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Case Report

Undifferentiated uterine sarcoma: case report

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ABSTRACT

Undifferentiated uterine sarcoma is a rare condition characterized by abnormal bleeding and pelvic pain, which may overlap with the clinical picture of uterine myomatosis. Its presentation is usually chronic and drastically affects the stage at the time of diagnosis. Since it is usually seen in postmenopausal patients, undifferentiated uterine sarcoma presentation in premenopausal patients is rare. We herein report a rare presentation of undifferentiated uterine sarcoma in a 58-year-old Mexican woman, who suffered from progressive pelvic pain for 3 months as well as a 5-day course of postmenopausal bleeding. Study images revealed a pedunculated uterine-mass suggestive of sarcomatous degeneration, accompanied by a left internal iliac necrotic adenopathy, as well as scarce pelvic free fluid. The patient underwent a pelvic tumor and implants resection, total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic lymphadenectomy, omentectomy and appendectomy via laparotomy. Despite the therapeutic options for the management of undifferentiated uterine sarcomas described in literature, its survival rate varies between 21% to 68%, when diagnosed on time and depending on if the cancer is localized, regional or distant.

Keywords: Uterine sarcoma, Endometrial sarcoma, Mesodermal tumors, Case report

INTRODUCTION

Uterine sarcomas can have a relative survival rate that varies between 21% to 68%, when diagnosed on time, depending on if the cancer is localized, regional or distant. The importance of identifying and treating uterine sarcomas defines the prognosis of the patients and since uterine sarcomas include a wide varied range of symptoms, they may overlap with the clinical picture of uterine myomatosis, which is why we present a case of a 58-year-old woman diagnosed with regional (pT3b) undifferentiated endometrial sarcoma and uterine myomatosis who died of pulmonary embolism despite the aggressive multidisciplinary therapeutic approach.

CASE REPORT

A 58-year-old woman was referred to the department of obstetrics and gynecology with her chief complaint being 3 months of progressive pelvic pain, as well as a 5-day course of postmenopausal bleeding. Her history included 3 cesarean sections and was otherwise unremarkable. A gynecological evaluation was performed, the speculoscopy was reported negative for transvaginal bleeding, but an irregular, enlarged uterus with a tumor of 6 cm approximately was found with the transvaginal ultrasound. A magnetic resonance with IV contrast confirmed a pedunculated uterine-mass suggestive of sarcomatous degeneration, accompanied by a left internal iliac necrotic adenopathy, as well as scarce pelvic free fluid. A total abdomen computed tomography (CT) with IV contrast was also performed which reported a heterogeneous uterus with multiple associated masses, retroperitoneal and pelvic cavity necrotized adenopathies, scarce free abdominal fluid, as well as lamellar atelectasis in the left lung base. She was scheduled for pelvic tumor and implants resection, total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic lymphadenectomy, omentectomy and appendectomy via laparotomy. Trans-surgical peritoneal lavage was

performed with a positive result for poorly differentiated stromal neoplastic cells. Surgical specimens were sent to the pathological- anatomy department, which reported an undifferentiated endometrial sarcoma. Grossly, a fragmented pelvic tumor and implants were analyzed, with an intact uterus. The tumor size of the intrauterine component was 8×5.5×5.4 cm (Figure 1).

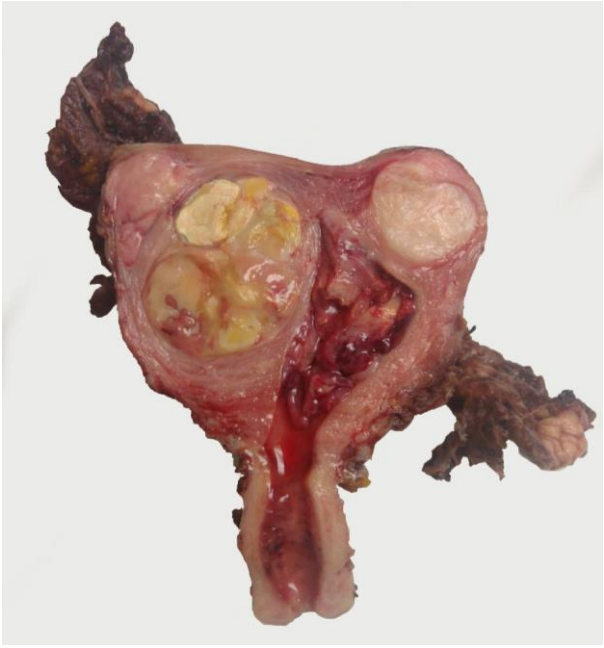

Figure 1. Gross anatomy: endometrial sarcoma.

