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Case Report

Rare case of undifferentiated uterine sarcoma with neuroectodermal differentiation and osteoclast-like giant cells

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ABSTRACT

Objective: We describe the first case of a rare undifferentiated uterine sarcoma exhibiting both neuroectodermal differentiation and osteoclast-like giant cells, and elaborate its morphology.
Case report: A 54-year-old woman presented with suprapubic pain, frequent urination, and perimenopausal abnormal vaginal bleeding. Computed tomography revealed a large heterogeneous uterine mass and multiple lung nodules. She received a staging surgery. The tumor pathology examination revealed an undifferentiated uterine sarcoma (UUS) with neuroectodermal differentiation and osteoclast-like giant cells (OGCs). The patient was managed with palliative hospice care; however, she died within 1.5 months of diagnosis.
Conclusion: UUSs are rare high-grade tumors observed in elderly women. These women typically present with postmenopausal bleeding and extrauterine diseases and have a poor prognosis. Neuroectodermal differentiation in UUSs has a müllerian origin. The presence of OGCs may suggest a poor prognosis; however, further studies are necessary to determine the exact nature of such neoplasms.

Introduction

Endometrial stromal tumors (ESTs) are rare neoplasms composed of cells resembling proliferative endometrial stroma cells. They comprise less than 2% of uterine tumors and less than 10% of uterine mesenchymal tumors. The 2014 World Health Organization (WHO) classification categorizes ESTs as endometrial stromal nodules (ESNs), low-grade endometrial stromal sarcomas (LGESSs), high-grade endometrial stromal sarcomas (HGESSs), and undifferentiated uterine sarcomas (UUSs) [1]. UUSs are highly aggressive tumors, typically presenting in elderly patients with postmenopausal bleeding. USSs exhibit pleomorphic morphology, but they do not show specific immunohistochemical differentiation. Although USSs exhibit complex karyotypes and genomic gains or losses, they lack specific translocations [1]. Uterine tumors with neuroectodermal differentiation are rare; approximately 64 cases have been reported in the literature in English. In the reported studies, the uterine tumors have been referred to as Ewing sarcomas, peripheral primitive neuroectodermal tumors (PNETs), unclassified, and uterine tumors with neuroectodermal differentiation [2]. Seventeen cases constituted a series in which PNETs were reported as admixtures with unclassified sarcomas in two cases [3].

Osteoclast-like giant cells (OGCs) are multinucleated giant cells resembling osteoclasts [4]. They have been reported in malignancies of various origins, including carcinomas and sarcomas. In uterine tumors, the presence of OGCs is most commonly reported in leiomyosarcomas of the corpus. The first case describing OGCs in association with undifferentiated sarcomas was reported in 2011 [5]. The neoplasm was reported to be high-grade, containing cells strongly positive for CD 10, and was most likely a UUS according to the new WHO classification. To our knowledge, this is the first case of a UUS exhibiting both neuroectodermal differentiation and OGCs.

Case report

A 54-year-old gravida 3/para 2/abortus 1 woman presented at our hospital because of suprapubic pain and frequent urination for 1 week. She had had menorrhagia (prolonged bleeding for up to 20 days per cycle) for 4 years, as well as had abdominal distension, a poor appetite, and recent weight loss. A physical examination

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revealed a heterogeneous lower abdominal mass, and a pelvic examination revealed a parous cervix with vaginal dark brown discharge. The right side indurable parametrium and partly movable uterine cervix was palpated. An ultrasonography examination of the pelvis revealed a uterine mass measuring 16 cm, and computed tomography revealed an enlarged uterus with a heterogeneous mass (Fig. 1), a right ovarian cystic lesion, and multiple nodular lesions in both the lungs. Her laboratory data clearly indicated microcytic anemia (hemoglobin level 5.4 g/dL).

She underwent staging surgery, including a total abdominal hysterectomy, bilateral salpingo-oophorectomy, bilateral pelvic lymph node dissection, and omentectomy, to relieve her abdominal distension and examine the pathology of the tumor. On postoperative day (POD) 3, the patient developed right-sided bloody pleural effusion. Although tissue proof was absent, the effusion was highly suspicious of metastatic malignant pleural effusion. The patient was managed by palliative hospice care according to her request, and died on POD 42.

Gross findings of the tumor

The uterus was enlarged, measuring 20 × 17 × 15 cm^3 and weighing 1850 g. Inspection revealed that the serosa over the posterior aspect of the lower uterine segment was perforated. Upon opening, a polypoid mass protruded into the endometrial cavity and protruded through the external os (Fig. 2). Sectioning revealed that the tumor grossly invaded the entire thickness of the myometrium and the width of the endocervix. The cut surface of the tumor was pink, fleshy, and soft with areas of hemorrhage and necrosis. Two small intramural leiomyomas were observed.

