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Case Report
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Low-Grade Endometrial Stromal Sarcoma of Endocervix: A Case Report

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Abstract

Background: Endometrial Stromal Sarcoma (ESS) are rare malignant tumors that make up approximately 10% of all uterine sarcomas. The primary can occur in extra-uterine sites like ovary, fallopian tube, cervix, vulva, vagina, omentum and retroperitoneum. Cervical sarcomas are still rare comprising 0.5% of all primary cervical malignancies. Primary low grade ESS occurring in endocervix is reported very rarely and it mimics polyp which poses a diagnostic dilemma.

Case Report: We report a case of 40 years perimenopausal lady with HCV positive who presented in gynec-oncol OPD with complaints of irregular bleeding per vaginum and something coming out P/V since 4 months. T2W MRI shows large lobulated pedunculated heterogenous hyperintense mass arising from anterior lip of cervix. Histological examination revealed epithelioid cell tumor lined by endocervical lining and tumor infiltrate to ectocervix and myometrium. The diagnostic challenge was to differentiate between epithelioid variant of leiomyoma and ESS for which immunohistochemistry was carried out which showed moderately positivity for Cluster of Differentiation 10 (CD10), focal positivity for ER (Estrogen Receptor) & PR (Progesterone Receptor) and negative for SMA, DESMIN, CALDESMON, CD34, CD117. Ki-67-was <2%.

Conclusion: The case report emphasizes the need to include ESS in the differential diagnosis of cervical polyp.

Keywords: Leiomyoma; Cervical polyp; Epithelioid cell tumor; Stromal sarcoma

Introduction

Cervical sarcomas comprise of 0.5% of all primary cervical malignancies [1,2]. The median age of women with low-grade ESS is between 45 and 57 years [3]. Low-Grade Endometrial Stromal Sarcoma (LG-ESS) comprises 50% to 60% of ESS and may arise in extra uterine sites i.e. ovary, fallopian tube, cervix, vagina, vulva, omentum, sigmoid colon, round ligament and retroperitoneum [4]. Primary low grade ESS occurring in endocervix is very rare. After extensive research only one case report has been found. This case highlights occurrence of extra uterine ESS in the endocervix, presenting as a cervical polyp.

Case Presentation

A 40 years old perimenopausal P2L2 patient with HCV positive status presented with bleeding per vaginum since 4 months duration with a polypoidal mass coming out of vagina. Her menstrual cycle was regular and no history of taking any hormonal pills. Local examination revealed 10 cm × 8 cm polypoidal growth arising from anterior lip of cervix lying outside vulva associated with cervical elongation. Cervical polyp had irregular surface with necrotic areas and had bleeding on touch. Per vaginum radiology shows growth was arising from anterior lip of cervix, uterus of normal size, B/L fornices and parametrium appeared free and rectal mucosa smooth. T2W MRI shows large lobulated pedunculated heterogenous hyperintense mass arising from anterior lip of cervix. A total abdominal hysterectomy with bilateral-salpingo-oopherectomy was carried out and the specimen was sent for histopathological examination.

Gross appearance

Gross examination revealed specimen of TAH+BSO in which uterus measures 10 cm × 4 cm × 4 cm with endometrial thickness was 2 mm and myometrial thickness was 25 mm and cervix measures 5 cm. On cut section of uterus, cervix, bilateral fallopian tube and ovary was unremarkable. An ulcero-proliferative globular polyp sent separately measures 10 cm × 8 cm × 4 cm (Figure 1). External surface was bosselated with ulceration and sloughing sat places. On cut section, fleshy tan

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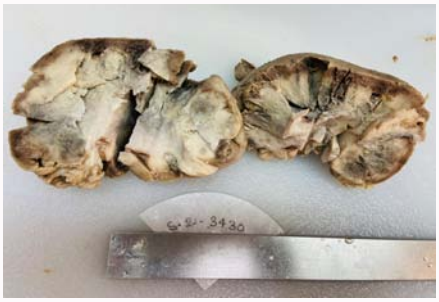

Figure 1: Cervical mass measuring 10 cm × 8 cm × 4 cm.

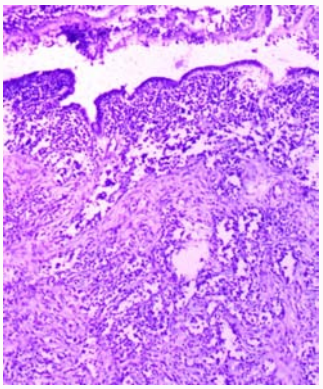

Figure 2: H&E 10x, diffusely infiltrating tumour with endocervical lining.

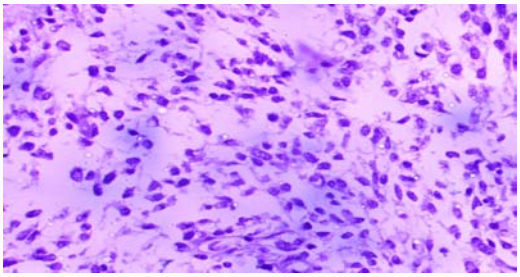

Figure 3: H&E 40x, sheets of mildly pleomorphic epitheloid cells having moderate amount of cytoplasm with round oval nuclei.

yellow homogenous area with few areas of hemorrhage.

