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Primary Carcinosarcoma of the Uterine Cervix: Case Report and Review of the Literature

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Abstract: *Carcinosarcoma of the uterine cervix are very rare tumors, constituting only 0.005% of all cervical malignancies (fewer than 70 cases of cervical carcinosarcoma have been reported in the English literature). It usually presents with abnormal vaginal bleeding in post-menopausal women. The tumor has high rates of metastasis, so careful management including surgery, chemotherapy and/or radiotherapy is done for these patients.*

Here, we present a case of a 60 year old woman who present with an ectocervical tumor. She underwent radical hysterectomy and bilateral pelvic lymphadenectomy; the tumor was diagnosed on histopathological examination as Carcinosarcoma of the uterine cervix. Following surgery, the patient was given radiotherapy with concurrent chemotherapy. Four months late, discovered a mesenteric recurrence. The patient was treated by surgery followed by palliative chemotherapy. This treatment had no effect and she died of peritoneal carcinomatosis three months after starting the palliative chemotherapy.

carcinosarcoma of the uterine cervix is a rare disease, mostly occurring in older age and advanced stage. Optimal treatment is unclear. Aggressive primary therapy can result in cure of early-stage tumors.

Keywords: *Carcinosarcoma; uterine cervix; Müllerian tumor mixed; Chemotherapy; Radiotherapy*

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I Introduction

Primary carcinosarcomas, also referred to as malignant mixed mullerian (mesodermal) tumors (MMMTs), are extremely rare tumors of the uterine cervix that constitute less than 0.005% of all cervical malignancies. Carcinosarcoma is a biphasic tumor including both epithelial and mesenchymal malign structures[1]. Cervical carcinosarcoma can be characterized by having two different origins: the Müllerian ducts and the mesonephric duct remnants[2].

Cervical carcinosarcoma (CCS) is an aggressive tumor. The optimal therapy, prognosis, prognostic factors, risk of recurrence, and survival are not clear because of the rare occurrence of primary CCS (Few case series and reports exist in the literature) [1]. In this study, we presented 1 case with primary CCS.

II Case Report

A 60-year-old woman consulted the gynaecologist because of vaginal bleeding, fetid leucorrhoea and pelvic pain. She was gravida 5, para 5, in menopause since 43 years. She had no medical history or familiar malignancy history. Vaginal exploration showed a friable cervical mass protruding into the vagina. The biopsy specimen revealed a carcinosarcoma.

The cervical mass measured about 9x8 cm on magnetic resonance imaging (Fig. 1 A and B).

A positron emission tomography scan, revealed only an isolated increased uptake of the radioactive tracer confined to the mass of the uterine cervix extended to the vagina with no uptake in the uterus and no evidence of metastatic disease.

