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Case Report

Dysuria Caused by Endometrial Stromal Sarcoma Arising from Uterine Cervical Endometriosis: A Case Report and Literature Review

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Abstract

Endometrial Stromal Sarcoma (ESS) arising from the uterine cervix is rare and always presents with symptoms of vaginal bleeding or spotting. We report the first case of uterine cervical ESS presenting with dysuria and review related literature.

Keywords: Case study; Dysuria; Endometrial stromal sarcoma; Uterine cervical endometriosis

Abbreviations: ESS: Endometrial Stromal Sarcoma; G2P2: Gravidity 2 Parity 2

Introduction

Endometriosis is a common benign gynecologic disease with endometrial tissue growing outside the uterine body. It affects approximately 10% of reproductive- age women, with rare malignant transformation occurring in only 0.1-0.7% of the cases [1,2]. Most malignant transformations of endometriosis are glandular origin, presenting as endometrioid adenocarcinoma and clear c. types, with stromal origin is being extremely rare. Furthermore, most uterine cervical sarcomas present with vaginal spotting. We report a rare case of ESS arising in the uterine cervical endometriosis presenting with dysuria and review related literature.

Case Report

A 45-year-old woman, G2P2, visited the urology outpatient department because of sudden voiding difficulty on January 2, 2020. She had a urine output of 1,400 ml after Foley catheterization, which was discontinued on January 6. However, she still complained of dysuria and urine retention that night. Trans-vaginal sonography showed an enlarged uterine cervical mass measuring 8.6 cm that was causing urethral compression and obstruction (Figure 1). The patient was referred to the gynecology department. Because uterine cervical myoma with compressed urethral obstruction was suspected, the patient underwent complicated myomectomy of the uterine cervix. A 9-cm -sized cervical stromal mass located on the posterior wall of the uterine cervix protruding to the posterior upper vaginal wall and causing upward urethral compression was observed. The mass appeared fleshy and friable with partial necrosis-like areas. A histologically examined intraoperative frozen section revealed “atypical cells,” and malignancy could not be excluded.

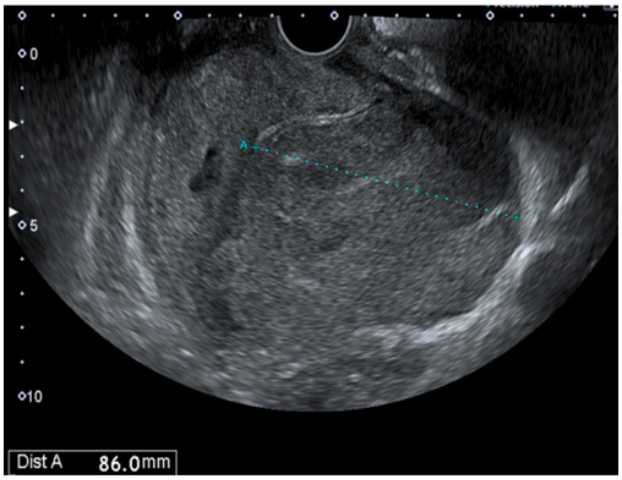

Figure 1: Trans vaginal sonography showing the enlarged uterine cervical mass (8.6 cm).

Total hysterectomy with tumor debulking and right ovarian cystectomy were performed. Pathologically, the submitted specimens included the totally resected uterus, (measuring 13.1×8.5×5.4 cm; weighing 282.5 g), and a bag of fragmented cervical stromal mass tissues (measuring 6.2×4.1×2.5 cm [9.5×5.5×5.0 cm-, when totally piled up]; weighing 202.0g) (Figure 2). Microscopically, the cervical stromal mass comprised proliferative tumor cells sheets, with ovoid nuclei -focal epithelioid cells concentrated around thickened small vessels, foci of patchy infarction, few focal endometrial glands, and focal increasing mitotic activity, up to 6/10 HPFs displaying an infiltrative pattern. Immunochemical staining results were positive for estrogen and progesterone receptors, desmin (partial), and CD10 (partial) but negative for actin-M851, CD117, cyclin D1, and inhibin (Figures 3 and 4).

