J Cases Obstet Gynecol, 2014 Jul;1(3):40-42

Case Report

Appendiceal carcinoid tumors concomitant with gynecologic cancers: Report of 3 cases

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Abstract

Carcinoid tumors of appendix are rare tumors and preoperative diagnosis may be challenging. The prevalance of appendiceal carcinoid is 0.3 to 0.9 % in patients undergoing appendectomy. Carcinoid of the appendix does not change the management of primary gynecologic disease in most cases. But if the surgical margins are not intact, size of the carcinoid is more than 2 cm, malignant histological appearance of the carcinoid or involvement of mesoappendix or lymph nodes requires further evaluation and treatment such as right hemicolectomy and/or chemotheraphy. We report two cases of endometrial carcinoma and one case of epithelial ovarian carcinoma undergoing staging surgery and found primary appendiceal tumor in the appendectomy specimens.

Key words:

Appendectomy, carcinoid tumor, endometrium cancer, ovarian cancer

Introduction

Carcinoid tumors of appendix are rare tumors and preoperative diagnosis may be challenging. The prevalance of appendiceal carcinoid is 0.3 to 0.9 per cent in patients undergoing appendectomy [1,2]. Even though most of the appendiceal carcinoids are benign, they may have metastatic potential [3]. In many cases, appendiceal carcinoids are found incidentally in appendiectomy specimens. Tumor size is often under 1 cm and lack of metastasis is probably due to small tumor size. Appendix carcinoids show slightly higher incidence among females compared to the males [4]. These neuroendocrin tumors are more often diagnosed in the second decade of life for females [5] and third decade of life for males [6].

We report two cases of endometrial carcinoma and one case of ovarian epithelial carcinoma undergoing staging surgery and found primary appendiceal carcinoid tumor in the appendectomy specimens.

Case presentation

Case 1: A 55-year-old woman was referred to our hospital with the complaint of postmenopausal bleeding. Medical history was unremarkable except type 2 diabetes mellitus regulated with oral anti-diabetics. Transvaginal sonography revealed endometrial polypoid lesion, 23x11 mm in size. Endometrial sampling showed endometrial adenocarcinoma. A staging procedure for endometrial carcinoma was planned for the patient. Total abdominal hysterectomy (TAH) with bilateral salphingo-oophorectomy (BSO), omentectomy, appendectomy, bilateral pelvic and para-aortic lymph node dissection (BPPLND) was performed. Appendix vermiformis had normal gross appereance. Histologic examination of the specimens revealed grade 1 and FIGO stage 1A endometrial adenocarcinoma of the endometrium (Fig. 1) and carcinoid tumor in the appendix vermiformis in a diameter of 2 mm. The carcinoid was lack of mitosis and no lympho-vascular invasion was noted (Fig. 2). Carcinoid tumor tissue showed strong positive immunostaining for synaptophysin, a marker for carcinoid tumors (Fig. 3). All resected lymph nodes were negative for tumor metastasis. Further adjuvant therapy was not necessary either for endometrial carcinoma, or appendiceal carcinoid. Surgical treatment for both endometrial and carcinoid tumors was adequate and sufficient. The patient is disease free after 7 years from the surgery.

Case 2: A 57 year-old-woman presented with endometrial thickness of 15 mm found in routine postmenopausal evaluation. Medical history was unremarkable. Endometrial sampling revealed endometrial adenocarcinoma. Preoperative evaluation of serum tumor markers were in normal range. A staging procedure for endometrial carcinoma was planned. TAH, BSO, omentectomy, appendectomy and BPPLND were performed. Histologic examination of the

