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.

Table 1

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

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

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

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

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

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

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

REFERENCES


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