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Primary Carcinosarcoma of the Uterine Cervix-Report of a case and review of the literature

Jessenia Guerrero, MD*, Tania Aftandilians MD**, Bernadette Cracchiolo, MD, MPH**, Debra Heller, MD*,**

From the Departments of Pathology & Laboratory Medicine*, and Obstetrics, Gynecology, and Women's Health**, Rutgers-New Jersey Medical School, Newark, NJ

Address Correspondence to:

Debra S. Heller, MD

Dept of Pathology-UH/E158

Rutgers-New Jersey Medical School

185 South Orange Ave

Newark, NJ, 07103

Tel 973-972-0751

Fax 973-972-5724

hellerds@njms.rutgers.edu

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Precis:

A rare case of cervical carcinosarcoma is presented, and the literature reviewed.

Keywords: Carcinosarcoma, uterine cervix, malignant mixed Müllerian tumors, radical hysterectomy

Abstract

Background: Cervical carcinosarcoma is extremely rare.

Case: A case of cervical carcinosarcoma is presented and the literature reviewed.

Conclusions: Due to the rarity of this lesion, there are no specific treatment protocols.

Spread from a uterine primary needs to be ruled out.

Introduction:

Carcinosarcoma , also known as malignant mixed Müllerian tumor (MMMT), is an uncommon entity that accounts for less than 5% of uterine malignancies [1].

Cervical carcinosarcomas are even rarer. Optimal treatment remains unclear due to its rarity and limited number of cases found in the literature. We describe a case and review the literature.

Case:

A multiparous woman in her mid-forties, with multiple medical comorbidities including BMI of 48, was referred to our hospital's gynecologic oncology service for adenocarcinoma identified on routine cervical cytology. The cytology was HPV 16/18 negative, raising concern for a primary endometrial rather than cervical carcinoma. Her gynecological history is significant for irregular menses and there is no known family history of malignancy.

On examination, a 3.0 cm exophytic lesion was seen replacing the cervix. An office cervical biopsy showed carcinosarcoma of endometrial vs. cervical origin. P16 immunostain was positive and CEA was focally positive, favoring a cervical primary, and the tumor stained for AE1/3, EMA, and CK7, and focally for vimentin in spindle cell areas. Chromogranin and synaptophysin were negative. The patient was unable to tolerate endometrial sampling to better determine the origin of this malignancy. She underwent a PET scan which revealed heterogeneous uptake in the mid- and left endocervix with no uptake in the uterus and no evidence of metastatic disease.

The patient underwent a robotic-assisted radical hysterectomy and pelvic lymph node dissection for stage IB1 cervical carcinosarcoma. Her postoperative course was complicated by ileus, which resolved spontaneously.

Pathology:

Gross examination showed an exophytic cervical lesion (figure 1). Histologic evaluation showed carcinosarcoma of the cervix (2.0 x 1.0 cm in greatest dimension) with < 1 mm invasion. The tumor was biphasic, with focal chondroid differentiation (figure 2). The epithelial areas stained for CK 5/6, p63, and p40 with some lesser staining of the spindle areas, which in turn were strongly positive for vimentin (figure 3). No lymphovascular invasion was noted. Additional findings included weakly proliferative endometrium, an endometrial polyp, adenomyosis, and unremarkable bilateral fallopian tubes and ovaries. Lymph nodes were negative.

Discussion

Carcinosarcoma of the uterine cervix is extremely rare. There were approximately 35 reported cases as of 2008, representing less than 0.5% of cervical cancers[2]. Reported cases have occurred in predominantly postmenopausal women, with median age at diagnosis between 62 and 69 years, and age range of 12 to 87 years[2]. The most common presenting symptom is vaginal bleeding and usually a detectable exophytic mass is seen on the cervix [3]. Other symptoms can include pain, abnormal cervical smear, passage of tissue, or a palpable mass[2]. Due to histologic similarity, the differential diagnosis includes sarcomatoid carcinoma. The principal characteristic feature of a carcinosarcoma is the mixture of carcinomatous and sarcomatous components that remain distinct from each other, whereas in sarcomatoid carcinoma, both components have the tendency to merge. Both neoplasms are thought to be due to metaplastic carcinoma, and may indeed be part of the same spectrum [4].

Uterine carcinosarcoma with spread to the cervix needs to be ruled out in cases of cervical involvement. In addition to the positive p16 immunostain in our case, the negative endometrium was confirmatory. Due to the rarity of cervical carcinosarcoma, there are no specific treatment protocols. Prognosis appears to be better than for the uterine counterpart, possibly due to early presentation[2], but the disease can be very aggressive. The best prognosis appears to be with complete excision, with possible adjuvant chemotherapy and/or radiation added by some[5].

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Abbreviations and Acronyms

HPV-Human papillomavirus

PET-positron emission tomography

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