Article history:
Received 14 March 2014
Accepted 15 June 2014

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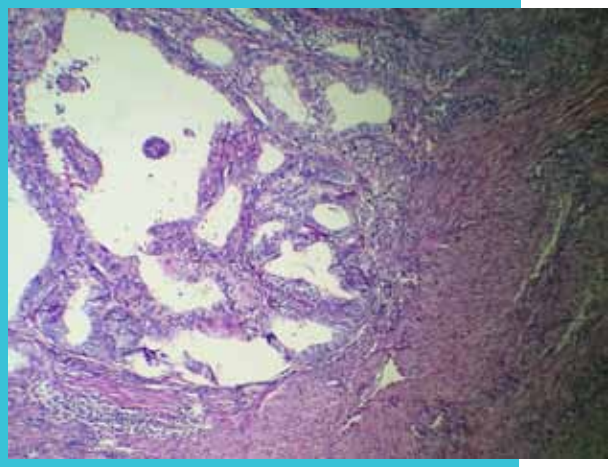
specimens showed well differantiated endometrium adenocarcinoma, showing myometrial invasion less than ½ of myometrium, compatible with FIGO stage 1A. Moreover, there was carcinoid tumor measuring 3x2 mm in size at the appendix vermiformis. Surgical borders of appendix were intact for tumoral invasion. The carcinoid was lack of mitosis and no lympho-vascular invasion was noted. All the 78 dissected lymph nodes were negative for tumoral involvement. But peritoneal cytology was positive for malignant cells. The patient was referred to medical oncology department for further treatment.

Figure 1.



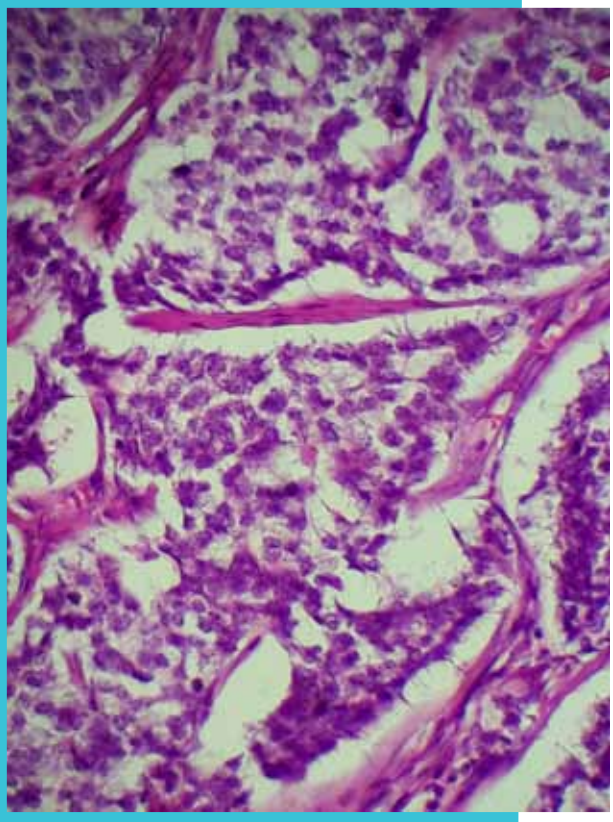
Grade 1 endometrioid type endometrium carcinoma (H&E, x200). Lower right quadrant of the figure shows myometrial invasion.

Case 3: A 73-year-old woman was referred to our department with the complaint of abdominal swelling and distension. Bimanual pelvic examination showed left adnexial mass. A transvaginal sonography was performed and revealed 20 mm fluid in the endometrial cavity. Left ovary had a normal sonographic appearance. An enlarged right ovary with a size of 127x115 mm with solid and cystic components was noted. Solid components had pathologic blood flow in doppler study. Pelvic magnetic resonance imaging showed 16x14 cm mass in the left adnexa with intra-abdominal and pelvic free fluid. Cervical cytology, endometrial sampling and endocervical curettage results were normal. Preoperative serum CA-125 was 271 U/mL, and CEA level was >100 ng/mL. Other tumor markers were in normal range. A staging procedure for endometrial carcinoma was planned for the patient. TAH, BSO, omentectomy, appendectomy and BPPLND were performed. Histologic examination of the specimens showed mucinous adenocarcinoma of the appendix infiltrating all layers of appendix and mesoappendiceal involvement, mucinous adenocarcinoma metastasis in the omentum, left ovarian mucinous epithelial carcinoma which showed large areas of carcinoid differantiation in immunohistochemical staining of the specimens. The patient was referred to medical oncology and gastroenterologic surgery departments for further treatment.

