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Bilateral Endometrial Stromal Sarcoma Originating from Ovarian Endometriosis: A Case Report

Endometrial stromal sarcomas usually develop in the uterine corpus whereas extrauterine sites are extremely rare. This tumor arising from extrauterine endometriosis is a rare and not easily diagnosed tumor. Only few reports are available of these tumors. The pathogenesis of this tumor is not so clear yet, but many authors believe that heterotopic mullerian ducts are the origin of these tumors.

Progesterone receptor A and B isoforms have been reported in low-grade extrauterine endometrial stromal sarcoma. This tumor has solid components at the peripheral and cystic components at the central part of the mass ultrasonographically. In addition, CT scan shows peripheral enhancement, so ovarian malignancy should be excluded. Besides it can be confused with several soft tissue neoplasms.

In every patient with endometriosis, malignant changes should be in differential diagnosis of a new pelvic mass. These tumors should not be confused with soft tissue adenocarcinomas like smooth muscle tumors and fibrous tumors. Recurrence and prognosis of low-grade endometrial stromal sarcoma are related by extraterine development but not by mitotic activity or DNA content.

Herein we report on a case of this extremely rare condition.

Case Presentation

A 57-year-old woman who was menopause from 5 years ago presented with a two–month history of pelvic pain and vaginal bleeding. Except for the mentioned symptoms there was no other positive medical history. She has had intermittent and repeating pelvic pain from about 6-7 years earlier without any specific evaluation and therapy, but her pelvic pain had been more obvious for the last 2 months. Vaginal examination revealed bilateral palpable pelvic masses about 10 cm in size. Physical examination was otherwise normal. Routine
laboratory exams and the chest radiogram were normal.

The uterus was atrophic with 2 mm endometrial thickness and no masses. There were large masses (10×9×6cm) in both ovaries with cystic and solid components detected in the transvaginal and transabdominal ultrasonography. Ascites and no other abnormality were detected in the abdomino-pelvic sonoography. Large masses (in the above measurements) with peripheral enhancement and central cystic components were seen in the CT scan of both ovaries with IV contrast and no other abnormalities were detected in the abdomino-pelvic CT scan (Fig. 1).

Our preoperative diagnosis was malignant neoplasm of the ovaries but an exact diagnosis was not possible. Also in the complementary laboratory tests, our patient’s CA125 level was above the normal limit.

The patient underwent total abdominal hysterectomy (TAH) and bilateral salpingo-oophorectomy (BSO) and complete excision was performed. In the pathologic evaluation, chronic cervicitis, atrophic endometrium, adenomyosis and endometriosis in both ovaries were noted. Malignant transformation of endometriosis was reported in both ovaries. The histologic features were typical of low-grade endometrial stromal sarcoma. The final diagnosis was extrauterine low-grade endometrial stromal sarcoma of the ovary (Figs. 2&3).

Post-operative ultrasonography of the abdomen and pelvis were normal. The patient remained free of any recurrence 15 months after surgery.

Discussion

Aggressive endometriosis raises important diagnostic and therapeutic difficulties and may correspond to misdiagnosed rare malignant neoplasm, which should be treated.2

Endometrial stromal sarcomas are low-grade malignant tumors that may pose a diagnostic challenge, especially when they are present in an extrauterine site.

Endometrial stromal sarcoma arising from extrauterine endometriosis is a rare and not easily diagnosed tumor.3-5 This tumor is of highly recurrent nature.3

It has been noted that cancers are more commonly found in ovaries when endometriosis is present in the ovary (5%) compared to when endometriosis is present at other sites (1%).

Only few reports are available on this type of tumor.3

In our patient, the clinical findings were nonspecific and consisted of both benign and malignant ovarian masses. Pathogenesis of endometrial stromal sarcoma is not so clear yet, but it may arise from heterotopic mullerian ducts.6

Solid components at the peripheral part, and cystic components at the central part of the mass are important for making the diagnosis ultrasonographically. Also, the peripheral enhancement in CT scans with IV contrast is diagnostic but can be confused with soft tissue neoplasms.8,9

Histologically, the tumor is characterized by infiltration and diffuse proliferation of uniform round and oval cells, abundant small vessels, low mitotic activity, presence of foam cells and vascular invasion.6

These tumors should not be confused with smooth muscle tumors and fibrous tumors.8

Recurrence and prognosis of low-grade endometrial stromal sarcomas are related to extrauterine development. Initial and recurrent tumors show the same configuration polycystic parts show in solid tumors.8,9

In a patient with a history of endometriosis, new symptoms, especially pelvic masses—with a frequency of 3-10%—must arise suspicion of extrauterine endometrial stromal sarcoma.
Although this tumor is very rare (the prevalence is approximately 0.2 per million women and about 75% are postmenopausal at diagnosis), the diagnosis and treatment is very important, because it can lead to long survival for the patient—a 5-year survival rate of 100%. Most patients require no further therapy after complete surgical resection. The recurrence rate is very low and the pelvic cavity is the main site of recurrence, followed by the vagina and the lung. In general, extraterine endometrial stromal sarcoma may develop from the ovary, fallopian tubes, pelvis, colon, or retroperitoneum.

The clinical and imaging appearances are similar to those of other ovarian and soft tissue masses, so in order for a correct diagnosis, pathologists should be alert to the previous history of the patients. Clear cell and endometrioid carcinomas are the malignancies most commonly seen in ovaries containing endometriosis, while clear cell adenocarcinoma and adenosarcoma are most commonly seen in conjunction with extraovarian endometriosis. The association of endometriosis with endometrial and clear cell carcinoma is much stronger than that of serous and mucinous tumors.

Both clinical and paraclinical findings are not definite for the diagnosis. All of the mentioned facts suggest the possibility of endometrial stromal sarcoma in the setting of a woman with a history of endometriosis and a pelvic mass, when there are masses with solid and cystic components in her imaging. With the suggestion of endometrial stromal sarcoma on sonography and CT scan, additional study is required to obtain a definite pathological diagnosis. This was a clinicopathological diagnosis with imaging correlation.

After taking the history, physical examination, ultrasonography, CT scan and then suspicion of a malignant bilateral ovarian mass, recurrent pathologic evaluation with an exact diagnosis was established.

References


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