CASE REPORT

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Endometrial stromal sarcoma developed on outer endometriosis foci

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Abstract

Introduction: Endometrial stromal tumor is a rare mesenchymal uterine tumor. Endometriosis, an affection of the women during her fertile period, is reported in the pertinent literature to have an incidence varying from 1% to 53%, which shows a wide difference from one study to another. The pathology that grafts onto the endometriosis foci is rare, and the malignant tumoral pathology is extremely rare. The most frequent malignant tumors quoted in the pertinent literature are of the adenocarcinoma type, on the endometriosis, and the alterations of the sarcomatous type can appear extremely rarely. Most sarcomas appear on the foci of the rectosigmoid endometriosis. These rare tumors should not be taken for the mesenchymal gastrointestinal tumors. The recurrence of the malignant alterations of the endometriosis foci is estimated at about 0.7–1% in the pertinent literature. *Materials and Methods*: We are presenting a case of endometrial stromal sarcoma, developed on foci of outer endometriosis (sigmoid and appendicular endometriosis), diagnosed in our laboratory on two operatory pieces taken from the same woman, at a 2-month interval. The macroscopic and microscopic study was done on operatory pieces, with the routine procedure, macroscopic examination, fixing in 10% formalin, embedding in paraffin and staining with Hematoxylin–Eosin and than we used immunohistochemical markers, for endometrial stroma. *Result*s: The endometrial stromal sarcoma developed on foci of endometriosis is very rare, has an incidence of 6% within the endometrial malignant tumors, with various degrees of malignancy, low malignancy, or high malignancy. *Conclusions*: We do not exclude the appearance of the endometrial sarcoma on the outer endometriosis foci. Moreover, the endometriosis foci can be considered as pre-malignant conditions.

***Keywords*:** endometrial stromal sarcoma, endometriosis, CD10, estrogen and progesterone receptors, vimentin, desmin.

Introduction

Endometriosis is an ectopic localization of endometrial tissue. The inner endometriosis (adenosis, adenomyosis), located in the uterine wall or the outer one (on the pelvic peritoneum, the uterine ligaments, the ischiorectal ligaments, the intestinal wall, the sigmoid, the rectum, the ovaries, the urinary bladder) are quite frequent. In appears very rare in lymph nodes, the umbilical area, post-operatory scars, hernia sacci, the appendix as well as in the lung and the pleura [1, 2]. The mechanisms of endometriosis appearance are varied:

- Tube endometrium retrograde seeding (during menses with tube reflux);
- Endometrium seeding via the blood or the lymph;
- Aberant differentiation of the coeloma epithelium;
- Adenosis is the presence of some remains from the Wolfian channel or from Garthner channel. For this pleads the presence of adenosis on the malformed ureters or of the newborns;
- The hereditary character of adenosis has also been considered in the specialty literature;
- Cullen claims that adenosis is the result of the invasion of the myometrium by the basal layer of the endometrium and is favored by hyperestrogenism and the uterine trauma [3, 4]).

Macroscopically, the endometriosis foci have an appearance of small, brown-reddish nodules, occasionally with a micro-cystic aspect and/or hemorrhage areas.

Microscopically, it is actually an endometrial tissue, made up of endometrial glands and cytogenous chorion. Recent hemorrhages, hyalinizations, fibroses, adherences may be noticeable around the lesion. The endometriosis foci suffer alterations related to the ovarian cycle, which are less obvious in the secretory stage and more obvious in the proliferation stage. There can appear glandular-cystic hyperplasias, decisive transformations during pregnancy [5]. It is frequently associated with uterine fibroleiomyoma but only occasionally with endometrium adenocarcinoma and sarcoma.

Materials and Methods

We study the operatory pieces from E.G., 51-year-old women, when she was operated for a sigmoid tumor, and 2 months later, on the second check-up, when a new appendicular tumor has been extirpated.

The patient's pathologic history includes two surgical interventions, performed 9 years before, at a distance of one month. The first surgery was performed for a bilateral ovarian tumor, and the second for a rectal one. Histopathologically, the diagnosis of endometrial stromal hyperplasia on endometriosis foci was set, which was plausible, considering the 9-year course since the first surgical intervention. The operatory pieces from the last two surgical interventions consisted of two tumors, a presigmoid one and a peri-appendicular one, the former of 5 cm, relatively encapsulated, on a whitish section, increased consistency, while the latter was an appendix with a wall-adherent tumor, encapsulated, of 3 cm, with the same macroscopic characteristics as the presigmoid tumor.

The pieces was fixed in 10% formalin, embedded in paraffin, and stained with Hematoxylin–Eosin, than, we used immunohistochemical markers for differential diagnosis with mesenchymal intestinal tumors (gastro-intestinal stromal tumors). In order to identify the pathology of the tumoral cells the immunostains was made: CD10, vimentin, desmin, estrogen and progesterone receptors, alpha-inhibin, CD99. CD10 immunoreactivity is a useful immunohistochemical marker of normal endometrial stroma and of endometrial stromal neoplasms. CD10 immunoreactivity is present in ectopic endometrial stroma, may be of value in confirming a diagnosis of endometriosis.

