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Original Research Article

Histopathological study of endometrial stromal sarcomas

Salma Bhat, Ambreen Beigh*, Summyia Farooq

Department of Pathology, GMC, Srinagar, Jammu and Kashmir, India

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***Correspondence:**
Dr. Ambreen Beigh,
E-mail: ambreenbeigh26@gmail.com

ABSTRACT

Background: Endometrial stromal sarcomas (ESSs) are rare malignant uterine tumours comparatively affecting younger women and the mean age is 42 to 58 years. The World Health Organization (WHO) classification categorises endometrial stromal neoplasms and related tumors as: endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma (UUS).
Methods: Present study is a retrospective one and includes 6 patients with histologically proven endometrial stromal sarcoma for a period of 3 years. Authors examined every slide available from each case and new HE-stained slides generated from formaline-fixed, paraffin-embedded tissue were reviewed to confirm the diagnoses. Demographic information, pathologic, and treatment information were collected from the clinic and hospital charts. All had primary surgical management in the form of total abdominal hysterectomy and salpingo-oophorectomy.
Results: The mean patient age was 41 years. All of the patients had presented with abnormal uterine bleeding. Diffuse growth of small cells closely resembling those of the normal proliferative endometrial stroma was the characteristic feature of these tumors. All of these patients had a low grade ESS on histopathology. They had regular follow-up visits until the end of study.
Conclusions: Endometrial stromal sarcomas are rare malignant tumors of the uterus and a proper preoperative diagnosis is difficult. Their differential diagnosis from typical submucosal uterine myomas or benign endometrial polyps can be difficult. The histological examination of the specimen is necessary to exclude malignancy and establish the final diagnosis.

Keywords: Endometrial, Histology, Sarcoma, Stromal

INTRODUCTION

Endometrial stromal sarcomas (ESSs) are rare malignant uterine tumors that make up approximately 10% of all uterine sarcomas but only around 0.2% of all uterine malignancies.[1] ESS comparatively affects younger women and the mean age is 42 to 58 years.[2] The World Health Organization (WHO) classification categorises endometrial stromal neoplasms and related tumors as: endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma (UUS).[3] The usual clinical presentation of ESS is abnormal uterine bleeding that occurs in about 90% of women and 70% cases show uterine enlargement. They can present with pelvic pain and dysmenorrhoea. An asymptomatic ESS occurs in 25% individuals.[4] About 30 to 50% of the ESS has extra uterine spread at the time of the diagnosis.[2] While LG-ESS is a clinically indolent neoplasm harbouring minimal

cytological atypia, infrequent mitotic figures, and numerous thin-walled small arteriolar type vessels, the undifferentiated endometrial sarcoma is a highly agressive tumor that lacks a plexiform vasculature, features severe cytological atypia, and has frequent often atypical mitotic figures.[5,6] In the uterine corpus, ESS characteristically shows prominent 'finger-like' myometrial infiltration with lymphovascular permeation. ESS in its 'classic' form is composed of a proliferation of small, round monomorphic cells with scanty cytoplasm and round to oval nuclei with smooth nuclear contours, resembling non-neoplastic proliferative-phase endometrial stroma. The mitotic index, size, stage, histological grade, positive surgical margins, menopause and age have been reported as potential prognostic parameters, but their use in ESS remains dubious.[7,8] In this study, the clinical and pathological features of endometrial stromal sarcomas are described to better define their characteristics and outcome.

METHODS

Inclusion criteria

- This observational retrospective study included patients with histologically proven endometrial stromal sarcoma for a period of 3 years (2015-2017).

Gross examination notes were found in the surgical pathology reports and clinical information was obtained from the Department of Obstetrics and Gynecology. Authors examined every slide available from each case and new HE-stained slides generated from formaline-fixed, paraffin-embedded tissue were reviewed to confirm the diagnoses. Endometrial stromal differentiation was recognized as small cells with scant cytoplasm and round to ovoid nuclei. The accompanying vasculature included spiral arteriole- like vessels, and staghorn vessels. Diagnosis of ESS were based on infiltrative margin and/or vascular invasion.