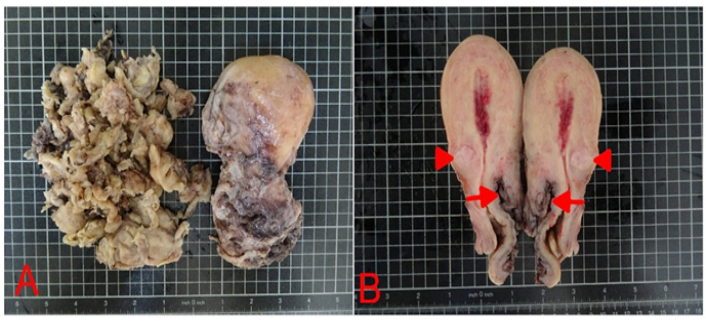

Figure 2: Grossly, the totally hysterectomized uterus shows an elongated gray- tan uterine cervix (right) and excisional tumor fragments from the uterine cervical stroma (left) **(A)**. The uterus shows an elongated uterine cervix with tumor space (S/P tumor excision) within the uterine stroma (arrow) and a leiomyoma at the low uterine segment (arrowhead) **(B)**.

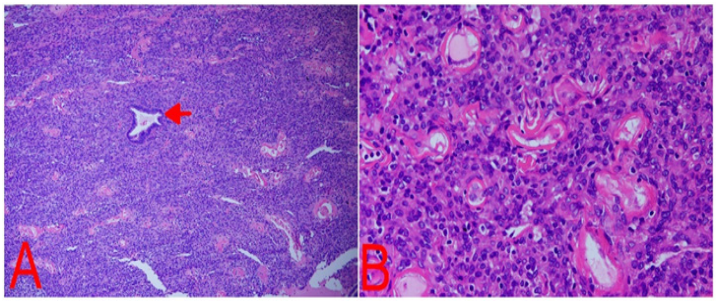

Figure 3: Microscopically, the tumor demonstrates a bland endometrial gland (arrow) within the proliferative sheets of uniform stromal tumor cells (hematoxylin-eosin stain, 40×) **(A)**. Tumor cells show ovoid nuclei surrounding the dilated hyalinized small vessels (hematoxylin-eosin stain, 200×) **(B)**.

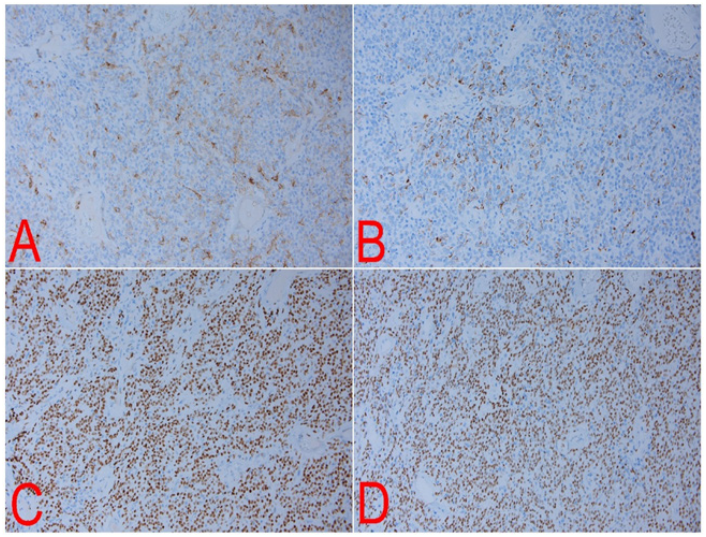

Figure 4: Immunohistochemical stain showing tumor cells are partially stained by CD10 antibody (**A**, 100×) and desmin antibody (**B**, 100×), and diffusely stained by estrogen receptor antibody (**C**, 100×) and progesterone receptor antibody (**D**, 100×).

As gross observation and microscopic images showed no connection between the mass and uterine cavity and cervical canal, a low-grade endometrial stromal sarcoma arising from the uterine cervical stromal endometriosis was finally diagnosed. The right ovarian cyst was an endometriotic cyst with endometrial stromal hyperplasia. Tumor markers, including CA-125, 112.18 U/ml (normal: 0-35 U/mL), and CEA, 1.67 ng/mL (normal: 0-7 ng/mL), were observed. Owing to inadequate surgical margins, adjuvant chemotherapy with docetaxel and gemcitabine was administered. Three-months later, follow-up pelvic computer tomography revealed lobulated tumor nodules-(measuring 7.8×6.0×3.5 cm), with invasion into the anterior rectal wall. As recurrent ESS was suspected, second-look tumor debulking was performed. Recurrent

ESS was histopathologically confirmed, and the patient received post-debulking combined with chemoradiotherapy. There was no tumor recurrence after at the six-months follow-up.