Results

The microscopic images of the two tumors ,with Hematoxylin–Eosin staining was similar, being represented by a tumoral proliferation of uniform, round (Figures 1 and 2), or fusiform cells, which reminds the cells of the endometrial stroma, with microscopic areas in which mitoses are rare, but also with areas in which mitoses overpass 10 per 10 microscopic fields. The rare microscopic fields with small vessels (Figures 3), hyaline areas, and areas in which glandular structures are present, similar to endometrial glands.

Immunoprofile

The neoplastic cells are immunoreactive for CD10 (Figure 4), vimentin and focal actin. They are negative for desmin. They are positive for estrogen and progesterone receptors (Figures 5 and 6).

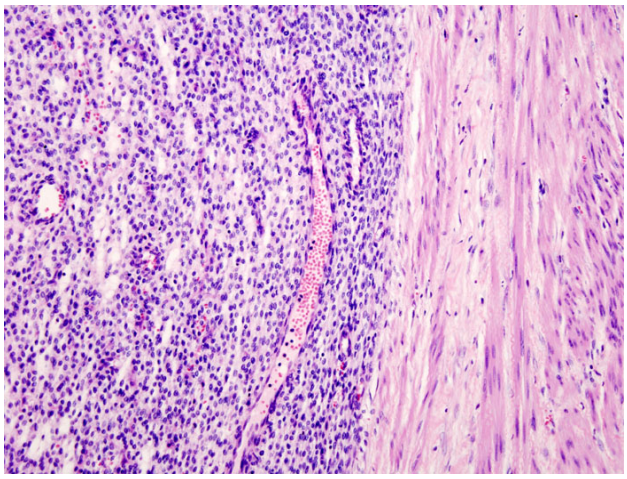

Figure 1 – ***Endometrial stromal sarcoma: low grade, uniform, round cells (HE stain, ×200).***

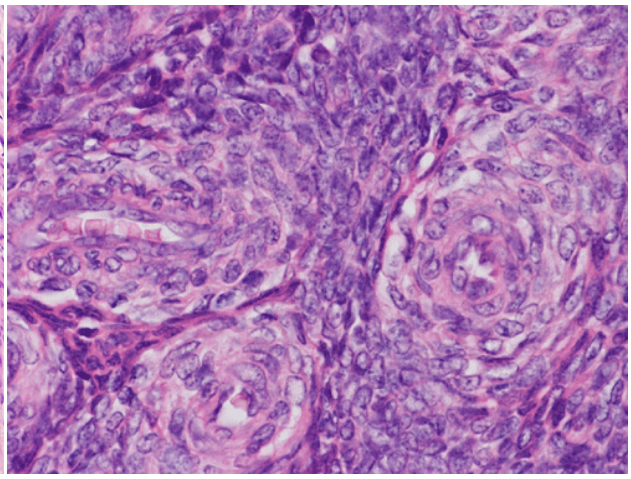

Figure 2 – ***Endometrial stromal sarcoma: low grade, hyaline vessels (HE stain, ×400).***

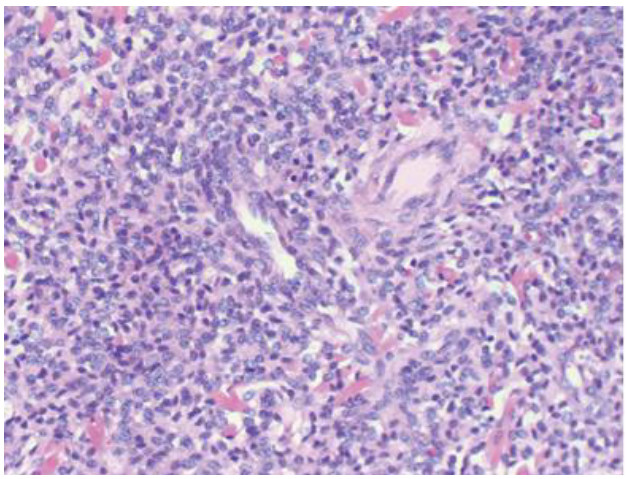

Figure 3 – ***Endometrial stromal sarcoma: round cells and small vessels (HE stain, ×200).***

