Pseudomyxoma peritonei and mucocele of vermiform appendix simulating right adnexal mass

Śluzak rzekomy otrzewnej oraz mucocele wyrostka robaczkowego przypominające zmianę w prawych przydatkach

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Summary

We present a case of a 51-year-old woman who suffered from persistent right lower abdominal pain. Ultrasound examination revealed two lesions in the area of right adnexa. A suspicion of right adnexal cyst was made. Laparoscopy showed that patient was affected by an early stage of pseudomyxoma peritonei (PMP) resulting from a ruptured mucocele of the vermiform appendix. This condition is very rare, however, due to its localization and not specific clinical manifestation it should be taken into consideration in the differential diagnosis of adnexal masses.

Key words: vermiform appendix, mucocele, pseudomyxoma peritonei.

Streszczenie

W pracy zaprezentowano przypadek 51-letniej pacjentki cierpiącej na uporczywe bóle zlokalizowane w okolicy prawego dołu biodrowego. Badanie ultrasonograficzne wykazało obecność dwóch patologicznych zmian w rzucie prawych przydatków, co nasunęło podejrzenie torbieli. Przeprowadzony zabieg laparoskopii pozwolił stwierdzić, że u pacjentki rozwinęła się wczesna postać rzekomego śluzaka otrzewnej. Przyczyną tego stanu była perforacja *mucocele* wyrostka robaczkowego zawierającego gruczolaka śluzowego.

Patologia ta jest spotykana rzadko, jednakże ze względu na swoją anatomiczną lokalizację i mało charakterystyczne objawy powinna być wzięta pod uwagę podczas diagnozy różnicowej zmian przydatków.

Słowa kluczowe: wyrostek robaczkowy, *mucocele*, śluzak rzekomy otrzewnej.

Introduction

Mucocele is a dilatation of a vermiform appendix caused by its obstruction and consequent accumulation of mucus in the lumen. This condition is found in 0.2-0.3% of all appendectomies [1]. It is more frequent in females and patients over 50 years of age [2]. Obliteration of appendiceal lumen can be a result of mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma and retention cysts (11%) [3]. There have been reported several cases of mucocele

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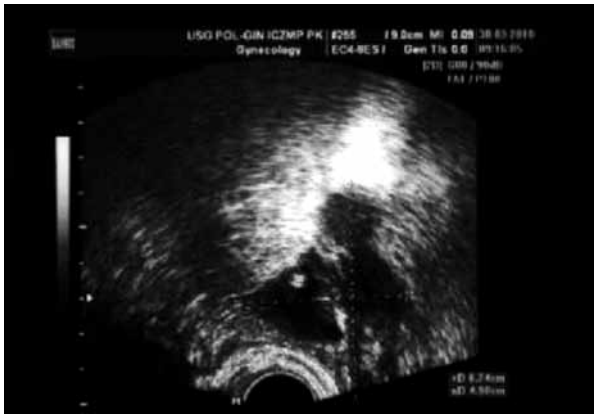

Fig. 1. Ultrasound scan of cystic mass (60 × 48 mm) localized in the region of right adnexa

secondary to diverticula, fecaliths, endometriosis and carcinoids [4-6]. Due to unspecific symptoms and localization in the lower abdominal quadrant, mucocele may mimic right adnexal mass [7] making the preoperative diagnosis very difficult.

Spontaneous perforation of a distended appendix or accidental rupture of its wall at the time of surgery may result in forming a condition known as pseudomyxoma peritonei (PMP). Epithelial cells originating from primary neoplasm proliferate and spread throughout the peritoneal cavity. The development of PMP manifests in accumulation of mucinous deposits in the abdomen which can be associated with peritoneal and omental implants. Ronnet et al. classified PMP into two categories: disseminated peritoneal mucinosis (DPAM) with epithelial cells presenting small atypia or mitotic activity and peritoneal mucinous carcinomatosis (PMC) characterized by epithelium with cytological features of carcinoma [8]. The estimated incidence of this disease is 1 to 2 per million per year [9].

Case report

A 51-year-old woman was admitted to our ward with episodes of right lower abdominal pain. Physical examination revealed a palpable pelvic mass on the same side. Ultrasound examination showed the presence of hypoechogenic area on the right side behind the uterus (34 × 24 mm) and irregular, partially fluid mass in the region of right adnexa (60 × 48 mm) (Fig. 1). There was also a suspicion of endometrial hyperplasia. The CA-125 tumor marker level was not elevated. Other biochemical studies as well as complete blood count were also normal.

In such circumstances diagnostic laparoscopy seemed to be the right decision. The surgery revealed presence of mucinous masses localized in the right pelvic region and dilated, perforated appendix covered by mucus. During laparoscopy appendectomy, total cytoreduction of mucinous lesions and irrigation of peritoneal cavity were performed. The surgeon also decided to resect ovaries with fallopian tubes in order to exclude the probable presence of malignancy involving their structure. All these specimens were sent for histological examination that showed a benign mucinous adenoma of the appendix and pseudomyxoma peritonei (Fig. 2, Fig. 3). Adnexa were not pathologically changed. Uterine curettage did not confirm the suspicion of endometrial hyperplasia. After the management, the patient recovered and was informed about the necessity of future follow-up.

