Synchronous Primary Heterorologous Type of Malignant Mixed Mullerian Tumor of the Uterus and Primary Ovarian Cancer

- an unusual case report -

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Abstract

The fetal face and neck represents one of the key anatomic regions in ultrasound investigation from a pscyhological and clinical standpoint point of view. Although the sonographic appearence of some masses is characteristic, a substantial overlap exist in echotexture of the masses of the face and neck. In addition to analysing the echotexture of the mass, attempts should be made to determine: the site of origin, the presence of distortion of surrounding anatomy, the presence or absence of other malformations and polyhidramnios. Cervical teratomas are very rare and account for 5% of all teratomas. The majority of them are benign, often large and produce airway obstruction. We present a rare case of a very large cervical teratoma diagnosed at a late gestational age.

Keywords: *cervical teratoma, sonographic diagnosis, polyhidramnios*

Introduction

Synchronous multiple primary tumors of the female genital tract is a well known event but occurs only 1% to 6% of all genital tract neoplasms[1]. The most frequent synchronous gynecological neoplasms reported are endometrial and ovarian cancer[2]. However synchronous malignant neoplasms of ovary and cervix, endometrium and cervix have also been reported. Coexistance of carcinosarcoma of uterus with other gynecological neoplasms is unusual. There are few case reports of primary carcinosarcoma concomitant with yolc sac tumor of uterus in the literature[3,4,5]. Isın et al.[6] reported a case of triple simultaneous primary gynecological malignancies consisting of ovarian mucinous adenocarcinoma, endometrial adenocarcinoma and leiomyosarcoma of uterus. As far as we know this is the first case of uterine carcinosarcoma synchronous with primary adenocarcinoma of ovary.

Case report

A 47 year old multiparous woman was referred to our department with 4-month history of malodorous vaginal discharge. Her past medical and surgical histories were unremarkable. Bimanual pelvic examination revealed that the uterus was slightly enlarged, the cervix was enlarged to 8 cm diameter and a solid-cystic mass was arising from the left lower pelvis. The parametrium were considered as bilaterally free. Pelvic ultrasound scan showed 30x30 mm polipoid solid mass in the endocervical canal, 63x48 mm sized solid and cystic complex mass having papillary projections in the left ovary, endometrial thickness was 13 mm. A pelvic MRI demonstrated contrast enhanced 5x7 cm solid mass in cervix and contrast enhanced 6x8 cm complex mass in the left ovary. Endometrial, endocervical biopsy and biopsy from cervical mass revealed well differentiated squamous cell carcinoma. Cancer antigen (CA) 125, CA 19-9, CA 15-3, alpha-feto protein and carcinoembryonic antigen levels were normal within referance range(17.6 U/mL, 35.35 U/mL, 19.1 U/mL, 5.12 U/mL, repectively). Chest radiography, laboratory tests including complete blood count and blood chemistry were normal.

The patient was subjected to operation with a diagnosis of cervical carcinoma and ovarian tumor. At laparotomy, a left ovarian tumor measuring approximately 6x5 cm was discovered, frozen section was performed and the result was adenocarcinoma. Type 3-Hysterectomy and bilateral salpyingo-oophorectomy, bilateral pelvic and para-aortic lymph node dissection, omentectomy and appendectomy were performed. Peritoneal washing fluid was collected.

On gross examination, a grayish white solid polipoid mass with partial hemorrhage and necrosis was detected. The tumor was measuring 5x4x3 cm and originating from uterine fundus, extending to cervical canal. Endometrium was 20mm except this area. The left ovary was

enlarged about 7x5,5x5,5 cm in size, contained a shiny white capsulated cystic mass with an irregular gelatinous tumoral area sized 6x5 cm. Microscopic examination of polipoid mass showed malignant mixed mullerian tumor with less than half of myometrial infiltration. Most of the tumor consisted of the epithelial component, well differentiated squamous cell carcinoma area was more than well differentiated endometrioid adenocarcinoma. On the other hand mesenchymal component contained poorly differentiated sarcoma, stromal sarcoma and osteosarcoma. Immunohistochemical analysis revealed that squamous cell carcinoma was positive for EMA, poorly differentiated sarcoma area was positive for vimentin, stromal sarcoma area was positive for CD10 (figure 1). Vascular invasion was prominent but regional lymph nodes were negative. Omentum and appendix were unremarkable. On microscopy of left ovary, endometrioid adenocarcinoma was detected. Endometrioid areas were presented in normal ovarian tissue which confirmed that primary endometrioid adenocarcinoma was associated with presence of ovarian endometriosis (figure 2).

Therefore the final diagnosis was a FIGO stage 1A endometrioid adenocarcinoma of left ovary and stage 1A carcinosarcoma of uterus. Postoperatively, the patient was planned to receive adjuvant chemotherapy and brachytherapy.

Discussion

The etiology of synchronous tumors remains unclear but there are several theories postulated to explain. It has been suggested that embryologically similar mullerian tissues may develop synchronous neoplasms when stimulated by same oncologenic carcinogens[7]. Others suggested that neoplasms originate in metaplasia, shared hormonal receptors may cause multiple primary tumors in histologically similar tissues[1]. Also it has been clearly demonstrated that some of synchronous neoplasms in female genital tract are related to familial cancer syndromes or genetic mutations.