Exclusion criteria

- Cases of histopathologically proven endometrial stromal nodule and undifferentiated uterine sarcomas were excluded from the study.

Demographic information, pathologic, and treatment information were collected from the clinic and hospital charts. All had primary surgical management in the form of total abdominal hysterectomy and salpingo-oophorectomy. They had regular follow-up visits until the end of study.

RESULTS

Clinical features

The mean patient age was 41 years. All of the patients had presented with abnormal uterine bleeding. Clinical presentation of two cases were submucosal and intramural myoma. One of the cases was found incidentally after TAH+BSO performed for other reasons.

Gross features

The tumor ranged 4 to 6 cm (mean 5 cm) in maximal dimension and each grossly presented as a well-circumscribed mass and resembled leiomyoma macroscopically. Four tumors were polypoid, projected into the endometrial cavity, and described as "submucosal" masses. Two cases were described as "intramural"mass. The cut surfaces were fleshy and uniformly yellow-white. None of them had hemorrage, necrosis, cyst and ulceration (Figure 1).

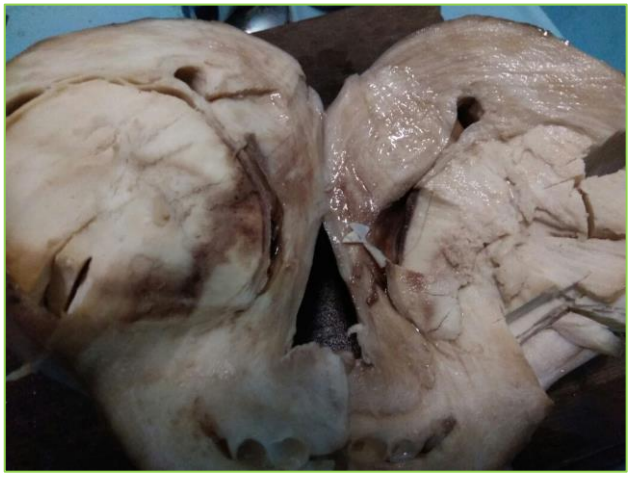

Figure 1: Endometrial stromal sarcoma presenting as a large mass occupying most of the endometrial cavity.

Microscopic features

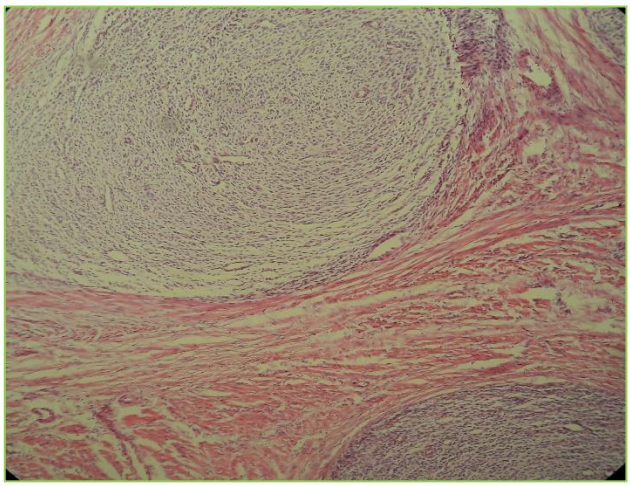

Figure 2: Nodules of neoplastic endometrial stromal cells in the full thickness of myometrium; the tumor cells demonstrate a tendency to make whorls around the arterioles.

Diffuse growth of small cells closely resembling those of the normal proliferative endometrial stroma was the characteristic feature of these tumors. The tumor cells were typicially oval to spindle shaped and small or medium with scant to occasionally more appreciable cytoplasm. The nuclei were uniformly oval to fusiform with inconspicuous nucleoli. No bizarre nuclei or mitotic figures were detected. Typical arterioles were numerous in two neoplasms. The tumor cells occasionally exhibited a tendency to make whorls around the arteioles (Figure 2). All of these patients had a low grade ESS on histopathology. Two LGESS cases were similar to ESN morphologically, despite showing myometrial and vascular invasion (Figure 3) and mitotic activity in one case.

