A case of uterine cervical carcinosarcoma with good prognosis

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Carcinosarcoma of the uterine cervix is rare and generally regarded as a very aggressive tumor with a poor prognosis. However, there could be very unusual reported cases in which cervical carcinosarcoma has a good prognosis. We present a case of cervical carcinosarcoma with invasion of the endometrium and fallopian tubes in a patient who is alive with no recurrence more than 3 years after the initial surgery. A 61-year-old nulligravida with post-menopausal genital bleeding visited our outpatient clinic. Transvaginal sonography and magnetic resonance imaging revealed an endometrial tumor and adnexal cysts with solid components on either side of the uterus. Endometrial malignancy was diagnosed, and she underwent total abdominal hysterectomy, bilateral salpingo-oophorectomy, and adjuvant chemotherapy with cisplatin and ifosfamide for 6 months. Gross examination of the surgically resected specimen revealed it to be a polypoid and bulky tumor (5 × 4.5 cm) arising from the endocervix. Apart from this, a polypoid sessile tumor adhering to the endometrium and both tubal lumina was detected. Histologically, the tumor had arisen exophytically from the endocervix; it had adenocarcinomatous and chondrosarcomatous components. Malignant epithelial tumors resembling the epithelial component of the endocervical tumor were also detected in the endometrium and fallopian tubes. Despite the predicted poor prognosis, the patient is currently alive and free of disease more than 36 months after the initial surgery. It is likely that the biological behavior and prognosis of these tumors are different than those expected. Therefore, cervical carcinosarcoma may show better prognosis than endometrial carcinosarcoma.

Keywords: carcinosarcoma, cervical cancer, chondrosarcoma, adenocarcinoma, prognosis

INTRODUCTION

Carcinosarcoma (CS) of the uterus is a rare neoplasm that usually arises from the endometrium. CS of the uterine cervix is extremely rare, generally progresses aggressively, and has a very poor prognosis. However, in rare cases, the prognosis of cervical CS may be relatively favorable. Here, we present a case of advanced cervical CS. The patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy and subsequently received chemotherapy with cisplatin and ifosfamide. She is currently free of disease more than 36 months after the operation. Here, we discuss the prognosis of cervical CS.

CASE REPORT

A 61-year-old Japanese nulligravida complaining of post-menopausal genital bleeding visited our outpatient clinic. Examination with a speculum did not reveal any abnormalities in the vaginal portion of the uterus. However, pelvic examination revealed enlargement of the uterus and the presence of a large pelvic tumor (5 × 6 cm). Transvaginal sonography revealed an endometrial tumor and huge adnexal cysts with solid components on either side...
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of the uterus. Cytological analysis of the endometrium revealed the tumor to be class 5 in the Papanicolaou test, suggestive of adenocarcinoma. The patient was admitted to our hospital for further examination. A pelvic examination on admission revealed a necrotic tumor protruding from the cervical os. Histological findings of these tumors that were easily resected indicated that adenocarcinoma accounted for the majority, but atypical mesenchymal component with positive vimentin staining was present in the focal area. Magnetic resonance imaging (MRI) revealed an endometrial tumor with no evidence of myometrial involvement. In addition, adnexal cysts with solid components were detected on either side of the uterus. No adenopathy was detected in the pelvic and para-aortic regions. The results of a complete blood count and serum biochemical analysis were unremarkable. The levels of tumor markers such as carbohydrate antigen (CA) 125, CA19-9, CA72-4, and the carcinoembryonic antigen were all within their respective normal ranges.

The patient was diagnosed with uterine endometrial cancer or carcinosarcoma with bilateral ovarian metastasis, and she underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. A small volume of bloody ascites was intraoperatively detected. Adnexal cysts were present in both fallopian tubes, and the condition progressed to the hydrosalpinx. These cysts had a smooth, unbroken surface, and had grown into the area of the uterine ligament. The left adnexal cyst extended to the xiphoid process. These cysts tightly adhered to adjacent retroperitoneal tissue. Neither pelvic nor para-aortic lymph node dissection could be performed because the retroperitoneal space could not be addressed for these severe adhesions. Gross examination did not reveal any dissemination of cancer cells in the abdominal cavity.

Gross examination of the surgically resected specimen revealed it to be a polypoid and bulky tumor (5 × 4.5 cm) arising from the endocervix. This endocervical tumor was mistakenly perceived as endometrial tumor by MRI. Apart from this, a polypoid sessile tumor (2 × 1.5 cm) adhering to the endometrium was detected. Furthermore, another polypoid tumor arising from both tubal lumina was detected. Light microscopy showed that the endocervical tumor was composed of a mixture of malignant epithelial and mesenchymal elements, the former being predominant. The epithelial element had serous and endometrioid adenocarcinomatous components, which had focally invaded the lymphatic canal and the endocervical stroma to a depth of 3 mm. No vascular canal invasion was detected. Each of these epithelial elements was grade 3. The mesenchymal element resembled chondrosarcoma (Figure 1, Figure 2).

Figure 1. Light microscopic images of the endocervical tumor stained with hematoxylin and eosin (HE) at low magnification. The tumor is composed of a mixture of malignant epithelial and mesenchymal elements.

Figure 2. Light microscopic images of the endocervical tumor stained with hematoxylin and eosin (HE) at high magnification. The epithelial element has an endometrioid adenocarcinoma component. The mesenchymal element resembles chondrosarcoma.
The sessile endometrial tumor and the polypoid tumor arising from the tubal lumina were solely composed of adenocarcinoma cells; the former was predominantly a serous adenocarcinoma while the latter was an endometrioid adenocarcinoma. These tumors had not invaded the submucosal layer. These epithelial malignancies, arising from endometrium and tubal lumina, closely resembled the epithelial malignancy arising from the endocervix (Figure 3, Figure 4).

