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REVIEW ARTICLE

Reappraisal of Endometrial Stromal Sarcoma: Report of Four Cases with Review of Literature

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ABSTRACT

Endometrial stromal sarcomas (ESSs) are morphologically heterogenous and diagnosed by light microscopy in most instances. The distinction between smooth muscle neoplasms, such as cellular leiomyoma and low-grade ESS can be problematic. The diagnoses of ESS on the basis of systematic assessment of gross and histological parameters are highlighted.

Hysterectomy from four patients for a clinical diagnosis of leiomyoma was studied. Grossly, three had polypoidal lesion and in one myometrial widening with obvious permeation was noted. Microscopy showed features of ESS in three cases. Other case was diagnosed as cellular leiomyoma thought to be endometrial polyp. Reticulin stain was employed to highlight the characteristic spiral arterioles in ESS and thick-walled vessels in cellular leiomyoma.

Keywords: Endometrial stromal sarcoma, Polypoidal lesion, Reticulin stain.

INTRODUCTION

Endometrial stromal tumors are uncommon mesenchymal neoplasms of uterus which belongs to a unique group of neoplasms that are composed of a spectrum from benign to highly malignant tumors.[1] Low-grade endometrial stromal sarcomas (ESSs) are rare malignant tumors that comprise only about 0.2% of all female genital tract malignancies.[2] These neoplasms histologically resemble the normal proliferative phase of endometrium and usually diagnosed by light microscopy.[3] Despite its well-known good prognostic nature, sometimes low-grade ESS might behave as an aggressive malignancy and in such cases thorough clinical and gynecological evaluation with integrated approach is required.[4,5]

CASE REPORT

Four cases of ESS were diagnosed over a period of one year. Patients were aged between 35 and 70 years and they presented with mass per abdomen. Clinical diagnosis of leiomyoma was made in all cases.

Hysterectomy with bilateral salpingo-oophorectomy was received. Grossly, three had polypoidal lesion and in one myometrial widening with obvious permeation was noted (Fig. 1). Bilateral adnexa and cervix were grossly unremarkable in all cases. Table 1 shows gross, microscopy and histopathological diagnoses of four cases.

Hematoxylin and eosin stained sections showed small tumor cells with scant cytoplasm and round to ovoid nucleus resembling proliferative endometrium with characteristic spiral arterioles. These arterioles were highlighted by reticulin stain (Fig. 2). Myometrial and vascular invasion were seen in three cases (Figs 3A and B). Two of them showed foci of hyalinization and fibromyxoid changes respectively. Involvement of the cervix and metastasis to ovary were noted in other case (Fig. 4). The dense cellular proliferation of spindle cells along with elongated nuclei associated with thick-walled vessels on

Table 1 Gross, microscopy and histological diagnoses of four cases

Sl. no	*Age (years)*	*Hysterectomy gross*	*Microscopy*	*Histological diagnosis*
1	35	Fundal polyp with myometrial widening and obvious permeation	Tumor cells infiltrating myometrium, extending to cervix with ovarian metastasis	ESS
2	43	Fundal polyp	Islands of endometrial stromal cells in the myometrium with characteristic spiral arterioles. No pleomorphism and no mitosis	ESS
3	36	Polypoidal mass from fundus projecting into cervix		
4	70	Fundal polyp	Oval to spindle cells arranged in fasciles with smooth muscle differentiation	Cellular leiomyoma

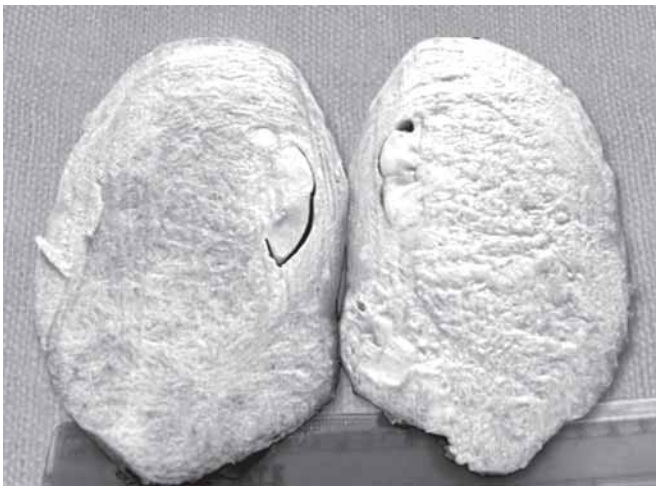

Fig. 1: Gross photograph showing endometrial polyp with myometrial widening and obvious permeation

