

# A rare case of primary extrauterine endometrial stromal sarcoma involving the sigmoid colon

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## Summary

Endometrial stromal sarcoma (ESS) is a rare type of endometrial mesenchymal neoplasm that commonly affects the uterus. The authors report an unusual case of sigmoid colonic ESS. A 46-year-old woman, complaining hematochezia and chronic abdominal pain, was diagnosed with ESS in the sigmoid colon. The patient underwent laparoscopic sigmoid colon resection and had no adjuvant therapy postoperatively. Two years later, the patient developed a new symptom of irregular vaginal bleeding with cervical myoma measuring about 9.0 cm, then a laparoscopic hysterectomy with a bilateral salpingo-oophorectomy was performed. Microscopically, the mass was diagnosed as adenomyosis and low-grade ESS. This case highlights the rarity of extrauterine ESS in the sigmoid colon, especially in the absence of a known history of primary uterine ESS. If the patient underwent prophylactic hysterectomy and adnexectomy after sigmoid colon resection, the recurrence would have been probably delayed.

**Key words:** Extrauterine endometrial stromal sarcoma; Sigmoid colon; Recurrent; Low grade; Primary.

## Introduction

Endometrial stromal sarcoma (ESS), a rare type of uterine mesenchymal neoplasm, accounts for less than 10% of uterine sarcomas [1]. ESS is classified into low- and high-grade based on differences in mitotic activity. Cells of the ESS are characterized by histological and genetic heterogeneities. Although ESSs commonly develop as primary endometrial tumors, several studies reported primary extrauterine ESS (EESS) occurring in the ovary, fallopian tube, vulva and vagina [2-4]. Very rarely does extrauterine endometrial stromal sarcoma (EESS) arise primarily in the gastrointestinal tract organs outside pelvis [5]. The clinical symptoms are non-specific, and it could be difficult to acquire a definite diagnosis for a rare site. Herein, we describe the rare case of primary EESS involving the sigmoid colon and no evidence of a primary ESS was established.

## Case Report

A 46-year-old female, gravida 3 para 1, with the history of cesarean and transabdominal myomectomy, was presented to hospital with hematochezia and chronic abdominal pain which had lasted for one year. The patient denied any symptoms or presentations of endometriosis and had no history of hormone replacement therapy. The colonoscopy showed polyps, but further evaluation with abdominal CT found an eccentric and irregular sigmoid wall thickening with mass measuring about 3 cm (Figure 1A). The tumor markers were negative. A laparotomy sigmoid colon resection was carried out and an exogenous polypoid mass measuring about 4 × 3 cm was identified. Intraoperatively, no enlarged lymph node was found and the other abdom-

inal organs had no metastasis. On immunohistochemical stain, the tumor cells were diffusely positive for CD10, vimentin, estrogen, and progesterone receptors, but negative for CD117, DOG1, desmin, CD34, and  $\alpha$ -inhibin. The Ki-67 labeling index was 30%. All the pathology results indicated a low-grade endometrial stromal sarcoma, involving the mucosa and muscularis (Figure 2A). The proximal and distal margins were negative for malignancy. After surgery, the patient had no additional treatment and was followed closely with examination, CT imaging, and endoscopy. Two years later, the patient developed a new symptom of irregular vaginal bleeding. CT showed adenomyosis and cervical myoma measuring about 9.0 cm (Figure 1B). Endometrial curettage was conducted and there was no abnormality of pathological result. Then, the laparoscopic hysterectomy with a bilateral salpingo-oophorectomy was performed. The uterus measured 8.0 × 5.0 × 4.0 cm, and upon cross-section, the uterine wall was thickened up to 2 cm and revealed a solid flaky tumor mass (2.0 × 1.5 cm). Microscopically, it was diagnosed as adenomyosis, an intramural leiomyoma, and a low-grade endometrial stromal sarcoma (Figure 2B). The cervical mass was diagnosed as myoma and the bilateral adnexa were normal. Postoperatively, the patient did not receive adjuvant therapy and was alive till now with no evidence of recurrence.

## Discussion

ESS occurs in middle-aged women, with a mean age ranging from 39 to 58 years [6]. ESS resembles endometrial stromal cells in their proliferative stage and is often of low-grade, slow-growing and indolent. Nevertheless, approx-

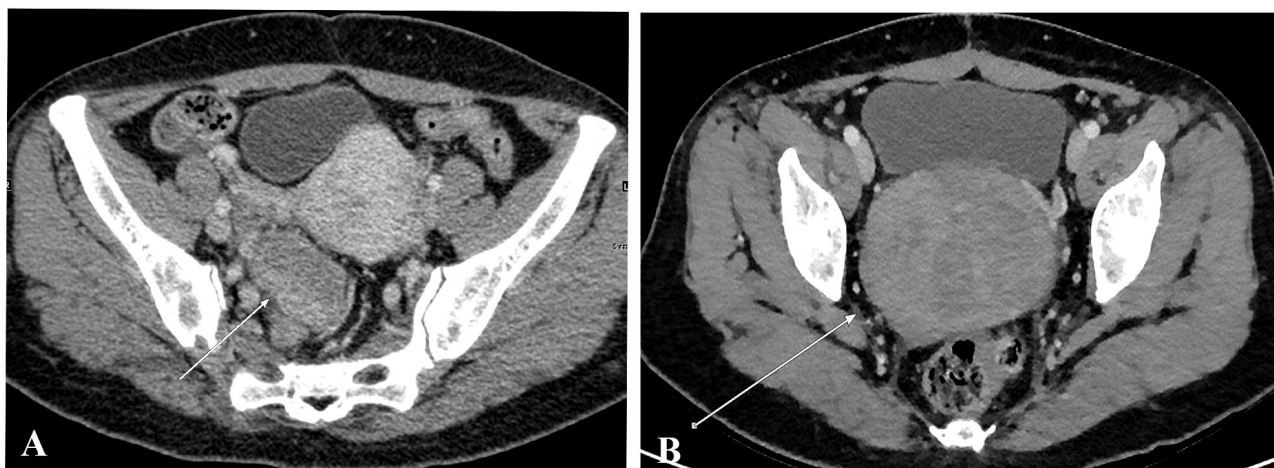


Figure 1. — (Greyscale and arrows). (A). Abdominal computed tomographic scan demonstrated thickening of sigmoid wall with inhomogeneous enhancement and a 3-cm mass involving the rectum and left ovary. (B). Pelvic computed tomography showing an enlarged uterus.