Histologically, the neoplasia was composed of an undifferentiated uterine sarcoma (previously known as endometrial sarcoma) (Figure 2). It was unable to determine the pelvic tumor margins because the mass was fragmented. The uterine margins were negative, but with a positive vascular and lymphatic invasion. The peritoneal lavage fluid was positive for malignancy (18C-8084), and the left pelvic lymph nodes were negative for metastasis (1 of 1). The tumor extension was found on the surface of the right and left ovary, right and left uterine tubes (mucosa, mural and serosa), omentum (4 cm implant) and cecal appendix (mesoappendix and serosa). The diagnosis of undifferentiated endometrial sarcoma pT3b, pN0 (AJCC 8th edition 2018), IIIC (FIGO 2009) was confirmed.

The auxiliary studies reported a positive CD10, CiclinD1, estrogen receptors (10%, weak), progesterone receptors (5%, weak), and a negative H-Caldesmon, smooth muscle actin, desmin and HMB-45. Additional findings were reported as mild chronic cervicitis, endocervicitis with squamous metaplasia, residual atrophic endometrium and 2 conventional leiomyomas.

For a multidisciplinary approach, surgery, internal medicine, gynecology, radiology, intensive care, medical oncology, and gynecological oncology services were consulted, however, on the third postoperative day, the patient died (two months later from the established diagnosis) before chemotherapy could be even started due to a pulmonary embolism originated by a deep venous thrombosis, which was the result of tumor metastasis. Verbal and written consent were given by the patient on admission and before the surgery.

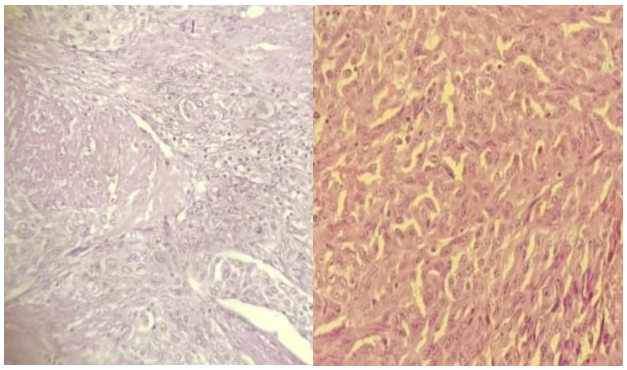

Figure 2: Histology: undifferentiated uterine sarcoma with nuclear pleomorphism, spindle cells and necrosis.

DISCUSSION

Uterine sarcomas represent 2 to 5% of all uterine cancers, which makes them rare. The term itself includes various types, such as leiomyosarcoma, carcinosarcoma, adenosarcoma, adenosarcoma with sarcomatous overgrowth, perivascular endothelial cell tumor, low grade endometrial stromal sarcoma, and high-grade undifferentiated sarcoma, which was our patient´s case.[6]

According to literature, this tumor usually presents in women over the age of 40 with a mean age of diagnosis at around 60 years old.[1] Our patient had 58 years when she was diagnosed, which correlates with a study published on the reports of practical oncology and radiotherapy from Fernandez et al in which the mean age was 62 years (range 51-69).[5] In another study from Denschlag et al, the age reported ranges between 62-67 years old.[7]

