

## Case Report

# Low-Grade Endometrial Stromal Sarcoma in a Postmenopausal Woman with Third-Degree Uterovaginal Prolapse: A Rare Case with Review of the Literature

Meenakshi Gothwal, Garima Yadav, Meenakshi Rao<sup>1</sup>, Pratibha Singh, Aasma Nalwa<sup>1</sup>

Departments of Obstetrics and Gynaecology and <sup>1</sup>Pathology, All India Institute of Medical Sciences, Jodhpur, Rajasthan, India

ABSTRACT

Endometrial stromal sarcomas (ESSs) are rare malignant tumors of the uterus that arise from the endometrial stroma, the annual incidence of ESS being 1–2/million women. Patients present with nonspecific complaints of abnormal uterine bleeding, pelvic pain, and pressure symptoms. Postmenopausal women rarely present with a low-grade ESS, as it is more common in younger women. ESS is usually confused with leiomyomas radiologically. Histopathological examination and immunohistochemistry are essential for the diagnosis. We, herein, report a case of a postmenopausal female who presented with something coming out per vagina. The clinical impression was that of a third-degree uterovaginal prolapse that on histopathological examination was diagnosed as a low-grade ESS.

**KEYWORDS:** *Endometrial stromal sarcoma, low grade, postmenopausal*

## INTRODUCTION

Uterine/endometrial stromal sarcoma (ESS) is a rare tumor of uterus. According to the literature, incidence is approximately 0.2% of all uterine malignancies and approximately 10% of all uterine sarcomas.<sup>[1]</sup> It can arise from smooth muscle, connective tissue, or endometrial stroma.<sup>[2,3]</sup> ESS usually occurs in the younger age group with mean age ranging between 45 and 55 years. It is very rare to have low-grade ESS in postmenopausal women. Patients usually present with heavy menstrual bleeding, pain abdomen, and enlarged uterus.<sup>[4]</sup> ESSs due to their low incidence are difficult to diagnose preoperatively and detected only after histopathological examination of the resected hysterectomy specimen. ESS is commonly associated with various chromosomal aberrations, of which deletion of chromosome 7p is most common.<sup>[1]</sup> It can occur in patients with polycystic ovarian syndrome and those who are exposed to drugs such as tamoxifen or unopposed estrogen.<sup>[2]</sup> We, herein, report a case of a postmenopausal female who presented with something coming out per vagina. The clinical impression was that of third-degree uterovaginal prolapse which on histopathological examination was diagnosed as a low-grade ESS.

## CASE REPORT

A 52-year-postmenopausal female reported with a complaint of something coming out per vagina since 16 years. She was para 4; with three normal vaginal deliveries and cesarean section for the last childbirth. She achieved menopause 1 year back, and there was no history of postmenopausal bleeding. She had menarche at the age of 14 years with regular cycles and normal flow. On clinical examination, abdomen was soft, nontender, midline scar of previous cesarean section was seen, and no mass was felt. Local examination showed third-degree uterovaginal prolapse with third-degree cystocele and rectocele. Per vaginal examination revealed uterus approximately 8–10 weeks' size, anteverted, and bilateral adnexa free.

Her routine investigations along with papanicolaou smear were within normal limits. Transvaginal ultrasound scan suggested a diffusely bulky uterus with heterogeneous

**Address for correspondence:** Dr. Meenakshi Gothwal, Department of Obstetrics and Gynaecology, All India Institute of Medical Sciences, Jodhpur - 342 005, Rajasthan, India. E-mail: meenakshigothwal@gmail.com

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myometrium with subendometrial echogenic and acoustic shadows suggestive of adenomyosis. Endometrial thickness was 2.4 mm and bilateral adnexa was normal. The patient underwent vaginal hysterectomy with pelvic floor repairs. The specimen sent for histopathological examination.

The resected hysterectomy specimen measured 14 cm × 12 cm × 6 cm. Sectioning showed a growth involving the myometrium, which was pushing into the endometrial cavity. The growth measured 9 cm × 5 cm × 4 cm and had a fleshy, gray-white trabeculated cut surface, with areas of necrosis and hemorrhage. The growth was 0.4 cm away from the serosal surface [Figure 1].

