, mobile, non tender, soft mass was palpated in the right lower abdominal quadrant. The pain had a dull character; there was a slow intestinal transit with constipation, but no abdominal distension nor alteration of urinary function.

The gynaecological examination revealed a uterus with no increase in volume, well definable and movable, with a sausage-shaped mass in the right adnexal region measuring about 7 cm in length; the left ovary was normal. There was no pain on vaginal examination, but atypical uterovaginal blood loss accompanied the abdominal pain episodes. Lymphadenopathy was not noted and cytological examination of the cervix and vagina showed no malignancy.

The ultrasound examination, performed with a pelvic and transvaginal probe, revealed a normal uterus with an echographic histerometry of 98 x 44 x 59 mm. A non-homogeneous mass with oblong morphology was found, measuring 71 x 53 mm, located on the site of the right adnexa (Figure 1). The right ovary was not visible. The left adnexa appeared normal and measured 34 x 23 mm. Color Doppler scanning of the mass revealed no color flow.

Explorative laparoscopy was performed revealing an elongated and enlarged vermiform appendix reaching the Douglas pouch with the right ovary lying on it. The mucocele had a soft consistency and the surface was brown and easily bled on touch. The uterus and bilateral tubes and ovaries were macroscopically normal and there were no signs of metastatic intraperitoneal disease or adnexal torsion; no ascites was noted.

The operative findings confirmed that the mass was located in the right side of the uterus with its origin from the appendix, rather than from the ovary. Because serosal invasion and regional lymph node enlargement were not evident, a standard laparotomic appendectomy was performed.

The women was an ASA I patient and totally intravenous general anaesthesia was performed with injected drugs: propofol was administered for sedation and hypnosis, the neuromuscular block was performed with atracurium besilate, and the narcotic analgesic was sufentanil. The patient had an uneventful postoperative course.

On gross examination, the appendiceal mass was an oblong structure separate and distinct from the normal right tube and ovary. The appendix was diffusely enlarged, measuring 8 cm in length and 4 cm in maximal diameter. On sectioning, the mass was cystic with a distended wall. The enlarged lumen contained large amounts of mucinous substance and had no internal proliferation. There was no gross evidence of malignancy.

Histological examination revealed a mucinous cystadenoma of the vermiform appendix. Microscopically, the cyst wall was almost completely lined by a columnar mucinous epithelium with tall, crowded, and basally located nuclei. Immunohistochemistry analysis described diffuse immunoreaction to cytokeratin 20 (+++).

Revised manuscript accepted for publication May 7, 2007

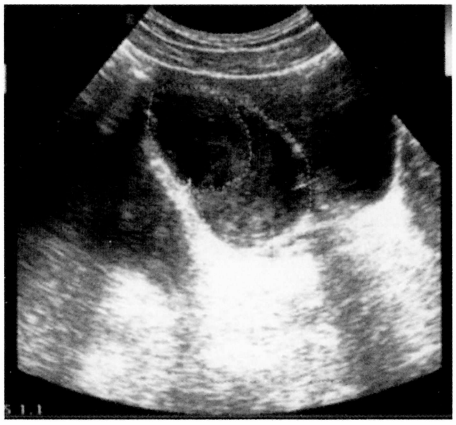

Figure 1. — Ultrasonographic appearance of appendiceal mucocele (transvaginal examination).

There was evidence of small areas (less than 10%) of mild atypical changes and a clinical and instrumental follow-up was started. Eighteen months after surgery the patient is alive with no evidence of disease and free of symptoms.

Discussion

Appendiceal mucocele is the enlargement of the vermiform appendix due to retention of a relatively large amount of mucus in its lumen. It is a rare disease and the incidence ranges between 0.2% and 0.3% of all appendectomies, with a higher frequency in females (4/1) and in people older than 50 years [1].

Mucus is retained either due to obstruction or hyperproduction. Microscopically, one of the following pathological entities can be found: retention cysts (retention mucocele or simple mucocele), mucosal hyperplasia, mucinous cystadenomas and cystadenocarcinomas [2], even if the term mucocele is used for a gross description, rather than histological diagnosis [3].

Symptomatology of appendiceal mucoceles is not specific and very large lesions are asymptomatic in 25% of patients. The most common presentation is right lower abdominal quadrant pain, similar to an acute appendicitis. A palpable mass can be found in 50% of cases, whereas urinary dysfunction or haematuria is rarely related [4].