Diagnosis can often be difficult for physicians given that the typical presentation is with non-specific symptoms, such as post-menopausal abnormal uterine bleeding, unusual vaginal discharge, abdominal distention and discomfort, pelvic pain, and pollakiuria.[1] In Fernandez et al study, the main presenting symptom was abnormal uterine bleeding (85.7%). And the time from symptom presentation to diagnosis was highly variable, ranging from 5 to 122 days; although in cases of postmenopausal bleeding, the time to diagnosis was shorter (mean: 33 days).[4] Correlating with our patient, she had a history of 3 months of pelvic pain and a 5-day course of postmenopausal bleeding. Radiological diagnosis is difficult, usually it is confirmed post-hysterectomy.[4]

Undifferentiated uterine sarcomas are aggressive tumors that can present destructive myometrial invasion, abrupt mitotic activity, vascular involvement as well as necrosis, and are considered diagnosis of exclusion. Its incidence

rate of distant metastasis is high. Usually dissemination occurs in the peritoneum, and it has an early hematogenous spread, lymphatic spread is unusual. Since leiomyosarcomas and endometrial sarcomas need to be ruled out first, this can be accomplished with immunohistochemical stains and morphologic evaluation.[2]

Previously undifferentiated endometrial sarcomas were classified all together, but now, we can divide sarcomas according to their morphology into undifferentiated endometrial sarcomas with nuclear uniformity (UES-U) and undifferentiated endometrial sarcomas with nuclear pleomorphism (UES-P). Patients with UES-P have a poor prognosis compared to patients with UES-U, which demonstrates the important role of pathology in these tumors. Our patient had an undifferentiated endometrial sarcoma with nuclear pleomorphism as well as spindle cells and necrosis (Figure 2).[5]

FIGO created a specific staging system for uterine sarcomas, which allows grouping patients by their prognosis. The main prognosis factors are stage, age, residual tumor after surgery, positive peritoneal cytology, lymphatic nodes involvement, vascular and lymphatic invasion, myometrial invasion, mitotic index, degree of differentiation, tumor size and histopathological type.[4] Our patient was classified as a IIIC (pT3b, pN0 from AJCC 8th edition 2018) according to the FIGO 2009 classification. In Fernández et al study, all patients were also classified initially as FIGO III, which contributed to the poor clinical outcome.[4]

The treatment of choice for endometrial sarcomas consists of a multidisciplinary approach, including surgery with or without chemotherapy or radiation depending on the tumor stage, being the hysterectomy with bilateral oophorectomy and adjuvant radiation the most effective.[4]

In an early-stage disease, the treatment of choice consists of a total abdominal hysterectomy with bilateral salpingo-oophorectomy and peritoneal washings; lymphadenectomy isn't necessary because of its unusual lymphatic dissemination.[4]

The adjuvant treatment consists of external pelvic radiation, decreasing the local recurrence rate without a significant impact on survival.[4] The EORTC study reported a local control improvement for patients with carcinosarcomas that received radiotherapy, but no difference in disease-free survival.[3] Hysterectomy with bilateral oophorectomy is one of the most effective treatment modality, that's why our patient underwent a pelvic tumor and implants resection, total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic lymphadenectomy, omentectomy and appendectomy via laparotomy. As we mentioned before, literature reports no difference in disease-free survival with radiotherapy and it hasn't been able to demonstrate better local control with brachytherapy.[7]

Even though this cancer can have a 5-year relative survival rate of 68%, the stage at which it is diagnosed dictates the course of the patient's life.[1] In Fernandez et al study, the overall 2-year survival estimate was 82.5% and the 2-year local control rate was 90% with a progression free survival at 2 years of 58%.[4] However, our patient, on the third postoperative day, died (two months later from the established diagnosis) before chemotherapy could be even started due to a pulmonary embolism originated by a deep venous thrombosis, which was the result of tumor metastasis.

CONCLUSION

Uterine cancer englobes a wide range of unspecific symptoms that can affect the prompt diagnosis in patients, and drastically affect the stage at the time of diagnosis. Further research needs to be made regarding the therapeutic approaches in order to achieve maximum benefits for the patient's outcome as well as regarding the role of aggressive surgical management and adjuvant treatment in advanced stages of this type of sarcoma, in order to gain a better understanding of the benefits and risks of the procedure performed. Even though it is not a newly discovered tumor, it is still a very rare one, which is why we hope this case report contributes to the literature regarding uterine sarcomas.

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