Discussion

The diagnosis of appendiceal mucocele is a challenging problem for gynecologists. This condition is asymptomatic in about 25% of patients and can be identified incidentally during radiological examination. Common clinical manifestations of the disease include right lower abdominal pain and palpable mass, whereas rare presentations of mucocele are intestinal obstruction or intestinal bleeding [10]. In some cases there is also an occurrence of increased levels of serum

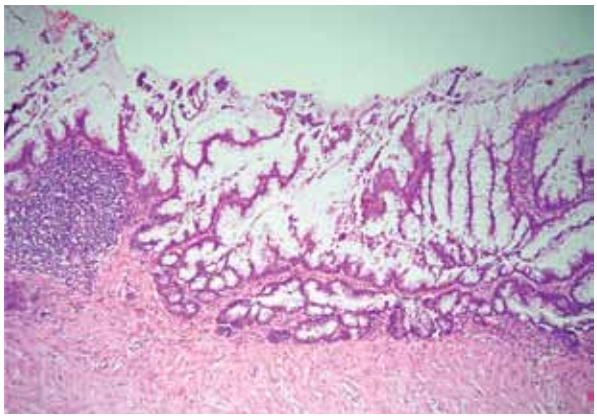

Fig. 2. Mucinous adenoma of the vermiform appendix, HE, magnification 100 ×

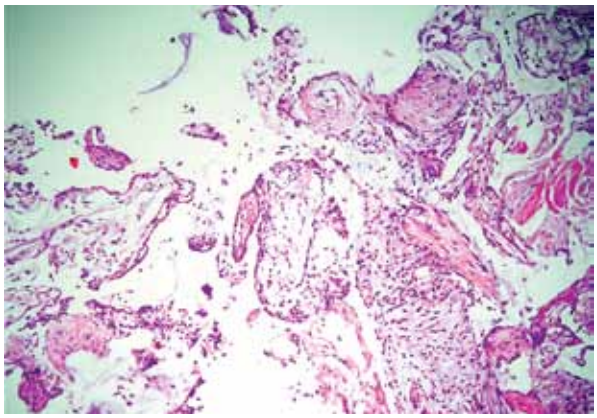

Fig. 3. Peritoneal mucin deposits containing inflammatory and mesothelial cells, HE, magnification 100 ×

tumor markers. Dragoumis et al. described an instance of mucocele simulating right adnexal mass with elevated CA125 serum levels [11]. Ultrasound evaluation is not always helpful in ascertaining the final diagnosis due to the fact that image may be quite miscellaneous (cystic lesions with anechoic fluid or with hypo/hyperechoic masses dependent on mucus density). Only the "onion skin" sign is believed to be the most distinctive finding for appendiceal mucocele [12]. Taking into consideration all the facts mentioned above and anatomic localization of appendix, it turns out that mucocele can be easily misdiagnosed. Balci et al. recently have reported a case of mucocele mimicking right adnexal cyst [13]. Their preoperative diagnosis was wrong and only laparotomy allowed to finally identify pathological masses. It was a similar situation to our case.

The most unwanted and problematic complication of mucocele is rupture of its wall and spillage of mucus within the peritoneal cavity. Perforation of distended appendix occurs usually spontaneously due to rising intraluminal pressure. This could be the beginning of a process leading to development of an advanced stage of PMP. In early stage of the disease the surgeon can find small deposits of free mucus in the peritoneal cavity and on surface of the appendix [14]. Over time mucus is redistributed in a specific way which is dependent on gravity and intraperitoneal fluid current. Epithelial cells continue to proliferate and form tumor implants especially on omental and diaphragmatic surface which leads consequently to mucinous ascites [15].

The origin of PMP has been a controversial issue for many years. It is believed that the most probable primary sites are ovaries and vermiform appendix. However, new evidence was discovered suggesting that the involvement of ovaries seems to be secondary to a primary origin in the appendix. Study performed by Ronnet et al. showed that samples collected from ovarian mucinous deposits in course of PMP were positive for cytokeratin 7, 18 , 20, human alveolar macrophage 56 and carcinoembryonic antigen [16]. These characteristics were also found in appendiceal lesions, whereas primary ovarian neoplasms without PMP were immunohistochemically different.

Clinical manifestation of the disease is quite variable and not specific. Patients with an advanced stage of PMP present with distended abdomen by mucinous ascites (so called "jelly belly") and intestinal obstruction [14]. In less advanced stages females are admitted to hospitals with lower abdominal pain, pelvic or ovarian masses, infertility and menstruation disturbances [17]. The most helpful tools for the diagnosis of the disease are ultrasound imaging and computer tomography which can visualize mucinous deposits in different abdominal regions. Serum tumour markers such as carcinoembryonic antigen or CA-19-9 can be useful fixtures related with PMP. The sensitivity of CA125 is approximately 60%, however, its levels may be also elevated in primary ovarian lesions [18]. It makes the differential diagnosis between adnexal masses and PMP caused by ruptured mucocele even more difficult.

The management of this condition depends on the stage of the disease. In more advanced PMP Sugarbaker suggests cytoreductive surgery consisting of parietal peritonectomy, debulking procedures and resection of all involved organs. This treatment should be followed by hyperthermic intraperitoneal chemotherapy (HIPEC) [14]. In the early stage of PMP there are no standard procedures. It is believed that appendectomy and total reduction of mucinous deposits is a sufficient treatment [14]. Our patient was treated this way as there were no signs of more advanced disease. After surgical management a close follow-up consisting of serum tumour markers control, computed tomography scan and physical examination are obligatory [14].

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