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Case Report

Uterine carcinosarcoma with heterologous mesenchymal element: a case report of a rare and aggressive tumor

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Abstract

Uterine carcinosarcoma (UCS), also known as malignant mixed Müllerian tumor, is a rare malignancy, which consists of both carcinomatous and sarcomatous elements, with a clinical picture resembling endometrial carcinoma. We report a case of a 74-year-old woman is reported with UCS, diagnosed after a 7 months history of vaginal bleeding and abdominal pain. Previous transvaginal sonography showed nonspecific findings, but a repeated one revealed a central uterine mass. Dilatation and curettage and several biopsies were performed. The initial histological report suggested high-grade endometrial stromal sarcoma. After total hysterectomy with salpingo-oophorectomy, pathology confirmed UCS whose sarcomatous element was heterologous type included osteosarcoma and chondrosarcoma. The patient is receiving adjuvant chemotherapy. This case highlights the importance of pathology evaluation after hysterectomy to raise the confidence of diagnosis with emphasis on prognostic outcomes that can be significantly affected in patients with this type of sarcomatous element.

INTRODUCTION

Uterine carcinosarcoma (UCS) -also known as malignant mixed Müllerian tumor- is a highly aggressive rare tumor of the female genital tract, that represents approximately 5% of all malignant uterine tumors [1]. It is responsible for 16% of deaths that are related to uterine malignancies and associated with poor prognosis [2]. This type of tumor is composed of two components: carcinomatous and sarcomatous components [3]. It is an uncommon type in young women and mostly arises in post-menopausal women [2]. The basic treatment is radical hysterectomy, especially in the early stages, and may need adjuvant therapy due to the high rate of relapsing and metastases [3].

We report a case of a 74-year-old postmenopausal female patient, diagnosed with high-grade UCS; whose sarcomatous component contained cartilage and bone foci.

CASE REPORT

A 74-year-old postmenopausal female patient was referred to our hospital with a history of 7 months of vaginal bleeding associated with abdominal pain. On her obstetrical history, she was gravid 9 para 9, while her past medical history included cholecystectomy during her childhood, and lumbar disk surgery a month before the bleeding started. She was diagnosed with recent arterial hypertension.

She had several consultations before coming to the hospital. A The vaginal speculum exam showed no abnormal appearance. Subsequently, a transvaginal ultrasound was performed, which revealed endometrial hyperplasia with a thickness of around 15 mm. Dilatation and curettage (D&C) procedure was performed, but no abnormal changes were detected. With an interval of about two months after the first transvaginal ultrasound, a repeated transvaginal ultrasound revealed a central mass, but it was difficult to determine if it was in the wall or the cavity. Color Doppler sonography demonstrated vascularity within the mass with central calcifications, and no ascites or pleural effusion was detected (Fig. 1).

Following the second transvaginal ultrasound, the patient presented to our hospital. Due to her symptoms and the results of a subsequent transvaginal ultrasound, several biopsies were taken from the cervix and endocervix, which revealed the presence of malignant endometrial cells. Consequently, the initial diagnosis was high-grade endometrial stromal sarcoma.

A total hysterectomy with salpingo-oophorectomy was performed. On pathology, the corpus uteri opening showed large, bulky, polypoid masses that filled the uterine cavity and prolapsed through the cervical canal. These masses were made up of malignant epithelial (glandular) and mesenchymal components (which included foci of cartilage and bone). The final diagnosis was UCS; high-grade; filled the entire uterus cavity, penetrated more than half of the myometrium, and protruded through the cervical canal

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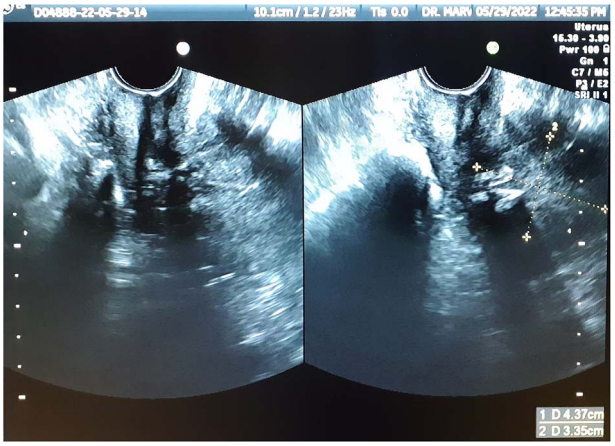

Figure 1. Uterine Sonography.

