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Endometrial stromal sarcoma: clinicopathological and immunophenotype study of 18 cases

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Abstract Malignant tumors of the uterine corpus are uncommon. They originate from the endometrial stroma, smooth muscle, blood vessels, or from a mixture of them. The objective of this article was to know the frequency and the clinical, morphologic, and immunophenotype characteristics of the endometrial stromal sarcoma (ESS). We reviewed the cases of ESS observed from 2002 to 2008 at the Pathology Unit of the General Hospital of Mexico. The following data were analyzed: age, clinical stage, degree of differentiation, and immunophenotype. We found 18 cases, and the average age of patients was 48.6 years; 66% were in clinical stages 1 and 2. Fifteen cases (83.3%) were classified as low-grade sarcomas and 3 (16.6%) as high-grade or undifferentiated sarcomas. We determined immunohistochemical markers in 17 cases; receptors to estrogens were positive in 5 (29.4%) and to progesterone in 9 (52.9%). CD10 was expressed in 10 (58.8%) and p53 in 11 cases (64.7%). Two cases were associated to primary tumors of the ovary (papillary cystadenocarcinoma). In conclusion, ESS was present at 0.6% in our institution; and most were low grade. Expression of markers, such as p53, CD10, and hormonal receptors, was positive.

Keywords: Endometrial stromal sarcoma

1. Introduction

Mesenchymal malignant tumors of the uterine corpus are rare; they present less than 1% of the neoplasms with this location [1]. The most frequent ones are the leiomyosarcomas and the tumors of the endometrial stroma. Sarcomas of the endometrial stroma are constituted by cells that resemble the proliferation stage of the endometrium, with numerous small plexiform arterioles [2]. Clinically, they present as an increase in the uterine size, abnormal bleeding, and/or pelvic pain. Their cause is unknown; racial hormonal factors as well as radiotherapy antecedents have been suggested to play a role in their development.

Histologically, the World Health Organization classifies them based on tumoral borders and the number of mitoses [3,4] in low-grade and high-grade sarcomas. The first are characterized by presenting nodules of well-delimited borders; malignant ones present infiltrating borders and are called *sarcomas of the stroma* [5].

Low-grade endometrial stromal sarcomas (ESSs) present an insidious clinical course in contrast to the undifferentiated endometrial sarcomas that are highly aggressive neoplasms.

The objective of the present article is to know the frequency and the clinical, morphologic, and immunophenotypic characteristics of ESSs studied in 18 patients.

2. Material and methods

We reviewed the surgical pathology files at the Pathology Unit of the General Hospital of Mexico corresponding to the years from 2002 to 2008. We found 18 cases of ESS; we analyzed in these cases the age of the patients, tumor size, clinical stage (TNM classification), and the clinical evolution of the patients. We reviewed in the hematoxylin-eosin–stained sections the characteristics of the tumoral borders,

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Table 1
Total number of uterine tumors analyzed per year and number of diagnosed ESSs

Year	No. of uterine tumors	No. of ESSs
2002	297	3
2003	261	4
2004	442	1
2005	505	1
2006	483	4
2007	370	4
2008	325	1
7 years	Total, 2683	18 (0.6%)

the number of mitoses, and the presence of necrosis and venous or lymphatic invasion. Endometrial stromal sarcomas were classified as low grade based on the number of mitoses when less than 10 mitoses to high power were found. High-grade or undifferentiated sarcomas contained more than 10 mitoses to high power. In 17 cases, we performed immunohistochemistry using the manual avidin-biotin-peroxidase technique. We used the following monoclonal antibodies: CD10 (1:20), estrogen receptors (1:50), progesterone receptors (1:50), and protein 53 (1:50), all from DAKO (Carpinteria, Calif).

The antibodies were subjected to antigenic recovery with citrate buffer at 99°C during 10 minutes in a pressure cooker, and diaminobenzidine was used as revealing stain.

3. Results

Tumors of the endometrial stroma are rare; they represent less than 1% of the total of all mesenchymal neoplasms of the uterine corpus (Table 1).