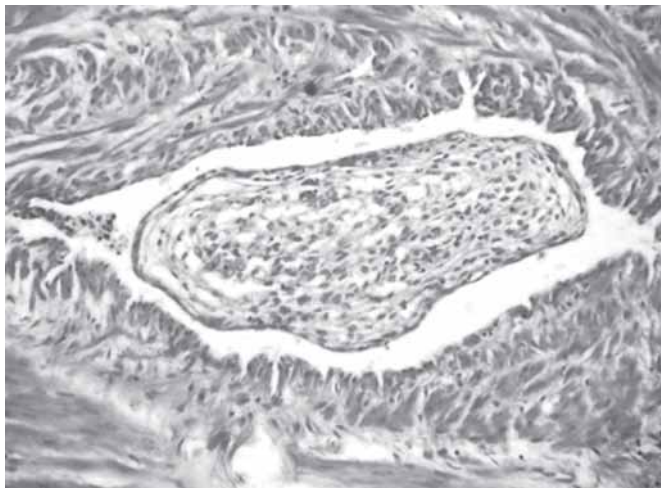

Fig. 3B: Microphotograph showing vascular invasion by tumor cells (H&E, ×100)

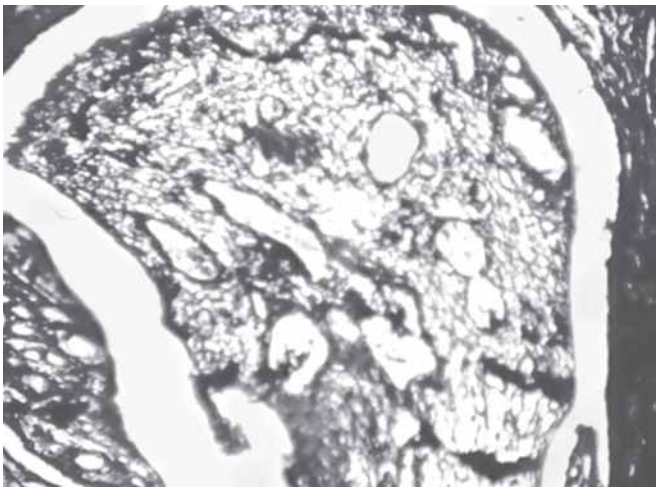

Fig. 2: Microphotograph highlighting spiral arterioles (reticulin stain, ×100)

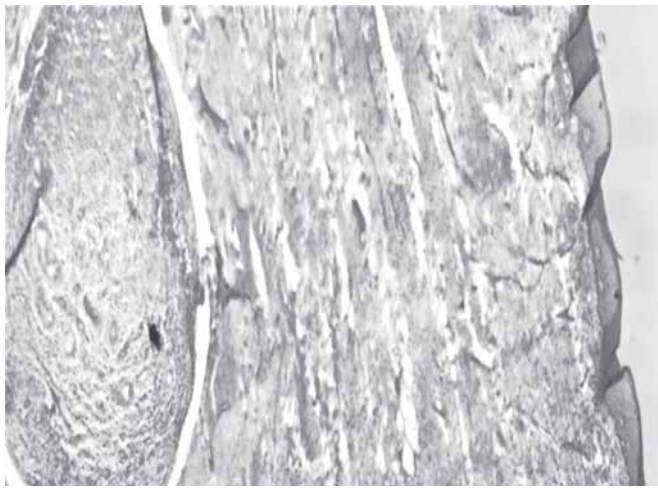

Fig. 4: Microphotograph showing tumor cells extending into cervix (H&E, ×100)

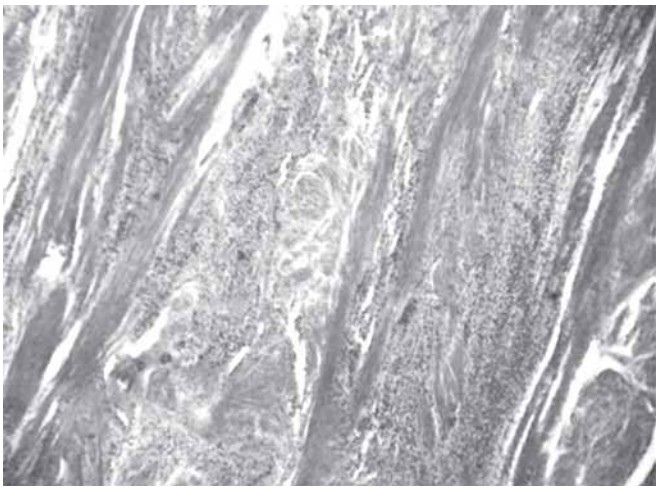

Fig. 3A: Microphotograph showing small tumor cells invading myometrium (H&E, ×100)