Discussion

Dermoid cysts are the most common type of ovarian tumCarcinoid tumors of appendix are rare tumors and preoperative diagnosis may be challenging.

Figure 2.

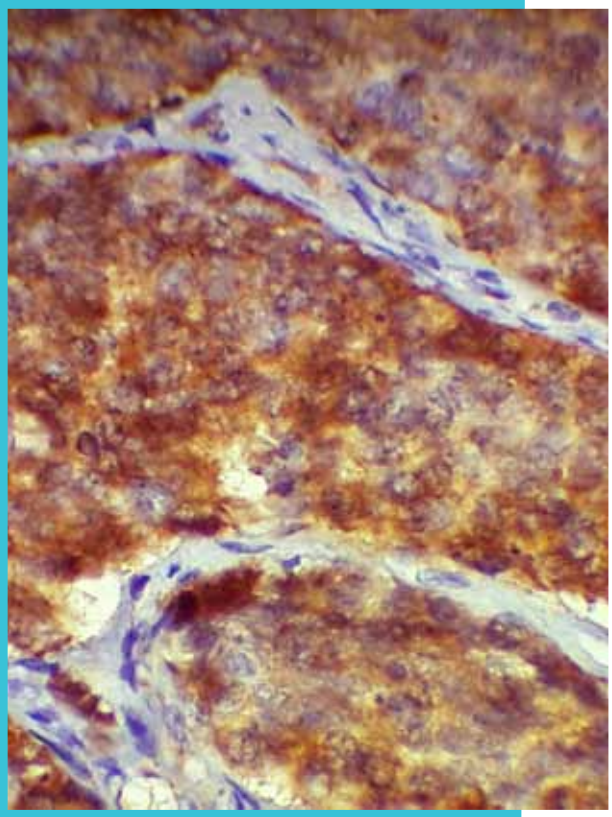


Carcinoid tumor of the appendix (H&E, x400).

Even though most of the appendiceal carcinoids are benign, they may have metastatic potential [3]. Tumor characteristics that predict aggressive behaviour include size, histological subtype and mesoappendiceal involvement. The authors considered that the risk of metastatic disease in tumours smaller than 2 cm was sufficiently low to treat them by local resection alone (appendcectomy) [7]. Lesions smaller than 1 cm require no staging procedure. Those with tumors larger than 2 cm, incomplete resections, and metastatic disease or goblet cell carcinoids warrant further investigation. Two of our cases had milimetric carcinoid focus in the appendix and no further

treatment was performed. In the third case, mucinous tumor infiltrated all layers of appendix and there was mesoappendiceal involvement with omental metastasis. For this third case, further treatment was necessary such as hemicolectomy and chemotherapy.

Figure 3.



Carcinoid tumor of the appendix. Tumor tissue showed positive immunhistochemical staining for Synaptophysin (x400).

Patients with appendiceal carcinoids have a good prognosis overall [7].

In the literature, appendix carcinoids concomitant with gynecologic malignancies were discovered incidentally at the time of histologic examination of appendiectomy specimens as a part of the staging procedure of endometrial or ovarian carcinomas. Meden et al. presented two cases with pathologic findings of the appendix vermiformis diagnosed during gynecological operations including carcinoid and adenocarcinoma [8]. Surmacki et al. presented a case of a carcinoid of the appendix, myoma of the uterus and adenoma of the uterine cavity [9]. Giesteira et al. reported two cases of carcinoid tumour of the appendix discovered at the time of abdominal hysterectomy [10].

Carcinoid of the appendix does not change the management of primary gynecologic disease in most cases, because the size of the carcinoid is less than 2 cm in these cases. But if the surgical margins are not intact, size of the carcinoid is more than 2 cm, malignant histological appearance of the carcinoid or involvement of mesoappendix or lymph nodes require further evaluation and treatment such as right hemicolectomy and/or chemotheraphy.

Conflict of interest statement

The authors declare no conflict of interest.

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