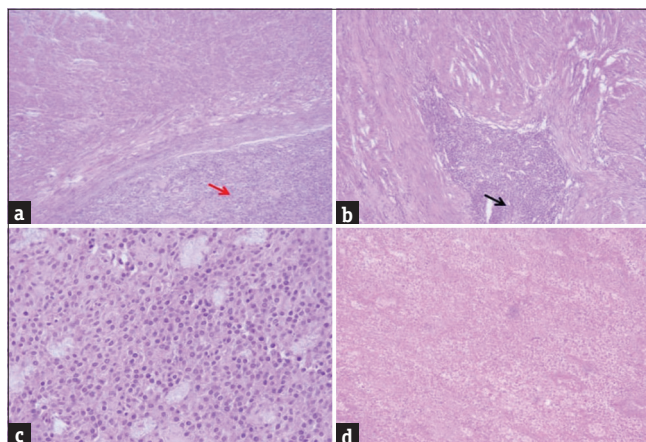
Microscopically, sections from the growth showed an invasive tumor involving the endometrium and infiltrating into the myometrium with invasive fronts. The tumor was composed of sheets of monomorphic to mildly pleomorphic cells having moderate eosinophilic cytoplasm, round-to-oval nuclei, fine chromatin, and inconspicuous nucleoli. Dispersed focal arterioles and crystals noted within the tumor. Vascular invasion was identified and an occasional mitotic figure was seen (<1 mitotic figures/10 high-power field [HPF]). Areas of necrosis, hemorrhage, and a few hemosiderophages were noted within the tumor. Hyalinization was also noted focally. Sections from the bilateral left fallopian tube stumps, cervix, and bilateral adnexa were unremarkable. Morphologically, the tumor cells resembled endometrial stromal cells, and the tumor had invasive fronts, with areas of necrosis; therefore, a morphological diagnosis of ESS rendered [Figure 2].

Immunohistochemistry (IHC) for CD10 was put up for confirmation, and IHC for smooth muscle actin (SMA), cytokeratin (CK), and epithelial membrane antigen (EMA) was also put up to rule out the close differential diagnoses, leiomyosarcoma, and carcinoma. Based on positivity of CD10 and negativity of SMA, CK, and EMA, a final diagnosis of low-grade ESS was given [Figure 3].

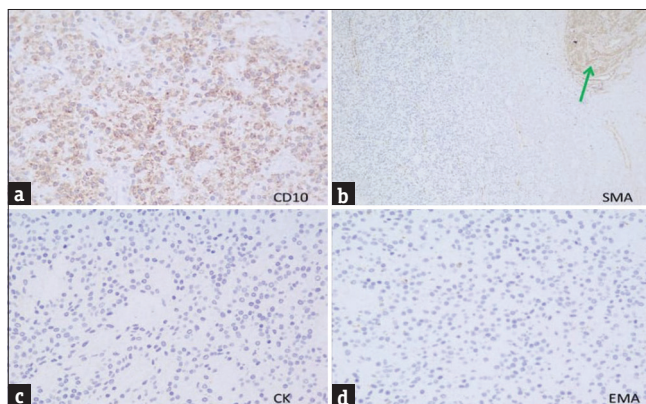
Based on the above histopathological findings, a decision for laparotomy considered following which bilateral salpingo-oophorectomy done on the day 8<sup>th</sup> of primary surgery. Intraoperatively, bilateral ovaries and tubes were normal with no palpable lymph nodes. After joint discussion with tumor board of our institute, it was decided that patient did not require further postoperative adjuvant therapy and was advised for follow-up visits. The patient discharged on the 7<sup>th</sup> postoperative day and called for follow-up every 3 monthly. During follow-up, proper clinical examination and transvaginal ultrasound were done. The patient is doing well 6-months post-follow-up.