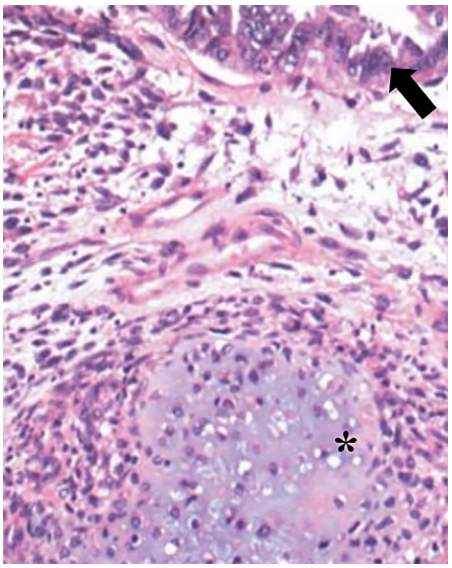

Figure 2. Histopathology of the tumor showing carcinomatous element: High-grade endometrioid carcinoma (Arrow) and heterologous type of sarcomatous element: Chondroid (*), Hematoxylin and eosin, ×20.

(T2b). However, the cervix, ovary, fallopian tubes, and surgical margins were free of tumor. The patient was diagnosed with stage II according to the international federation of obstetrics and gynecology (FIGO) (Fig. 2, Fig. 3, Fig. 4).

After the surgery, the CA125 value decreased. However, an X-ray image revealed increased infiltrates at the base of the lungs (Fig. 5), which was later confirmed by a CT scan indicating the presence of pulmonary metastases at the lung base.

The patient underwent adjuvant chemotherapy consisting of 6 cycles of paclitaxel with carboplatin. Following the completion of adjuvant chemotherapy, a contrast CT of the chest, abdomen, and pelvis was conducted (Fig. 6), revealing no pulmonary masses or metastases and normal organ shape, size, and density. This improvement is currently under continuous monitoring.

DISCUSSION

UCS is a rare tumor and represents less than 5% of all uterine malignancies [1]. patient with UCS is a postmenopausal female. Additional characteristics may include a history of tamoxifen use, and comorbidities such as obesity, hypertension, nulliparity,

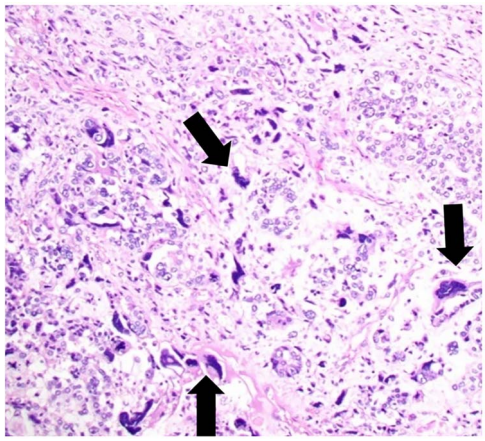

Figure 3. Histopathology of the tumor showing sarcomatous element: Spindle and pleomorphic cells (Arrows), Hematoxylin and eosin, ×20.

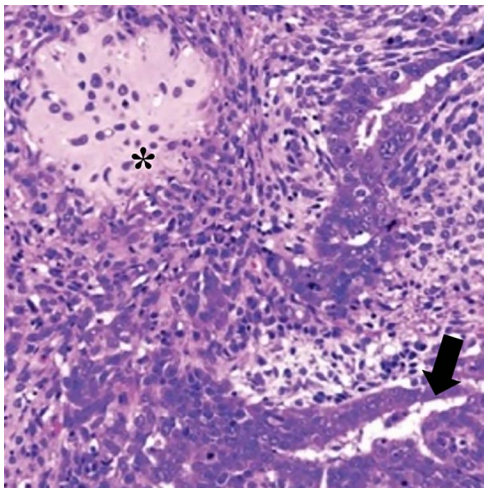

Figure 4. Histopathology of the tumor showing the heterologous type of Sarcomatous element: Chondroid (*) and carcinomatous element: High-grade endometrioid carcinoma (Arrow), Hematoxylin and eosin, ×20.