However, none of the three tumors, arising from the endocervix, endometrium, or tubal lumina, was contiguous. The cervical lesion was the largest and the only one to show stromal invasion among the three lesions. On the basis of these findings from light microscopy, we conclusively diagnosed the condition as endocervical carcinosarcoma (CS) of stage pT1b2, pNx, pM1, with invasion of the endometrium and tubal lumina.7

The patient was administered post-operative chemotherapy with cisplatin and ifosfamide for 6 months. As of 36 months after the surgery, the patient is alive with no evidence of tumor recurrence, as assessed by computed tomography.

DISCUSSION

Cervical CS is characterized by an admixture of malignant epithelial and mesenchymal elements. The median age of affected patients is 62.8 years, and the most common early manifestation is post-menopausal genital bleeding.2,4 On gross examination, cervical CS appears as a polypoid or bulky tumor growing over the cervix.5,8 Uterine CS accounts for less than 1.5% of all uterine malignancies and usually arises from the endometrium. Since the first description of cervical CS in 1973, less than 50 cases have been reported in the English literature.1,2,8-10

The histogenesis of cervical CS remains unclear, but is suggested to be analogous to that of endometrial CS.6 Three hypotheses have been proposed to explain the histogenesis of uterine CS: composition, collision, and combination theories. The combination theory, which is most favored, describes both the epithelial and the mesenchymal components originating from a single stem cell clone.9 However, in some cases, the tumor may originate from 2 distinct cell lines. This is the basis of the collision theory, according to which synchronous biclonal tumor cells merge to acquire the histology of CS. In all probability, both histogenetic mechanisms exist and may be applicable in certain cases.8 The principal factors that influence the progression-free interval (PFI) in CS patients are lymph node involvement, cell type, adnexal involvement, and the sarcoma grade. The presence of a sarcomatous com-

![Figure 3](image1.png)

**Figure 3.** Light microscopic images of the bilateral tubal luminal tumor stained with HE at high magnification. Serous adenocarcinoma of tubal lumina resembles an epithelial malignancy arising from the endocervix.

![Figure 4](image2.png)

**Figure 4.** Light microscopic images of the endometrial tumor stained with HE at high and low magnification. Serous and endometrioid adenocarcinoma of endometrium resembles an epithelial malignancy arising from the endocervix.
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Cervical carcinosarcoma (CS) is thought to be a very aggressive tumor, metastasizing to other organs even in early stages. In fact, 40–50% of cases are already at an advanced stage and refractory to treatment at the time of diagnosis. The median survival time is 18 months for patients with any stage of cervical CS. However, because of the aggressiveness of the tumor, the interval from the onset of symptoms, usually post-menopausal genital bleeding, to diagnosis is relatively short (median, 3.7 months). This facilitates early diagnosis and improves the prognosis. In fact, morality associated with cervical CS is lower than that associated with endometrial CS, possibly because the former has a better prognosis.

In our opinion, the paradox of the aggressiveness of cervical CS facilitating early diagnosis causes confusion regarding the prognosis of this condition. In the present case, the cervical tumor was bulky and metastasized very aggressively to the endometrium and tubal lumina; therefore, the predicted prognosis was very poor. Nevertheless, the patient has been disease-free for more than 36 months after the initial operation. Therefore, the question remains as to whether there are specific differences between cervical and endometrial CS in terms of biological behavior and prognosis. In a previous study, human papilloma virus DNA was detected in all 8 cases investigated, in 3 of which HPV-16 DNA was detected in both epithelial and mesenchymal cells. Thus, a relationship may exist between HPV infection and the histogenesis of cervical CS and may contribute to the behavioral and prognostic differences between cervical and endometrial CS. Unfortunately, we could not obtain HPV staining in this case.

In summary, we present a rare case of cervical CS with invasion of the endometrium and tubal lumina. The patient underwent total abdominal hysterectomy, bilateral salpingo-oophorectomy, and subsequently received chemotherapy with cisplatin and ifosfamide. Although cervical CS is generally thought to have poor prognosis, similar to endometrial CS, the patient has survived disease-free as of more than 36 months after the surgery. Thus, the histogenesis and prognosis of cervical CS remain unclear and warrant further investigation.

REFERENCES

予後良好であった子宮頚部原発癌肉腫の1例

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子宮頚部由来の癌肉腫は非常にまれな腫瘍であり、予後不良であると一般的には考えられている。今回我々は、子宮頚部由来の癌肉腫が子宮内膜と卵管に進展していたが、初回治療後3年間無病生存している症例を経験したため、若干の文献的考察を加え報告する。

症例は61歳、未経妊女性。閉経後不正性器出血を主訴に当科外来を初診。初診時の経腔超音波検査およびMRIでは、子宮内膜腫瘍および充実部分を伴う両側付属器腫瘍を指摘、卵巢転移を伴う子宮内膜癌との術前診断にて子宮及び両側付属器切除術を施行した。病理組織学的検査にて、子宮内頚部より外向発育性の腫瘍を認め、同部には腺癌成分と軟骨肉腫成分を認めた。腺癌成分と類似した癌腫成分を子宮内膜および卵管内膜に認めた。子宮頚部由来の癌肉腫、および両腫瘍の子宮内膜、卵管転移と診断。シスプラチンおよびイホマイドによる術後全身化学療法を施行。非常に厳しい予後が予想されたが、初回治療後36か月の時点で無病生存中である。子宮頚部由来の癌肉腫は子宮内膜由来の癌肉腫とは生物学的特性が異なる可能性が考えられた。

キーワード：癌肉腫、子宮頚癌、軟骨肉腫、腺癌、予後