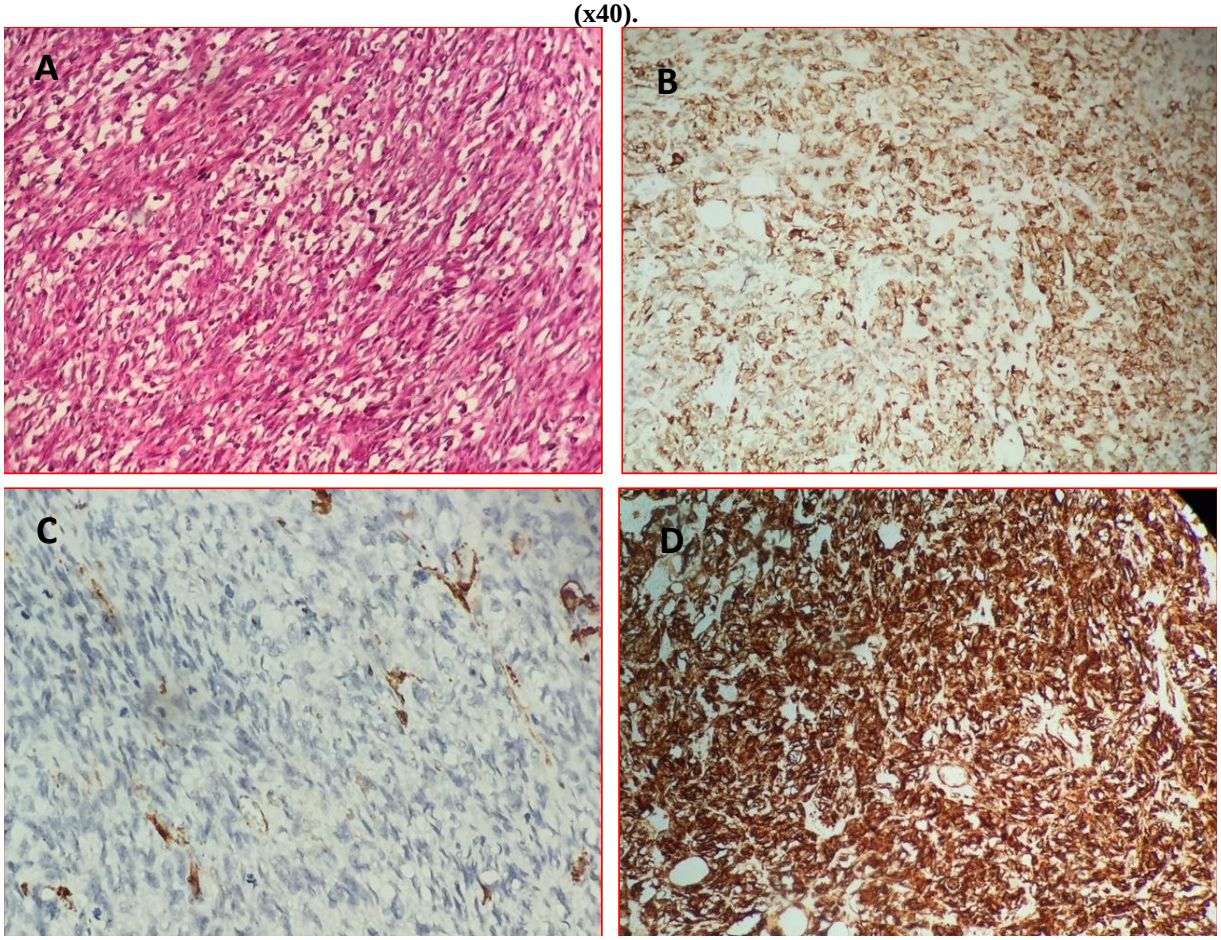
She underwent radical hysterectomy, bilateral salpingo-oophorectomy, and bilateral pelvic lymphadenectomy.

Grossly, there was a solid mass in the uterine cervix measuring 11x7x.6 cm.

Microscopically, the cervical mass revealed a malignant tumor with epithelial and mesenchymal components (Fig. 2 A). Immunohistochemical studies were performed, the epithelial component was positive for epithelial membrane antigen and cytokeratins AE1-AE3 (Fig. 2 B). On the other hand, the malignant mesenchymal component was positive for vimentin (Fig. 2 D). Absence of expression of smooth muscle actin, expressed by vascular walls (Fig. 2 C).

There was no parametrial involvement, no endometrial involvement and no lymph node metastasis, and the patient was free of surgical margins.

Fig 1: (A) Malignant tumoral proliferation of sarcomatous appearance (HEx40). Immunohistochemical staining: (B) Diffuse and intense expression of cytokeratin AE1-AE3 (x40), (C) Absence of expression of smooth muscle actin, expressed by vascular walls (x40), (D) Diffuse and intense expression of vimentin (x40).