Microscopic examination

Uterus showed normal endometrial glands in proliferative phase. Section from polyp shows sheets of monotonous population round to oval cells covered by cuboidal to low columnar lining epithelium (endocervical lining) (Figure 2). High power of tumor proper shows round to oval nucleus with mild anisonucleosis, vesicular nuclear chromatin and mostly inconspicuous nucleoli with scant to moderate eosinophilic cytoplasm (Figure 3). Atypical mitosis was negligible (<2 mitosis/10 hpf). Section from ectocervix shows stratified squamous epithelium with underlying tissue shows infiltration by tumor cells (Figure 4). Multiple section from myometrium shows sheets of tumor cells separated by muscle fibers (Figure 5). On the basis of microscopy differential diagnosis of Leiomyoma (Epithelioid variant) and Endometrial stromal tumour was considered. Hence, a panel of immunohistochemical markers were put. Moderately positive staining was noted for CD10, ER and PR shows focal positivity. SMA, DESMIN, CALDESMON, CD34 and CD117 was negative. Ki67 was <2% (Figure 6). Hence, a final diagnosis of Low grade endometrial

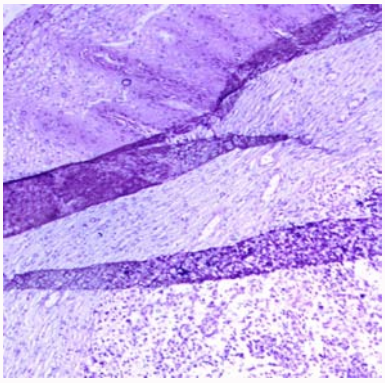

Figure 4: H&E 10x, diffusely infiltrating tumour with ectocervical lining.

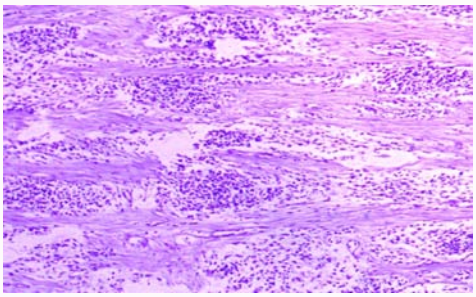

Figure 5: H&E 10x, myometrium shows sheets of tumor cells separated by muscle fibers.

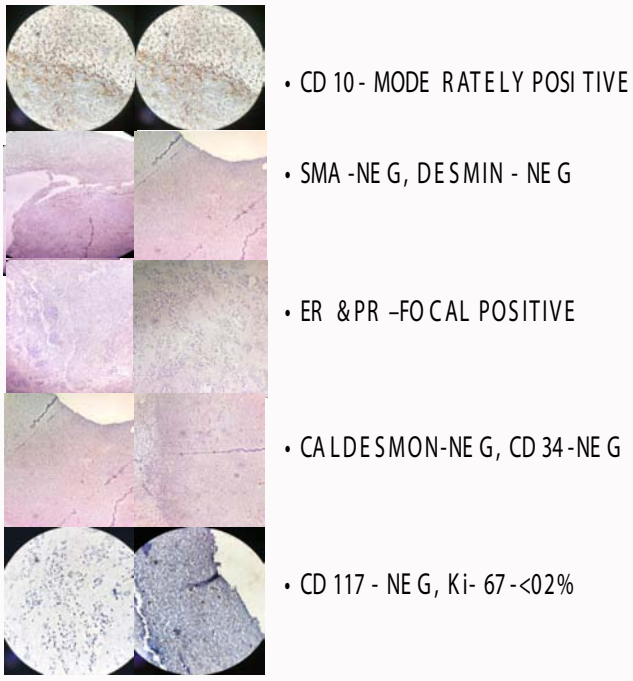

Figure 6: 10x, IHC.

stromal sarcoma of endocervix locally infiltrating ectocervix and myometrium was given.

Discussion

There was very few reported cases of LGESS in the cervix. The origin and biology of stromal sarcomas are not well understood. There is a relation between chromosomal aberrations and endometrial

sarcomas. Chromosomal deletion on 7p was the most common finding (55.6%) in ESS [5]. The fusion of the two zinc-finger genes JAZF1 and SUZ1 at the 7p15 and 17q21 breakpoints, respectively was described in EST by Koontz in 2001 [6]. The prevalence of the associated translocation, t(7;17) (p15;q21) in low grade endometrial stromal sarcoma by RT-PCR [8,9] and FISH [7,10] ranges from 23 to 80%. The pathogenesis of ESS is unknown, but exposure to tamoxifene, unopposed estrogens and conditions such as polycystic disease of ovary are implicated [11]. One fourth of patients are asymptomatic. Most patients present with abnormal vaginal bleeding, polypoidal vaginal mass and dysmenorrhea. The other symptoms could be urinary urgency, retention of urine or constipation. LG-ESS cells resemble proliferative phase endometrium. Cells have abundant clear to eosinophilic cytoplasm with poorly defined cell borders, round to oval nucleus, finely granular chromatin and small inconspicuous nucleoli. About one-third of patients with ESS show recurrent local disease [1,12]. The standard surgical treatment is TAH with bilateral-salphingo-oophrectomy. As the tumor is hormonally responsive, hormone replacement therapy containing estrogen and tamoxifene is contraindicated postoperatively [13].

Conclusion

We report a case of ESS which occurred at an unusual location and suggest that ESS should be included in the differential diagnosis of cervical sarcoma.

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