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Collision Tumor of the Appendix: Mucinous Cystadenoma and Carcinoid. A Case Report

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Rezumat

Tumoră de coliziune apendiculară: chistadenom mucinos și tumoră carcinoidă. Prezentare de caz

Introducere: Chistadenomul mucinous este cea mai comună formă de neoplazie benignă a apendicelui, iar carcinoidul este cel mai comun tip de leziune primară malignă apendiculară. Raportăm cazul rar al unei paciente în vârstă de 57 de ani cu prezență simultană de chistadenom mucinous și tumoră carcinoidă a apendicelui. Neoplazia dual carcinoidă și epitelială este un eveniment rar întâlnit la nivelul apendicelui. *Prezentare de caz:* O femeie de rasă caucaziană în vârstă de 57 de ani s-a prezentat la medic după palparea întâmplătoare a unei formațiuni în fosa iliacă dreaptă. Mijloacele imagistice au revelat o tumoră chistică la nivelul fosei iliace drepte cu diametrul de cca. 8 cm, cu originea fie în ovarul drept, fie în apendice. Pacienta a fost supusă explorării laparoscopice chirurgicale, ce a detectat prezența unui mucocel appendicular, procedura finalizându-se cu apendicectomie și excizia unui chist ovarian drept. Examenul patologic a decelat o tumoră de coliziune, constând în chistadenom mucinous și tumoră carcinoidă apendiculară. Datorită dimensiunii și extensiei tumorii carcinoide, revelate de raportul anatomopatologic, pacienta a fost supusă din nou unei explorări chirurgicale și colectomiei drepte laparoscopice.
Discuții: Chistadenomul mucinous este o entitate rară, dar este cea mai frecventă tumoră apendiculară benignă, reprezentând 0,6% din probele operatorii post-apendicectomie. Se poate manifesta similar apendicitei, unui mucocel sau, dacă tumora este ruptă, asemănător unui pseudomyxoma peritonei. Pe de altă parte, carcinoidul este cea mai frecventă leziune malign primară apendiculară și reprezintă 0,3-0,9% din probele operatorii postapendicectomie, cu o ușoară predominanță în rândul sexului feminine. Cazuri rare de prezență simultană a tumorii carcinoide și chistadenomului mucos au mai fost raportate în literatură, dar în cazul acesta a fost vorba de o tumoră de coliziune, între cele două tumori neexistând nicio zonă de tranziție. De asemenea, forma clinică de prezentare a pacientei și necesitatea unui diagnostic diferențial între leziunile ovariene și tumorile apendiculare merită menționate.

Cuvinte cheie: apendice, chistadenom mucos, tumoră carcinoidă, tumoră de coliziune

Abstract

Introduction: Mucinous cystadenoma is the most common of benign neoplasms of the appendix and carcinoid is the most common type of primary malignant lesions of the appendix. We report a rare case of a 57-year-old female with combined mucinous cystadenoma and carcinoid tumor of the appendix. Dual carcinoid and epithelial neoplasia is a rare occurrence in the appendix.
Case report: A 57-year-old Caucasian woman presented after incidentally palpating a mass on her right iliac fossa. Imaging modalities revealed a cystic tumor in the right iliac fossa with a diameter of about 8 cm, originating either from the right ovary or the appendix. She underwent laparoscopic surgical

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exploration, which revealed appendiceal mucocele and appendicectomy was finally performed as well as excision of a right ovarian cyst. Pathological examination showed a collision tumor consisting of mucinous cystadenoma and carcinoid tumor of the appendix. Because of the size and extension of the carcinoid tumor, which the pathology report revealed, she underwent re-exploration and laparoscopic right colectomy.

Discussion: Mucinous cystadenoma is rare, but it is the commonest of benign appendiceal tumours accounting for 0.6% of appendectomy specimens. It can present as appendicitis, mucocele or if the tumour ruptures, as pseudomyxoma peritonei. On the other hand, carcinoid is the most common type of primary malignant lesion of the appendix and 0.3-0.9% of appendectomy specimens, with small predominance in female patients. Rare cases of mucinous cystadenomas of the appendix coexisting with carcinoid tumors were reported before, but in our case it was a collision tumor with no transitional zone between them. Also, the clinical presentation of our patient and the differential diagnosis of ovarian lesions from appendiceal tumors is worth being mentioned.

Key words: appendix, mucinous cystadenoma, carcinoid tumour, collision tumors

Introduction

Mucinous cystadenoma is the most common of benign neoplasms of the appendix, with an incidence of 0.6% in recent series of appendectomy specimens (1). Carcinoid tumor of the appendix is the most common type of primary malignant lesion of the appendix and one of the most common single presentations of this type of tumor and is incidentally found in 0.3-0.9% of patients undergoing appendectomy (2). We report a rare case of a 57-year-old female with combined mucinous cystadenoma and carcinoid tumor of the appendix. Dual carcinoid and epithelial neoplasia is a rare occurrence in the appendix.

