Large mucinous neoplasm of the appendix mimicking adnexal mass in a postmenopausal woman

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1. Introduction

None of the different diagnostic investigation modalities is alone sufficient to characterize an adnexal mass. History, physical examination, imaging methods and plasma tumor markers have to be used together in the preoperative investigation of an adnexal mass. Appendiceal tumors are first diagnosed as acute appendicitis or may be a chance finding in the course of surgery for unrelated conditions. Mucinous neoplasms with malignant potential are seldom encountered; their course is generally silent and their identification is often by chance finding.

2. Case report

An 81-year-old postmenopausal woman admitted with abdominal pain during the last few months. She had an unremarkable history. Physical examination showed tenderness and semi-mobile palpable mass with 10 cm in diameter in the lower right abdominal quadrant. On vaginal examination, subtotal uterine prolapse was seen and a semi-mobile, semi-solid, smooth bordered mass with 10 cm in diameter was palpated on right adnexal region. She had no fever. Her laboratory tests for renal function, liver function, complete blood count, electrolyte status and blood sugar levels were within normal range. Also, serum tumors markers were within normal limits (Ca125: 13.18 U/ml, Ca19.9: 20.8 U/ml, Ca15.3: 6.7 U/ml and CEA: 1.76 ng/ml). Preoperative ultrasonographic examination was revealed normal uterus and left adnexa, and a 11 cm × 9 cm heteroechogenic cystic mass in right adnexal location. A magnetic resonance imaging revealed a 12 cm × 10 cm cystic, heteroechogenic lesion that did not exhibit contrast enhancement or solid components (Fig. 1). Then, laparotomy was performed. Inspection of the abdominal cavity revealed an approximately 10 cm mucinous tumor mass arising in the appendix (Fig. 2a and b). The uterus and ovaries appeared to be normal. An appendectomy and a right hemicolectomy with ileo-transverse anastomosis were performed. A total abdominal hysterectomy and bilateral salpingo-oophorectomy had to be performed before the end of surgery due to the presence of a subtotal uterine prolapse. Histopathological examination was revealed the appendiceal mucinous neoplasm, measuring 12 cm in its largest diameter, disclosed a largely mucocel-like lining epithelium and acellular mucus, with occasional thicker, columnar-looking epithelial portions (Fig. 3a). No cytological atypia or complexities of the glandular structures were observed. The lesion wall disclosed a rupture and the
passage of mucus in the surrounding fat, in an area with occasional calcifications and otherwise minor leakage (Fig. 3b). This area was surrounded by soft tissue reaction. The postoperative period was uneventful. The patient has been followed for one year and there are no clinical or biochemical findings suggesting a recurrence.

3. Discussion

Appendiceal tumors constitute less than 0.4% of all gastrointestinal neoplasms. Less than 2% of all appendectomies result in a finding of tumor; approximately 20% of these cases, however, have a perforated appendix. Mucinous appendiceal neoplasms with low malignancy potential are rare. Cystadenomas are reported to be found in approximately 1% of all appendectomies. Mucinous cystadenomas are the most frequent of mucinous appendiceal neoplasms. Appendiceal tumors are generally accepted as being benign. Appendiceal tumors are generally diagnosed as acute appendicitis, or they may be incidental in the course of surgery for unrelated conditions. The clinical importance of such tumors is related to the pseudomyxoma peritonei that may result from mucin leakage in the peritoneal cavity following appendix rupture. This complication negatively affects treatment outcomes as compared to unruptured cases. While 5-year survival of benign mucocele is 100%, that of its malignant variety is only 45%. A mucocele lesion is accepted as neoplasia if its diameter is larger than 2 cm. Recent publications on appendiceal tumors are not uniform as to their diagnostic criteria or treatment recommendations. Surgery is unique therapy for the appendix mucocele. Also, laparoscopic approach must be performed by experienced surgeons because of the risk of rupture. The type of surgical treatment is related to the histopathological features of the mucocele. Appendectomy can be performed for simple mucocele or for cystadenoma, when the appendiceal base is intact. Performing a right colectomy seems to be of particular importance in the case of malignant mucinous tumors and of tumor invasion of the appendiceal base. 11–20% of patients with appendiceal mucinous tumors have associated colonic and ovarian tumors. Therefore, careful exploration of the other abdominal organs, in particular the colon and ovaries, are important during the surgery. Cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy and early postoperative intraperitoneal chemotherapy are recommended for the patients with pseudomyxoma peritonei. Adnexal masses are seen in women of every age group; at high risk for ovarian malignancy are those, especially postmenopausal patients with a tumor morphologically characterized as solid or complex by ultrasound and with increased CA-125 levels (>35 U/mL). The most important characteristics differentiating benign formations from malignant adnexal masses are thick septa,
papillary projections, and the presence of non-hyperechogenic (vascularized) tissue. McDonald et al. evaluated 395 women with adnexal mass preoperatively by transvaginal ultrasonography and serum CA-125 and reported that the combination of these two tests is successful in evaluating malignity.12 In a case report by Dragoumis et al., a patient with a CA-125 level of 120U/mL evaluated as having a right adnexal cyst had a final diagnosis of appendiceal mucocoe.13 Similarly, Yildiz and Abbasoglu presented two cases with a periappendicular abscess and mucocoe and the preoperative diagnosis of the cases was an adnexal mass.14 Balci et al. report identifying a 6 cm × 8 cm appendiceal mucocoe during laparotomy in a patient operated on for adnexal mass.15

In this presented case, the patient was evaluated preoperatively by ultrasonography, magnetic resonance imaging, physical examination, and serum tumor markers and scheduled for surgery with a tentative diagnosis of adnexal mass. The diagnosis of appendiceal tumor was intra-operative. While a simple appendectomy is sufficient treatment for patients with benign mucocoe, mucinous neoplasms additionally require resection of the cecum or a hemicolectomy, as also practiced on our patient. Although the patient have mucinous appendiceal tumor with a diameter of 12 cm × 9 cm, neither acute signs of appendicitis nor pseudomyxoma peritonei were seen.

In conclusion, appendiceal tumors are rare, late-diagnosed neoplasms that may not be differentiated from adnexal masses even by advanced imaging methods and other diagnostic procedures. They may be asymptomatic and remain undiagnosed until surgery. Gastrointestinal tumors such as appendiceal tumors kept in mind in a patient with diagnosed adnexal mass.

Conflict of interest

Authors have no conflict of interest to declare.

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No commercial support was obtained for this study.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

All authors were involved in the researching and editing of the manuscript. The surgery was done by LA, BSH, KO and TS. LA, IH and BSH wrote the case report. BDA was the pathologist.

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