The right ovary was received as an opened cystic tissue coated with thick chocolate-colored semiliquid substances.

Histological findings of the tumor

Histological sections of the tumor revealed that most areas of the tumor were composed of pleomorphic or spindle-shaped cells, with hyperchromatic nuclei, that infiltrated into the myometrium. Numerous multinucleated OGCs were scattered among the neoplastic cells (Fig. 3A). The mitotic index was between 50 and 100/10HPF, including atypical mitoses, and areas of coagulative necrosis and lymphovascular invasion were abundant. The morphology gradually transformed into uniform small blue cells, with oval- and spindle-shaped nuclei arranged in rosettes (Fig. 3B) at the lower uterine segment and endocervix. A transformation zone with biphasic features was observed (Fig. 3C).

Immunohistochemically, both pleomorphic cells and small blue cells were strongly and diffusely positive, respectively, for CD 10 (Fig. 4A and B) and negative for Wilms tumor protein WT-1, α-smooth muscle actin (α-SMA), desmin, estrogen receptors, progesterone receptors, cyclin D1, and cytokeratin. The small blue cells also expressed CD 56 (Fig. 4C), synaptophysin, and CD 99 (cytoplasmic) (Fig. 4D); additionally, the OGCs were positive for CD 68 (Fig. 4E). We diagnosed the tumor as a UUS with neuroectodermal differentiation and OGCs.

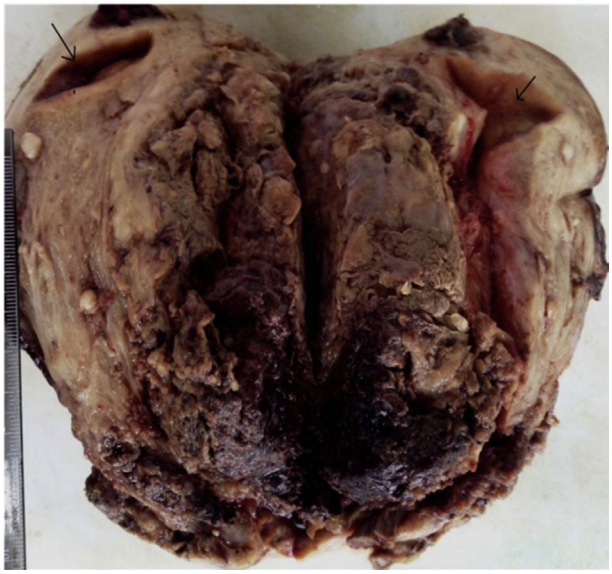

Fig. 2. Gross appearance of the tumor (arrows indicate the endometrial cavity).

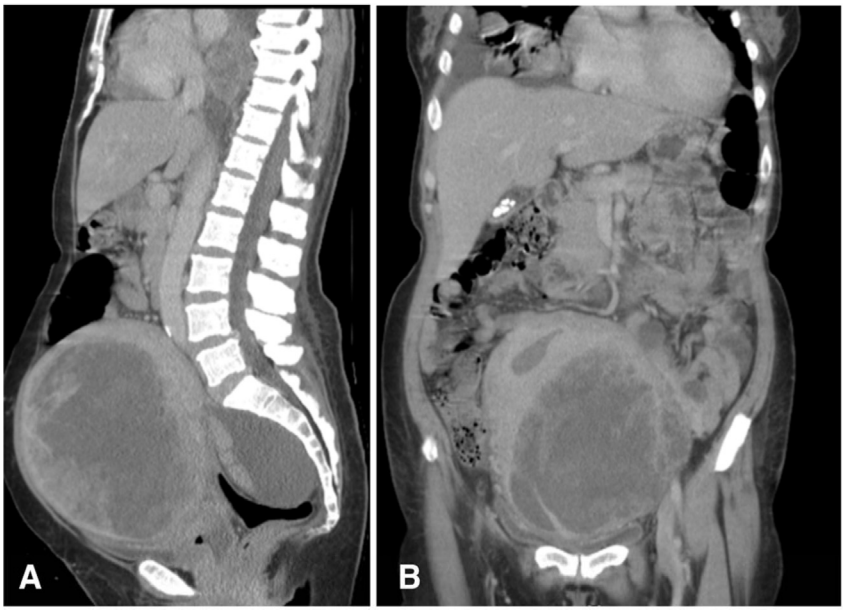

Fig. 1. Abdominal computed tomography showing a heterogeneous uterine mass. (A) Sagittal plane. (B) Coronal plane.