case report

Vulvar extrauterine endometrial stromal sarcoma: A case report and literature review

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Endometrial stromal sarcoma (ESS) is an extremely rare neoplasm accounting for only 0.2% of all uterine malignancies and for 15–26% of primary uterine sarcomas. The annual incidence of ESS is 1–2 per million women. Herein, to the best of our knowledge, we present the first reported case of ESS of the vulva in a 50-year-old female presenting with per vaginal spotting over a period of three months. Her past surgical history included a subtotal hysterectomy and left salpingo-oophorectomy for uterine fibroids ten years previously. On examination, a $3.5 \times 3 \times 2$ cm cystic mass was found in the right labia majora. The mass was excised and the diagnosis of endometrial stromal sarcoma was made. Subsequent metastatic workup was negative and the patient was started on megestrol acetate. She has remained disease free with no signs or symptoms of recurrent or advanced disease for 28 months.

KEYWORDS: Endometrial stromal sarcoma; Vulva; Uterine sarcoma

INTRODUCTION

Endometrial stromal sarcoma (ESS) is an extremely rare neoplasm accounting for only 0.2% of all uterine malignancies and for 15–26% of primary uterine sarcomas.[1] The annual incidence of ESS is 1–2 per million women. Compared to other uterine malignancies, ESS affects younger women with a mean age of 42–58 years.[2] ESS resembles stromal cells in the proliferative stage of the normal endometrium and are often low-grade, indolent, but metastatic exhibiting myometrial and/or vascular invasion.[3] The incidence of ESS in extrauterine locations is exceedingly rare especially in the absence of metastasis or extension of a primary neoplasm. Principal extrauterine sites of ESS include the ovary, bowel wall, abdomen, peritoneum, pelvis, and vagina.[4–6]

Herein, to the best of our knowledge, we present the first reported case of ESS of the vulva in a 50-year-old female presenting with per vaginal spotting of three months' duration. A literature review of ESS is also presented.

CASE REPORT

A 50-year-old female, para 8+0 was referred to our service with per vaginal spotting. Her past medical history was unremarkable; her previous surgical history included a subtotal hysterectomy and left salpingo-oophorectomy for uterine fibroids ten years previously. On clinical examination, a soft yellowish lesion with hemorrhagic foci, measuring $3.5 \times 3 \times 2$ cm, was found and a Bartholin gland cyst was suspected. On February 2012, an excisional biopsy was performed and the rendered histopathological diagnosis was low-grade endometrial stromal sarcoma. Microscopy showed a diffuse cellular infiltrate composed of monotonous bland looking oval to spindle cells (simulating endometrial stromal cells) surrounding arterioles resembling endometrial spiral

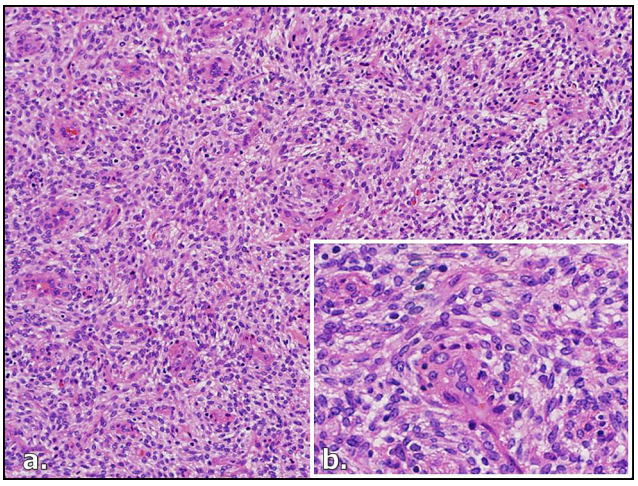

Figure 1. Endometrial stromal sarcoma. (a) A cellular neoplasm composed of bland oval cells surrounding arterioles resembling endometrial spiral arterioles (hematoxylin and eosin stain, ×200 magnification). (b) A higher magnification microphotograph showing the spindle cells surrounding a blood vessel (hematoxylin and eosin stain, ×400 magnification).

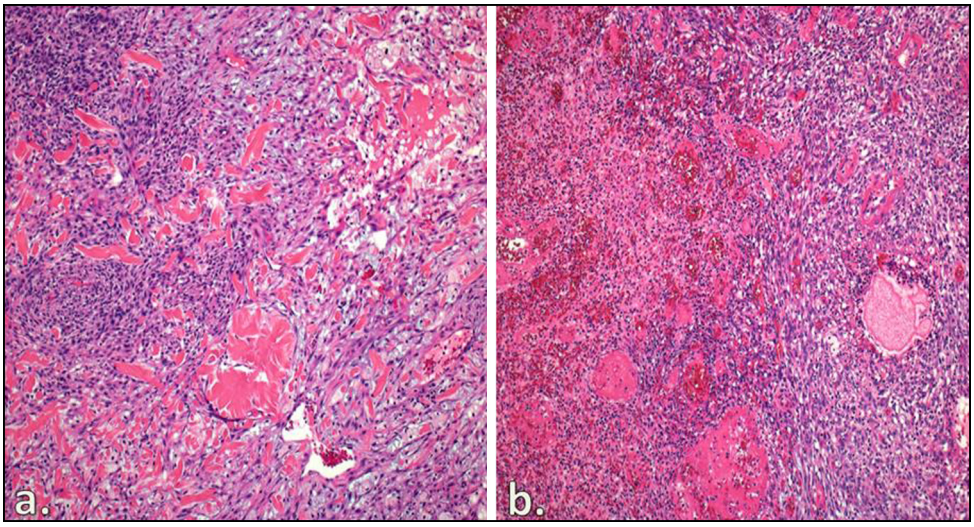

Figure 2. Endometrial stromal sarcoma. (a) Plaques of hyalinized fibrosis (hematoxylin and eosin stain, ×200 magnification). (b) Areas of tumor necrosis on the left of the image in contrast to the viable tumor on the right (hematoxylin and eosin stain, ×100 magnification).

arterioles (Figure 1). Few mitoses and large foci of tumor necrosis and hemorrhage were seen, as well as plaques of hyaline fibrosis (Figure 2). The lesion was infiltrative with poorly defined margins. The surgical resection margins were negative for tumor and there was no evidence of associated endometriosis. Moreover, immunohistochemical staining was strongly positive for CD10, vimentin, estrogen receptor and progesterone receptor (ER and PR) (Figures 3 and 4). Cytokeratin cocktail (CKAE1/AE3) was moderately positive, whereas h-caldesmon, desmin, CD34, SMA and S-100 were all negative (Figure 5). The patient was started on megestrol acetate. In May 2012, surveillance computed tomography of the chest, abdomen, and pelvis displayed an unremarkable cervix and no signs of metastasis or recurrence. In January 2013, the patient had a positron emission tomography (PET) scan which was unremarkable; the patient was therefore kept on megestrol. In July 2013, magnetic resonance imaging (MRI) revealed an ill-defined lesion in the right side of her perineum and lower vagina, encasing the urethra, with no lymph node involvement (Figure 6). The patient was referred to urology, and a cystoscopy was performed with unremarkable results. An excisional biopsy was performed showing a benign polypoid piece of endocervical mucosa with chronic inflammation. Subsequent cervical smears throughout the patient's