Case Report

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

Murat Oz1,*, Emre Ozgu1, Hakan Yalcin1, Mengü Turker2, Salim Erkaya1, Tayfun Gungor4

1 Department of Gynecologic Oncology and 2 Department of Pathology, Zekai Tahir Burak Women’s Health, Education and Research Hospital, Ankara, Turkey

Abstract
Carcinoid tumors of appendix are rare tumors and preoperative diagnosis may be challenging. The prevalence 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 chemotherapy. 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 prevalence 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 appendectomy 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 salpingo-oophorectomy (BSO), omentectomy, appendectomy, bilateral pelvic and para-aortic lymph node dissection (BPPLND) was performed. Appendix vermiformis had normal gross appearance. 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...
specimens showed well differentiated endometrium adenocarcinoma, showing myometrial invasion less than ⅔ 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.

Discussion

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

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 (appendectomy) [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.

Patients with appendiceal carcinoids have a good prognosis overall [7]. In the literature, appendiceal carcinoids concomitant with gynecologic malignancies were discovered incidentally at the time of histologic examination of appendicectomy 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]. Giesteria 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 chemotherapy.

Conflict of interest statement
The authors declare no conflict of interest.

References