The age of patients, the clinical stage, and the anatomopathological characteristics are summarized in Table 2. Average age of patients was of 48.6 years, ranging from 32 to 71 years. Clinical stage 1 was found in 9 cases (50%); stage 2, 1 case; stage 3, 2 cases; stage 4, 2 cases; and unknown, 4 cases (Table 2).

Size of the tumors varied from 1 to 54 cm, and half of them measured more than 8 cm. Adenomyosis was observed in 3 cases. Atypical nuclei and pleomorphism were present in 3 cases (27%) of the low-grade sarcomas.

Fifteen cases (83.3%) were classified as low-grade ESSs (Fig. 1), and 3 (16.6%) were undifferentiated because they had more than 10 mitoses high power in 10 fields (Fig. 2). Hemorrhage and necrosis were present in 10 cases (8 low-grade ESS and 2 undifferentiated) (Figs. 3 and 4), and venous and lymphatic invasion was observed in 1 low-grade ESS case.

Invasion to the myometrium was present in the 18 studied cases; in 7 (38.8%), an almost 50% invasion to the uterine muscle was observed; in 2, the invasion was up to 80%; 2 cases showed a 90% invasion reaching the endocervix (Fig. 5); and in 7 more, minimal myometrial invasion was observed.

Metastases were found in 3 patients in the iliac lymph nodes, the sacrum bone, and the lung. Five tumors (29.4%) were positive to estrogen receptors (4 low grade and 1 undifferentiated); positivity to progesterone receptors was found in 9 (52.9%) (8 low grade and 1 undifferentiated) (Fig. 6). CD10 was positive in 10 (58.8%), 8 low grade and 2 undifferentiated (Figs. 7 and 8). p53 was positive in 11 cases (64.7%), 10 low grade and 1 undifferentiated (Table 3).

In 9 patients, no tumor activity was present for a period of 1 to 5 years; 5 patients presented tumor activity 1 year after the diagnosis, and 2 died due to a primary tumor of the ovary

Table 2
Clinicopathological characteristics of ESSs

Age (y)	Tumor size (cm)	AD	P/A	M	G/H	Ne/He	V/L	IM (%)	Mt	EC	Other findings
64	8	No	Yes	13	High	Yes	No	50	No	1	Without AT, 36 mo
40	12.5	No	Yes	7	Low	Yes	Yes	80	No	2	Without AT, 48 mo
46	D	Yes	Yes	12	High	Yes	No	90	No	1	With AT, 24 mo
58	D	No	Yes	1	Low	No	D	M	D	D	D
50	4	No	No	4	Low	Yes	D	M	D	D	Without AT, 18 mo
57	3.4	No	No	0	Low	Yes	No	90	No	1	With, AT 12 mo
68	13	No	No	20	High	No	D	M	D	4	With AT, 12 mo
34	4	No	No	0	Low	No	No	50	No	1	Without AT, 5 mo
36	8	Yes	No	0	Low	No	No	50	No	1	Without AT, 24 mo
71	4.3	No	No	3	Low	Yes	No	50	GL	2	With AT, 24 mo
46	2	No	No	2	Low	Yes	No	M	Sacrum bone	3	Without AT, 12 mo
45	1	No	No	0	Low	No	D	M	D	D	D
33	D	No	No	0	Low	Yes	D	M	Lung	4	With AT, 12 mo
51	9	No	No	2	Low	Yes	No	M	No	1	CACO death
54	2.1	Yes	No	1	Low	No	No	50	No	1	CACO death
32	1.3	Yes	No	2	Low	Yes	No	50	No	1	Without AT
54	7	No	No	0	Low	no	No	50	No	1	Without AT, 12 mo
37	D	Yes	Yes	1	Low	no	No	80	No	1	Without AT, 12 mo

Cases 14 and 15 died due to ovarian tumor. AD indicates adenomyosis; P/A, pleomorphism and atypical nuclei; M, mitosis; G/H, histological grade; Ne/He, necrosis and hemorrhage; V/L, vascular and lymphatic invasion; IM, muscular invasion; EC, clinical stage; AT, tumoral activity; D, unknown; M, minimal invasion; Mt, metastases; GL, lymph nodes; CACO, ovarian carcinoma.