In synchronous tumors if the histologic types are different, it is easy to identify the coexistance of primary cancers. On the other hand in the cases of synchronous detection of neoplasms with similar histologic features, the distinction between two primary independent primary tumors or metastasis may be difficult. In endometrioid tumors of endometrium and ovary, Ulbright and Roth[8] and Scully et al.[9] created clinicopathologic fetaures to differentiate primary cancers from metastasis. Presenting ovarian endometriosis is a criteria for primary endometrioid adenocarcinoma of ovary that suggested in our case. Only superficial myometrial invasion, no distant metastasis, no direct extention between tumors are also criteria for synchronous primary cancers in our case also.

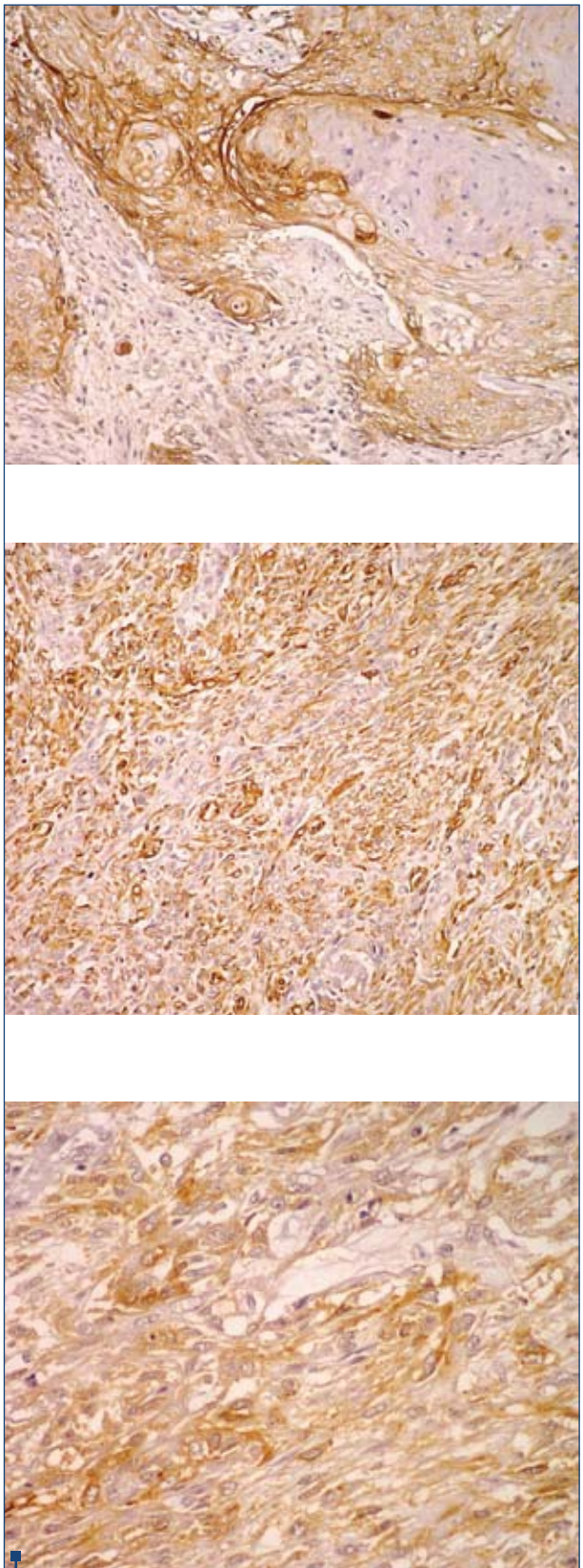

Figure 1. (A) Squamous cell carcinoma component positive for EMA; (B) Poorly differentiated sarcoma area positive for vimentin; (C) stromal sarcoma area positive for CD10

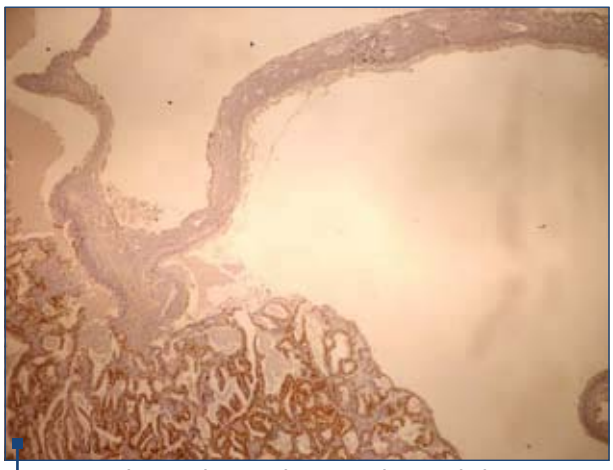

Figure 2. Endometrioid areas and primary endometrioid adenocarcinoma in ovarian tissue

Although the most common reported syncronous malignancies were the coexistance of endometrial and ovarian cancers[2], the main difference in this case was coexistance of carcinosarcoma. Carcinosarcomas accounts for almost half of uterine sarcomas and most cases are postmenopausal. Carcinosarcomas are typically large, bulky polipoid masses, filling the uterine cavity and prolapsing through the cervical os[10]. Rarely tumors may arise from cervix. Microscopically epithelial components mainly composed of serous(two-thirds of cases) or endometrioid (one-third of the cases) carcinomas. Sarcomatous components are heterogenous. This case was also different in the aspect of the prominent epithelial component which was well differentiated squamous cell carcinoma.

In conclusion we described a rare case of synchronous uterine carcinosarcoma and primary adenocarcinoma of ovary which is an unique event and may guide for future studies. ■

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