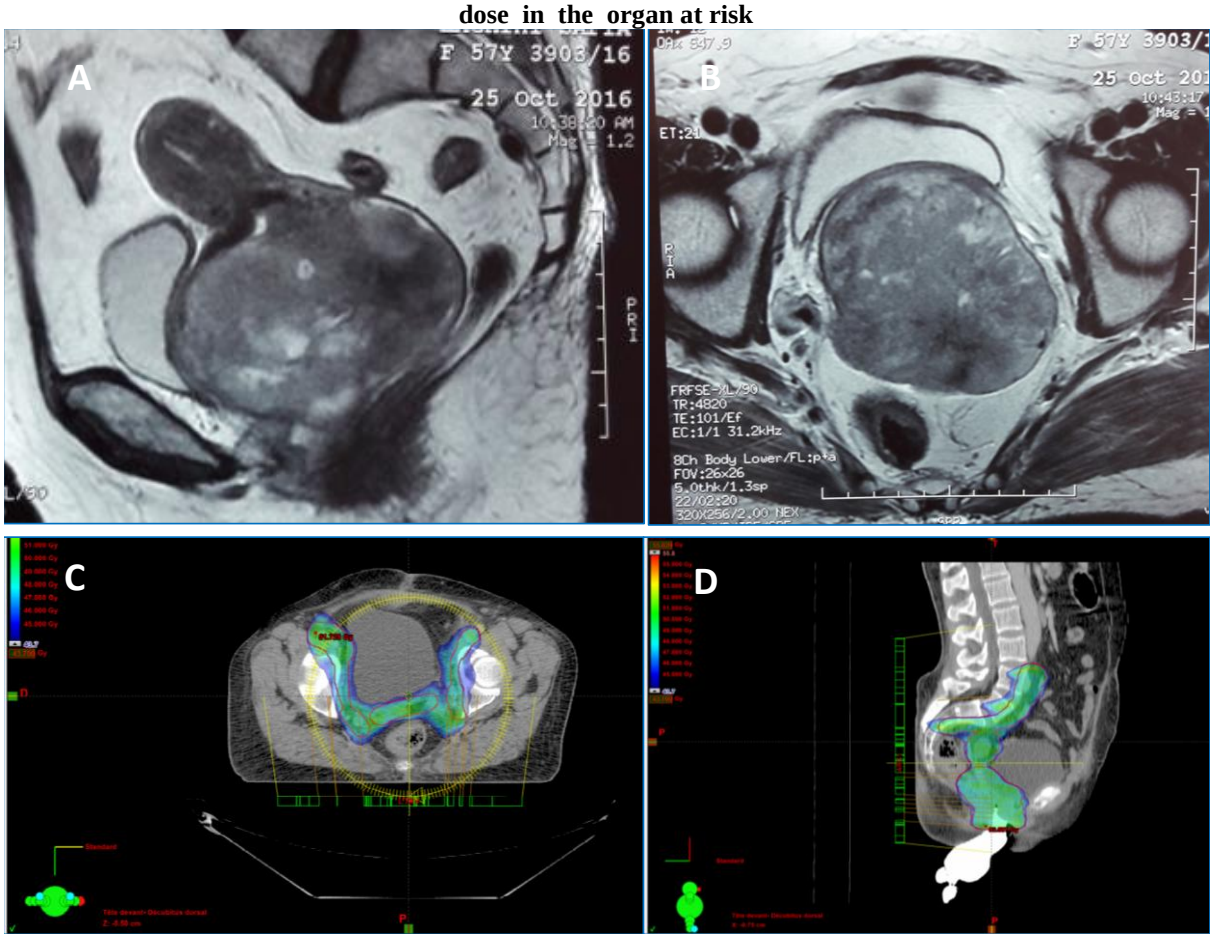


Following surgery, the patient was given an external pelvic radiotherapy 46 Gy in a 2 Gy daily fraction, 5 days a week. Radiotherapy was delivered using volumetric modulated arc radiation therapy VMAT technique (rapidarc) (Fig. 1 C and D) with concurrent chemotherapy (cisplatin 40 mg/m2 weekly) followed by 7 Gy × 2 fractions of high dose rate brachytherapy.

Computed tomography (CT) performed 4 months after completion of the adjuvant treatment showed a mesenteric mass of the left flank measuring 30x34 cm. A positron emission tomography scan, revealed an hypermetabolic mass of the left flank.

The patient underwent a surgical exeresis of the mass followed by chemotherapy consisting of Ifosfamide and cisplatin. This treatment had no effect and she died of peritoneal carcinomatosis three months after starting the palliative chemotherapy.