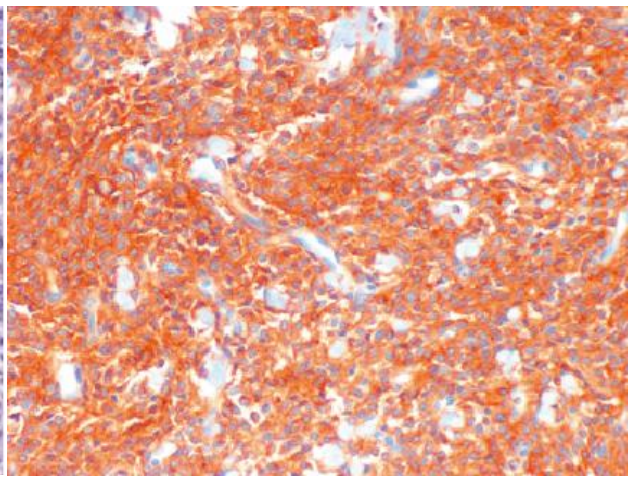

Figure 4 – ***Endometrial stromal sarcoma: CD10 (diffuse positivity).***

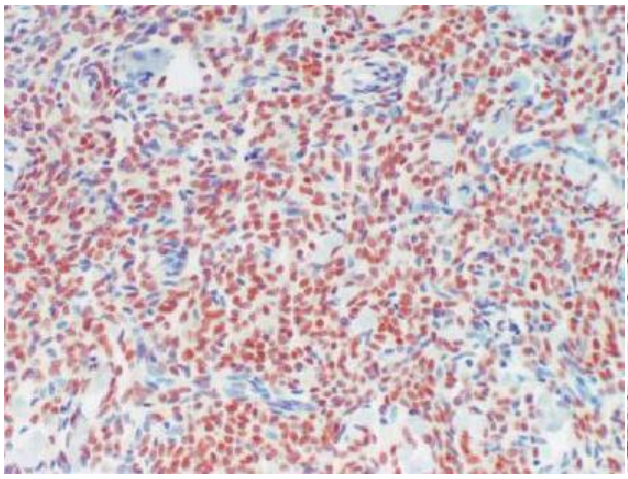

Figure 5 – ***Endometrial stromal sarcoma: estrogen receptors (×200).***

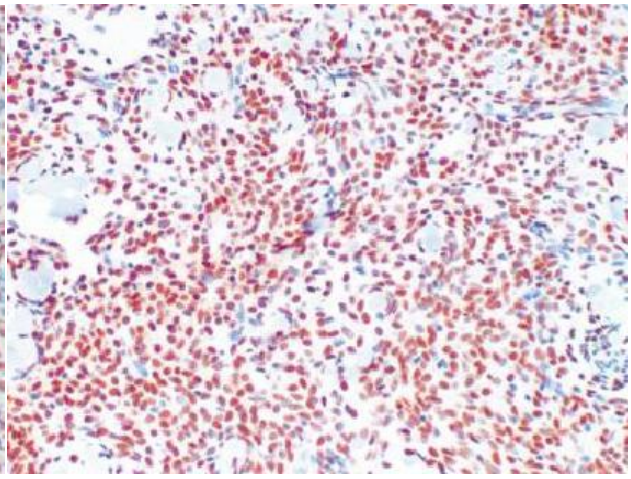

Figure 6 – ***Endometrial stromal sarcoma: progesterone receptors (×200).***

Discussion

The tumors of the endometrial stroma are specific to the age of over 45 years [6]. The benign variant is the stromal nodule. Malignancy of the sarcoma type of the endometrial stroma can have two degrees: low grade and high. Diagnosis of undifferentiated endometrial stromal sarcoma is made on pleomorphism and necrosis intratumoral. Endometrial stromal sarcomas are composed of cells resembling those of proliferative endometrial stroma, subdivided into benign and malignant groups based on the type of tumor margin [7, 8]. Those with pushing margins are benign stromal nodules and those with infiltrating margins qualify as stromal sarcoma. Mitotic index of 10 or more per 10 high power fields is an adverse prognostic finding [9]. Immunoprofile of neoplastic cells in stromal nodule and low grade of endometrial stromal sarcoma are immuno-reactive for vimentin, CD10, and focal actin. Usually, they are negative for desmin and h-caldesmin. Low grade of endometrial stromal sarcoma is almost positive for estrogen and progesterone receptors. Rarely, low grade displaying a sex cord pattern may be positive for alpha-inhibin, CD99, desmin [10–13].

In our case, the presence of endometrial glands without periglandular stromal condensation and IHC characteristics such as negative staining for smooth muscle actin, S-100 protein, CD34 were not consistent with GIST or müllerian adenocarcinoma with sarcomatous overgrowth.

Yantiss RK *et al.* [14] described a large series of neoplastic and pre-neoplastic lesion of gastrointestinal endometriosis among the seventeen cases (endometrial stromal sarcoma was only one).

Our case is different because the tumoral pathology appears on multiple foci of endometriosis. In their turn, these foci have had a progressive pathological course, namely, in 9 years they passed from aspects of nodular stromal hyperplasia with ovarian and rectal localization to sarcoma of the endometrial stroma, with various degrees of histological differentiation.

Conclusions

The case presented by us seems to confirm the opinion of some authors who suggest that endometriosis should be considered a pre-malignant lesion.

The endometrial stromal sarcoma of the intestinal tract arising in endometriosis should always be included in the differential diagnosis of small round cell or spindle cell tumors of the intestinal tract. The distinction from GIST should be considered.

The rectosigmoid is an area of bowel having the highest incidence of endometriosis.

Immunoreactivity for vimentin, CD10, estrogen and progesterone receptors confirm the diagnosis of endometrial stromal sarcoma.

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