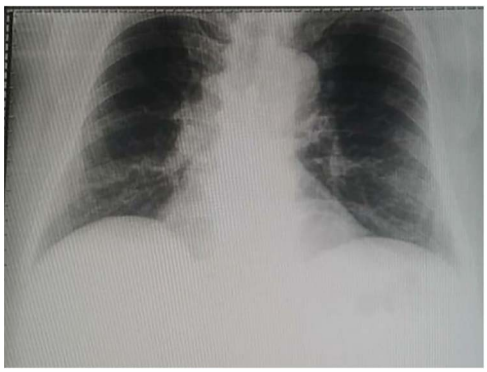

Figure 5. Chest X-ray.

or diabetes. The symptoms include vaginal bleeding, abdominal pain, and uterus enlargement [3]. The most common symptom is vaginal bleeding [1].

Microscopically, UCS is composed of two distinct components: carcinomatous and sarcomatous components. The majority of cases involve only one sarcomatous component. While 33% of

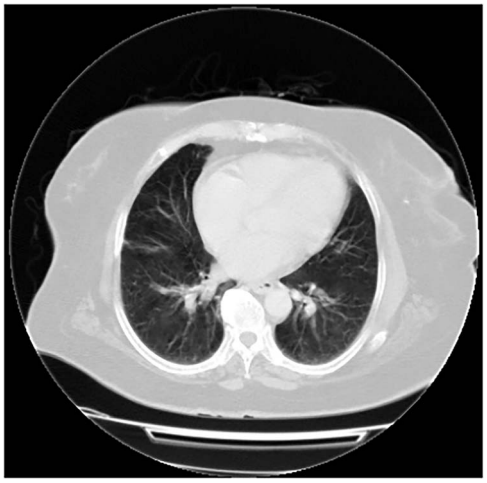

Figure 6. Axial lung base CT Slice.

cases involve two or more sarcomatous components [2]. sarcomatous component has two types heterologous and homologous. The heterologous type comprises rhabdomyosarcoma (18%), chondrosarcoma (10%), osteosarcoma (5%), and liposarcoma (1%). Meanwhile, the predominant form of the homologous type is high-grade stromal sarcoma [3]. Carcinosarcoma is classified based on the type of sarcomatous component [2]. In this case, the sarcomatous component was osteosarcoma and chondrosarcoma. The carcinomatous component in both types of sarcomatous component is typically papillary serous (66%) or endometrioid (42%), or can also be other histotypes [3].

UCS was previously classified as a subtype of uterine sarcoma. UCS is now considered uterine carcinomas. Uterine carcinoma is metaplastic and dedifferentiated [4]. As a result, it is staged similarly to endometrial carcinomas and treated similarly to type II endometrial cancer [2].

Sonography can detect dilated endometrial cavity with or without a mass, or a heterogeneous mass replaces the entire uterus or slight homogeneous echogenic thickening of the endometrium in patients. Echogenic thickening in postmenopausal patients with bleeding is considered abnormal and may indicate the presence of a serious condition, including UCS, and require further evaluation [5]. Echogenic thickening was observed in our case during the first Transvaginal sonography. However, subsequent sonography and other findings suggested the presence of a mass.

Endometrial sampling is commonly performed in women suspected of having uterine malignancy via endometrial biopsy or D&C before definitive surgery. However, it may not accurately diagnose UCS. The complete diagnosis of this condition is typically based on pathologic evaluation after a hysterectomy [6]. Approximately 25% of cases have cervical involvement. Half of those cases have a clinically normal cervix [7]. In our case, the endometrial sample was initially believed to be consistent with endometrial sarcoma. We discovered that it was UCS after the hysterectomy.

The gold standard of surgical management for patients with early-stage UCS is hysterectomy and bilateral salpingo-oophorectom with pelvic lymphadenectomy [4].

Adjuvant therapy is recommended for all stages of the disease and has demonstrated a more favorable prognosis given the highly aggressive nature and poor prognosis of UCS [8]. The presence of the heterologous type should be considered when deciding on the best treatment options for UCS patients [9].

For adjuvant treatment the carboplatin/paclitaxel combination is now the recommended initial treatment for endometrial carcinosarcoma due to its comparable effectiveness and lower toxicity compared to ifosfamide/paclitaxel. However, if a patient is hypersensitive to carboplatin, ifosfamide/paclitaxel and cisplatin/paclitaxel regimens can be considered as alternative options [10]. In our case, the patient received 6 cycles of paclitaxel with carboplatin. The CT scan after adjuvant therapy showed that the metastasis in the lung base have disappeared.