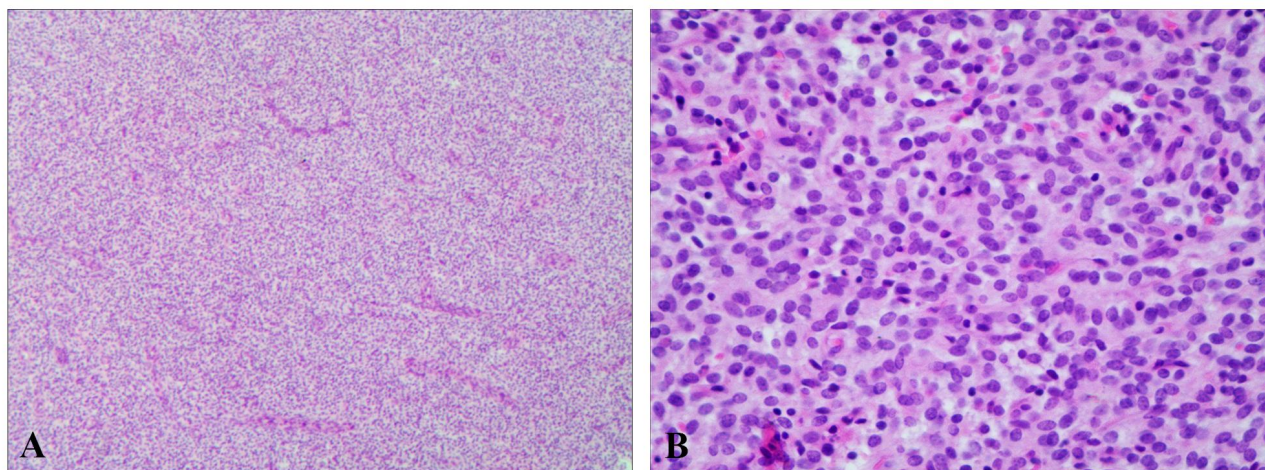


Figure 2. — (Colour images). (A). H&E stain at  $\times 40$  reveals that the tumor cells presents a widespread and deranged distribution, and conglomeration in sigmoid. (B). H&E stain at  $\times 400$  showing tumor cell nuclei that are large, irregular, and hyperchromatic.

imately 50% of women are diagnosed with recurrent disease. Primary EESS was more common in premenopausal women, suggesting a hormonal influence. The patients usually suffer from abnormal bleeding. However, in our study, the patient presented with chronic abdominal pain and was diagnosed with EESS. Abnormal pain was found to have the EESS in sigmoid colon. According to previous estimates, ovary was the most common involved area of EESS (76%), whereas the colorectum was involved in only 5% of cases [7]. Cho *et al.* illustrated that ESS of the sigmoid colon was in close relation with endometriosis, and other studies also supported this conclusion [8-11]. In 1925, Sampson described three criteria to aid in the diagnosis of malignancy arising in endometriosis: (1) close proximity of benign endometriosis to tumor, (2) no primary site of malignancy, and (3) tumor histology suggestive of an endometrial origin. Uniquely, this paper showed an un-expected case of

EESS which was not relevant to endometriosis.

Given the rarity of EESSs, there are no specific treatment guidelines in the clinical practice. Therefore, current treatment strategies are mainly based on previous case reports and treatment guidelines of ESS. Complete cytoreductive surgery was recommended, especially in patients for whom this surgery could result in being residual-disease-free. Tumor-free margin is important in prognosis. Adjuvant treatment for EESS is controversial with no prospective studies showing a survival advantage. The roles of radiation, chemotherapy, and hormonal therapy remained relatively unknown [12]. Despite this, some oncologists suggest that adjuvant therapy may be offered to patients with metastatic or recurrent ESS. Due to the near ubiquitous expression of estrogen and progesterone receptors, hormonal therapy is commonly used to suppress stromal proliferation [2]. Radiotherapy may serve a palliative role to relieve pain,

bleeding, and compression of surrounding organs [12]. The prognostic predictor of ESS is unclear. It is reported that low-grade ESS is associated with good prognosis and long overall and disease-free survival [13].

In the present case, the patient was premenopausal and only accepted partial sigmoid colon resection. Following the strict examination, the recurrence occurred in two years after the surgery. The authors conducted a secondary cytoreductive surgery and the patient refused further adjuvant therapy. Thus, the patient's outcome may reflect the natural history of EESS. Retrospectively, if the patient underwent prophylactic hysterectomy and adnexectomy after sigmoid colon resection, the recurrence would have been probably delayed. However, further research studies including larger cohorts of patients are warranted.

#### Ethics Approval and Consent to Participate

Informed consent was obtained from participant. The study was conducted in accordance with the Declaration of Helsinki and the protocol was approved by the Ethics Committee of Peking University People's Hospital (approval number was 184-01).

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#### Conflict of interest

The authors declare no conflict of interest.

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