**Figure 1:** Gross image of specimen



**Figure 2:** (a) Tumor (lower half, red arrow) with adjacent myometrium (upper half), H and E, ×10. (b) Invasive tumor front (black arrow) into the myometrium, H and E, ×10. (c) Tumor composed of uniform round-to-oval cells, resembling endometrial stromal cells, H and E, ×40. (d) Tumor necrosis, H and E, ×10



**Figure 3:** (a) CD10 positivity in tumor cells, ×40. (b) Smooth muscle actin negativity in tumor cells, adjacent myometrium in upper right half shows positivity in the myometrial smooth muscle cells (green arrow), ×10. (c) Cytokeratin negativity in tumor cells, ×40. (d) Epithelial membrane antigen negativity in tumor cells, ×40

## DISCUSSION

Endometrial stromal tumors are rare malignant tumors of the uterus. They are also known as interstitial

endometrioma, endolymphatic stromal myosis, or low-grade ESS. ESS has an indolent course and even many years after diagnosis recurrent disease can occur.<sup>[1,5]</sup> According to mitosis and degree of invasion, it is classified into three groups, that is, endometrial stromal nodule, low-grade ESS, and high-grade stromal sarcoma.<sup>[6]</sup> Low-grade ESS shows <10 mitoses per 10 HPF whereas a high-grade ESS shows more than 10 mitotic figures per 10 HPF.<sup>[4]</sup>

The patients usually present with symptoms of abnormal uterine bleeding or pelvic pain or pressure symptoms. Some of the patients can be completely asymptomatic and are diagnosed incidentally when examined for some other complaint.<sup>[6]</sup> As the patient survival is directly proportional to the stage of tumor, an early diagnosis is essential for a better prognostic outcome. It is difficult to diagnose ESS in the preoperative phase as in approximately 75% of the cases; it is diagnosed as benign leiomyoma clinically and radiologically.<sup>[2,7]</sup> Imaging studies such as ultrasonography and magnetic resonance imaging are often inconclusive as it is diagnosed as a leiomyoma or a benign pelvic mass.<sup>[4]</sup> Endometrial curettage scrapes the superficial lining only and myometrium is not included; hence, the procedure is of little diagnostic help for definitive diagnosis of stromal tumors. ESS has a propensity to grow intramurally in the uterus rather than intracavitary portion as in endometrial carcinomas. Histologically, it is extremely difficult to differentiate between leiomyoma and ESS, especially low-grade ESS with a low mitotic index and a high-grade ESS from leiomyosarcoma. IHC using CD10 is essential and often helpful in diagnosis as it is strongly immunopositive in ESS and negative for later.<sup>[2]</sup>

According to the English literature, 5-year survival rate of low-grade ESS and high-grade ESS is 100 and 55%, respectively. ESS is malignant in nature and can spread to fallopian tubes, ovaries, bladder, vagina, ureters, and/or metastasize to distant organs such as lung, liver, or even heart. Approximately 30% cases of low-grade ESS have an extrauterine disease at the time of presentation; rarely, these tumors may initially present at other extrauterine sites, most commonly ovary.<sup>[4]</sup>

Total abdominal hysterectomy with bilateral salpingo-oophorectomy with pelvic and periaortic selective lymphadenectomy is the treatment of choice.<sup>[4]</sup> For stage 3/4 low-grade ESS and recurrent disease, hormonal therapy with medroxyprogesterone, tamoxifen, gonadotropin-releasing hormone analogs, and aromatase inhibitors has been suggested.<sup>[2,8]</sup> There will be no reoccurrence in 75% patients with stage 1 disease if treated with adjuvant medroxyprogesterone acetate as compared with 29% with same stage patients

in which adjuvant therapy with medroxyprogesterone is not given.<sup>[9]</sup> Long-term follow-up is essential in these tumors have a tendency of late recurrence and distant metastasis.<sup>[1]</sup>

## CONCLUSION

ESS is a rare uterine malignancy. Definitive diagnosis achieved only after histopathological examination. A clinical diagnosis of endometrial stromal tumors should be considered whenever a patient presents with bulky uterus without any symptoms. Early diagnosis and timely intervention are necessary for patient survival.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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