Early diagnosis is important since it might be associated with poor prognosis due to cystadenocarcinoma of the appendix or pseudomyxoma peritonei for mucocele rupture, if left untreated.

Preoperative diagnosis of an appendiceal mucocele has rarely been reported, but it is possible with colonscopy or with the use of ultrasound and computed tomography (CT). Elevated CEA levels have been described in neoplastic mucoceles. A typical CT scan finding of an appendiceal mucocele is a round, low-density, thin-walled, encapsulated mass, communicating with the cecum. Ultrasonography shows a nonspecific cystic lesion, encapsulated, firmly attached to the cecum, with liquid content and an internal variable echogenicity related to mucus density [5]. There is no ultrasonographic pattern typical for an appendiceal mucocele, since various ultrasonographic patterns have been previously described: a typical cystic structure with thin walls, cyst with internal echoes and septations along with acoustic shadowing, a complex mass with acoustic enhancement, a cystic mass with layered walls and calcifications in the walls, and an elongated echo-poor mass without posterior echo enhancement, with the cyst wall less distinct than expected for a cyst.

The anatomic location of an appendiceal mucocele in the right lower quadrant of the abdomen includes it in the differential diagnosis of masses in this region. In women, mucocele of the vermiform appendix can sometimes present as a mass of the right adnexa. The clinical and ultrasonographic appearance can be misleading, resulting in major gynaecologic surgery to remove a potentially malignant adnexal mass, only to find a lesion originating from the appendix.

The type of surgical treatment is related to the dimensions and to the histology of the mucocele. Appendectomy is used for simple mucoceles or for cystadenomas. Right hemicolectomy is recommended for cystadenocarcinoma. The laparoscopic approach is not advised by some authors because of the risk of rupture and dissemination [6]. Moreover, an accurate exploration of the abdomen during laparotomy is advised because of the presence of malignant mucinous cystadenocarcinomas that represent 11-20% of cases [7], or the association between the appendiceal mucocele and other tumours, particularly carcinoma of the colon (11-20%) and tumours of the ovary [8].

The gynaecologist should also be familiar with the clinical occurrence of synchronous mucinous tumors of the ovary and appendix. Alternatively, when the imaging findings suggest an ovarian cystic tumour with pseudomyxoma peritonei, the radiologist should be alerted to the probability of a clinically unsuspected appendiceal mucocele and should search for it. Ovarian tumours and appendiceal mucocele have clearly been demonstrated in some cases, e.g., right ovarian mucinous cystadenoma and villous adenoma of the appendix, right ovarian and appendiceal mucinous cystadenocarcinoma, and bilateral metastatic ovarian implants of appendiceal mucinous cystadenocarcinoma [9].

We have presented a rare case of an appendiceal mucinous cystadenoma mimicking ultrasonographically a tumor of the right adnexa, with increased preoperative serum levels of CEA and CA19-9 and treated by laparotomic appendectomy.

The operation was started by laparoscopy under a totally intravenous general anaesthesia and converted to laparotomy after general surgeon consultation because of the risk of malignancy and for an accurate exploration of the abdomen. As serosal invasion and regional lymph node enlargement were not evident, a standard appendectomy was performed.

Our case suggests that appendiceal mucocele should be considered as a differential diagnosis for an adnexal mass in subjects without previous appendectomy. Since serum CEA possibly increases in subjects with tumours, pathol-

ogists should examine the whole resected specimen in this type of tumour to completely rule out any coexisting carcinoma [10].

Although primary adenocarcinoma of the appendix is uncommon, it should be emphasized that a preoperative diagnosis of a mucocele is important for patient management to choose the best surgical approach (laparotomic or laparoscopic); however, it is difficult on imaging studies [11].

In our opinion in the surgical approach to appendiceal mucocele, in accordance with some other authors [12], it is possible to avoid a first-line laparotomy and execute a laparoscopic appendectomy after accurate abdominal exploration and extemporaneous exams that exclude any suspicion of malignant pathology after the definitive histological diagnosis. A general surgeon should be on standby for consultation and for the possibility of intestinal surgery. Pseudomyxoma peritonei can be prevented by insertion of the mass in a laparoscopic endo-bag.

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