The heterologous type of sarcomatous component in early-stage UCS is an independent negative prognostic factor. The heterologous type can significantly impact patient outcomes when compared to the homologous type. It has a 23.0% rate of 3-year progression-free survival [9].

Irrespective of the treatment given, the 5-year disease-specific survival rates for women with stage I/II, III, and IV disease were 59%, 22%, and 9%, respectively [8].

The recommended follow-up program for UCS is Physical and gynecological examinations every 3–4 months for the first 2 years and every 6 months until 5 years. A CT scan every 12 months for the first 3–5 years. Regular physical and radiological assessments are also recommended [10].

CONCLUSION

Uterine carcinosarcoma is a highly aggressive tumor that is classified treated, and monitored similarly to uterine carcinomas. The outcome is affected by the pathological type of the sarcomatous component. The optimal treatment is hysterectomy and bilateral salpingo-oophorectom. In cases of metastasis, adjuvant therapy with paclitaxel/carboplatin is an effective therapy.

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CONFLICT OF INTEREST STATEMENT

None declared.

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ETHICAL APPROVAL

No approval was required.

PATIENT CONSENT

Written informed consent was obtained.

REFERENCES

1. Denschlag D, Ulrich UA. Uterine carcinosarcomas—diagnosis and management. *Oncol Res Treat* 2018;**41**:675–9. Epub 2018 Oct 13. PMID: 30317231.
2. Artioli G, Wabersich J, Ludwig K, Gardiman MP, Borgato L, Garbin F. Rare uterine cancer: carcinosarcomas. Review from histology

to treatment. *Crit Rev Oncol Hematol* 2015 Apr;**94**:98–104. Epub 2014 Nov 7. PMID: 25468677.

3. Kanthan R, Senger JL. Uterine carcinosarcomas (malignant mixed müllerian tumours): a review with special emphasis on the controversies in management. *Obstet Gynecol Int* 2011;**2011**:470795. Epub 2011 Oct 5. PMID: 22007228; PMCID: PMC3189599.
4. Denschlag D, Thiel FC, Ackermann S, Harter P, Juhasz-Boess I, Mallmann P. *et al.* Sarcoma of the uterus. Guideline of the DGGG (S2k-Level, AWMF Registry No. 015/074, August 2015). *Geburtshilfe Frauenheilkd* 2015;**75**:1028–42. Erratum in: Geburtshilfe Frauenheilkd. 2015 Oct;75(10):e3. PMID: 26640293; PMCID: PMC4651298.
5. Teo SY, Babagbemi KT, Peters HE, Mortele KJ. Primary malignant mixed mullerian tumor of the uterus: findings on sonography, CT, and gadolinium-enhanced MRI. *AJR Am J Roentgenol* 2008;**191**:278–83. PMID: 18562759.
6. Sagae S, Yamashita K, Ishioka S, Nishioka Y, Terasawa K, Mori M. *et al.* Preoperative diagnosis and treatment results in 106 patients with uterine sarcoma in Hokkaido, Japan. *Oncology* 2004;**67**:33–9. PMID: 15459493.
7. Callister M, Ramondetta LM, Jhingran A, Burke TW, Eifel PJ. Malignant mixed Müllerian tumors of the uterus: analysis of patterns of failure, prognostic factors, and treatment outcome. *Int J Radiat Oncol Biol Phys* 2004;**58**:786–96. PMID: 14967435.
8. Gonzalez Bosquet J, Terstriep SA, Cliby WA, Brown-Jones M, Kaur JS, Podratz KC. *et al.* The impact of multi-modal therapy on survival for uterine carcinosarcomas. *Gynecol Oncol* 2010;**116**: 419–23. Epub 2009 Nov 5. PMID: 19896181.
9. Rosati A, Vargiu V, Certelli C, Arcieri M, Vizza E, Legge F. *et al.* Is the sarcomatous component (homologous vs heterologous) the prognostic "driving force" in early-stage uterine carcinosarcomas? A retrospective multicenter study. *J Cancer Res Clin Oncol* 2023;**149**:6479–88. Epub ahead of print. PMID: 36773091.
10. Bogani G, Ray-Coquard I, Concin N, Ngoi NYL, Morice P, Caruso G. *et al.* Endometrial carcinosarcoma. *Int J Gynecol Cancer* 2023;**33**: 147–74. PMID: 36585027.