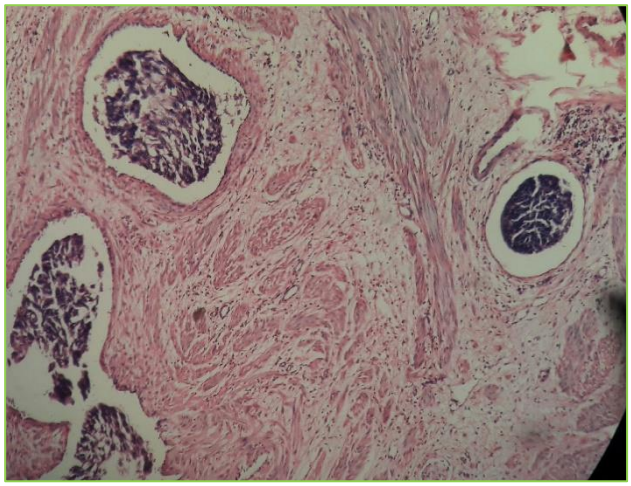

Figure 3: Lymphovascular invasion as seen in ESS.

DISCUSSION

Endometrial stromal sarcoma (ESS) is the second most common type of uterine sarcoma. In the latest World Health Organization classification, ESSs are classified into low-grade and high-grade subtypes.[3] Although infrequent, uterine sarcomas are among the most lethal of all uterine malignancies. The 5-year survival rate reportedly ranges from 30% to 68%.[7] ESS can be mistaken for a leiomyoma and the diagnosis is often made postoperatively after histological examination.[8] Authors report six cases of ESS in present study. Three patients in present study presented with irregular menstrual bleeding with hysterectomy performed for a leiomyoma. Two of our patients presented with postmenopausal bleeding. One of the cases was found incidentally after TAH+BSO performed for other reasons. All of these patients had a low grade ESS on histopathology. The mean age at presentation in present study was 41 years. Women with LGESS are younger than women with other uterine sarcomas, with a median age between 45 and 57 years.[1] Majority of low grade ESS occur in the perimenopausal period, occasionally arising in young women and adolescents. Patients usually present with vaginal bleeding or pelvic pain.[9]

Low grade endometrial stromal sarcoma is an infrequent tumor of the uterus. Its gross appearance may be that of a submucous polyp, or an intra-myometrial mass.[10] The classical gross appearance of an intra-myometrial mass is either a single nodule, multiple solid-cystic masses, or a poorly demarcated solid-cystic lesion.[11]

In the uterine corpus, ESS characteristically shows prominent 'finger-like' myometrial infiltration with lympho-vascular permeation (Figure 1). ESS in its 'classic' form is composed of a proliferation of small, round monomorphic cells with scanty cytoplasm and round to oval nuclei with smooth nuclear contours, resembling non-neoplastic proliferative-phase endometrial stroma (Figure 2 and 3).The mitotic index, size, stage, histological grade, positive surgical margins, menopause and age have been reported as potential prognostic parameters, but their use in ESS remains dubious.[12,13] The results of a study of 831 women with ESSs showed that age, race, stage, and grade of disease are important independent prognostic factors for survival. The survival of more than 90% in patients with grades 1 and 2 disease compared with only 42% in those with grade 3 disease supports the concept that low-grade ESSs have a significantly different clinical behaviour from high-grade tumors.[14] All our patients are doing well and are on follow-up.

CONCLUSION

Endometrial stromal sarcomas are rare malignant tumors of the uterus and a proper preoperative diagnosis is difficult. Their differential diagnosis from typical submucosal uterine myomas or benign endometrial polyps can be difficult. The histological examination of the specimen is necessary to exclude malignancy and establish the final diagnosis.

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Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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