Discussion

Mesenchymal tumors are rare uterine neoplasms with stromal differentiation and are classified as ESS. ESS accounts for 0.2% of all malignant uterine tumors and represents 10-15% of all uterine sarcomas. There are four types of endometrial stromal tumors: endometrial stromal nodule, low-grade ESS, high-grade ESS, and undifferentiated uterine sarcoma. Of these, 50-60% are low-grade ESS that mainly present in younger patients (aged 45-57 years) and ESS symptoms include abnormal vaginal bleeding, abdominal pain, and progressive menorrhagia. Slow-growing, ESS is malignant with an approximately 50% recurrence rate. Histologically, ESS can present with variable differentiation of proliferative-phase stromal cells with invasion into the myometrium. The origin of extra-uterine ESS is unknown, but is postulated to arise from endometriosis or the secondary Mullerian system. A part of the extra-uterine ESS combined with endometriosis was considered as arising from the endometriosis, so the secondary Mullerian system could be considered as the origin site.

It is plausible that the secondary Mullerian system is distributed from the low peritoneal cavity to the pelvic peritoneum, possibly causing metaplastic changes to the derivatives of the Mullerian system, including the endometrial glands and stroma. Extra uterine ESS arising from the uterine cervix is very rare, with only eight previously reported cases, including our case (Table 1) [3-10]. The tumor usually occurs in peri-menopausal women (aged 44-54 years), but two cases in patients aged 20 years have been reported. The symptoms of all cervical ESS patients were vaginal bleeding or spotting, with the exception of our case, who complained of dysuria. Owing to its rarity, the preoperative diagnosis of cervical ESS is challenging. Five of eight patients were preoperatively diagnosed with leiomyoma. Only two cases were accurately diagnosed preoperatively as endometrial stromal neoplasm and ESS, based on tissue biopsy. This indicates that issue histology is mandatory for accurate preoperative diagnosis. Tumors measured 2.8 -11.9 cm and had low grade and occasional polypoid features. Owing to the rarity and short follow-up in previous cases, the prognosis of cervical ESS remains unclear.

Case (year)	Age	symptom	Pre-OP Dx	Tumor size (cm)	OP method/ treatment	Grade (endometriosis)	recurrence	Death of disease
1 (2000)	54	Vaginal spotting	Uterine stromal neoplasm (by biopsy)	2.8x2	RH + BSO + BPLND	Low (-)	NA	NA
2 (2007)	44	Vaginal bleeding/ secretion	Degenerated leiomyoma	7x4x3	Excision + D&C, then, RH + BSO + BPLND	Low (+)	No (F/U 1 yr)	No
3 (2012)	22	Vaginal bleeding	NA	NA	RH + PLND, then C/T	Low (+)	No (F/U 7 yrs)	No
4 (2014)	20	Vaginal bleeding	Infected leiomyomatous polyp	4x2.2x2 (polypoid)	polypectomy	Low (-)	NA	NA
5 (2014)	51	Vaginal bleeding	ESS (by biopsy)	11.9x8.1	CCRT	Low (NA)	No	NA
6 (2015)	48	Vaginal bleeding	Degenerated fibroid mass	11.0x13.0 (polypoid)	ATH + BSO	Low (-)	NA	NA
7 (2017)	43	Vaginal bleeding	Cervical fibroid	4x3x2 (polypoid)	ATH + polypectomy	Low (-)	NA	NA
8 (2020)	45	Dysuria	Degenerated myoma	9.5x5.5x5.0	ATH + tumor excision	low (+)	Yes (6 month later)	NA

ATH: abdominal total hysterectomy; BSO: bilateral salpingooophorectomy; BPLND: bilateral pelvic lymph node dissection; C/T: chemotherapy; CCRT: concomitant chemo-radiotherapy; D&C: dilation and curettage; F/U: follow-up; NA: not available; OP: operation; Pre-OP Dx: pre-operation diagnosis; RH: radical hysterectomy.

Table 1: Endometrial stromal tumor arising from uterine cervix.

However, this is the first reported case of rapid tumor recurrence, 3 months after the initial operation. This is likely owing to large tumor size and inadequate tumor excision and further confirms that a safe surgical margin is necessary to prevent tumor recurrence. Extra-uterine ESS presenting with dysuria has not been reported previously. The differential diagnosis of a female with non-inflammatory dysuria should include anatomic, drug-related, endocrine, lithiasis, traumatic, iatrogenic, neoplastic, and idiopathic causes. In our case, the urethra was markedly compressed posteriorly by a large cervical ESS, causing total urethral obstruction. Thus, a cervical ESS may be a rare cause of female non-inflammatory dysuria and should be considered in the differential diagnosis of female dysuria. Although chemotherapy, radiotherapy, or hormone therapy were previously used, the role of postoperative adjuvant treatment in cervical ESS remains unknown, with no standard guidelines currently available and complete tumor excision may be the first choice to reduce recurrence rates [3]. Moreover, recurrent translocation is a frequent phenomenon observed in both low-grade and high-grade ESSs. The tumor-specific translocation fusion protein might be a target site for therapy besides immunotherapeutic options [10].

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