Fig 2: (A-B) Magnetic resonance imaging (MRI): tumor involving the uterine cervix, measuring 9x8 cm. (C-D) treatment by volumetric modulated arc radiation therapy :the use of two arcs allowed the optimal coverage of the target volume (red) by isodoseline 95%(blue) without exceeding the permissible level dose in the organ at risk



III Discussion

Carcinosarcoma is uncommon malignancy in female reproductive system, composed of carcinomatous component and sarcomatous component. Uterus corpus is the most common location of carcinosarcoma, however, it can be also located in uterine cervix, ovary, fallopian tube, vagina, Douglas pouch or even peritoneum. Cervical carcinosarcoma is rare: Including this case, fewer than 70 cases of CCS have been reported in the English literature[3].

Reported cases have occurred in predominantly postmenopausal women, with a median age at diagnosis between 62 and 69 and an age range of 12–87. 2 The most common presenting symptom is vaginal bleeding and usually a detectable exophytic mass is seen on the cervix. Other symptoms can include pain, passage of tissue, or a palpable mass[4].

Grossly, cervical tumors range in size from 1.1 to 10 cm in maximal dimension. The tumors are usually large, soft, broad-based and polypoid with a fleshy cut surface, often with areas of hemorrhage and necrosis[5]. Microscopically, MMMTs are biphasic tumors, composed of distinctive and separate, but admixed, malignant appearing epithelial and mesenchymal elements[6]. The epithelial component is usually poorly differentiated and represents a variety of different subtypes, alone or in combination, including squamous cell carcinoma, basaloid squamous carcinoma, adenocarcinoma, adeno-squamous carcinoma, adenoid basal carcinoma, adenoid cystic carcinoma and undifferentiated carcinoma[7]. The sarcomatous component may be homologous or heterologous. The homologous sarcoma has the appearance of a spindle cell sarcoma, often poorly differentiated, and contains fibroblasts or smooth muscle cells. Heterologous tumors contain one or more of the following elements in descending order of frequency: rhabdomyoblasts, mature appearing cartilage or chondrosarcoma, osteoid, bone or osteosarcoma and liposarcoma. Rarely, it may contain neural or neuroendocrine elements[8]. On immunohistochemical examination, both epithelial and sarcomatous components may show positivity for broad spectrum cytokeratins, low molecular weight cytokeratins, high molecular weight cytokeratins and epithelial

membrane antigen. Immunoreactivity for a variety of muscle specific markers such as actin, desmin, myosin and myoglobin are almost invariably confined to the sarcomatous cells[7].

Evidence-based guidelines for treatment of CCS are not available due to its rarity [5]: CCS are managed with surgery, chemotherapy and radiotherapy. Surgery should include hysterectomy, bilateral salpingo-oophorectomy, and pelvic lymphadenectomy. Carcinosarcomas also require comprehensive peritoneal surgical staging including peritoneal cytology and biopsies[5, 9, 10]. The role of adjuvant chemotherapy and radiotherapy are not well defined. Radical radiotherapy with or without chemotherapy is recommended for locally advanced disease. Patients with metastatic disease are treated with palliative chemotherapy and have a poor prognosis[5].

Data about adjuvant therapies are limited and mainly obtained from studies on sarcomas of the uterine corpus. Radiotherapy has been employed in patients with pelvic disease, but the impact on survival is uncertain. In the review of Iida et al., 5/10 patients submitted to surgery and adjuvant radiotherapy, and 3/3 patients treated with radiotherapy alone died of disease within 18 months. Similar data are reported by Wright et al in a study on five cervical carcinosarcomas[11].

Ifosfamide and cisplatin have shown activity in carcinosarcomas of the corpus and have been employed in palliative treatment of metastatic carcinosarcomas of the cervix. Although some activity seems warranted, the benefit of adjuvant chemotherapy in early-stage disease is difficult to be definitively established, given the rarity of the disease[11].

IV Conclusion

Carcinosarcoma of the uterine cervix is a rare disease, mostly occurring in older age and advanced stage. Aggressive primary therapy can result in cure of early-stage tumors. Extracervical disease is usually associated with a poor prognosis.

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