Case report

A 57-year-old white woman presented after incidentally palpating a mass in her right iliac fossa. Her laboratory examinations were unremarkable and tumor markers were between normal limits range. Abdominal ultrasound and computed tomography (CT-scan) revealed a cystic tumor in the right iliac fossa with a diameter of about 8 cm, originating either from the right ovary or the appendix. She underwent laparoscopic surgical exploration and an appendiceal mucocele and a small right ovarian cyst were found (Fig. 1), so laparoscopic appendectomy as well as excision of the ovarian cyst were finally performed. Histopathological examination showed a cystically dilated appendix measuring 8.5 cm in diameter. The luminal content consisted of viscid mucin and the base of the appendix was thickened and firm. Microscopically, the specimen had features of mucinous cystadenoma with adenomatous epithelium extending all over the thickened appendiceal wall. In that area the wall was involved by a well differentiated neuroendocrine neoplasm WHO grade I, a carcinoid. The neoplastic cells infiltrated the entire thickness of the appendiceal wall, the mesoappendix and reached the serosal surface (Fig. 2). The two neoplasms were separate and there was no transitional zone between them, categorizing them as collision tumors of the appendix. Because of the size (>2 cm) and extension of the carcinoid tumor in the mesoappendix, re-exploration and laparoscopic right colectomy was performed. The patient was discharged on the fifth postoperative day, and remains disease free on her follow-up 12 months later.

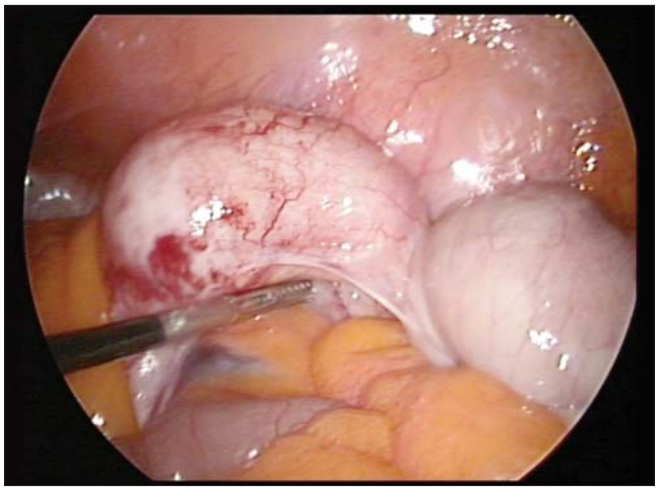


Figure 1. *Laparoscopic view of appendiceal mucocele*

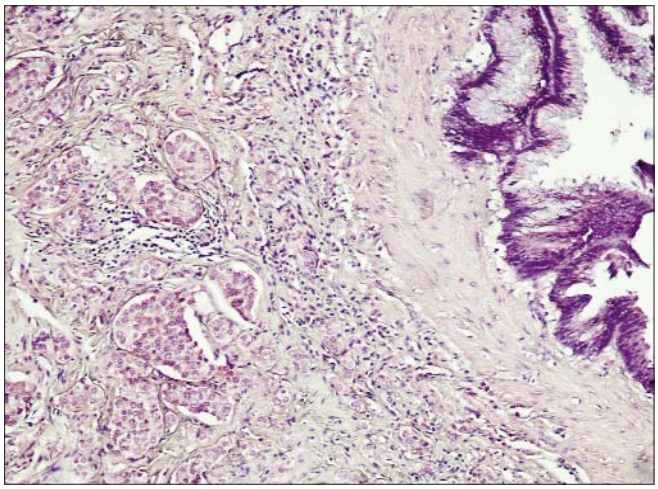


Figure 2. *Histopathological section of both mucinous cystadenoma and carcinoid tumor*

Discussion

Mucinous cystadenoma is rare, but is the commonest of benign appendiceal tumours accounting for 0.6% of appendectomy specimens. It can present as appendicitis, mucocele or if the tumour ruptures, as pseudomyxoma peritonei. It is now widely accepted that mucinous cystadenoma is the appendiceal counterpart of villous adenomas occurring elsewhere in the intestine, differing from the latter only because they are restricted to the appendix (3). Treatment is usually appendectomy with care taken not to disperse the mucin intraperitoneally.

On the other hand, carcinoid is the most common type of primary malignant lesion of the appendix and is found in 0.3-0.9% of appendectomy specimens with small predominance in female patients with mean age 38-49 years old (4). Patients usually present with acute appendicitis and diagnosis of carcinoid is revealed in pathology examination. Carcinoid tumors of the appendix rarely metastasise (5). In a recent review, Goede et al reports that acceptable indications for reintervention and right colectomy consisted in all lesions larger than 2 cm in diameter, histological evidence of meso-appendiceal extension, tumors at the base of the appendix with positive margins or involvement of the cecum, high-grade malignant carcinoids and goblet cell adenocarcinoids (6,7). It is also worth mentioning that patients with carcinoid lesions have a notable risk of developing a synchronous or metachronous colorectal neoplasm up to 33% (8).

Rare cases of mucinous cystadenomas of the appendix coexisting with carcinoid tumors were reported before (9-11), but in our case it was a collision tumor with no transitional zone between them. Also, the clinical presentation of our patient and the differential diagnosis of ovarian lesions from appendiceal tumors have to be highlighted.

Competing interests

The authors declare that they have no competing interests.

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