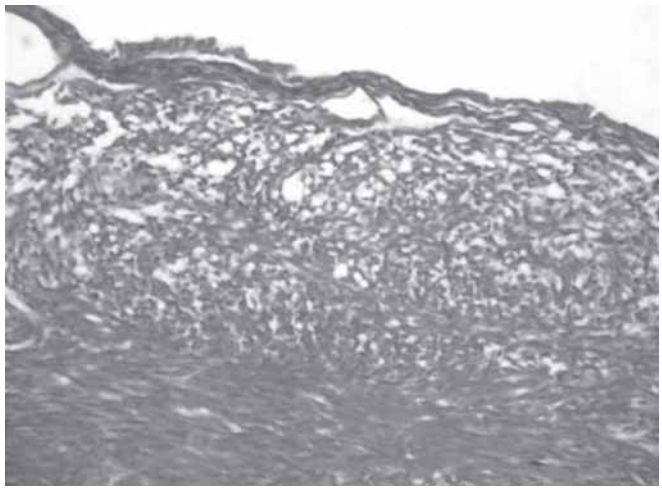

Fig. 5: Microphotograph showing polypoidal lining and spindle cells arranged in fascicles consistent with cellular leiomyoma (H&E, ×100)

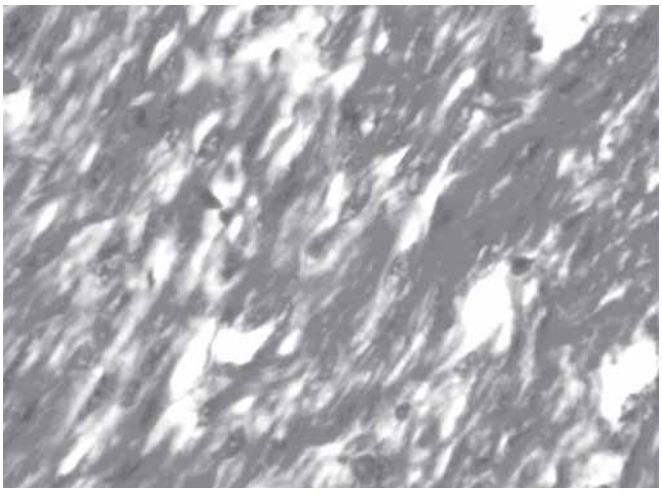

Fig. 6: Microphotograph showing smooth muscle differentiation consistent with leiomyoma (Massion trichrome, ×400)

reticulin stain suggested cellular leiomyoma in one case though to be endometrial polyp (Fig. 5). Masson trichrome stain (MTS) was also done to highlight muscle bundles (Fig. 6). No tubules, glandular structures or rhabdoid cells were identified. So, three cases were diagnosed as low-grade endometrial stromal sarcoma, fourth case as cellular leiomyoma.

DISCUSSION

Uterine sarcomas, such as leiomyosarcoma, endometrial stromal sarcoma and mixed mullerial tumor constitute 2 to 5% of all uterine malignancies.[5] ESS is a neoplastic process developing from endometrial stromal cells. Neoplastic cells may originate from endometrial tissue but they may also originate from pathologic processes, such as adenomyosis and endometriosis. It can be de novo, derived from pluripotent mullerial cells. In our case, ESS was not associated with either adenomyosis or endometriosis. This is in contrast to case reported by Berkowitz et al where low-grade ESS was associated with endometriosis.[6] Depending on the mitotic activity, endometrial stromal sarcoma is classified as low- or high-grade ESS. Although, half the low-grade ESSs are limited to the endometrium, the other half shows focal worm-like, or diffuse multiple nodular permeations in the myometrium from endometrial foci as in one of our cases.[4] Low-grade ESS tends to occur in a younger age group (mean 39 years) as in our cases. It leads to same symptoms as those of any other uterine sarcoma or endometrial carcinoma, such as mass per abdomen, abnormal bleeding per vagina.[5]

Although uterine sarcomas are described as aggressive neoplasm, low-grade ESS has a low potential for spreading. It can spread to the vagina, fallopian tubes, uterine ligaments, ovaries, bladder and ureter.[5] In one of our cases diffuse myometrial permeation of fallopian tube, ovary and cervix without any interruption was seen.

Morphological variations and histological novelties have been described in ESSs include smooth muscle, sex cord-like differentiation, fibrous and fibromyxoid changes. Other uncommon findings include endometrioid type glandular structures, skeletal muscle differentiation, rhabdoid cells, clear cells, ossification and osteoclast-like giant cells. There have been comparatively fewer descriptions of ESSs with fibrous and myxoid features.[7] In two of our cases foci of hyalinization and extensive fibromyxoid changes were seen.

In one of our cases, thought to be ESS, more than 70% of the tumor was showing smooth muscle differentiation, so diagnosis of cellular leiomyoma was considered. Reticulin stain highlighted thick-walled vessels. MTS showed smooth muscle differentiation. Our findings support those of Baker et al.[8] The clinical, gross pathologic and microscopic features of our cases are compatible with those reported in the literature.

CONCLUSION

Endometrial stromal sarcomas are clinically indolent malignancies with minimal cytologic atypia and proliferative activity with infiltrative margins. They may manifest as polyps. Histologic features recapitulate the gross appearance. A characteristic vascular pattern is helpful in differentiating from cellular leiomyoma. Simple special stains can be employed. Thus, in most instances diagnosis of ESS may be established on morphology alone by paying attention to diagnostic features. Integrated approach should be employed only in difficult situations.

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